

**UNITED STATES
SECURITIES AND EXCHANGE COMMISSION
Washington, D.C. 20549**

**FORM S-1
REGISTRATION STATEMENT**

*UNDER
THE SECURITIES ACT OF 1933*

AKEBIA THERAPEUTICS, INC.

(Exact name of registrant as specified in its charter)

Delaware
(State or other jurisdiction of
incorporation or organization)

2834
(Primary Standard Industrial
Classification Code Number)
245 First Street, Suite 1100
Cambridge, MA 02142
(513) 985-1920

20-8756903
(I.R.S. Employer
Identification Number)

(Address, including zip code, and telephone number, including area code, of registrant's principal executive offices)

John P. Butler
President and Chief Executive Officer
Akebia Therapeutics, Inc.
245 First Street, Suite 1100
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(513) 985-1920

(Name, address, including zip code, and telephone number, including area code, of agent for service)

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Approximate date of commencement of proposed sale to public: As soon as practicable after this Registration Statement is declared effective.

If any of the securities being registered on this Form are to be offered on a delayed or continuous basis pursuant to Rule 415 under the Securities Act of 1933, check the following box.

If this Form is filed to register additional securities for an offering pursuant to Rule 462(b) under the Securities Act, check the following box and list the Securities Act registration statement number of the earlier effective registration statement for the same offering.

If this Form is a post-effective amendment filed pursuant to Rule 462(c) under the Securities Act, check the following box and list the Securities Act registration statement number of the earlier effective registration statement for the same offering.

If this Form is a post-effective amendment filed pursuant to Rule 462(d) under the Securities Act, check the following box and list the Securities Act registration statement number of the earlier effective registration statement for the same offering.

Indicate by check mark whether the registrant is a large accelerated filer, an accelerated filer, a non-accelerated filer, or a smaller reporting company. See the definitions of "large accelerated filer," "accelerated filer" and "smaller reporting company" in Rule 12b-2 of the Exchange Act.

Large accelerated filer

Non-accelerated filer (Do not check if a smaller reporting company)

Accelerated filer

Smaller reporting company

CALCULATION OF REGISTRATION FEE

Title of each class of securities to be registered	Proposed maximum aggregate offering price ⁽¹⁾	Amount of registration fee ⁽²⁾
Common stock, \$0.00001 par value	\$	\$

(1) Estimated solely for the purpose of calculating the registration fee in accordance with Rule 457(o) of the Securities Act of 1933, amended.

(2) Calculated pursuant to Rule 457(o) based on an estimate of the proposed maximum aggregate offering price.

The Registrant hereby amends this Registration Statement on such date or dates as may be necessary to delay its effective date until the Registrant shall file a further amendment which specifically states that this Registration Statement shall thereafter become effective in accordance with Section 8(a) of the Securities Act of 1933 or until the Registration Statement shall become effective on such date as the Securities and Exchange Commission, acting pursuant to said Section 8(a), may determine.

The information in this preliminary prospectus is not complete and may be changed. We may not sell these securities until the registration statement filed with the Securities and Exchange Commission is effective. This preliminary prospectus is not an offer to sell these securities and it is not soliciting an offer to buy these securities in any jurisdiction where the offer or sale is not permitted.

SUBJECT TO COMPLETION, DATED DECEMBER 20, 2013

PRELIMINARY PROSPECTUS

Shares



AkebiaTM
THERAPEUTICS

Akebia Therapeutics, Inc.
Common Stock

This is the initial public offering of shares of common stock of Akebia Therapeutics, Inc.

We are offering _____ shares of our common stock. Prior to this offering, there has been no public market for our common stock. We intend to apply to have our common stock listed on the NASDAQ Global Market under the trading symbol "AKBA."

We are an emerging growth company under the federal securities laws and are subject to reduced public company reporting requirements.

Investing in our common stock involves a high degree of risk. See "[Risk Factors](#)" beginning on page 11.

	<u>Per share</u>	<u>Total</u>
Initial public offering price	\$ _____	\$ _____
Underwriting discounts and commissions(1)	\$ _____	\$ _____
Proceeds, before expenses, to Akebia	\$ _____	\$ _____

(1) See "Underwriting" for additional disclosure regarding underwriting discounts, commissions and expenses.

To the extent that the underwriters sell more than _____ shares of common stock, the underwriters have an option to purchase up to an additional _____ shares from us at the initial public offering price, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

The underwriters expect to deliver the shares against payment in New York, New York on _____, 2014.

Neither the Securities and Exchange Commission nor any state securities commission has approved or disapproved of these securities or passed on the adequacy or accuracy of this prospectus. Any representation to the contrary is a criminal offense.

Morgan Stanley

UBS Investment Bank

Credit Suisse

Nomura

, 2014

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We are responsible for the information contained in this prospectus and in any free-writing prospectus we prepare or authorize. We have not, and the underwriters have not, authorized anyone to provide you with different information, and we take no responsibility for any other information others may give you. We are not, and the underwriters are not, making an offer to sell these securities in any jurisdiction where the offer or sale is not permitted. You should not assume that the information contained in this prospectus is accurate as of any date other than the date on the cover page of this prospectus.

Summary

This summary highlights information contained in other parts of this prospectus and does not contain all of the information that you should consider in making your investment decision. Before investing in our common stock, you should carefully read this entire prospectus, including our financial statements and the related notes and the information set forth under the sections titled “Risk Factors,” “Cautionary Note Regarding Forward-Looking Statements” and “Management’s Discussion and Analysis of Financial Condition and Results of Operations.” Unless the context requires otherwise, references in this prospectus to “Akebia,” “we,” “us,” “our,” the “Company” and similar designations refer to Akebia Therapeutics, Inc.

Overview

We are a biopharmaceutical company focused on the development of novel proprietary therapeutics based on hypoxia inducible factor, or HIF, biology and the commercialization of these products for patients with kidney disease. HIF is the primary regulator of the production of red blood cells, or RBCs, in the body and a potentially novel mechanism of treating anemia. Our lead product candidate, AKB-6548, is being developed as a once-daily oral therapy that has successfully completed a Phase 2a proof of concept study demonstrating that AKB-6548 safely and predictably raised hemoglobin levels in patients with anemia secondary to chronic kidney disease, or CKD, not requiring dialysis.

We are conducting a Phase 2b trial for AKB-6548 in patients with anemia secondary to CKD who are not dependent on dialysis and expect data to be available in the fourth quarter of 2014. We have also initiated a development program for patients dependent on dialysis. If the results of our Phase 2b trial are positive, we would expect to initiate Phase 3 trials for anemia secondary to CKD in 2015, and would anticipate submitting a New Drug Application, or NDA, for AKB-6548 in the United States by 2018 if the Phase 3 data are favorable.

We own worldwide rights to our HIF-based product candidates, including AKB-6548. If approved by regulatory authorities, we plan to commercialize AKB-6548 in the United States ourselves and intend to seek one or more collaborators to commercialize the product candidate in additional markets.

Anemia is a serious medical condition in which blood is deficient in RBCs and hemoglobin, both of which are critical in delivering oxygen to tissue. Anemia generally exists when hemoglobin, a protein in RBCs that carries oxygen, is less than 13 g/dL in men or 12 g/dL in women. Untreated anemia is associated with chronic fatigue, increased risk of progression of multiple diseases and death. Anemia is common in patients with CKD, cancer, heart failure, inflammatory diseases and other critical illnesses, as well as in the elderly.

More than 30 million people in the United States have CKD, with estimates that over 1.8 million of these patients suffer from anemia. Anemia from these indications is currently treated by injectable recombinant protein erythropoiesis stimulating agents, or rESAs—including Epogen, Aranesp and Procrit—with iron supplementation or an RBC transfusion. Based on the reported revenues of companies that market and sell rESAs, we estimate that global sales of injectable rESAs were \$6.3 billion in 2012, the vast majority of which were for renal indications.

rESAs are designed to stimulate production of RBCs by binding directly to and saturating erythropoietin, or EPO, receptors. While injectable rESAs and transfusions may be effective in raising hemoglobin levels, they carry significant potential side effects and also need to be delivered subcutaneously or intravenously. In particular, injectable rESAs may lead to thrombosis, stroke, myocardial infarction and death, and these risks are described in black box warnings on the prescribing information of all products marketed in this class. These

safety concerns, which became evident starting in 2006, have led to a significant reduction in the use of injectable rESAs. Today anemia is either not treated or inadequately treated in the majority of CKD patients, and we believe that a safe, effective, oral therapeutic option will take significant market share and meaningfully grow the market in patients not requiring dialysis.

AKB-6548 works by a differentiated mechanism of action that we believe has the potential to be safer than that of injectable rESAs. This novel mechanism of action is referred to as HIF prolyl-hydroxylase, or HIF-PH, inhibition. Instead of binding directly to the EPO receptors on cells in the bone marrow, AKB-6548 leads to activation of critical pathways for hemoglobin and RBC production. This approach mimics the physiological adjustment made by the body when exposed to reduced oxygen levels at higher altitudes.

To date, AKB-6548 has been studied in eight clinical trials across four separate patient populations: healthy volunteers and patients with CKD stages 3, 4 and 5 (non-dialysis). Our largest study was a Phase 2a trial in 91 patients with anemia secondary to CKD, which showed significantly increased hemoglobin levels among subjects taking AKB-6548 compared to baseline in a dose-dependent manner across all treatment arms ($p < 0.0001$). No drug-related serious adverse events were reported, and dosing was well-tolerated. In addition, AKB-6548 was also shown to stabilize the iron supply to the bone marrow while improving hemoglobin production.

Our ongoing Phase 2b trial explores a dosing approach for AKB-6548 to enable subjects with anemia secondary to CKD to appropriately and safely raise hemoglobin levels. As of December 16, 2013, we had enrolled over 50% of our targeted 200 patients in this study at investigational sites in the United States, with data expected in the fourth quarter of 2014. With positive data, we plan to progress to Phase 3 global registration studies for AKB-6548 in patients with anemia secondary to CKD. We anticipate the design of the Phase 3 studies will mirror the Phase 2b study, except that they will be longer and larger in size, positioning us to file for approval in the United States by 2018.

Given the burdens of the current standard of care and costs associated with administering an injectable rESA, we believe AKB-6548 is a promising alternative for the overall cost-effective treatment of anemia. We intend to commercialize AKB-6548 ourselves in the United States for the treatment of anemia in patients with CKD. These patients are primarily treated by approximately 7,000 nephrologists, and we believe we can reach most of this market with a specialty sales force of approximately 125 people. We intend to seek one or more commercial collaborators for the development and commercialization of AKB-6548 outside of the United States. We may also explore opportunities to expand AKB-6548 into additional markets not adequately addressed by injectable rESAs because of safety or dosing delivery issues, including idiopathic anemia of aging, or IAA, and anemia of congestive heart failure.

We are led by a team of experienced biopharmaceutical executives with a background in developing and commercializing drugs for the treatment of renal and metabolic disorders. John P. Butler, our CEO, was former President of Genzyme Corp.'s renal division which grew to over \$1 billion in annual revenue under his leadership, and is current Chairman of the Board of the American Kidney Fund, the leading patient advocacy organization for kidney disease patients. Earlier in his career, Mr. Butler held sales and marketing positions at Amgen, working on the early commercial launch of injectable rESAs in the renal anemia market. Our executive team also includes Robert Shalwitz, M.D., CMO and co-founder of Akebia. Dr. Shalwitz is an academic pediatric endocrinologist and has extensive industry experience developing novel pharmaceuticals at Abbott Laboratories and Reliant Pharmaceuticals. He has developed extensive knowledge of HIF biology over his career, particularly over the past seven years in leading development at Akebia.

Our Strategy

Our strategy is to develop novel therapeutics for patients based on HIF biology and to commercialize products for patients with kidney disease, beginning with AKB-6548 for patients with anemia secondary to CKD. The key elements of our strategy are to:

- **Complete the development of AKB-6548 for anemia secondary to CKD.** We plan to complete the Phase 2b trial that is currently enrolling in the United States. We intend to initiate a Phase 3 development program in 2015 following our end of Phase 2 meeting with the United States Food and Drug Administration, or FDA.
- **Obtain regulatory approval of AKB-6548 for anemia secondary to CKD in the United States, Europe and other markets.** We plan to complete an end of Phase 2 meeting with the FDA and seek scientific advice from the European Medicines Agency, or EMA, to define the Phase 3 development program necessary to secure regulatory approval to market AKB-6548. We would expect to initiate Phase 3 trials for anemia secondary to CKD in 2015, and would anticipate submitting an NDA for AKB-6548 in the United States by 2018 if the Phase 3 data are favorable.
- **Commercialize AKB-6548 in the United States and other territories.** We will establish a specialty sales and marketing organization to commercialize AKB-6548 in the United States. Outside of the United States, we intend to seek one or more commercial collaborators.
- **Continue to develop AKB-6548 for further indications.** We plan to initiate, in the first half of 2014, a Phase 2 study for AKB-6548 in dialysis patients with anemia, the second indication we intend to pursue. Additionally, we plan to evaluate the product candidate in IAA and other indications.
- **Advance our earlier stage pipeline asset.** We plan to advance AKB-6899, a second HIF-PH inhibitor product candidate, which we believe, based on preclinical testing, has the ability to increase EPO levels while reducing vascular endothelial growth factor, or VEGF, levels. We intend to file an Investigational New Drug, or IND, application and begin Phase 1 trials to determine its potential use in oncology and ophthalmology.
- **Acquire or in-license additional nephrology products.** If we are able to successfully launch AKB-6548, we will look to leverage our commercial infrastructure with additional products that would be prescribed by nephrologists.

We may enter into strategic collaborations to fully realize all of the elements of our strategy.

AKB-6548 as a Potential Solution

We are developing our lead product candidate, AKB-6548, to be a best in class HIF-PH inhibitor for the treatment of anemia secondary to CKD. We expect AKB-6548 to offer:

- Predictable, meaningful and sustained improvements in hemoglobin levels;
- Once a day therapy delivered orally;
- A dosing regimen that restores the normal diurnal EPO pattern;
- Robust pharmacodynamics and substantially lower peak EPO levels than with injectable rESAs; and
- Reduced administration of IV or oral iron supplementation to patients treated for anemia secondary to CKD.

Potential Best in Class Profile

We believe AKB-6548 has compelling clinical data demonstrating a best in class profile with several potential safety and efficacy advantages over current injectable rESA therapy in the treatment of anemia secondary to CKD.

- *AKB-6548 significantly increases hemoglobin in anemic CKD patients.* We have successfully completed a Phase 2a trial, in which AKB-6548 significantly increased hemoglobin levels compared to baseline in a dose-dependent manner across all treatment arms ($p < 0.0001$). Further, AKB-6548 provides a physiologic reticulocyte, or newly formed RBC, response, which leads to a more gradual and consistent increase in hemoglobin levels than what is seen with injectable rESA therapies, meaning that these improvements occur without causing patients' hemoglobin to rise to levels that cause concern.
- *AKB-6548 may have the potential to restore the normal diurnal variation of EPO for a patient with anemia in a way that an injectable rESA cannot.* Instead of binding directly to and saturating the EPO receptor for prolonged periods of time as is the case with current injectable rESA treatments, AKB-6548 acts by simulating the body's natural response to hypoxia that is carried out by stabilization of HIFa.
- *Oral, once-daily dosing.* Once-daily, oral dosing of AKB-6548 offers improved convenience for patients as compared to injectable rESAs. This convenience may increase access to anemia therapy for the largely underserved population of patients with anemia secondary to CKD who are not yet on dialysis and for patients with other forms of anemia, such as idiopathic anemia of aging. AKB-6548 offers the potential of flexible oral dosing that provides a more gradual and reliable means of titration than that of injectable rESAs.
- *Ability to stabilize the iron supply to the bone marrow while improving hemoglobin production.* In clinical trials, AKB-6548 has demonstrated a dose-related increase in total iron binding capacity. These results indicate that AKB-6548 will stabilize the iron supply to the bone marrow while improving hemoglobin production and should improve EPO responsiveness. As a result, unlike injectable rESAs, which have no effect on iron mobilization, AKB-6548 offers the added potential benefit of reducing the amount of supplemental iron required by anemia patients.
- *Differentiated safety profile.* AKB-6548's novel mechanism of action and dosing profile offer the opportunity to potentially avoid the black box label ascribed to injectable rESAs. In our recently completed Phase 2a study, no drug-related serious adverse events were reported. Dosing was well-tolerated and there was no evidence of undesirable vascular response.

Risk Associated with Our Business

An investment in our common stock involves a high degree of risk. Any of the factors set forth under "Risk Factors" may limit our ability to successfully execute our business strategy. You should carefully consider all of the information set forth in this prospectus and, in particular, should evaluate the specific factors set forth under "Risk Factors" in deciding whether to invest in our common stock. These risk factors include, among others:

- We depend heavily on the success of one product candidate, AKB-6548, which is in a Phase 2b clinical trial. Even if we obtain favorable clinical results, we may not be able to obtain regulatory approval for, or successfully commercialize, AKB-6548.
- We have incurred significant losses since inception and anticipate that we will continue to incur significant losses for the foreseeable future and may never achieve or maintain profitability.
- We will require substantial additional financing. A failure to obtain this necessary capital when needed could force us to delay, limit, reduce or terminate our product development or commercialization efforts.

- We have not obtained agreement with the FDA, the EMA, or other regulatory authorities on the design of our Phase 3 development program.
- Clinical drug development is a lengthy and expensive process with an uncertain outcome, and positive results from Phase 1 and Phase 2a clinical trials of AKB-6548 are not necessarily predictive of the results of our current Phase 2b and any future clinical trials of AKB-6548. If we cannot replicate the positive results from our Phase 1 and Phase 2a clinical trials of AKB-6548 in our Phase 2b and subsequent clinical trials, we may be unable to successfully develop, obtain regulatory approval for and commercialize AKB-6548.
- Even if we receive regulatory approval for our product candidates, such drug products will be subject to ongoing regulatory review, which may result in significant additional expense. Additionally, our product candidates, if approved, could be subject to labeling and other restrictions, and we may be subject to penalties if we fail to comply with regulatory requirements or experience unanticipated problems with our products.
- If our efforts to protect our proprietary technologies are not adequate, we may not be able to compete effectively in our market. We are currently involved in an opposition proceeding involving one of our European patents, and the outcome of that proceeding may affect our ability to establish a competitive advantage in the market or successfully commercialize our lead product candidate in the European Union.
- Third-party claims of intellectual property infringement may be costly and time consuming, and may delay or harm our drug discovery and development efforts. We are currently involved in an opposition proceeding involving the granted European patent of one of our potential competitors.

Implications of Being an Emerging Growth Company

As a company with less than \$1 billion in revenue during our most recently completed fiscal year, we qualify as an “emerging growth company” as defined in Section 2(a) of the Securities Act of 1933, as amended, or the Securities Act, as modified by the Jumpstart Our Business Startups Act of 2012, or the JOBS Act. As an emerging growth company, we may take advantage of specified reduced disclosure and other requirements that are otherwise applicable, in general, to public companies that are not emerging growth companies. These provisions include:

- Reduced disclosure about our executive compensation arrangements;
- Exemption from the non-binding shareholder advisory votes on executive compensation or golden parachute arrangements;
- Exemption from the auditor attestation requirement in the assessment of our internal control over financial reporting; and
- Reduced disclosure of financial information in this prospectus, such as being permitted to include only two years of audited financial information and two years of selected financial information in addition to any required unaudited interim financial statements, with correspondingly reduced “Management’s Discussion and Analysis of Financial Condition and Results of Operations” disclosure.

We may take advantage of these exemptions for up to five years or such earlier time that we are no longer an emerging growth company. We would cease to be an emerging growth company if we have more than \$1 billion in annual revenues as of the end of a fiscal year, have more than \$700 million in market value of our capital stock held by non-affiliates as of any June 30 or if we issue more than \$1 billion of non-convertible debt over a three-year-period. We may choose to take advantage of some, but not all, of the available exemptions. We have taken advantage of some reduced reporting burdens in this prospectus. Accordingly, the information contained herein may be different than the information you receive from other public companies in which you hold stock.

The JOBS Act permits an emerging growth company to take advantage of an extended transition period to comply with new or revised accounting standards applicable to public companies. We have irrevocably elected not to avail ourselves of this extended transition period and, as a result, we will adopt new or revised accounting standards on the relevant dates on which adoption of such standards is required for non-emerging growth companies.

Corporate Information

We were incorporated under the laws of the state of Delaware in February 2007. In December 2011, we spun out our programs focused on the treatment of diabetic eye disease and inflammatory bowel disease into Aerpio Therapeutics, Inc., or Aerpio, which has since operated as a stand-alone company. Our principal executive office is located at 245 First Street, Suite 1100, Cambridge MA 02142, and our telephone number is 513-985-1920. Our website address is www.akebia.com. We have included our website address in this prospectus solely as an inactive textual reference. The information on, or that can be accessed through, our website is not part of this prospectus, and you should not rely on any such information in making the decision whether to purchase our common stock.

This prospectus contains trademarks and tradenames of other businesses that are the property of their respective owners. We have omitted the ® and ™ designations, as applicable, for the trademarks named in this prospectus.

The Offering

Common stock offered by us	shares
Common stock to be outstanding immediately following this offering	
	shares
Underwriters' over-allotment option	The underwriters have an option to purchase up to additional shares of common stock to cover over-allotments as described in "Underwriting."
Use of proceeds	<p>We estimate that the net proceeds from the issuance of our common stock in this offering will be approximately \$ million or approximately \$ million if the underwriters exercise their over-allotment option in full, assuming an initial public offering price of \$ per share, which is the midpoint of the price range set forth on the cover page of this prospectus, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.</p> <p>We intend to use the net proceeds from this offering to continue clinical development of AKB-6548 in renal indications, including the preparation for and initiation of the Phase 3 development program in anemia secondary to CKD; to conduct a Phase 2 clinical trial of AKB-6548 in idiopathic anemia of aging; to advance our preclinical candidate, AKB-6899, through Phase 1 development in oncology; and for working capital and other general corporate purposes. See "Use of Proceeds" for additional information.</p>
Risk factors	See "Risk Factors" and the other information included in this prospectus for a discussion of factors you should carefully consider before deciding to invest in our common stock.
Proposed NASDAQ Global Market symbol	We intend to apply for listing of our common stock on the NASDAQ Global Market under the symbol "AKBA."

The number of shares of common stock to be outstanding after this offering is based on 7,214,193 shares of common stock outstanding as of September 30, 2013, including 244,513 shares of restricted stock and 6,790,149 shares of our common stock issuable upon the automatic conversion of all outstanding shares of our preferred stock, and excludes the following:

- 702,625 shares of common stock issuable upon exercise of stock options outstanding as of September 30, 2013 at a weighted-average exercise price of \$1.08 per share;
- 64,408 shares of common stock reserved for future issuance under our Amended and Restated 2008 Equity Incentive Plan as of September 30, 2013; and
- shares of common stock reserved for future issuance under our 2014 Equity Incentive Plan.

Unless otherwise indicated, all information in this prospectus reflects or assumes the following:

- the amendment and restatement of our certificate of incorporation and bylaws, which will occur immediately prior to the closing of this offering;
- the conversion of all of our outstanding shares of our preferred stock into 6,790,149 shares of common stock, which will occur automatically upon the closing of this offering;
- no exercise of stock options on or after September 30, 2013; and
- no exercise by the underwriters of their option to purchase up to an additional _____ shares of common stock in this offering.

Summary Financial Data

The following summary financial data for the years ended December 31, 2011 and 2012, the nine months ended September 30, 2013, the period from February 27, 2007 (inception) to September 30, 2013 and as of September 30, 2013 are derived from our audited financial statements included elsewhere in this prospectus. The summary financial data for the nine months ended September 30, 2012 have been derived from our unaudited financial statements included elsewhere in this prospectus. These unaudited financial statements have been prepared on a basis consistent with our audited financial statements and, in our opinion, contain all adjustments, consisting only of normal and recurring adjustments, necessary for a fair presentation of such financial data. You should read this data together with our financial statements and related notes included elsewhere in this prospectus and the information under the captions “Selected Financial Data” and “Management’s Discussion and Analysis of Financial Condition and Results of Operations.” Our historical results for any prior period are not necessarily indicative of results to be expected in any future period, and our interim period results are not necessarily indicative of results to be expected for a full year or any other interim period.

	<u>Year Ended December 31,</u>		<u>Nine Months Ended</u> <u>September 30,</u>		<u>Period from</u> <u>February 27,</u> <u>2007 (Date of</u> <u>Inception) to</u> <u>September 30,</u> <u>2013</u>
	<u>2011⁽¹⁾</u>	<u>2012</u>	<u>2012</u> <u>(unaudited)</u>	<u>2013</u>	
Consolidated statements of operations data:					
Revenue	\$ —	—	\$ —	\$ —	\$ —
Expenses:					
Research and development	12,976	5,632	5,065	7,591	48,557
General and administrative	2,567	2,891	1,594	2,141	12,258
Total expenses	15,543	8,523	6,659	9,732	60,815
Loss from operations	(15,543)	(8,523)	(6,659)	(9,732)	(60,815)
Other income, net	246	327	866	2,513	3,721
Net loss	<u>\$ (15,297)</u>	<u>\$ (8,196)</u>	<u>\$ (5,793)</u>	<u>\$ (7,219)</u>	<u>\$ (57,094)</u>
Net loss per share applicable to common stockholders—basic and diluted ⁽²⁾	\$ (109.36)	\$ (48.68)	\$ (35.80)	\$ (206.32)	\$ (827.64)
Weighted-average number of common shares used in net loss per share applicable to common stockholders—basic and diluted	167,039	236,633	230,748	291,206	146,688
Pro forma net loss per share applicable to common stockholders—basic and diluted (unaudited) ⁽²⁾		<u>\$ (2.26)</u>		<u>\$ (1.35)</u>	
Pro forma weighted-average number of common shares used in net loss per share applicable to common stockholders—basic and diluted (unaudited)		<u>3,622,838</u>		<u>5,362,450</u>	

- (1) In December 2011, we spun out our programs focused on the treatment of diabetic eye disease and inflammatory bowel disease into Aerpio, which has since operated as a stand-alone company. See “Management’s Discussion and Analysis of Financial Condition and Results of Operations” appearing elsewhere in this prospectus for further information.
- (2) See Note 2 within the notes to our financial statements appearing elsewhere in this prospectus for a description of the method used to calculate basic and diluted net loss per share of common stock. Pro forma basic and diluted net loss per share of common stock is calculated by dividing net loss attributable to common stockholders, excluding the impact of gains (losses) on the extinguishment of preferred stock and accretion of preferred stock by the pro forma weighted-average number of common shares outstanding, which assumes the automatic conversion of all outstanding shares of our preferred stock into an aggregate of 6,790,149 shares of common stock upon the closing of this offering.

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The pro forma balance sheet data set forth below give effect to the automatic conversion of all outstanding shares of our preferred stock into an aggregate of 6,790,149 shares of our common stock upon the closing of this offering.

The pro forma as adjusted balance sheet data set forth below give further effect to our issuance and sale of _____ shares of our common stock in this offering at an assumed initial public offering price of \$ _____ per share, which is the midpoint of the price range listed on the cover page of this prospectus, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

The pro forma as adjusted information presented in the summary balance sheet data are illustrative only and will change based on the actual initial public offering price and other terms of this offering determined at pricing. Each \$1.00 increase or decrease in the assumed initial public offering price of \$ _____ per share, which is the midpoint of the price range set forth on the cover page of this prospectus, would increase or decrease each of cash and cash equivalents, total assets and total stockholders' deficit on a pro forma as adjusted basis by approximately \$ _____, assuming that the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same and after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us. Similarly, each increase or decrease of 1,000,000 shares offered by us at the assumed initial public offering price would increase or decrease each of cash and cash equivalents, total assets and total stockholders' deficit on a pro forma as adjusted basis by approximately \$ _____, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

	As of September 30, 2013		Pro forma as adjusted
	Actual	Pro forma (in thousands)	
Balance Sheet Data:			
Cash and cash equivalents	\$ 24,603	\$ 24,603	\$
Working capital (deficit)	35,566	35,566	
Total assets	38,470	38,470	
Redeemable convertible preferred stock	154,803	—	
Accumulated deficit	(119,231)	(57,691)	
Total stockholders' (deficit) equity	(119,231)	35,572	

Risk Factors

Investing in our common stock involves a high degree of risk. You should carefully consider the risks and uncertainties described below together with all of the other information contained in this prospectus, including our financial statements and the related notes appearing at the end of this prospectus, before deciding to invest in our common stock. If any of the following risks actually occur, our business, prospects, operating results and financial condition could suffer materially, the trading price of our common stock could decline and you could lose all or part of your investment. The risks and uncertainties described below are not the only ones we face. Additional risks and uncertainties not presently known to us or that we currently believe to be immaterial may also adversely affect our business.

Risks Related to Our Financial Position and Need for Additional Capital

We have incurred significant losses since inception and anticipate that we will continue to incur significant losses for the foreseeable future and may never achieve or maintain profitability.

We have incurred net losses each year since our inception, including net losses of \$15.3 million for the year ended December 31, 2011, \$8.2 million for the year ended December 31, 2012, and \$7.2 million for the nine months ended September 30, 2013. As of September 30, 2013, we had an accumulated deficit of \$119.2 million. To date, we have not commercialized any products or generated any revenues from the sale of products, and we do not expect to generate any product revenues in the foreseeable future. We do not know whether or when we will generate revenue or become profitable.

We have devoted most of our financial resources to research and development, including our clinical and preclinical development activities. To date, we have financed our operations primarily through private placements of our preferred stock. The amount of our future net losses will depend, in part, on the rate of our future expenditures, and our financial position will depend, in part, on our ability to obtain funding through equity or debt financings, strategic collaborations or grants. Our lead product candidate, AKB-6548, is currently in an ongoing Phase 2b clinical trial, and our other product candidate is in preclinical development. As a result, we expect that it will be several years, if ever, before we have a product candidate ready for commercialization. Even if we obtain regulatory approval to market AKB-6548, our future revenues will depend upon the size of any markets in which AKB-6548 has received approval, our ability to achieve sufficient market acceptance, reimbursement from third-party payors and other factors.

We expect to continue to incur significant expenses and increasing operating losses for the foreseeable future. We anticipate that our expenses will increase significantly if and as we:

- continue our Phase 2b trial and prepare for a future Phase 3 development program of AKB-6548 for the treatment of anemia secondary to CKD;
- seek regulatory approvals for our product candidates that successfully complete clinical studies;
- have our product candidates manufactured for clinical trials and for commercial sale;
- establish a sales, marketing and distribution infrastructure to commercialize any products for which we may obtain marketing approval;
- initiate additional preclinical, clinical or other studies for AKB-6548, AKB-6899 and other product candidates that we may develop or acquire;
- seek to discover and develop additional product candidates;
- acquire or in-license other commercial products, product candidates and technologies;
- make royalty, milestone or other payments under any future in-license agreements;

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- maintain, protect and expand our intellectual property portfolio;
- attract and retain skilled personnel; and
- create additional infrastructure to support our operations as a public company.

Because of the numerous risks and uncertainties associated with pharmaceutical product development, we are unable to accurately predict the timing or amount of increased expenses or when, if at all, we will be able to achieve profitability. If we are required by the United States Food and Drug Administration, or FDA, the EMA or other regulatory authorities to perform studies in addition to those currently expected, or if there are any delays in completing our clinical trials or the development of any of our product candidates, our expenses could increase.

The net losses we incur may fluctuate significantly from quarter to quarter and year to year, such that a period-to-period comparison of our results of operations may not be a good indication of our future performance. In any particular quarter or quarters, our operating results could be below the expectations of securities analysts or investors, which could cause our stock price to decline.

To become and remain profitable, we must succeed in developing and commercializing our product candidates, which must generate significant revenue. This will require us to be successful in a range of challenging activities, including completing preclinical testing and clinical trials of our product candidates, discovering additional product candidates, obtaining regulatory approval for these product candidates and manufacturing, marketing and selling any products for which we may obtain regulatory approval. We are only in the preliminary stages of most of these activities. We may never succeed in these activities and, even if we do, may never generate revenues that are significant enough to achieve profitability.

Even if we do achieve profitability, we may not be able to sustain or increase profitability on a quarterly or annual basis. Our failure to become and remain profitable could depress the value of our company and could impair our ability to raise capital, expand our business, maintain our research and development efforts, diversify our product offerings or even continue our operations. A decline in the value of our company could cause you to lose all or part of your investment.

We will require substantial additional financing. A failure to obtain this necessary capital when needed could force us to delay, limit, reduce or terminate our product development or commercialization efforts.

As of September 30, 2013, our cash and cash equivalents were \$24.6 million. We believe that we will continue to expend substantial resources for the foreseeable future developing AKB-6548, AKB-6899 and any other product candidates that we may develop or acquire. These expenditures will include costs associated with research and development, potentially obtaining regulatory approvals and having our products manufactured, as well as marketing and selling products approved for sale, if any. In addition, other unanticipated costs may arise. Because the outcome of our current and anticipated clinical trials is highly uncertain, we cannot reasonably estimate the actual amounts necessary to successfully complete the development and commercialization of our product candidates.

Our future capital requirements depend on many factors, including:

- the rate of progress, results and cost of completing our Phase 2b clinical trial of AKB-6548 and our operating costs incurred as we conduct these trials and through our end of Phase 2 meeting with the FDA, and equivalent meetings with the EMA and other regulatory authorities;
- assuming AKB-6548 advances to Phase 3 clinical trials, the scope, size, rate of progress, results and costs of initiating and completing our Phase 3 development program of AKB-6548;
- assuming favorable clinical results, the cost, timing and outcome of our efforts to obtain marketing approval for AKB-6548 in the United States, Europe and in other jurisdictions, including to fund the preparation and filing of regulatory submissions for AKB-6548 with the FDA, the EMA and other regulatory authorities;

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- the scope, progress, results and costs of preclinical development, laboratory testing and clinical trials for AKB-6899 and any other product candidates that we may develop or acquire;
- the timing of, and the costs involved in, obtaining regulatory approvals for AKB-6899 if clinical trials are successful;
- the cost and timing of future commercialization activities for our products, if any of our product candidates are approved for marketing, including product manufacturing, marketing, sales and distribution costs;
- the revenue, if any, received from commercial sales of our product candidates for which we receive marketing approval;
- the cost of having our product candidates manufactured for clinical trials in preparation for regulatory approval and in preparation for commercialization;
- our ability to establish and maintain strategic collaborations, licensing or other arrangements and the financial terms of such agreements;
- the costs involved in preparing, filing, prosecuting patent applications, maintaining, defending and enforcing our intellectual property rights, including litigation costs and the outcome of such litigation; and
- the extent to which we acquire or in-license other products or technologies.

Based on our current operating plan, we believe that the net proceeds we receive from this offering, and our existing cash and cash equivalents and investments will be sufficient to fund our projected operating expenses and capital expenditure requirements through at least . However, our operating plan may change as a result of many factors currently unknown to us, and we may need additional funds sooner than planned. In addition, we may seek additional capital due to favorable market conditions or strategic considerations even if we believe we have sufficient funds for our current or future operating plans. Additional funds may not be available when we need them on terms that are acceptable to us, or at all. If adequate funds are not available to us on a timely basis, we may be required to delay, limit, reduce or terminate preclinical studies, clinical trials or other development activities for AKB-6548, AKB-6899 or any other product candidates that we develop or acquire, or delay, limit, reduce or terminate our establishment of sales and marketing capabilities or other activities that may be necessary to commercialize our product candidates.

Raising additional capital may cause dilution to our existing stockholders, restrict our operations or require us to relinquish rights to product candidates on unfavorable terms to us.

Until such time, if ever, as we can generate substantial product revenues, we expect to finance our cash needs through a combination of equity offerings, debt financings and license, development and commercialization agreements with collaborators. To the extent that we raise additional capital through the sale of equity or convertible debt securities, your ownership interest will be diluted, and the terms may include liquidation or other preferences and anti-dilution protections that adversely affect your rights as a stockholder. Debt financing, if available, may involve agreements that include covenants limiting or restricting our ability to take certain actions, such as incurring additional debt, making capital expenditures or declaring dividends. If we raise additional funds through strategic collaborations with third parties, we may have to relinquish valuable rights to our product candidates, future revenue streams, research programs or product candidates or grant licenses on terms that are not favorable to us. If we are unable to raise additional funds through equity or debt financing when needed, we may be required to delay, limit, reduce or terminate our product development or commercialization efforts for AKB-6548, AKB-6899 or any other product candidates that we develop or acquire, or grant rights to develop and market product candidates that we would otherwise prefer to develop and market ourselves.

Our limited operating history may make it difficult for you to evaluate the success of our business to date and to assess our future viability.

We commenced active operations in 2007, and our operations to date have been limited to organizing and staffing our company, business planning, raising capital, identifying potential product candidates, undertaking preclinical studies and conducting clinical trials. We currently have two product candidates, one of which is in preclinical development. Biopharmaceutical product development is a highly speculative undertaking and involves a substantial degree of risk. Only a small fraction of biopharmaceutical development programs ultimately result in commercial products or even product candidates and a number of events could delay our development efforts and negatively impact our ability to obtain regulatory approval for, and to manufacture, market and sell, a drug product. We have not yet demonstrated our ability to successfully complete later stage clinical trials, obtain regulatory approvals, manufacture a commercial scale product, or arrange for a third party to do so on our behalf, or conduct sales and marketing activities necessary for successful product commercialization. Consequently, any predictions you make about our future success or viability may not be as accurate as they could be if we had a longer operating history.

In addition, as a young business, we may encounter unforeseen expenses, difficulties, complications, delays and other known and unknown factors. We will need to expand our capabilities to support commercial activities. We may not be successful in adding such capabilities.

Risks Related to Our Business and the Clinical Development, Regulatory Review and Approval of AKB-6548 and AKB-6899

We depend heavily on the success of one product candidate, AKB-6548, which is in a Phase 2b clinical trial. Even if we obtain favorable clinical results, we may not be able to obtain regulatory approval for, or successfully commercialize, AKB-6548.

We currently have only one product candidate, AKB-6548, in clinical development, and our business depends almost entirely on the successful clinical development, regulatory approval and commercialization of that product candidate, which may never occur. We currently have no drug products for sale, generate no revenues from sales of any drugs, and may never be able to develop marketable drug products. AKB-6548, which is currently in an ongoing Phase 2b clinical trial, will require substantial additional clinical development, testing, manufacturing process development, and regulatory approval before we are permitted to commence its commercialization. Our other product candidate, AKB-6899, is in preclinical development. None of our product candidates has advanced into a pivotal study, and it may be years before such study is initiated, if ever. The clinical trials of our product candidates are, and the manufacturing and marketing of our product candidates will be, subject to extensive and rigorous review and regulation by numerous government authorities in the United States and in other countries where we intend to test and, if approved, market any product candidates. Before obtaining regulatory approval for the commercial sale of any product candidate, we must demonstrate through extensive preclinical testing and clinical trials that the product candidate is safe and effective for use in each target indication. This process can take many years. Of the large number of drugs in development in the United States, only a small percentage successfully complete the FDA regulatory approval process and are commercialized. Accordingly, even if we are able to obtain the requisite capital to continue to fund our development and clinical programs, we may be unable to successfully develop or commercialize AKB-6548.

We are not permitted to market AKB-6548 in the United States until we receive approval of an NDA from the FDA, or in any foreign countries until we receive the requisite approval from such countries. As a condition to submitting an NDA to the FDA for AKB-6548 regarding its ability to treat patients with anemia secondary to CKD, we must complete our ongoing Phase 2b clinical trial, Phase 3 studies, and any additional non-clinical or clinical studies required by the FDA. To date, we have only commenced the Phase 2b clinical trial. AKB-6548 may not be successful in clinical trials or receive regulatory approval. Further, AKB-6548 may not receive regulatory approval even if it is successful in clinical trials. Obtaining approval of an NDA is a complex, lengthy, expensive and uncertain process that typically takes many years following the commencement of clinical trials

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and depends upon numerous factors, including the substantial discretion of the regulatory authorities. In addition, the safety concerns associated with injectable rESAs and the black box warnings in their prescribing information may affect the FDA's review of the safety results of AKB-6548. Further, the policies or regulations, or the type and amount of clinical data necessary to gain approval, may change during the course of a product candidate's clinical development and may vary among jurisdictions. We have not obtained regulatory approval for any product candidate and it is possible that AKB-6548 will never obtain regulatory approval. The FDA may delay, limit or deny approval of AKB-6548 for many reasons, including, among others:

- we may not be able to demonstrate that AKB-6548 is safe and effective in treating anemia secondary to CKD to the satisfaction of the FDA;
- the results of our clinical trials may not meet the level of statistical or clinical significance required by the FDA for marketing approval;
- the FDA may disagree with the number, design, size, conduct or implementation of our clinical trials;
- the FDA may not approve the formulation, labeling or specifications of AKB-6548;
- the FDA may require that we conduct additional clinical trials;
- the contract research organizations, or CROs, that we retain to conduct our clinical trials may take actions outside of our control that materially adversely impact our clinical trials;
- we may fail to perform in accordance with the FDA's good clinical practice, or GCP, requirements;
- the FDA may disagree with our interpretation of data from our preclinical studies and clinical trials;
- the FDA may not approve the manufacturing processes or facilities of third-party manufacturers with which we contract; or
- the policies or regulations of the FDA may significantly change in a manner that renders our clinical data insufficient for approval, or requiring that we amend or submit new clinical protocols.

In addition, similar reasons may cause the EMA or other regulatory authorities to delay, limit or deny approval of AKB-6548 outside the United States.

Any of these factors, many of which are beyond our control, could jeopardize our ability to obtain regulatory approval for and successfully market AKB-6548. Because our business is almost entirely dependent upon AKB-6548, any such setback in our pursuit of regulatory approval would have a material adverse effect on our business and prospects.

Alternatively, even if we obtain regulatory approval, that approval may be for indications or patient populations that are not as broad as we intend or desire or may require labeling that includes significant use or distribution restrictions or safety warnings. We may also be required to perform additional, unanticipated clinical trials to obtain approval or be subject to additional post marketing testing requirements to maintain regulatory approval. In addition, regulatory authorities may withdraw their approval of a product or the FDA may require a risk evaluation and mitigation strategy, or REMS, for a product, which could impose restrictions on its distribution. Any of the foregoing scenarios could materially harm the commercial prospects for our product candidates.

We have not obtained agreement with the FDA, EMA or other regulatory authorities on the design of our Phase 3 development program.

As we have not completed our Phase 2b clinical trial, we have not obtained agreement with the FDA on the design of our Phase 3 development program. We plan to hold an end of Phase 2 meeting with the FDA upon successful completion of our Phase 2b clinical trial. If the FDA determines that the Phase 2b trial results do not support moving into a pivotal program, we would be required to conduct additional Phase 2 studies.

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Alternatively, the FDA could disagree with our proposed design of our Phase 3 development program and could suggest a larger number of subjects or a longer course of treatment than our current expectations. If the FDA takes such positions, the costs of our AKB-6548 development program could increase materially and the potential market introduction of AKB-6548 could be delayed or we could risk not obtaining FDA approval even if the Phase 3 trials meet their primary endpoints. The FDA also may require that we conduct additional clinical, nonclinical or manufacturing validation studies and submit that data before it will consider an NDA application.

We have not yet sought guidance for the regulatory path for AKB-6548 with the EMA or other regulatory authorities. We cannot predict what additional requirements may be imposed by these regulatory authorities or how such requirements might delay or increase costs for our planned Phase 3 development program. Because our business is almost entirely dependent upon the successful development, regulatory approval, and commercialization of AKB-6548, any such delay or increase costs would have an adverse effect on our business.

We may find it difficult to enroll patients in our clinical studies, which could delay or prevent clinical studies of our product candidates.

Identifying and qualifying patients to participate in clinical studies of our product candidates is critical to our success. The timing of our clinical studies depends on the speed at which we can recruit patients to participate in testing our product candidates. Patients may be unwilling to participate in our clinical trials for AKB-6548 because of negative publicity from adverse events observed in injectable rESAs, other investigational agents and commercial products in CKD or for other reasons, including competitive clinical studies for similar patient populations. In addition, patients controlling their disease with current injectable rESAs may be reluctant to participate in a clinical trial with an investigational drug. As a result, the timeline for recruiting patients, conducting studies and obtaining regulatory approval of potential products may be delayed. These delays could result in increased costs, delays in advancing our development of AKB-6548 or termination of the clinical studies altogether.

We may not be able to identify, recruit and enroll a sufficient number of patients, or those with required or desired characteristics to achieve diversity in a study, to complete our clinical studies in a timely manner. Patient enrollment is affected by factors including:

- severity of the disease under investigation;
- design of the study protocol;
- size and nature of the patient population;
- eligibility criteria for and design of the study in question;
- perceived risks and benefits of the product candidate under study;
- proximity and availability of clinical study sites for prospective patients;
- availability of competing therapies and clinical studies and clinicians' and patients' perceptions as to the potential advantages of AKB-6548 in relation to available therapies or other products under development;
- efforts to facilitate timely enrollment in clinical studies;
- patient referral practices of physicians; and
- ability to monitor patients adequately during and after treatment.

We may not be able to initiate or continue clinical studies if we cannot enroll a sufficient number of eligible patients to participate in the clinical studies required by regulatory agencies. If we have difficulty enrolling a sufficient number of patients to conduct our clinical studies as planned, we may need to delay, limit or terminate on-going or planned clinical studies, any of which would have an adverse effect on our business.

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We may not be able to comply with requirements of foreign jurisdictions in conducting trials outside of the United States. In addition, we may not be able to obtain regulatory approval in foreign jurisdictions.

We currently expect to seek regulatory approval for AKB-6548 for the treatment of anemia secondary to CKD in major markets outside the United States, including the European Union. Our ability to successfully initiate, enroll and complete a clinical study in any foreign country, should we attempt to do so, is subject to numerous risks unique to conducting business in international markets, including:

- difficulty in establishing or managing relationships with qualified CROs and physicians;
- different local standards for the conduct of clinical studies;
- the potential burden of complying with a variety of foreign laws, medical standards and regulatory requirements, including the regulation of pharmaceutical and biotechnology products and treatments; and
- the acceptability of data obtained from studies conducted in the United States to the EMA and other regulatory authorities.

If we fail to successfully meet requirements for the conduct of clinical trials outside of the United States, we may be delayed in obtaining, or be unable to obtain, regulatory approval for AKB-6548 in countries outside of the United States.

Regulatory authorities outside the United States will require compliance with numerous and varying regulatory requirements. The approval procedures vary among jurisdictions and may involve requirements for additional testing, and the time required to obtain approval may differ from that required to obtain FDA approval. In addition, in many countries outside the United States, a drug product must be approved for reimbursement before it can be approved for sale in that country. In some cases, the price that we intend to charge for our drug product is also subject to approval. Approval by the FDA does not ensure approval by regulatory authorities in other countries or jurisdictions, and approval by one foreign regulatory authority does not ensure approval by regulatory authorities in other foreign countries or by the FDA. However, the failure to obtain approval in one jurisdiction may negatively impact our ability to obtain approval in another jurisdiction. The foreign regulatory approval process may include all of the risks associated with obtaining FDA approval. We may not obtain foreign regulatory approvals on a timely basis, if at all. We may not be able to file for regulatory approvals and may not receive necessary approvals to commercialize our drug products in any market.

Clinical drug development is a lengthy and expensive process with an uncertain outcome, and positive results from Phase 1 and Phase 2a clinical trials of AKB-6548 are not necessarily predictive of the results of our current Phase 2b and any future clinical trials of AKB-6548. If we cannot replicate the positive results from our Phase 1 and Phase 2a clinical trials of AKB-6548 in our Phase 2b and subsequent clinical trials, we may be unable to successfully develop, obtain regulatory approval for and commercialize AKB-6548.

Clinical testing is expensive and can take many years to complete, and its outcome is inherently uncertain. Failure can occur at any time during the clinical trial process. Success in preclinical studies may not be predictive of similar results in humans during clinical trials, and successful results from early or small clinical trials may not be replicated in later and larger clinical trials. For example, our early encouraging preclinical and clinical results for AKB-6548 do not ensure that the results of our ongoing Phase 2b clinical trial or any future clinical trials will demonstrate similar results. Our current Phase 2b clinical trial and our planned Phase 3 development program will enroll a larger number of subjects and will treat subjects for longer periods than our prior trials, which will result in a greater likelihood that adverse events may be observed. Many companies in the pharmaceutical and biotechnology industries have suffered significant setbacks in late-stage clinical trials after achieving positive results in early stage development, and we may face similar setbacks. If the results of our ongoing or future clinical trials for AKB-6548 are inconclusive with respect to efficacy, if we do not meet our clinical endpoints with statistical significance, or if there are safety concerns or adverse events, we may be prevented or delayed in obtaining marketing approval for AKB-6548.

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We may experience delays in our ongoing Phase 2b clinical trial for AKB-6548 and we do not know whether planned clinical trials will begin on time, need to be redesigned, enroll patients on time or be completed on schedule, if at all.

Clinical trials can be delayed or aborted for a variety of reasons, including delay or failure to:

- obtain regulatory approval to commence a clinical trial;
- reach agreement on acceptable terms with prospective CROs and clinical trial sites, the terms of which can be subject to extensive negotiation and may vary significantly among different CROs and trial sites;
- obtain institutional review board, or IRB, approval at each site;
- recruit, enroll and retain patients through the completion of clinical trials;
- maintain clinical sites in compliance with trial protocols through the completion of clinical trials;
- address any patient safety concerns that arise during the course of the trial;
- initiate or add a sufficient number of clinical trial sites; or
- manufacture sufficient quantities of our product candidate for use in clinical trials.

We could encounter delays if a clinical trial is suspended or terminated by us, by the relevant IRBs in which such trials are being conducted, by the Data Safety Monitoring Board, or DSMB, for such trial or by the FDA or other regulatory authorities. Such authorities may impose such a suspension or termination due to a number of factors, including failure to conduct the clinical trial in accordance with regulatory requirements or our clinical protocols, inspection of the clinical trial operations or trial site by the FDA or other regulatory authorities resulting in the imposition of a clinical hold, unforeseen safety issues or adverse side effects, changes in laws or regulations or lack of adequate funding to continue the clinical trial. Any delays in completing our clinical trials will increase our costs, slow down our product candidate development and approval process and jeopardize our ability to commence product sales and generate revenues. Any of these occurrences may harm our business, financial condition and prospects significantly.

Even if we receive regulatory approval for our product candidates, such drug products will be subject to ongoing regulatory review, which may result in significant additional expense. Additionally, our product candidates, if approved, could be subject to labeling and other restrictions, and we may be subject to penalties if we fail to comply with regulatory requirements or experience unanticipated problems with our products.

Any regulatory approvals that we receive for our product candidates may be subject to limitations on the approved indicated uses for which the product may be marketed or to conditions of approval, or contain requirements for potentially costly post-marketing testing, including Phase 4 clinical trials, and surveillance to monitor the safety and efficacy of the drug product. In addition, if the FDA approves any of our product candidates, the manufacturing processes, labeling, packaging, distribution, adverse event reporting, storage, advertising, promotion and recordkeeping for the product will be subject to extensive and ongoing regulatory requirements. These requirements include submissions of safety and other post-marketing information and reports, registration, as well as continued compliance with current Good Manufacturing Practice, or cGMP, requirements and GCP requirements for any clinical trials that we conduct post-approval.

Post-approval discovery of previously unknown problems with an approved drug product, including adverse events of unanticipated severity or frequency or relating to manufacturing operations or processes, or failure to comply with regulatory requirements, may result in, among other things:

- restrictions on the marketing or manufacturing of the drug product, withdrawal of the drug product from the market, or drug product recalls;
- fines, warning letters, or holds on clinical trials;

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- refusal by the FDA to approve pending applications or supplements to approved applications filed by us, or suspension or revocation of product approvals;
- product seizure or detention, or refusal to permit the import or export of products;
- a REMS program; and
- injunctions or the imposition of civil or criminal penalties.

The FDA's policies may change and additional government regulations may be enacted. We cannot predict the likelihood, nature or extent of government regulation that may arise from future legislation or administrative action, either in the United States or abroad. If we are slow or unable to adapt to changes in existing requirements or the adoption of new requirements or policies, or not able to maintain regulatory compliance, we may lose any marketing approval that may have been obtained and we may not achieve or sustain profitability, which would adversely affect our business.

Risks Related to Our Reliance on Third Parties

We rely on third parties to conduct preclinical studies and clinical trials for our product candidates, and if they do not properly and successfully perform their obligations to us, we may not be able to obtain regulatory approvals for our product candidates.

We rely on third party CROs, and other third parties to assist in managing, monitoring and otherwise carrying out our ongoing Phase 2b trial of AKB-6548. We expect to continue to rely on third parties, such as CROs, clinical data management organizations, medical institutions and clinical investigators to conduct our clinical trials in the future, including our Phase 3 development program. We compete with many other companies for the resources of these third parties. The third parties on whom we rely may terminate their engagements at any time, and having to enter into alternative arrangements would delay development and commercialization of our product candidates.

Our reliance on these third parties for research and development activities will reduce our control over these activities but will not relieve us of our responsibilities. For example, the FDA and foreign regulatory authorities require compliance with regulations and standards, including GCP requirements, for designing, conducting, monitoring, recording, analyzing, and reporting the results of clinical trials to ensure that the data and results are credible and accurate and that the rights, integrity, and confidentiality of trial participants are protected. Although we rely on third parties to conduct our clinical trials, we are responsible for ensuring that each of these clinical trials is conducted in accordance with its general investigational plan and protocol under legal and regulatory requirements. Regulatory authorities enforce these GCP requirements through periodic inspections of trial sponsors, principal investigators and trial sites. If we or any of our CROs fail to comply with applicable cGCP requirements, the clinical data generated in our clinical trials may be deemed unreliable and the FDA, EMA or other regulatory authorities may require us to perform additional clinical trials before approving our marketing applications. We cannot assure you that upon inspection by a given regulatory authority, such regulatory authority will determine that any of our clinical trials comply with GCP requirements. In addition, our clinical trials must be conducted with product produced under applicable cGMP regulations. Failure to comply with these regulations may require us to repeat clinical trials, which would delay the regulatory approval process.

If these third parties do not successfully carry out their duties under their agreements, if the quality or accuracy of the data they obtain is compromised due to their failure to adhere to clinical trial protocols or to regulatory requirements, or if they otherwise fail to comply with clinical trial protocols or meet expected deadlines, the clinical trials of our product candidates may not meet regulatory requirements. If clinical trials do not meet regulatory requirements or if these third parties need to be replaced, preclinical development activities or clinical trials may be extended, delayed, suspended or terminated. If any of these events occur, we may not be able to obtain regulatory approval of our product candidates on a timely basis or at all.

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We also expect to rely on other third parties to store and distribute drug supplies for our clinical trials. Any performance failure on the part of our distributors could delay clinical development or marketing approval of our product candidates or commercialization of our products, producing additional losses and depriving us of potential product revenue.

We intend to rely on third parties to conduct some or all aspects of our product manufacturing, and these third parties may not perform satisfactorily.

We do not have any manufacturing facilities and do not expect to independently conduct our product manufacturing for research and preclinical and clinical testing. We currently rely, and expect to rely, on third parties to manufacture and supply drug products for our AKB-6548 clinical trials, and we expect to continue to rely on third parties for the manufacture of clinical and, if necessary, commercial quantities of our product candidates. This reliance on third parties increases the risk that we will not have sufficient quantities of our product candidates or such quantities at an acceptable cost or quality, which could delay, prevent or impair our development or commercialization efforts.

Any of these third parties may terminate their engagement with us at any time. While we believe we have sufficient drug product to complete our ongoing Phase 2b trial of AKB-6548, we do not currently have arrangements in place for the manufacturing of drug substance or drug product for the Phase 3 development program or a second source for bulk drug substance for AKB-6548. Prior to the commencement of the Phase 3 development program for AKB-6548, we plan to engage identified third party manufacturers. If those contract manufacturers cannot perform as agreed, we may be required to replace such manufacturers. Although we believe that there are several potential alternative manufacturers who could manufacture our product candidates, we may incur significant delays and added costs in identifying, qualifying and contracting with any such replacement, as well as producing the drug product. In addition, we have to enter into technical transfer agreements and share our know-how with the third-party manufacturers, which can be time-consuming and may result in delays. These delays could result in a suspension of our clinical trials or, if AKB-6548 is approved and marketed, a failure to satisfy patient demand.

Reliance on third party manufacturers entails risks to which we would not be subject if we manufactured the product candidates ourselves, including:

- the inability to negotiate manufacturing agreements with third parties under commercially reasonable terms;
- reduced control as a result of using third party manufacturers for all aspects of manufacturing activities, including regulatory compliance and quality assurance;
- termination or nonrenewal of manufacturing agreements with third parties in a manner or at a time that is costly or damaging to us;
- the possible misappropriation of our proprietary information, including our trade secrets and know-how; and
- disruptions to the operations of our manufacturers or suppliers caused by conditions unrelated to our business or operations, including the bankruptcy of the manufacturer or supplier or a catastrophic event affecting our manufacturers or suppliers.

Any of these events could lead to clinical study delays or failure to obtain regulatory approval, or affect our ability to successfully commercialize future products. Some of these events could be the basis for FDA action, including injunction, recall, seizure or total or partial suspension of production.

The facilities used by our contract manufacturers to manufacture our product candidates must be approved by the FDA pursuant to inspections that will be conducted after we submit our NDA to the FDA. We do not control the manufacturing process of, and are completely dependent on, our contract manufacturers for

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compliance with cGMP requirements for manufacture of both drug substance and finished drug product. If our contract manufacturers cannot successfully manufacture material that conforms to our specifications and the strict regulatory requirements of the FDA, we will not be able to secure and/or maintain regulatory approval for our product candidates. In addition, we have no control over the ability of our contract manufacturers to maintain adequate quality control, quality assurance and qualified personnel. If the FDA, EMA or other regulatory authorities do not approve these facilities for the manufacture of our product candidates or if they withdraw any such approval in the future, we may need to find alternative manufacturing facilities, which would significantly impact our ability to develop, obtain regulatory approval for or market our product candidates, if approved. Moreover, our failure, or the failure of our third party manufacturers, to comply with applicable regulations could result in sanctions being imposed on us, including clinical holds, fines, injunctions, civil penalties, delays, suspension or withdrawal of approvals, license revocation, seizures or recalls of product candidates or products, operating restrictions and criminal prosecutions, any of which could significantly and adversely affect supplies of our products or product candidates.

In addition, our product candidates and any products that we may develop may compete with other product candidates and products for access to manufacturing facilities. Certain of these manufacturing facilities may be contractually prohibited from manufacturing our product due to non-compete agreements with our competitors. There are a limited number of manufacturers that operate under cGMP regulations and that might be capable of manufacturing for us.

Our current and anticipated future dependence upon others for the manufacture of our product candidates or products may adversely affect our future profit margins and our ability to commercialize any products that receive marketing approval on a timely and competitive basis.

If we are unable to manufacture our product candidates in sufficient quantities, at sufficient yields, we may experience delays in product development, clinical trials, regulatory approval and commercial distribution.

Completion of our clinical trials and commercialization of our product candidates require access to, or development of, facilities to manufacture our product candidates at sufficient yields and at commercial scale. We have limited experience manufacturing, or managing third parties in manufacturing, any of our product candidates in the volumes that will be necessary to support large-scale clinical trials or commercial sales. Efforts to establish these capabilities may not meet initial expectations as to scheduling, scale-up, reproducibility, yield, purity, cost, potency or quality.

Our reliance on contract manufacturers may adversely affect our operations or result in unforeseen delays or other problems beyond our control. Because of contractual restraints and the limited number of third-party manufacturers with the expertise, required regulatory approvals and facilities to manufacture our bulk drug product on a commercial scale, replacement of a manufacturer may be expensive and time-consuming and may cause interruptions in the production of our drug product. A third-party manufacturer may also encounter difficulties in production. These problems may include:

- difficulties with production costs, scale-up and yields;
- availability of raw materials and supplies;
- quality control and assurance;
- shortages of qualified personnel;
- compliance with strictly enforced federal, state and foreign regulations that vary in each country where product might be sold; and
- lack of capital funding.

Any delay or interruption in our supply of product candidates could have a material adverse effect on our business, financial condition, results of operations and cash flows.

We may not be successful in establishing and maintaining strategic collaborations, which could adversely affect our ability to develop and commercialize our product candidates, negatively impacting our operating results.

We plan to commercialize AKB-6548 in the United States and will likely seek one or more strategic collaborators to commercialize AKB-6548 in additional markets. We face competition in seeking appropriate collaborators for our product candidates, and the negotiation process is time-consuming and complex. In order for us to successfully collaborate with a third party on our product candidates, potential collaborators must view these product candidates as economically valuable. Even if we are successful in our efforts to establish strategic collaborations, the terms that we agree upon may not be favorable to us, and we may not be able to maintain such strategic collaborations if, for example, development or approval of a product is delayed or sales of an approved product are disappointing. Any delay in entering into strategic collaboration agreements related to our product candidates could delay the development and commercialization of our product candidates and reduce their competitiveness even if they reach the market.

In addition, our strategic collaborators may terminate any agreements they enter into with us, and we may not be able to adequately protect our rights under these agreements. Furthermore, our strategic collaborators will likely negotiate for certain rights to control decisions regarding the development and commercialization of our product candidates, if approved, and may not conduct those activities in the same manner as we do.

If we fail to establish and maintain strategic collaborations related to our product candidates, we will bear all of the risk and costs related to the development and commercialization of any such product candidate, and we may need to seek additional financing, hire additional employees and otherwise develop expertise. This could negatively affect the development of any unpartnered product candidate.

Risks Related to Our Intellectual Property

If our efforts to protect our proprietary technologies are not adequate, we may not be able to compete effectively in our market. We are currently involved in an opposition proceeding involving one of our European patents, and the outcome of that proceeding may affect our ability to establish a competitive advantage in the market or successfully commercialize our lead product candidate in the European Union.

We rely upon a combination of patents, trade secret protection and confidentiality agreements to protect the intellectual property related to our technologies. We will only be able to protect our product candidates, proprietary technologies and their uses from unauthorized use by third parties to the extent that valid and enforceable patents or trade secrets cover them. Any disclosure to or misappropriation by third parties of our confidential proprietary information could enable competitors to quickly duplicate or surpass our technological achievements, thus eroding our competitive position in our market.

In July of 2011, a third party filed an opposition to one of our issued European patents, European Patent No. 2044005, which we refer to as the '005 Patent. During the oral proceedings, which took place on April 10, 2013, the Opposition Division of the European Patent Office decided to maintain certain claims of the patent directed to a compound chosen from a group of eight compounds, including AKB-6548, as well as claims to compositions and methods for treating various diseases, including, but not limited to anemia. Both parties have appealed the decision of the Opposition Division and final resolution of the opposition proceeding will likely take a number of years. We cannot be assured of the breadth of the claims that will remain in the '005 Patent or that the patent will not be revoked in its entirety. If the European Patent Office decides to narrow the scope of the claims or revoke the '005 Patent, we may not be able to establish a competitive advantage in the European Union in our market or successfully commercialize our lead product candidates in the European Union, which could materially adversely affect our business, operating results and financial condition.

Composition-of-matter patents on the active pharmaceutical ingredient are generally considered to be the strongest form of intellectual property protection for pharmaceutical products, as such patents provide protection

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without regard to any method of use. Method-of-use patents protect the use of a product for the specified method. This type of patent does not prevent a competitor from making and marketing a product that is identical to our product for an indication that is outside the scope of the patented method. Moreover, even if competitors do not actively promote their product for our targeted indications, physicians may prescribe these products “off-label.” Although off-label prescriptions may infringe or contribute to the infringement of method-of-use patents, the practice is common and such infringement is difficult to prevent or prosecute.

The strength of patents in the biotechnology and pharmaceutical field involves complex legal and scientific questions and can be uncertain. The patent applications that we own or license may fail to result in issued patents in the United States or in other foreign countries. Even if the patents do successfully issue, third parties may challenge the validity, enforceability, inventorship, or scope thereof, which may result in such patents being narrowed, invalidated or held unenforceable. Furthermore, even if they are unchallenged, our patents and patent applications may not adequately protect our intellectual property or prevent others from designing around our claims. If the breadth or strength of protection provided by the patent applications we hold with respect to our product candidates is threatened, it could dissuade companies from collaborating with us to develop, and threaten our ability to commercialize, our product candidates. Further, if we encounter delays in our clinical trials, the period of time during which we could market our product candidates under patent protection would be reduced. Since patent applications in the United States and most other countries are confidential for a period of time after filing, we cannot be certain that we were the first to file any patent application related to our product candidates. Furthermore, for applications in which all claims are entitled to a priority date before March 16, 2013, an interference proceeding can be provoked by a third-party or instituted by the USPTO, to determine who was the first to invent any of the subject matter covered by the patent claims of our applications. For applications containing a claim not entitled to priority before March 16, 2013, there is greater level of uncertainty in the patent law with the passage of the America Invents Act (2011), which brings into effect significant changes to the U.S. patent laws and which introduces new procedures for challenging pending patent applications and issued patents. A primary change under this reform is creating a “first to file” system in the U.S. This will require us to be cognizant of the time from invention to filing of a patent application.

In addition to the protection afforded by patents, we seek to rely on trade secret protection and confidentiality agreements to protect proprietary know-how that is not patentable, processes for which patents are difficult to enforce and any other elements of our drug discovery and development processes that involve proprietary know-how, information or technology that is not covered by patents. Although we require all of our employees to assign their inventions to us, and require all of our employees, consultants, advisors and any third parties who have access to our proprietary know-how, information or technology to enter into confidentiality agreements, we cannot be certain that our trade secrets and other confidential proprietary information will not be disclosed or that competitors will not otherwise gain access to our trade secrets or independently develop substantially equivalent information and techniques. Furthermore, the laws of some foreign countries do not protect proprietary rights to the same extent or in the same manner as the laws of the United States. As a result, we may encounter significant problems in protecting and defending our intellectual property both in the United States and abroad. If we are unable to prevent unauthorized material disclosure of our intellectual property to third parties, we will not be able to establish or maintain a competitive advantage in our market, which could materially adversely affect our business, operating results and financial condition.

Our reliance on third parties requires us to share our trade secrets, which increases the possibility that a competitor will discover them or that our trade secrets will be misappropriated or disclosed.

Because we rely on third parties to research and develop and to manufacture our product candidates, we must, at times, share trade secrets with them. We seek to protect our proprietary technology in part by entering into confidentiality agreements and, if applicable, material transfer agreements, consulting agreements or other similar agreements with our advisors, employees, third-party contractors and consultants prior to beginning research or disclosing proprietary information. These agreements typically limit the rights of the third parties to use or disclose our confidential information, including our trade secrets. Despite the contractual provisions

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employed when working with third parties, the need to share trade secrets and other confidential information increases the risk that such trade secrets become known by our competitors, are inadvertently incorporated into the technology of others, or are disclosed or used in violation of these agreements. Given that our proprietary position is based, in part, on our know-how and trade secrets, a competitor's discovery of our trade secrets or other unauthorized use or disclosure would impair our competitive position and may have a material adverse effect on our business.

In addition, these agreements typically restrict the ability of our advisors, employees, third-party contractors and consultants to publish data potentially relating to our trade secrets, although our agreements may contain certain limited publication rights. For example, any academic institution that we may collaborate with in the future will usually expect to be granted rights to publish data arising out of such collaboration, provided that we are notified in advance and given the opportunity to delay publication for a limited time period in order for us to secure patent protection of intellectual property rights arising from the collaboration, in addition to the opportunity to remove confidential or trade secret information from any such publication. In the future we may also conduct joint research and development programs that may require us to share trade secrets under the terms of our research and development or similar agreements. Despite our efforts to protect our trade secrets, our competitors may discover our trade secrets, either through breach of our agreements with third parties, independent development or publication of information by any of our third-party collaborators. A competitor's discovery of our trade secrets would impair our competitive position and have an adverse impact on our business.

Third-party claims of intellectual property infringement may be costly and time consuming, and may delay or harm our drug discovery and development efforts. We are currently involved in an opposition proceeding involving the granted European patent of one of our potential competitors.

Our commercial success depends in part on our avoiding infringement of the patents and proprietary rights of third parties. The pharmaceutical and biotechnology industries are characterized by extensive litigation over patent and other intellectual property rights. We may become a party to, or threatened with, future adversarial litigation or other proceedings regarding intellectual property rights with respect to our drug candidates. As the pharmaceutical and biotechnology industries expand and more patents are issued, the risk increases that our drug candidates may give rise to claims of infringement of the patent rights of others.

While our product candidates are in preclinical studies and clinical trials, we believe that the use of our product candidates in these preclinical studies and clinical trials in the U.S. falls within the scope of the exemptions provided by 35 U.S.C. Section 271(e), which provides that it shall not be an act of infringement to make, use, offer to sell, or sell within the U.S. or import into the U.S. a patented invention solely for uses reasonably related to the development and submission of information to the FDA. As our product candidates progress toward commercialization, the possibility of a patent infringement claim against us increases. We attempt to ensure that our product candidates and the methods we employ to manufacture them, as well as the methods for their use we intend to promote, do not infringe other parties' patents and other proprietary rights. There can be no assurance they do not, however, and competitors or other parties may assert that we infringe their proprietary rights in any event.

Third parties may hold or obtain patents or other intellectual property rights and allege in the future that the use of our technologies infringes these patents or intellectual property rights or that we are employing their proprietary technology without authorization. For example, we are aware of certain patents that have been acquired by FibroGen, Inc., or FibroGen, directed to certain heterocyclic carboxamide compounds that are described as inhibitors of prolyl-4-hydroxylase. Those patents, however, expire as of December 2014, before we anticipate receiving regulatory approval for our product candidates. In addition, we are aware of subsequent U.S. patents issued to FibroGen directed to purportedly new methods of using such previously known heterocyclic carboxamide compounds for purposes of treating or affecting certain specified conditions in subjects. Such method-of-use patents protect the use of a product for the specified method. This type of patent does not prevent a competitor from making and marketing the product for an indication that is outside the scope of the patented

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method. We are not aware of any valid U.S. Patents issued to FibroGen that claim methods of using any of our product candidates for purposes of inhibiting hypoxia-inducible factor prolyl-hydroxylases, or HIF-PHs, for the treatment of anemia secondary to CKD. In June 2013, the European Patent Office granted European Patent No. 1463823, or the '823 patent, to FibroGen. The '823 patent claims, among other things, the use of a heterocyclic carboxamide compound selected from the group consisting of pyridine carboxamides, quinoline carboxamides, isoquinoline carboxamides, cinnoline carboxamides, and beta-carboline carboxamides that inhibits HIF-PH enzyme activity in the manufacture of a medicament for increasing endogenous erythropoietin, or EPO, in the prevention, pretreatment, or treatment of anemia. On December 5, 2013, we filed an opposition to the '823 patent requesting that the '823 patent be revoked in its entirety. While, for the reasons set forth in our opposition, we believe the '823 patent should be revoked in its entirety, the ultimate outcome of the opposition remains uncertain. If the European Patent Office decides not to revoke the '823 patent in its entirety, or only certain claims of the '823 patent, and any surviving claims are determined to encompass our intended use of our lead product candidate, we may not be able to commercialize our lead product candidate in the European Union for its intended use, which could materially adversely affect our business, operating results and financial condition. FibroGen filed patent applications related to the '823 patent in other countries and some of these applications have since issued as patents. FibroGen is also pursuing other patent applications in the United States and other countries, and some of these have issued as patents. To the extent any such patents issue or have been issued, we may initiate opposition or other legal proceedings with respect to those patents.

There may be patents of third parties, including FibroGen, of which we are currently unaware with claims to materials, formulations, methods of manufacture or methods for treatment related to the use or manufacture of our drug candidates. Also, because patent applications can take many years to issue, there may be currently pending patent applications which may later result in issued patents that our product candidates may infringe. Notwithstanding the above, third parties, including FibroGen, may in the future claim that our product candidates and other technologies infringes upon these patents and may file suit against us.

Parties making claims against us may seek and obtain injunctive or other equitable relief, which could effectively block our ability to further develop and commercialize AKB-6548 or AKB-6899. If any third-party patents were held by a court of competent jurisdiction to cover the manufacturing process of any of our product candidates, any molecules formed during the manufacturing process or any final product itself, the holders of any such patents may be able to block our ability to commercialize such product candidate unless we obtained a license under the applicable patents, or until such patents expire or they are finally determined to be held invalid or unenforceable. Similarly, if any third-party patent were held by a court of competent jurisdiction to cover aspects of our formulations, processes for manufacture or our intended methods of use, including patient selection methods, the holders of any such patent may be able to block or impair our ability to develop and commercialize the applicable product candidate unless we obtained a license or until such patent expires or is finally determined to be held invalid or unenforceable. We may also elect to enter into a license in order to settle litigation or in order to resolve disputes prior to litigation. Furthermore, even in the absence of litigation, we may need to obtain licenses from third parties to advance our research or allow commercialization of our product candidates. Should a license to a third party patent become necessary, we cannot predict whether we would be able to obtain a license, or if a license were available, whether it would be available on commercially reasonable terms. If such a license is necessary and a license under the applicable patent is unavailable on commercially reasonable terms, or at all, our ability to commercialize our drug candidates may be impaired or delayed, which could in turn significantly harm our business.

Further, defense of infringement claims, regardless of their merit, would involve substantial litigation expense and would be a substantial diversion of employee resources from our business. In the event of a successful claim of infringement against us, we may have to pay substantial damages, including treble damages and attorneys' fees for willful infringement, pay royalties or redesign our products, which may be impossible or require substantial time and monetary expenditure.

We are currently involved in opposition proceedings and may in the future be involved in lawsuits or administrative proceedings to protect or enforce our patents, which could be expensive, time consuming, and unsuccessful.

We are currently involved in two opposition proceedings in the European Patent Office. These proceedings may be ongoing for a number of years and may involve substantial expense and diversion of employee resources from our business. For more information, see the other risk factors under “—Risks Related to Intellectual Property.”

Competitors may infringe our patents or misappropriate our trade secrets or confidential information. To counter infringement or unauthorized use, we may be required to file infringement or misappropriation claims, which can be expensive and time-consuming. We may not be able to prevent infringement of our patents or misappropriation of our trade secrets or confidential information, particularly in countries where the laws may not protect those rights as fully as in the United States. In addition, in an infringement proceeding, a court may decide that a patent of ours is not valid or is unenforceable, or may refuse to stop the other party from using the technology at issue on the grounds that our patents do not cover the technology in question. An adverse result in any litigation or defense proceedings could put one or more of our patents at risk of being invalidated, held unenforceable, or interpreted narrowly and could put our patent applications at risk of not issuing. In addition, there may be a challenge or dispute regarding inventorship or ownership of patents or applications currently identified as being owned by or licensed to us. Defense of these claims, regardless of their merit, would involve substantial litigation expense and would be a substantial diversion of employee resources from our business.

Various administrative proceedings are also available for challenging patents, including interference, reexamination, *inter partes* review, and post-grant review proceedings before the USPTO or oppositions and other comparable proceedings in foreign jurisdictions. Interference proceedings provoked by third parties or brought by the USPTO may be necessary to determine the priority of inventions with respect to our patents or patent applications. An unfavorable outcome could require us to cease using the related technology or to attempt to license rights to it from the prevailing party. Our business could be harmed if the prevailing party does not offer us a license on commercially reasonable terms or at all. Even if we are successful, participation in interference or other administrative proceedings before the USPTO or a foreign patent office may result in substantial costs and distract our management and other employees.

Furthermore, because of the substantial amount of discovery required in connection with intellectual property litigation and some administrative proceedings, there is a risk that some of our confidential information could be compromised by disclosure. In addition, there could be public announcements of the results of hearings, motions or other interim proceedings or developments. If securities analysts or investors perceive these results to be negative, it could have a substantial adverse effect on the price of our common stock.

Obtaining and maintaining our patent protection depends on compliance with various procedural, document submission, fee payment and other requirements imposed by governmental patent agencies, and our patent protection could be reduced or eliminated for non-compliance with these requirements.

Periodic maintenance fees on any issued patent are due to be paid to the USPTO and foreign patent agencies in several stages over the lifetime of the patent. The USPTO and various foreign governmental patent agencies also require compliance with a number of procedural, documentary, fee payment (such as annuities) and other similar provisions during the patent application process. While an inadvertent lapse can in many cases be cured by payment of a late fee or by other means in accordance with the applicable rules, there are situations in which noncompliance can result in abandonment or lapse of the patent or patent application, resulting in partial or complete loss of patent rights in the relevant jurisdiction. Non-compliance events that could result in abandonment or lapse of a patent or patent application include, but are not limited to, failure to respond to official actions within prescribed time limits, non-payment of fees and failure to properly legalize and submit formal documents. In such an event, our competitors might be able to enter the market, which would have a material adverse effect on our business.

We may be subject to claims that our employees, consultants or independent contractors have wrongfully used or disclosed confidential information of third parties.

We have received confidential and proprietary information from collaborators, prospective licensees and other third parties. In addition, we employ individuals who were previously employed at other biotechnology or pharmaceutical companies. We may be subject to claims that we or our employees, consultants or independent contractors have inadvertently or otherwise used or disclosed confidential information of these third parties or our employees' former employers. We may also be subject to claims that former employees, collaborators or other third parties have an ownership interest in our patents or other intellectual property. We may be subject to ownership disputes in the future arising, for example, from conflicting obligations of consultants or others who are involved in developing our drug candidates. Litigation may be necessary to defend against these claims. If we fail in defending any such claims, in addition to paying monetary damages, we may lose valuable intellectual property rights, such as exclusive ownership of, or right to use, valuable intellectual property. Such an outcome could have a material adverse effect on our business. Even if we are successful in defending against these claims, litigation could result in substantial cost and be a distraction to our management and employees.

We may not be able to protect our intellectual property rights throughout the world.

Filing, prosecuting and defending patents on product candidates in all countries throughout the world would be prohibitively expensive, and our intellectual property rights in some countries outside the United States can be less extensive than those in the United States. In addition, the laws of some countries do not protect intellectual property rights to the same extent as laws in the United States. Consequently, we may not be able to prevent third parties from practicing our inventions in all countries outside the United States, or from selling or importing products made using our inventions in and into the United States or other countries. Competitors may use our technologies in countries where we have not obtained patent protection to develop their own products and further, may infringe our patents in territories where we have patent protection, but enforcement is not as strong as in the United States. These products may compete with our products and our patents or other intellectual property rights may not be effective or sufficient to prevent them from competing.

Many companies have encountered significant problems in protecting and defending intellectual property rights in certain countries. The legal systems of certain countries, particularly certain developing countries, do not favor the enforcement of patents, trade secrets and other intellectual property, particularly those relating to pharmaceutical and biotechnology products, which could make it difficult for us to stop the infringement of our patents or marketing of competing products in violation of our proprietary rights generally. Proceedings to enforce our patent rights in foreign countries could result in substantial costs and divert our efforts and attention from other aspects of our business, could put our patents at risk of being invalidated or interpreted narrowly and our patent applications at risk of not issuing and could provoke third parties to assert claims against us. We may not prevail in any lawsuits that we initiate and the damages or other remedies awarded, if any, may not be commercially meaningful. Accordingly, our efforts to enforce our intellectual property rights around the world may be inadequate to obtain a significant commercial advantage from the intellectual property that we develop or license.

Risks Related to Commercialization

Our future commercial success depends upon attaining significant market acceptance of our product candidates, if approved, among physicians, patients, third-party payors and others in the medical community.

Even if we obtain marketing approval for AKB-6548, AKB-6899 or any other product candidates that we may develop or acquire in the future, these product candidates may not gain market acceptance among physicians, third-party payors, patients and others in the medical community. In addition, market acceptance of any approved products depends on a number of other factors, including:

- the efficacy and safety of the product, as demonstrated in clinical trials;
- the clinical indications for which the product is approved and the label approved by regulatory authorities for use with the product, including any warnings that may be required on the label;

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- acceptance by physicians and patients of the product as a safe and effective treatment and the willingness of the target patient population to try new therapies and of physicians to prescribe new therapies;
- the cost, safety and efficacy of treatment in relation to alternative treatments;
- the availability of adequate coverage and reimbursement by third party payors and government authorities;
- the ability to contract with dialysis providers;
- relative convenience and ease of administration;
- the prevalence and severity of adverse side effects;
- the effectiveness of our sales and marketing efforts; and
- the restrictions on the use of our products together with other medications, if any.

For example, two of the largest operators of dialysis clinics in the United States, DaVita and Fresenius, account for more than half of the injectable rESA sales in the U.S. dialysis market and have entered into long-term sales agreements with Amgen that began in January 2012. We believe that it may be challenging to enter into or expand upon long or short-term supply agreements with DaVita, Fresenius or other operators of dialysis clinics.

Market acceptance is critical to our ability to generate significant revenue. In addition, any product candidate, if approved and commercialized, may be accepted in only limited capacities or not at all. If any approved products are not accepted by the market to the extent that we expect, we may not be able to generate significant revenue and our business would suffer.

If we are unable to establish sales, marketing and distribution capabilities or to enter into agreements with third parties to market and sell our product candidates, we may not be successful in commercializing our product candidates if and when they are approved.

We do not have a sales or marketing infrastructure and have no experience in the sale, marketing or distribution of pharmaceutical products. To achieve commercial success for any product for which we obtain marketing approval, we will need to establish a sales and marketing organization or make arrangements with third parties to perform these services.

There are risks involved with establishing our own sales, marketing and distribution capabilities. For example, recruiting and training a sales force are expensive and time consuming and could delay any product launch. If the commercial launch of a product candidate for which we recruit a sales force and establish marketing capabilities is delayed or does not occur for any reason, we would have prematurely or unnecessarily incurred these commercialization expenses. This may be costly, and our investment would be lost if we cannot retain or reposition our sales and marketing personnel.

Factors that may inhibit our efforts to commercialize our products on our own include:

- our inability to recruit, train and retain adequate numbers of effective sales and marketing personnel;
- the inability of sales personnel to obtain access to physicians or persuade adequate numbers of physicians to prescribe any future products;
- our inability to effectively manage geographically dispersed sales and marketing team;
- the lack of complementary products to be offered by sales personnel, which may put us at a competitive disadvantage relative to companies with more extensive product lines; and
- unforeseen costs and expenses associated with creating an independent sales and marketing organization.

If we are unable to establish our own sales, marketing and distribution capabilities and have to enter into arrangements with third parties to perform these services, our profitability, if any, is likely to be lower than if we

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were to market, sell and distribute any products that we develop ourselves. In addition, we may not be successful in entering into arrangements with third parties to sell, market and distribute our product candidates or may be unable to do so on terms that are favorable to us. We likely will have little control over such third parties, and any of them may fail to devote the necessary resources and attention to sell and market our products effectively. If we do not establish sales, marketing and distribution capabilities successfully, either on our own or in collaboration with third parties, we will not be successful in commercializing our product candidates.

Coverage and reimbursement may be limited or unavailable in certain market segments for any approved products, which could make it difficult for us to sell our products profitably.

Market acceptance and sales of any approved products will depend significantly on the availability of adequate coverage and reimbursement from third-party payors and may be affected by existing and future healthcare reform measures. Government authorities and third-party payors decide which drugs they will pay for and establish formularies and reimbursement levels. Coverage and reimbursement by a third-party payor may depend upon a number of factors, including the third-party payor's determination that use of a product is:

- a covered benefit under its health plan;
- safe, effective and medically necessary;
- appropriate for the specific patient;
- cost-effective; and
- neither experimental nor investigational.

Obtaining coverage and reimbursement approval for a product from a government or other third party payor is a time consuming and costly process that could require us to provide supporting scientific, clinical and cost-effectiveness data for the use of our products to the payor. Additionally, we may be required to enter into contracts with third-party payors to obtain favorable formulary status. We may not be able to provide data sufficient to gain acceptance with respect to coverage and reimbursement. We cannot be sure that coverage or adequate reimbursement will be available for any of our product candidates. Even if we obtain coverage for our product candidates, third-party payors may not establish adequate reimbursement amounts, which may reduce the demand for, or the price of, our products. If reimbursement is not available or is available only to limited levels, we may not be able to commercialize certain of our products. In addition, in the United States third-party payors are increasingly attempting to contain healthcare costs by limiting both coverage and the level of reimbursement of new drugs. As a result, significant uncertainty exists as to whether and how much third-party payors will reimburse patients for their use of newly approved drugs, which in turn will put pressure on the pricing of drugs.

In addition, if AKB-6548 is used in an outpatient dialysis facility, such facilities often receive fixed reimbursement for all dialysis services furnished to patients with end-stage renal disease, or ESRD. For example, Medicare payments to ESRD facilities for such services are based on a prospective payment system known as the basic case-mix adjusted composite payment system. These payments cover a bundle of items and services routinely required for dialysis treatments furnished to Medicare beneficiaries in Medicare-certified ESRD facilities or at their home, including the cost of certain routine drugs such as our product candidates. Patient and treatment provider access to adequate coverage and reimbursement by government and private insurance plans is central to the acceptance of any products for which we receive regulatory approval. We may be unable to sell AKB-6548, if approved, to dialysis providers on a profitable basis if third-party payors reduce their current levels of payment, or if our costs of production increase faster than increases in reimbursement levels.

Price controls may be imposed, which may adversely affect our future profitability.

In some countries, particularly member states of the European Union, the pricing of prescription pharmaceuticals is subject to governmental control. In these countries, pricing negotiations with governmental authorities can take considerable time after receipt of marketing approval for a product. In addition, there can be

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considerable pressure by governments and other stakeholders on prices and reimbursement levels, including as part of cost containment measures. Political, economic and regulatory developments may further complicate pricing negotiations, and pricing negotiations may continue after reimbursement has been obtained. Reference pricing used by various European Union member states and parallel distribution, or arbitrage between low-priced and high-priced member states, can further reduce prices. In some countries, we may be required to conduct a clinical trial or other studies that compare the cost-effectiveness of our product candidates to other available products in order to obtain or maintain reimbursement or pricing approval. Publication of discounts by third-party payors or authorities may lead to further pressure on the prices or reimbursement levels within the country of publication and other countries. If reimbursement of our products is unavailable or limited in scope or amount, or if pricing is set at unsatisfactory levels, our business could be adversely affected.

The impact of recent healthcare reform and other changes in the healthcare industry and in healthcare spending is currently unknown, and may adversely affect our business model.

Our revenue prospects could be affected by changes in healthcare spending and policy in the United States and abroad. We operate in a highly regulated industry and new laws, regulations or judicial decisions, or new interpretations of existing laws, regulations or decisions, related to healthcare availability, the method of delivery or payment for healthcare products and services could negatively impact our business, operations and financial condition.

In the United States, the Medicare Prescription Drug, Improvement, and Modernization Act of 2003, also called the MMA, changed the way Medicare covers and pays for pharmaceutical products. The legislation expanded Medicare coverage for drug purchases by the elderly and introduced a new reimbursement methodology based on average sales prices for physician-administered drugs. In addition, this legislation provided authority for limiting the number of drugs that will be covered in any therapeutic class. As a result of this legislation and the expansion of federal coverage of drug products, we expect that there will be additional pressure to reduce costs. For example, the Centers for Medicare and Medicaid Services, or CMS, has enacted regulations that reduced capitated payments to dialysis providers. These cost reduction initiatives and other provisions of this legislation could decrease the scope of coverage and the price that we receive for any approved products and could seriously harm our business. While the MMA applies only to drug benefits for Medicare beneficiaries, private payors often follow Medicare coverage policies and payment limitations in setting their own reimbursement rates, and any reduction in reimbursement that results from the MMA may cause a similar reduction in payments from private payors. Similar regulations or reimbursement policies may be enacted in international markets which could similarly impact our business.

In addition, the Patient Protection and Affordable Care Act, as amended by the Health Care and Education Reconciliation Act, or collectively PPACA, was enacted in 2010 with a goal of reducing the cost of healthcare and substantially changing the way healthcare is financed by both government and private insurers. The PPACA, among other things, increases the minimum Medicaid rebates owed by manufacturers under the Medicaid Drug Rebate Program and extends the rebate program to individuals enrolled in Medicaid managed care organizations, establishes annual fees and taxes on manufacturers of certain branded prescription drugs, and creates a new Medicare Part D coverage gap discount program, in which manufacturers must agree to offer 50% point-of-sale discounts off negotiated prices of applicable brand drugs to eligible beneficiaries during their coverage gap period as a condition for the manufacturer's outpatient drugs to be covered under Medicare Part D. In addition, other legislative changes have been proposed and adopted in the United States since the PPACA was enacted. On August 2, 2011, the Budget Control Act of 2011 created measures for spending reductions by Congress. A Joint Select Committee on Deficit Reduction, tasked with recommending a targeted deficit reduction of at least \$1.2 trillion for the years 2013 through 2021, was unable to reach required goals, thereby triggering the legislation's automatic reduction to several government programs. This includes aggregate reductions of Medicare payments to providers of up to 2% per fiscal year, which went into effect on April 1, 2013.

It is likely that federal and state legislatures within the United States and foreign governments will continue to consider changes to existing healthcare legislation. We cannot predict the reform initiatives that may be

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adopted in the future or whether initiatives that have been adopted will be repealed or modified. The continuing efforts of the government, insurance companies, managed care organizations and other payors of healthcare services to contain or reduce costs of healthcare may adversely affect:

- the demand for any drug products for which we may obtain regulatory approval;
- our ability to set a price that we believe is fair for our products;
- our ability to obtain coverage and reimbursement approval for a product;
- our ability to generate revenues and achieve or maintain profitability; and
- the level of taxes that we are required to pay.

If our product candidates obtain marketing approval, we will be subject to healthcare laws, regulation and enforcement and our failure to comply with those laws could have a material adverse effect on our results of operations and financial conditions.

If we obtain approval for any of our product candidates and begin commercializing them, our operations may be directly, or indirectly through our customers, subject to additional healthcare regulation and enforcement by the federal government and the states and foreign governments in which we conduct our business. The laws that may affect our ability to operate include:

- the federal Anti-Kickback Statute, which prohibits, among other things, persons from knowingly and willfully soliciting, receiving, offering or paying remuneration, directly or indirectly, to induce, or in return for, the purchase or recommendation of an item or service reimbursable under a federal healthcare program, such as the Medicare and Medicaid programs;
- federal civil and criminal false claims laws and civil monetary penalty laws, which prohibit, among other things, individuals or entities from knowingly presenting, or causing to be presented, claims for payment from Medicare, Medicaid, or other third-party payors that are false or fraudulent;
- the federal Health Insurance Portability and Accountability Act of 1996, or HIPAA, which created new federal criminal statutes that prohibit executing a scheme to defraud any healthcare benefit program and making false statements relating to healthcare matters;
- HIPAA, as amended by the Health Information Technology and Clinical Health Act, or HITECH, and its implementing regulations, which imposes certain requirements relating to the privacy, security, and transmission of individually identifiable health information;
- the federal physician sunshine requirements under PPACA, which requires manufacturers of drugs, devices, biologics, and medical supplies to report annually to the CMS information related to payments and other transfers of value to physicians, other healthcare providers, and teaching hospitals, and ownership and investment interests held by physicians and other healthcare providers and their immediate family members;
- state law equivalents of each of the above federal laws, such as anti-kickback and false claims laws that may apply to items or services reimbursed by any third-party payor, including commercial insurers; state laws that require pharmaceutical companies to comply with the pharmaceutical industry's voluntary compliance guidelines and the applicable compliance guidance promulgated by the federal government, or otherwise restrict payments that may be made to healthcare providers and other potential referral sources; state laws that require drug manufacturers to report information related to payments and other transfers of value to physicians and other healthcare providers or marketing expenditures; and state laws governing the privacy and security of health information in certain circumstances, many of which differ from each other in significant ways, thus complicating compliance efforts.

Because of the breadth of these laws and the narrowness of the statutory exceptions and safe harbors available, it is possible that some of our business activities could be subject to challenge under one or more of

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such laws. In addition, recent healthcare reforms have strengthened these laws. For example, the PPACA, among other things, amends the intent requirement of the federal anti-kickback and criminal healthcare fraud statutes. A person or entity no longer needs to have actual knowledge of the statute or specific intent to violate it. The PPACA also amended the False Claims Act, such that violations of the anti-kickback statute are now deemed violations of the False Claims Act. To constitute a false claim prior to this amendment, an anti-kickback violation had to be accompanied by a false statement, such as false certification of compliance.

If our operations are found to be in violation of any of such laws or any other governmental regulations that apply to us, we may be subject to penalties, including civil and criminal penalties, damages, fines, the curtailment or restructuring of our operations, the exclusion from participation in federal and state healthcare programs and imprisonment, any of which could materially adversely affect our ability to operate our business and our financial results.

We face substantial competition, which may result in others discovering, developing or commercializing products before, or more successfully, than we do.

The development and commercialization of new drug products is highly competitive. Our future success depends on our ability to demonstrate and maintain a competitive advantage with respect to the development and commercialization of our product candidates. Our objective is to develop and commercialize new products with superior efficacy, convenience, tolerability and safety. In many cases, the products that we commercialize will compete with existing, market-leading products.

If AKB-6548 is approved and launched commercially, competing drugs may include EPOGEN and Aranesp, commercialized by Amgen, Procrit and Eprex, commercialized by Johnson & Johnson, and Mircera, commercialized by Roche outside of the United States. We may face competition from potential new anemia therapies. There are several other HIF product candidates in various stages of active development for anemia indications that may be in direct competition with AKB-6548 if and when it is approved and launched commercially. These candidates are being developed by such companies as FibroGen/AstraZeneca, GlaxoSmithKline and Bayer. FibroGen, in particular, is currently in Phase 3 clinical development of its product candidate, FG-4592 (roxadustat). Some of these product candidates may enter the market as early as 2017. In addition, certain companies are developing potential new therapies for renal-related diseases that could potentially reduce rESA utilization and thus limit the market for AKB-6548 if and when it is approved and launched commercially.

Since rESAs are biologic products, the introduction of biosimilars into the rEPO market in the United States will constitute additional competition for AKB-6548 if we are able to obtain approval for and commercially launch our product. A biosimilar product is a follow-on version of an existing, branded biologic product. The patents for the existing, branded product must expire in a given market before biosimilars may enter that market without risk of being sued for patent infringement. In addition, an application for a biosimilar product cannot be approved by the FDA until 12 years after the existing, branded product was approved under a Biologics License Application, or BLA. The patents for epoetin alfa, a version of rEPO, expired in 2004 in the European Union, and the remaining patents have expired or will expire between 2012 and 2015 in the United States. Several biosimilar versions of rEPO are available for sale in the European Union and biosimilar versions of rEPO are currently being studied in clinical trials in the United States.

Many of our potential competitors have significantly greater financial, manufacturing, marketing, drug development, technical and human resources than we do. Large pharmaceutical companies, in particular, have extensive experience in clinical testing, obtaining regulatory approvals, recruiting patients and in manufacturing pharmaceutical products. Large and established companies such as Amgen and Roche, among others, compete in the market for drug products to treat anemia. In particular, these companies have greater experience and expertise in securing government contracts and grants to support their research and development efforts, conducting testing and clinical trials, obtaining regulatory approvals to market products, manufacturing such products on a broad scale and marketing approved products. These companies also have significantly greater research and marketing

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capabilities than we do and may also have products that have been approved or are in late stages of development, and have collaborative arrangements in our target markets with leading companies and research institutions. Established pharmaceutical companies may also invest heavily to accelerate discovery and development of novel compounds or to in-license novel compounds that could make the product that we develop obsolete. As a result of all of these factors, our competitors may succeed in obtaining patent protection and/or FDA approval or discovering, developing and commercializing products before, or more effectively than, we do. In addition, any new product that competes with an approved product must demonstrate compelling advantages in efficacy, convenience, tolerability and safety in order to overcome price competition and to be commercially successful. If we are not able to compete effectively against potential competitors, our business will not grow and our financial condition and operations will suffer.

Our products may cause undesirable side effects or have other properties that delay or prevent their regulatory approval or limit their commercial potential.

Undesirable side effects caused by our products or even competing products in development that utilize a common mechanism of action could cause us or regulatory authorities to interrupt, delay or halt clinical trials and could result in the denial of regulatory approval by the FDA or other regulatory authorities and potential products liability claims. We are currently conducting a Phase 2b clinical trial for AKB-6548. Serious adverse events deemed to be caused by our product candidates could have a material adverse effect on the development of our product candidates and our business as a whole. The most common drug-related adverse events to date in the clinical trial evaluating the safety and tolerability of AKB-6548 have been gastro-intestinal disorders. Our understanding of the relationship between AKB-6548 and these events, as well as our understanding of adverse events in future clinical trials of other product candidates, may change as we gather more information, and additional unexpected adverse events may be observed.

If we or others identify undesirable side effects caused by our product candidates either before or after receipt of marketing approval, a number of potentially significant negative consequences could result, including:

- our clinical trials may be put on hold;
- patient recruitment could be slowed, or enrolled patients may not want to complete a clinical trial;
- we may be unable to obtain regulatory approval for our product candidates or regulatory authorities may withdraw approvals of product candidates;
- regulatory authorities may require additional warnings on the label;
- a medication guide outlining the risks of such side effects for distribution to patients may be required;
- we could be sued and held liable for harm caused to patients; and
- our reputation may suffer.

Any of these events could prevent us from achieving or maintaining market acceptance of our products and could substantially increase commercialization costs.

Risks Related to Our Business and Industry

If we fail to attract and keep senior management and key scientific personnel, we may be unable to successfully develop our products, conduct our clinical trials and commercialize our product candidates.

We are highly dependent on members of our senior management, including John Butler, our President and Chief Executive Officer and Robert Shalwitz, our Chief Medical Officer. The loss of the services of either of these persons could impede the achievement of our research, development and commercialization objectives.

Recruiting and retaining qualified scientific, clinical, manufacturing and sales and marketing personnel will also be critical to our success. The loss of the services of our executive officers or other key employees could

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impede the achievement of our research, development and commercialization objectives and seriously harm our ability to successfully implement our business strategy. Furthermore, replacing executive officers and key employees may be difficult and may take an extended period of time because of the limited number of individuals in our industry with the breadth of skills and experience required to successfully develop, gain regulatory approval of and commercialize products. We may be unable to hire, train, retain or motivate these key personnel on acceptable terms given the intense competition among numerous biopharmaceutical companies for similar personnel.

In addition, certain of our current employees, including an executive officer, also provide services to Aerpio Therapeutics, Inc., or Aerpio, a company we spun out in 2011. As a result, these employees devote some of their time to activities relating to Aerpio's business. Moreover, some of these employees may become full-time employees of Aerpio and we will be forced to hire additional personnel to replace them.

We also experience competition for the hiring of scientific and clinical personnel from universities and research institutions. In addition, we rely on consultants and advisors, including scientific and clinical advisors, to assist us in formulating our research and development and commercialization strategy. Our consultants and advisors may be employed by employers other than us and may have commitments under consulting or advisory contracts with other entities that may limit their availability to us. If we are unable to continue to attract and retain high quality personnel, our ability to pursue our growth strategy will be limited.

Our employees, independent contractors, principal investigators, CROs, consultants and vendors may engage in misconduct or other improper activities, including noncompliance with regulatory standards and requirements and insider trading.

We are exposed to the risk that our employees, independent contractors, principal investigators, CROs, consultants and vendors may engage in fraudulent conduct or other illegal activity. Misconduct by these parties could include intentional, reckless and/or negligent conduct or unauthorized activities that violate: (1) FDA regulations, including those laws that require the reporting of true, complete and accurate information to the FDA, (2) manufacturing standards, (3) federal and state healthcare fraud and abuse laws and regulations, or (4) laws that require the reporting of true and accurate financial information and data. Specifically, sales, marketing and business arrangements in the healthcare industry are subject to extensive laws and regulations intended to prevent fraud, kickbacks, self-dealing and other abusive practices. These laws and regulations may restrict or prohibit a wide range of pricing, discounting, marketing and promotion, sales commission, customer incentive programs and other business arrangements. It is not always possible to identify and deter misconduct by employees and other third parties, and the precautions we take to detect and prevent this activity may not be effective in controlling unknown or unmanaged risks or losses or in protecting us from governmental investigations or other actions or lawsuits stemming from a failure to be in compliance with such laws or regulations. If any such actions are instituted against us, and we are not successful in defending ourselves or asserting our rights, those actions could have a significant impact on our business, including the imposition of civil, criminal and administrative penalties, damages, monetary fines, possible exclusion from participation in Medicare, Medicaid and other federal healthcare programs, contractual damages, reputational harm, diminished profits and future earnings, and curtailment of our operations, any of which could adversely affect our ability to operate our business and our results of operations.

We may encounter difficulties in managing our growth and expanding our operations successfully.

As we seek to advance our product candidates through clinical trials and commercialization, we will need to expand our development, regulatory, manufacturing, marketing and sales capabilities or contract with third parties to provide these capabilities for us. As our operations expand, we expect that we will need to manage additional relationships with various strategic collaborators, suppliers and other third parties. Future growth will impose significant added responsibilities on members of management. Our future financial performance and our ability to commercialize AKB-6548, if approved, and any other product candidates and to compete effectively will depend, in part, on our ability to manage any future growth effectively. To that end, we must be able to

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manage our development efforts and clinical trials effectively and hire, train and integrate additional management, administrative and, if necessary, sales and marketing personnel. We may not be able to accomplish these tasks, and our failure to accomplish any of them could prevent us from successfully growing our company.

If product liability lawsuits are brought against us, we may incur substantial liabilities and may be required to limit commercialization of our product candidates.

We face an inherent risk of product liability as a result of the clinical testing of our product candidates and will face an even greater risk if we commercialize any products. For example, we may be sued if any product we develop allegedly causes injury or is found to be otherwise unsuitable during product testing, manufacturing, marketing or sale. Any such product liability claims may include allegations of defects in manufacturing, defects in design, a failure to warn of dangers inherent in the product, negligence, strict liability and a breach of warranties. Claims could also be asserted under state consumer protection acts. If we cannot successfully defend ourselves against product liability claims, we may incur substantial liabilities or be required to limit commercialization of our product candidates. Even a successful defense would require significant financial and management resources. Regardless of the merits or eventual outcome, liability claims may result in:

- decreased demand for any product candidates or products that we may develop;
- injury to our reputation and significant negative media attention;
- withdrawal of clinical trial participants;
- significant costs to defend the related litigation;
- a diversion of management's time and our resources;
- substantial monetary awards to trial participants or patients;
- product recalls, withdrawals, or labeling, marketing or promotional restrictions;
- loss of revenue;
- the inability to commercialize any product candidates that we may develop; and
- a decline in our stock price.

Failure to obtain and retain sufficient product liability insurance at an acceptable cost to protect against potential product liability claims could prevent or inhibit the commercialization of products we develop. We currently carry product liability insurance covering our clinical trials in the amount of \$10 million in the aggregate. Although we maintain product liability insurance, any claim that may be brought against us could result in a court judgment or settlement in an amount that is not covered, in whole or in part, by our insurance or that is in excess of the limits of our insurance coverage. Our insurance policies also have various exclusions, and we may be subject to a product liability claim for which we have no coverage. We will have to pay any amounts awarded by a court or negotiated in a settlement that exceed our coverage limitations or that are not covered by our insurance, and we may not have, or be able to obtain, sufficient capital to pay such amounts.

If we fail to comply with environmental, health and safety laws and regulations, we could become subject to fines or penalties or incur costs that could harm our business.

We are subject to numerous environmental, health and safety laws and regulations, including those governing laboratory procedures and the handling, use, storage, treatment and disposal of hazardous materials and wastes. Our operations involve the use of hazardous and flammable materials, including chemicals and biological materials. Our operations also produce hazardous waste products. We generally contract with third parties for the disposal of these materials and wastes. We cannot eliminate the risk of contamination or injury from these materials. In the event of contamination or injury resulting from our use of hazardous materials, we could be held liable for any resulting damages, and any liability could exceed our resources. We also could incur significant costs associated with civil or criminal fines and penalties for failure to comply with such laws and regulations.

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Although we maintain workers' compensation insurance to cover us for costs and expenses we may incur due to injuries to our employees resulting from the use of hazardous materials, this insurance may not provide adequate coverage against potential liabilities. We do not maintain insurance for environmental liability or toxic tort claims that may be asserted against us in connection with our storage or disposal of biological, hazardous or radioactive materials.

In addition, we may incur substantial costs in order to comply with current or future environmental, health and safety laws and regulations. These current or future laws and regulations may impair our research, development or production efforts. Our failure to comply with these laws and regulations also may result in substantial fines, penalties or other sanctions.

We may not be able to win government, academic institution or non-profit contracts or grants.

From time to time, we may apply for contracts or grants from government agencies, non-profit entities and academic institutions. Such contracts or grants can be highly attractive because they provide capital to fund the on-going development of our product candidates without diluting our stockholders. However, there is often significant competition for these contracts or grants. Entities offering contracts or grants may have requirements to apply for or to otherwise be eligible to receive certain contracts or grants that our competitors may be able to satisfy that we cannot. In addition, such entities may make arbitrary decisions as to whether to offer contracts or make grants, to whom the contracts or grants will be awarded and the size of the contracts or grants to each awardee. Even if we are able to satisfy the award requirements, there is no guarantee that we will be a successful awardee. Therefore, we may not be able to win any contracts or grants in a timely manner, if at all.

Risks Related to Our Common Stock and This Offering

We are eligible to be treated as an "emerging growth company" as defined in the Jumpstart Our Business Startups Act of 2012, and we cannot be certain if the reduced disclosure requirements applicable to emerging growth companies will make our common stock less attractive to investors.

We are an "emerging growth company", as defined in the Jumpstart Our Business Startups Act of 2012, or the JOBS Act. For as long as we continue to be an emerging growth company, we may take advantage of exemptions from various reporting requirements that are applicable to other public companies that are not emerging growth companies. These exemptions include:

- being permitted to provide only two years of audited financial statements, in addition to any required unaudited interim financial statements, with correspondingly reduced "Management's Discussion and Analysis of Financial Condition and Results of Operations" disclosure;
- not being required to comply with the auditor attestation requirements in the assessment of our internal control over financial reporting;
- not being required to comply with any requirement that may be adopted by the Public Company Accounting Oversight Board;
- reduced disclosure obligations regarding executive compensation; and
- exemptions from the requirements of holding a nonbinding advisory vote on executive compensation and shareholder approval of any golden parachute payments not previously approved.

We have taken advantage of reduced reporting burdens in this prospectus. In particular, in this prospectus, we have provided only two years of audited financial statements and have not included all of the executive compensation related information that would be required if we were not an emerging growth company. Investors may find our common stock less attractive if we continue to rely on these exemptions. If some investors find our common stock less attractive as a result, there may be a less active trading market for our common stock and our stock price may be more volatile. In addition, the JOBS Act provides that an emerging growth company can take advantage of an extended transition period for complying with new or revised accounting standards. This allows an emerging growth company to

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delay the adoption of these accounting standards until they would otherwise apply to private companies. We have irrevocably elected not to avail ourselves of this exemption and, therefore, we will be subject to the same new or revised accounting standards as other public companies that are not emerging growth companies.

We could be an emerging growth company for up to five years, although circumstances could cause us to lose that status earlier, including if the market value of our common stock held by non-affiliates exceeds \$700 million as of any June 30 before that time or if we have total annual gross revenue of \$1 billion or more during any fiscal year before that time, in which cases we would no longer be an emerging growth company as of the following December 31 or, if we issue more than \$1 billion in non-convertible debt during any three-year period before that time, we would cease to be an emerging growth company immediately. Even after we no longer qualify as an emerging growth company, we may still qualify as a “smaller reporting company” if the market value of our common stock held by non-affiliates is below \$75 million as of June 30 in any given year, which would allow us to take advantage of many of the same exemptions from disclosure requirements, including exemption from the auditor attestation requirements of Section 404 of the Sarbanes-Oxley Act and reduced disclosure obligations regarding executive compensation in our periodic reports and proxy statements.

We do not know whether a market will develop for our common stock or what the market price of our common stock will be and, as a result, it may be difficult for you to sell your shares of our common stock.

Before this offering, there was no public trading market for our common stock. If a market for our common stock does not develop or is not sustained, it may be difficult for you to sell your shares of common stock at an attractive price, or at all. Further, an inactive market may also impair our ability to raise capital by selling shares of our common stock and may impair our ability to enter into strategic collaborations or acquire companies or products by using our shares of common stock as consideration. We cannot predict the prices at which our common stock will trade. It is possible that in one or more future periods our results of operations may be below the expectations of public market analysts and investors and, as a result of these and other factors, the price of our common stock may fall.

The market price of our common stock may be highly volatile, and you may not be able to resell your shares at or above the initial public offering price.

The initial public offering price for our shares will be determined by negotiations between us and the representatives of the underwriters and may not be indicative of prices that will prevail in the trading market. The market price of shares of our common stock could be subject to wide fluctuations in response to many risk factors listed in this section, and others beyond our control, including:

- results of clinical trials of our product candidates;
- the timing of the release of results of our clinical trials;
- results of clinical trials of our competitors’ products;
- safety issues with respect to our products or our competitors’ products;
- regulatory actions with respect to our products or our competitors’ products;
- actual or anticipated fluctuations in our financial condition and operating results;
- publication of research reports by securities analysts about us or our competitors or our industry;
- our failure or the failure of our competitors to meet analysts’ projections or guidance that we or our competitors may give to the market;
- additions and departures of key personnel;
- strategic decisions by us or our competitors, such as acquisitions, divestitures, spin-offs, joint ventures, strategic investments or changes in business strategy;

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- the passage of legislation or other regulatory developments affecting us or our industry;
- fluctuations in the valuation of companies perceived by investors to be comparable to us;
- sales of our common stock by us, our insiders or our other stockholders;
- speculation in the press or investment community;
- announcement or expectation of additional financing efforts;
- changes in accounting principles;
- terrorist acts, acts of war or periods of widespread civil unrest;
- natural disasters and other calamities;
- changes in market conditions for biopharmaceutical stocks; and
- changes in general market and economic conditions.

In addition, the stock market has recently experienced significant volatility, particularly with respect to pharmaceutical, biotechnology and other life sciences company stocks. The volatility of pharmaceutical, biotechnology and other life sciences company stocks often does not relate to the operating performance of the companies represented by the stock. As we operate in a single industry, we are especially vulnerable to these factors to the extent that they affect our industry or our products, or to a lesser extent our markets. In the past, securities class action litigation has often been initiated against companies following periods of volatility in their stock price. This type of litigation could result in substantial costs and divert our management's attention and resources, and could also require us to make substantial payments to satisfy judgments or to settle litigation.

Our principal stockholders and management own a significant percentage of our stock and will be able to exercise significant influence over matters subject to stockholder approval.

As of September 30, 2013, our executive officers, directors and principal stockholders, together with their respective affiliates, owned approximately 80% of our common stock, including shares subject to outstanding options that are exercisable within 60 days after such date, and we expect that upon completion of this offering that same group will continue to hold at least % of our outstanding common stock. Accordingly, even after this offering, these stockholders will be able to exert a significant degree of influence over our management and affairs and over matters requiring stockholder approval, including the election of our board of directors and approval of significant corporate transactions. This concentration of ownership could have the effect of entrenching our management and/or the board of directors, delaying or preventing a change in our control or otherwise discouraging a potential acquirer from attempting to obtain control of us, which in turn could have a material and adverse effect on the fair market value of our common stock.

A significant portion of our total outstanding shares may be sold into the public market in the near future, which could cause the market price of our common stock to drop significantly, even if our business is doing well.

Sales of a substantial number of shares of our common stock in the public market could occur at any time after the expiration of the lock-up agreements described in the "Underwriting" section of this prospectus. These sales, or the market perception that the holders of a large number of shares intend to sell shares, could reduce the market price of our common stock. After this offering, we will have shares of common stock outstanding. This includes the shares that we are selling in this offering, which may be resold in the public market immediately subject to any restrictions imposed on our affiliates under Rule 144. The remaining shares, or % of our outstanding shares after this offering, are currently restricted as a result of securities laws or lock-up agreements but will be able to be sold, subject to any applicable volume limitations under federal securities laws with respect to affiliate sales, in the near future as set forth below.

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In addition, as of September 30, 2013, there were 702,625 shares subject to outstanding options that will become eligible for sale in the public market to the extent permitted by any applicable vesting requirements, the lock-up agreements and Rules 144 and 701 under the Securities Act. Moreover, after this offering, holders of an aggregate of 7,381,221 shares of our common stock will have rights, subject to some conditions, to require us to file registration statements covering their shares or to include their shares in registration statements that we may file for ourselves or other stockholders. If such holders, by exercising their registration rights, cause a large number of securities to be registered and sold into the public market, these sales could have an adverse effect on the market price for our common stock. We also intend to register all shares of common stock that we may issue under our employee benefit plans, including our 2014 Equity Incentive Plan. Once we register these shares and they are issued in accordance with the terms of the plans, they can be freely sold in the public market upon issuance, subject to the lock-up agreements and the restrictions imposed on our affiliates under Rule 144. For more information, see “Shares Eligible for Future Sale—Rule 144”.

You will incur immediate and substantial dilution as a result of this offering.

The initial public offering price of our common stock will be substantially higher than the net tangible book value per share of our common stock. Therefore, if you purchase common stock in this offering, you will pay a price per share that substantially exceeds our pro forma adjusted net tangible book value per share after this offering. To the extent shares subsequently are issued under options, you will incur further dilution. Based on an initial public offering price of \$, the midpoint of the range set forth on the cover page of this prospectus, you will incur immediate and substantial dilution of \$ per share, representing the difference between our pro forma net tangible book value per share, after giving effect to this offering, and the assumed initial public offering price. In addition, purchasers of common stock in this offering will have contributed approximately % of the aggregate price paid by all purchasers of our stock but will own approximately % of our common stock outstanding after this offering.

We have broad discretion in the use of net proceeds from this offering and may not use them effectively.

We currently intend to use the net proceeds from this offering for continuing clinical development of AKB-6548 in renal indications, including the preparation for and initiation of the Phase 3 development program in anemia secondary to CKD; for conducting a Phase 2 clinical trial of AKB-6548 in idiopathic anemia of aging, or IAA; for advancing AKB-6899 through Phase 1 development in oncology; and for working capital and other general corporate purposes. See the section of this prospectus entitled “Use of Proceeds.” Although we currently intend to use the net proceeds from this offering in such a manner, we will have broad discretion in the application of the net proceeds. Our failure to apply these funds effectively could affect our ability to continue to develop and commercialize our product candidates. Pending their use, we may invest the net proceeds from this offering in a manner that does not produce income or loses value.

We will incur increased costs as a result of being a public company and our management expects to devote substantial time to public company compliance programs.

As a public company, we will incur significant legal, insurance, accounting and other expenses that we did not incur as a private company. In addition, our administrative staff will be required to perform additional tasks. We intend to invest resources to comply with evolving laws, regulations and standards, and this investment will result in increased general and administrative expenses and may divert management’s time and attention from product development activities. If our efforts to comply with new laws, regulations and standards differ from the activities intended by regulatory or governing bodies due to ambiguities related to practice, regulatory authorities may initiate legal proceedings against us and our business may be harmed. In connection with this offering, we are increasing our directors’ and officers’ insurance coverage, which will increase our insurance cost. In the future, it will be more expensive for us to obtain director and officer liability insurance, and we may be required to accept reduced coverage or incur substantially higher costs to obtain coverage. These factors could also make it more difficult for us to attract and retain qualified members of our board of directors, particularly to serve on our audit committee and compensation committee, and qualified executive officers.

In addition, in order to comply with the requirements of being a public company, we may need to undertake various actions, including implementing new internal controls and procedures and hiring new accounting or internal audit staff. The Sarbanes-Oxley Act requires that we maintain effective disclosure controls and procedures and internal control over financial reporting. We are continuing to develop and refine our disclosure controls and other procedures that are designed to ensure that information required to be disclosed by us in the reports that we file with the SEC is recorded, processed, summarized and reported within the time periods specified in the SEC's rules and forms, and that information required to be disclosed in reports under the Securities Exchange Act of 1934 as amended, or the Exchange Act, is accumulated and communicated to our principal executive and financial officers. Any failure to develop or maintain effective controls could adversely affect the results of periodic management evaluations. In the event that we are not able to demonstrate compliance with the Sarbanes-Oxley Act, that our internal control over financial reporting is perceived as inadequate, or that we are unable to produce timely or accurate financial statements, investors may lose confidence in our operating results and the price of our ordinary shares could decline. In addition, if we are unable to continue to meet these requirements, we may not be able to remain listed on NASDAQ.

We are not currently required to comply with the SEC's rules that implement Section 404 of the Sarbanes-Oxley Act, and are therefore not yet required to make a formal assessment of the effectiveness of our internal control over financial reporting for that purpose. Upon becoming a public company, we will be required to comply with certain of these rules, which will require management to certify financial and other information in our quarterly and annual reports and provide an annual management report on the effectiveness of our internal control over financial reporting commencing with our second annual report. This assessment will need to include the disclosure of any material weaknesses in our internal control over financial reporting identified by our management or our independent registered public accounting firm. To achieve compliance with Section 404 within the prescribed period, we will be engaged in a process to document and evaluate our internal control over financial reporting, which is both costly and challenging. In this regard, we will need to continue to dedicate internal resources, potentially engage outside consultants and adopt a detailed work plan to assess and document the adequacy of internal control over financial reporting, continue steps to improve control processes as appropriate, validate through testing that controls are functioning as documented and implement a continuous reporting and improvement process for internal control over financial reporting. Despite our efforts, there is a risk that we will not be able to conclude, within the prescribed timeframe or at all, that our internal control over financial reporting is effective as required by Section 404. If we identify one or more material weaknesses, it could result in an adverse reaction in the financial markets due to a loss of confidence in the reliability of our financial statement.

Our independent registered public accounting firm will not be required to formally attest to the effectiveness of our internal control over financial reporting until the later of our second annual report or the first annual report required to be filed with the SEC following the date we are no longer an "emerging growth company" as defined in the JOBS Act. We cannot assure you that there will not be material weaknesses or significant deficiencies in our internal controls in the future.

Our independent registered public accounting firm has identified material weaknesses in our internal control over financial reporting which will require remediation.

Our independent registered public accounting firm issued a letter to our board of directors and management in which they identified certain matters that they consider to constitute material weaknesses in the design and operation of our internal control over financial reporting as of December 31, 2012 and September 30, 2013. A deficiency in internal control over financial reporting exists when the design or operation of a control does not allow management or employees, in the normal course of performing their assigned functions, to prevent or detect misstatements on a timely basis. A significant deficiency is a deficiency, or a combination of deficiencies, in internal control over financial reporting that is less severe than a material weakness, yet important enough to merit attention by those responsible for the oversight of the company's financial reporting. A material weakness is a deficiency, or a combination of deficiencies, in internal control over financial reporting, such that there is a reasonable possibility that a material misstatement of the company's annual or interim financial statements will not be prevented or detected on a timely basis.

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The material weaknesses identified by our auditors relate to deficiencies with our disclosure controls and procedures, including inadequate review and approval procedures with respect to financial information generated to prepare our consolidated financial statements, coupled with a lack of segregation of duties as a result of our size and overall lack of resources, including appropriate technical accounting resources, in the accounting department. This resulted in not ensuring appropriate segregation of duties between incompatible functions, and made it more difficult to ensure review of financial reporting issues.

We have taken recent steps to remediate these material weaknesses by hiring significant positions within our company which relate to internal control over financial reporting, such as our Chief Financial Officer on September 23, 2013 and our Corporate Controller on November 25, 2013, both of whom have significant experience with the internal control, compliance and financial reporting requirements of public companies. If we fail to continue to remediate these material weaknesses, we may fail to meet our future reporting obligations, our financial statements may contain material misstatements and our operational results may be harmed. Any such failure could also adversely affect the results of the periodic management evaluations and, to the extent we are no longer an emerging growth company, the annual auditor attestation reports regarding the effectiveness of our internal control over financial reporting that will be required under Section 404 of the Sarbanes-Oxley Act of 2002. Internal control deficiencies could also cause investors to lose confidence in our reported financial information.

Provisions in our charter documents and Delaware law may have anti-takeover effects that could discourage an acquisition of us by others, even if an acquisition would be beneficial to our stockholders, and may prevent attempts by our stockholders to replace or remove our current management.

Provisions in our amended and restated certificate of incorporation and amended and restated by-laws that will become effective upon the closing of this offering contain provisions that may have the effect of discouraging, delaying or preventing a change in control of us or changes in our management. These provisions could also limit the price that investors might be willing to pay in the future for shares of our common stock, thereby depressing the market price of our common stock. In addition, because our board of directors is responsible for appointing the members of our management team, these provisions may frustrate or prevent any attempts by our stockholders to replace or remove our current management by making it more difficult for stockholders to replace members of our board of directors. Among other things, these provisions:

- authorize “blank check” preferred stock, which could be issued by our board of directors without stockholder approval and may contain voting, liquidation, dividend and other rights superior to our common stock;
- create a classified board of directors whose members serve staggered three-year terms;
- specify that special meetings of our stockholders can be called only by our board of directors pursuant to a resolution adopted by a majority of the total number of directors;
- prohibit stockholder action by written consent;
- establish an advance notice procedure for stockholder approvals to be brought before an annual meeting of our stockholders, including proposed nominations of persons for election to our board of directors;
- provide that our directors may be removed only for cause;
- provide that vacancies on our board of directors may be filled only by a majority of directors then in office, even though less than a quorum;
- require a supermajority vote of the holders of our common stock or the majority vote of our board of directors to amend our amended and restated by-laws; and
- require a supermajority vote of the holders of our common stock to amend the classification of our board of directors into three classes and to amend certain other provisions of our certificate of incorporation.

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These provisions, alone or together, could delay or prevent hostile takeovers and changes in control or changes in our management.

Moreover, because we are incorporated in Delaware, we are governed by the provisions of Section 203 of the Delaware General Corporation Law, which prohibits a person who owns in excess of 15% of our outstanding voting stock from merging or combining with us for a period of three years after the date of the transaction in which the person acquired in excess of 15% of our outstanding voting stock, unless the merger or combination is approved in a prescribed manner.

Any provision of our amended and restated certificate of incorporation, our amended and restated by-laws or Delaware law that has the effect of delaying or deterring a change in control could limit the opportunity for our stockholders to receive a premium for their shares of our common stock, and could also affect the price that some investors are willing to pay for our common stock.

Our ability to use net operating losses to offset future taxable income may be subject to certain limitations.

In general, under Section 382 of the Internal Revenue Code of 1986, as amended, or the Code, a corporation that undergoes an “ownership change” is subject to limitations on its ability to utilize its pre-change net operating losses, or NOLs, to offset future taxable income. Our existing NOLs may be subject to substantial limitations arising from previous ownership changes, and if we undergo an ownership change in connection with or after this offering, our ability to utilize NOLs could be further limited by Section 382 of the Code. In addition, future changes in our stock ownership, many of which are outside of our control, could result in an ownership change under Section 382 of the Code. Our NOLs may also be impaired under state law. Accordingly, we may not be able to utilize a material portion of our NOLs. Furthermore, our ability to utilize our NOLs is conditioned upon our attaining profitability and generating U.S. federal taxable income. As described above under “—Risks related to our financial position and need for additional capital,” we have incurred significant net losses since our inception and anticipate that we will continue to incur significant losses for the foreseeable future; thus, we do not know whether or when we will generate the U.S. federal taxable income necessary to utilize our NOLs. A full valuation allowance has been provided for the entire amount of our NOLs.

Our amended and restated certificate of incorporation designates the state or federal courts located in the State of Delaware as the sole and exclusive forum for certain types of actions and proceedings that may be initiated by our stockholders, which could limit our stockholders’ ability to obtain a favorable judicial forum for disputes with us or our directors, officers or employees.

Our amended and restated certificate of incorporation that will become effective upon the closing of this offering provides that, subject to limited exceptions, the state and federal courts located in the State of Delaware will be the sole and exclusive forum for (1) any derivative action or proceeding brought on our behalf, (2) any action asserting a claim of breach of a fiduciary duty owed by any of our directors, officers or other employees to us or our stockholders, (3) any action asserting a claim against us arising pursuant to any provision of the Delaware General Corporation Law, our amended and restated certificate of incorporation or our amended and restated by-laws, or (4) any other action asserting a claim against us that is governed by the internal affairs doctrine. Any person or entity purchasing or otherwise acquiring any interest in shares of our capital stock shall be deemed to have notice of and to have consented to the provisions of our amended and restated certificate of incorporation described above. This choice of forum provision may limit a stockholder’s ability to bring a claim in a judicial forum that it finds favorable for disputes with us or our directors, officers or other employees, which may discourage such lawsuits against us and our directors, officers and employees. Alternatively, if a court were to find these provisions of our amended and restated certificate of incorporation inapplicable to, or unenforceable in respect of, one or more of the specified types of actions or proceedings, we may incur additional costs associated with resolving such matters in other jurisdictions, which could adversely affect our business and financial condition.

Because we do not anticipate paying any cash dividends on our capital stock in the foreseeable future, capital appreciation, if any, will be your sole source of gain.

You should not rely on an investment in our common stock to provide dividend income. We do not anticipate that we will pay any cash dividends to holders of our common stock in the foreseeable future. Instead, we plan to retain any earnings to maintain and expand our operations. In addition, any future debt financing arrangement may contain terms prohibiting or limiting the amount of dividends that may be declared or paid on our common stock. Accordingly, investors must rely on sales of their common stock after price appreciation, which may never occur, as the only way to realize any return on their investment. As a result, investors seeking cash dividends should not purchase our common stock.

Cautionary Note Regarding Forward-Looking Statements

This prospectus contains forward-looking statements. These statements include all matters that are not related to present facts or current conditions or that are not historical facts, including statements regarding our strategy, future operations, future financial position, future revenue, projected costs, prospects, plans, objectives of management and expected market growth. The words “anticipate,” “believe,” “could,” “estimate,” “expect,” “intend,” “may,” “plan,” “will,” “would,” or the negative of these terms or other similar expressions are intended to identify forward-looking statements, although not all forward-looking statements contain these identifying words.

The forward-looking statements in this prospectus include, among other things, statements about:

- the timing of data from our pending Phase 2b trial of AKB-6548, the timing of commencement of our Phase 3 development program of AKB-6548 and the timing of our submission of an NDA for AKB-6548;
- our plans to commercialize AKB-6548, if it is approved;
- our development plans with respect to AKB-6899;
- the timing or likelihood of regulatory filings and approvals, including any required post-marketing testing or any labeling and other restrictions;
- the implementation of our business model and strategic plans for our business, product candidates and technology;
- our commercialization, marketing and manufacturing capabilities and strategy;
- the rate and degree of market acceptance and clinical utility of our products;
- our competitive position;
- our intellectual property position;
- developments and projections relating to our competitors and our industry;
- our ability to establish collaborations or obtain additional funding;
- our expectations regarding the time during which we will be an emerging growth company under the JOBS Act;
- our expectations related to the use of proceeds from this offering; and
- our estimates regarding expenses, future revenue, capital requirements and needs for additional financing.

We may not actually achieve the plans, intentions or expectations disclosed in our forward-looking statements, and you should not place undue reliance on our forward-looking statements. Actual results or events could differ materially from the plans, intentions and expectations disclosed in the forward-looking statements we make. We have included important factors in the cautionary statements included in this prospectus, particularly in the “Risk Factors” section, that we believe could cause actual results or events to differ materially from the forward-looking statements that we make. Our forward-looking statements do not reflect the potential impact of any future acquisitions, mergers, dispositions, joint ventures or investments we may make.

You should read this prospectus and the documents that we reference in this prospectus and have filed as exhibits to the registration statement of which this prospectus is a part completely and with the understanding that our actual future results may be materially different from what we expect. The forward-looking statements in this prospectus represent our views as of the date of this prospectus. We anticipate that subsequent events and developments will cause our views to change. However, while we may elect to update these forward-looking

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statements at some point in the future, we have no current intention of doing so except to the extent required by applicable law. You should, therefore, not rely on these forward-looking statements as representing our views as of any date subsequent to the date of this prospectus.

Industry and Market Data

This prospectus includes statistical and other industry and market data that we obtained from industry publications and research, surveys and studies conducted by third parties. Industry publications and third party research, surveys and studies generally indicate that their information has been obtained from sources believed to be reliable, although they do not guarantee the accuracy or completeness of such information. While we believe these industry publications and third party research, surveys and studies are reliable, we have not independently verified such data.

Use of Proceeds

We estimate that the net proceeds from the sale of _____ shares of common stock in this offering will be approximately \$ _____ million at an assumed initial public offering price of \$ _____ per share, the midpoint of the price range set forth on the cover page of this prospectus, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us. If the underwriters exercise their option to purchase additional shares in full, we estimate that the net proceeds will be approximately \$ _____ million after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

Each \$1.00 increase or decrease in the assumed initial public offering price of \$ _____ per share, the mid-point of the price range set forth on the cover page of this prospectus, would increase or decrease our net proceeds by \$ _____ million, assuming the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same and after deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us. An increase or decrease of 1,000,000 shares in the number of shares offered by us would increase or decrease the net proceeds to us by \$ _____ million, assuming no change in the assumed initial public offering price of \$ _____ per share and after deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us.

We intend to use the net proceeds from this offering as follows:

- to continue clinical development of AKB-6548 in renal indications, including the preparation for and initiation of the Phase 3 development program in anemia secondary to CKD;
- to conduct a Phase 2 clinical trial of AKB-6548 in idiopathic anemia of aging;
- to advance our preclinical candidate, AKB-6899, through Phase 1 development in oncology; and
- for working capital and other general corporate purposes.

Our expected use of net proceeds from this offering represents our intentions based upon our present plans and business conditions, which could change in the future as our plans and business conditions evolve. The amount and timing of our actual expenditures will depend upon numerous factors, including the results of our research and development efforts, the timing and success of preclinical studies, our ongoing clinical studies or clinical studies we may commence in the future, the timing of regulatory submissions and the feedback from regulatory authorities. As a result, our management will have broad discretion over the use of the net proceeds from this offering.

Pending the use of the proceeds from this offering, we intend to invest the net proceeds in short-term, interest-bearing, investment-grade securities, certificates of deposit or government securities.

Dividend Policy

We have never declared or paid cash dividends on our common stock. We currently intend to retain all available funds and any future earnings, if any, to fund the development and expansion of our business and we do not anticipate paying any cash dividends in the foreseeable future. Any future determination to pay dividends will be made at the discretion of our board of directors after taking into account various factors, including our financial condition, operating results, current and anticipated cash needs, the requirements of any then-existing debt instruments and other factors the board of directors deems relevant.

Capitalization

The following table sets forth our cash and cash equivalents and capitalization as of September 30, 2013:

- on an actual basis;
- on a pro forma basis to give effect to the automatic conversion of all outstanding shares of our preferred stock into an aggregate of 6,790,149 shares of common stock upon the closing of this offering and the filing of our amended and restated certificate of incorporation upon the closing of this offering; and
- on a pro forma as adjusted basis to give further effect to the sale of _____ shares of our common stock offered in this offering, assuming an initial public offering price of \$ _____ per share, the midpoint of the price range set forth on the cover page of this prospectus, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

You should read this information together with our financial statements and related notes appearing elsewhere in this prospectus and the information set forth under the heading “Selected Financial Data” and “Management’s Discussion and Analysis of Financial Condition and Results of Operations”.

	As of September 30, 2013		
	Actual	Pro Forma	Pro Forma, as Adjusted
Cash and cash equivalents	\$ 24,603,252	\$ 24,603,252	\$
Series A redeemable convertible preferred stock, par value \$0.00001 per share; 734,538 shares authorized, issued and outstanding, actual; no shares authorized, issued or outstanding, pro forma and pro forma as adjusted	\$ 38,785,314	\$ —	\$
Series B redeemable convertible preferred stock, par value \$0.00001 per share; 1,287,525 shares authorized, shares issued and outstanding, actual; no shares authorized, issued or outstanding, pro forma and pro forma as adjusted	20,720,557	—	
Series C redeemable convertible preferred stock, par value \$0.00001 per share; 3,428,572 shares authorized, 3,302,885 shares issued and outstanding, actual; no shares authorized, issued or outstanding, pro forma and pro forma as adjusted	95,296,787	—	
Common stock, par value \$0.00001 per share; 8,400,000 shares authorized, 424,044 shares issued and outstanding, actual; _____ shares authorized, 7,214,193 shares issued or outstanding, pro forma and _____ shares authorized, _____ shares issued and outstanding, pro forma as adjusted	3	71	
Additional paid-in capital	—	93,263,055	
Accumulated deficit	(119,230,682)	(57,691,147)	
Total stockholders’ (deficit) equity	(119,230,679)	35,571,979	
Total capitalization	\$ (94,627,427)	\$ 60,175,231	\$

A \$1.00 increase or decrease in the assumed initial public offering price of \$ _____ per share, the mid-point of the price range set forth on the cover page of this prospectus, would increase or decrease each of cash and cash equivalents, additional paid-in capital, total stockholders’ deficit (equity) and total capitalization on a pro forma as adjusted basis by approximately \$ _____ million, assuming that the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same and after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us. An increase or decrease of 1,000,000 shares in the

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number of shares offered by us would increase or decrease each of cash and cash equivalents, additional paid-in capital, total stockholders' deficit (equity) and total capitalization on a pro forma as adjusted basis by approximately \$ million, assuming no change in the assumed initial public offering price of \$ per share and after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

The actual, pro forma and pro forma as adjusted information set forth in the table above excludes the following:

- 702,625 shares of our common stock issuable upon the exercise of stock options outstanding as of September 30, 2013 at a weighted-average exercise price of \$1.08 per share;
- 64,408 shares of common stock reserved for issuance pursuant to future equity awards under our Amended and Restated 2008 Equity Incentive Plan; and
- shares of common stock reserved for future issuance under our 2014 Equity Incentive Plan.

Dilution

If you invest in our common stock in this offering, your ownership interest will be diluted to the extent of the difference between the initial public offering price per share of our common stock in this offering and the pro forma as adjusted net tangible book value per share of our common stock after this offering.

We had a historical net tangible book value of \$(119.2) million, or \$(281.0) per share of common stock, as of September 30, 2013. Our historical net tangible book value represents total tangible assets less total liabilities and redeemable convertible preferred stock. Our historical net tangible book value per share is our historical net tangible book value, divided by the number of shares of our common stock outstanding as of September 30, 2013.

The pro forma net tangible book value of our common stock as of September 30, 2013 was \$35.6 million, or \$5.0 per share of our common stock. Pro forma net tangible book value represents the amount of our total tangible assets less our total liabilities, divided by the pro forma number of shares of our common stock outstanding after giving effect to the automatic conversion of our outstanding preferred stock into an aggregate of 6,790,149 shares of common stock upon the closing of this offering.

After giving further effect to the sale of _____ shares of common stock in this offering, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us, at an assumed initial public offering price of \$ _____ per share, the midpoint of the price range set forth on the cover page of this prospectus, our pro forma as adjusted net tangible book value as of September 30, 2013 would have been approximately \$ _____ million, or approximately \$ _____ per share of common stock. This represents an immediate increase in pro forma as adjusted net tangible book value of \$ _____ per share to our existing stockholders and an immediate dilution of \$ _____ per share to investors participating in this offering. Dilution per share to new investors is determined by subtracting pro forma net tangible book value per share after this offering from the initial public offering price per share paid by new investors. The following table illustrates this dilution on a per share basis:

Assumed initial public offering price per share		\$
Historical net tangible book value per share as of September 30, 2013	\$ (281.0)	
Pro forma net tangible book value per share as of September 30, 2013	5.0	
Increase in net tangible book value per share attributable to new investors		
Pro forma as adjusted net tangible book value per share after this offering		
Dilution per share to new investors		\$

Each \$1.00 increase or decrease in the assumed initial public offering price of \$ _____ per share, the mid-point of the price range set forth on the cover page of this prospectus, would increase or decrease our pro forma as adjusted net tangible book value by approximately \$ _____ million, the pro forma as adjusted net tangible book value per share by approximately \$ _____ and the dilution to investors purchasing shares in this offering by approximately \$ _____ per share, assuming the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same and after deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us. An increase or decrease of 1,000,000 shares in the number of shares offered by us would increase or decrease our pro forma as adjusted net tangible book value by approximately \$ _____ million, the pro forma as adjusted net tangible book value per share by approximately \$ _____ and the dilution to investors purchasing shares in this offering by approximately \$ _____ per share, assuming no change in the assumed initial public offering price of \$ _____ per share and after deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us. If the underwriters exercise their option to purchase additional shares, you will experience further dilution.

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The following table summarizes, on a pro forma as adjusted basis as of September 30, 2013, the number of shares of common stock purchased from us, the total consideration and the average price per share paid by existing stockholders (giving effect to the conversion of all outstanding shares of our preferred stock into 6,790,149 shares of common stock upon the completion of this offering) and by investors participating in this offering, after deducting estimated underwriting discounts and commissions and estimated offering expenses, at an assumed initial public offering price of \$ _____ per share, the midpoint of the price range set forth on the cover page of this prospectus. As the table illustrates, new investors purchasing shares in this offering will pay an average price per share substantially higher than our existing stockholders paid.

	Shares purchased		Total consideration		Average price per share
	Number	Percent	Amount	Percent	
Existing stockholders		%	\$	%	\$
New investors		%		%	\$
Total		100%		\$ 100%	

A \$1.00 increase or decrease in the assumed initial public offering price of \$ _____ per share, the mid-point of the price range set forth on the cover page of this prospectus, would increase or decrease the total consideration paid by new investors by \$ _____ million and increase or decrease the percentage of total consideration paid by new investors by approximately _____%, assuming that the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same. An increase or decrease of 1,000,000 shares in the number of shares offered by us would increase or decrease the total consideration paid by new investors by \$ _____ million and increase or decrease the percentage of total consideration paid by new investors by approximately _____%, assuming no change in the assumed initial public offering price of \$ _____ per share.

If the underwriters exercise their option to purchase additional shares in full, pro forma as adjusted net tangible book value as of September 30, 2013 will increase to \$ _____ million, or \$ _____ per share, representing an increase to existing stockholders of \$ _____ per share, and there will be an immediate dilution of an additional \$ _____ per share to new investors.

The number of shares of common stock to be outstanding after this offering is based on the number of shares outstanding as of September 30, 2013 and excludes the following:

- 702,625 shares of common stock issuable upon the exercise of outstanding stock options having a weighted-average exercise price of \$1.08 per share;
- 64,408 shares of common stock reserved for issuance pursuant to future equity awards under our Amended and Restated 2008 Equity Incentive Plan; and
- _____ shares of common stock reserved for future issuance under our 2014 Equity Incentive Plan.

New investors will experience further dilution if any of our outstanding options are exercised, new options are issued and exercised under our equity incentive plans or we issue additional shares of common stock, other equity securities or convertible debt securities in the future.

Selected Financial Data

The selected statements of operations data for the years ended December 31, 2011 and 2012, the nine months ended September 30, 2013, the period from February 27, 2007 (inception) to September 30, 2013 and the balance sheet data as of December 31, 2011 and 2012 and September 30, 2013 are derived from our audited financial statements included elsewhere in this prospectus. The selected statements of operations data for the nine months ended September 30, 2012 have been derived from our unaudited financial statements included elsewhere in this prospectus. These unaudited financial statements have been prepared on a basis consistent with our audited financial statements and, in our opinion, contain all adjustments, consisting only of normal and recurring adjustments, necessary for a fair presentation of such financial data. You should read this data together with our financial statements and related notes included elsewhere in this prospectus and the information under the captions “Selected Financial Data” and “Management’s Discussion and Analysis of Financial Condition and Results of Operations.” Our historical results for any prior period are not necessarily indicative of results to be expected in any future period, and our interim period results are not necessarily indicative of results to be expected for a full year or any other interim period.

	<u>Year Ended December 31,</u>		<u>Nine Months Ended September 30,</u>		<u>Period from</u>
	<u>2011⁽¹⁾</u>	<u>2012</u>	<u>2012</u>	<u>2013</u>	<u>February 27, 2007</u>
			<u>(unaudited)</u>		<u>(Date of Inception)</u>
					<u>to September 30,</u>
					<u>2013</u>
	<u>(dollars in thousands, except per share data)</u>				
Consolidated statements of operations data:					
Revenue	\$ —	—	\$ —	\$ —	\$ —
Expenses:					
Research and development	12,976	5,632	5,065	7,591	48,557
General and administrative	2,567	2,891	1,594	2,141	12,258
Total expenses	15,543	8,523	6,659	9,732	60,815
Loss from operations	(15,543)	(8,523)	(6,659)	(9,732)	(60,815)
Other income, net	246	327	866	2,513	3,721
Net loss	<u>\$ (15,297)</u>	<u>\$ (8,196)</u>	<u>\$ (5,793)</u>	<u>\$ (7,219)</u>	<u>\$ (57,094)</u>
Net loss per share applicable to common stockholders—basic and diluted ⁽²⁾	\$ (109.36)	\$ (48.68)	\$ (35.80)	\$ (206.32)	\$ (827.64)
Weighted-average number of common shares used in net loss per share applicable to common stockholders—basic and diluted	167,039	236,633	230,748	291,206	146,688
Pro forma net loss per share applicable to common stockholders—basic and diluted (unaudited) ⁽²⁾		<u>\$ (2.26)</u>		<u>\$ (1.35)</u>	
Pro forma weighted-average number of common shares used in net loss per share applicable to common stockholders—basic and diluted (unaudited)		<u>3,622,838</u>		<u>5,362,450</u>	

(1) In December 2011, we spun out our programs focused on the treatment of diabetic eye disease and inflammatory bowel disease into Aerpio, which has since operated as a stand-alone company. See “Management’s Discussion and Analysis of Financial Condition and Results of Operations” appearing elsewhere in this prospectus for further information.

(2) See Note 2 within the notes to our financial statements appearing elsewhere in this prospectus for a description of the method used to calculate basic and diluted net loss per share of common stock. Pro forma basic and diluted net loss per share of common stock is calculated by dividing net loss attributable to common stockholders, excluding the impact of gains (losses) on the extinguishment of preferred stock and accretion of preferred stock by the pro forma weighted-average number of common shares outstanding, which assumes the automatic conversion of all outstanding shares of our preferred stock into an aggregate of 6,790,149 shares of common stock upon the closing of this offering.

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	December 31,		September 30,
	2011	2012	2013
	(in thousands)		
Balance Sheet Data:			
Cash and cash equivalents	\$ 5,011	\$ 1,641	\$ 24,603
Working capital (deficit)	5,394	(2,679)	35,566
Total assets	7,211	2,244	38,470
Redeemable convertible preferred stock	53,586	56,909	154,803
Accumulated deficit	(48,192)	(59,588)	(119,231)
Total stockholders' deficit	(48,192)	(59,588)	(119,231)

**Management's Discussion and Analysis of
Financial Condition and Results of Operations**

You should read the following discussion and analysis of our financial condition and results of operations together with our consolidated financial statements and related notes appearing in this prospectus. Some of the information contained in this discussion and analysis or set forth elsewhere in this prospectus, including information with respect to our plans and strategy for our business, includes forward-looking statements that involve risks and uncertainties. As a result of many factors, including those factors set forth in the "Risk Factors" section of this prospectus, our actual results could differ materially from the results described in or implied by the forward-looking statements contained in the following discussion and analysis.

Overview

We are a biopharmaceutical company focused on the development of novel proprietary therapeutics based on hypoxia inducible factor, or HIF, biology and the commercialization of these products for patients with kidney disease. HIF is the primary regulator of the production of red blood cells, or RBCs, in the body and a potentially novel mechanism of treating anemia. Our lead product candidate, AKB-6548, is being developed as a once-daily oral therapy that has successfully completed a Phase 2a proof of concept study demonstrating that AKB-6548 safely and predictably raised hemoglobin levels in patients with anemia secondary to chronic kidney disease, or CKD, not requiring dialysis.

We are conducting a Phase 2b trial for AKB-6548 in patients with anemia secondary to CKD who are not dependent on dialysis and expect data to be available in the fourth quarter of 2014. We have also initiated a development program for patients dependent on dialysis. If the results of our Phase 2b trial are positive, we would expect to initiate Phase 3 trials for anemia secondary to CKD in 2015, and would anticipate submitting an NDA for AKB-6548 in the United States by 2018 if the Phase 3 data are favorable.

We own worldwide rights to our HIF-based product candidates, including AKB-6548. If approved by regulatory authorities, we plan to commercialize AKB-6548 in the United States ourselves and intend to seek one or more collaborators to commercialize the product candidate in additional markets.

Since our inception in 2007, we have devoted substantially all of our resources to our development efforts relating to AKB-6548, including preparing for and conducting clinical studies of AKB-6548, providing general and administrative support for these operations and protecting our intellectual property. We do not have any products approved for sale and have not generated any revenue from product sales. We have funded our operations primarily through the private placement of preferred stock, common stock and convertible notes.

In December 2011, we spun out our programs focused on the treatment of diabetic eye disease and inflammatory bowel disease into Aerpio which has since operated as a stand-alone company. We have administrative services agreements with Aerpio under which we obtain from and provide to Aerpio certain services including consulting services and use of premises.

We have never been profitable and have incurred net losses in each year since inception. Our net losses were \$15.3 million and \$8.2 million for the years ended December 31, 2011 and 2012, and \$5.8 million and \$7.2 million for the nine months ended September 30, 2012 and 2013, respectively. Substantially all our net losses resulted from costs incurred in connection with our research and development programs and from general and administrative costs associated with our operations.

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We expect to continue to incur significant expenses and increasing operating losses for at least the next several years. We expect our expenses will increase substantially in connection with our ongoing activities, as we:

- continue our Phase 2b trial and prepare for a future Phase 3 development program of AKB-6548 for the treatment of anemia secondary to CKD;
- seek regulatory approvals for our product candidates that successfully complete clinical trials;
- have our product candidates manufactured for clinical trials and for commercial sale;
- establish a sales, marketing and distribution infrastructure to commercialize any products for which we may obtain marketing approval;
- initiate additional preclinical, clinical or other studies for AKB-6548, AKB-6899 and other product candidates that we may develop or acquire;
- seek to discover and develop additional product candidates;
- acquire or in-license other commercial products, product candidates and technologies;
- make royalty milestone or other payments under any future in-license agreements;
- maintain, protect and expand our intellectual property portfolio;
- attract and retain skilled personnel; and
- create additional infrastructure to support our operations as a public company.

We do not expect to generate revenue from product sales unless and until we successfully complete development and obtain regulatory approval for one or more of our product candidates, which we expect will take a number of years and is subject to significant uncertainty. We have no manufacturing facilities and all of our manufacturing activities are contracted out to third parties. Additionally, we currently utilize third-party CROs to carry out our clinical development activities and we do not yet have a sales organization. If we obtain regulatory approval for any of our product candidates, we expect to incur significant commercialization expenses related to product sales, marketing, manufacturing and distribution. Accordingly, we will seek to fund our operations through public or private equity or debt financings or other sources. However, we may be unable to raise additional funds or enter into such other arrangements when needed on favorable terms or at all. Our failure to raise capital or enter into such other arrangements as and when needed would have a negative impact on our financial condition and our ability to develop our products.

Financial Operations Overview

Revenue

To date, we have not generated any revenues from the sales of products or other means.

Our ability to generate product revenue and become profitable depends upon our ability to successfully develop and commercialize products. We expect to incur losses for the foreseeable future, and we expect these losses to increase as we continue our development of, and seek regulatory approvals for, our product candidates and begin to commercialize any approved products. Because of the numerous risks and uncertainties associated with product development, we are unable to predict the timing or amount of increased expenses or when or if we will be able to achieve or maintain profitability. Even if we are able to generate revenues from the sale of our products, we may not become profitable. If we fail to become profitable or are unable to sustain profitability on a continuing basis, then we may be unable to continue our operations at planned levels and be forced to reduce our operations.

Research and Development Expenses

Research and development expenses consist primarily of costs incurred for the development of our product candidates, which include:

- employee-related expenses, including salaries, benefits, travel and stock-based compensation expense;
- expenses incurred under agreements with the CROs and investigative sites that will conduct our clinical studies;
- the cost of acquiring, developing and manufacturing clinical study materials;
- facilities, depreciation and other expenses, which include direct and allocated expenses for rent and maintenance of facilities, insurance and other supplies; and
- costs associated with preclinical and clinical activities.

Research and development costs are expensed as incurred. Costs for certain development activities are recognized based on an evaluation of the progress to completion of specific tasks using information and data provided to us by our vendors and our clinical sites.

We cannot determine with certainty the duration and completion costs of the current or future clinical studies of our product candidates or if, when, or to what extent we will generate revenues from the commercialization and sale of any of our product candidates that obtain regulatory approval. We may never succeed in achieving regulatory approval for any of our product candidates. The duration, costs and timing of clinical studies and development of our product candidates will depend on a variety of factors, including:

- the rate of progress, results and cost of completing our Phase 2b clinical trial of AKB-6548;
- assuming AKB-6548 advances to Phase 3, the scope, size, rate of progress, results and costs of initiating and completing our Phase 3 development program of AKB-6548;
- the scope, progress, results and costs of preclinical development, laboratory testing and clinical trials for AKB-6899 and any other product candidates that we may develop or acquire;
- the cost of having our product candidates manufactured for clinical trials;
- difficulties or delays in enrolling patients in our clinical trials;
- unanticipated changes to laws or regulations applicable to our clinical trials; and
- the timing of, and the costs involved in, obtaining regulatory approval for AKB-6548 and any other product candidates, if clinical trials are successful.

A change in the outcome of any of these variables with respect to the development of a product candidate could mean a significant change in the costs and timing associated with the development of that product candidate. For example, if the FDA, EMA or another regulatory authority were to require us to conduct clinical studies beyond those that we currently anticipate will be required for the completion of clinical development of a product candidate, or if we experience significant delays in enrollment in any of our clinical studies, we could be required to expend significant additional financial resources and time on the completion of clinical development.

From inception through September 2013, we have incurred \$48.6 million in research and development expenses. We plan to increase our research and development expenses for the foreseeable future as we continue the development of our AKB-6548 product candidate. Our current and planned research and development activities include the following:

- We plan to complete a Phase 2b clinical study during 2014 to examine the safety and efficacy of AKB-6548 in patients with anemia secondary to CKD.
- We plan to initiate a Phase 3 development program for AKB-6548 in 2015 for anemia secondary to CKD.

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- We have begun an efficacy study for AKB-6548 in dialysis patients with anemia, the second indication we will pursue.
- We intend to conduct a Phase 2 clinical trial of AKB-6548 in IAA.
- We intend to file an Investigational New Drug, or IND, and begin Phase 1 trials for AKB-6899 and explore its use in oncology and ophthalmology.

Our direct research and development expenses consist principally of external costs, such as startup fees paid to investigators, consultants, central laboratories and CROs in connection with our clinical studies, and costs related to acquiring and manufacturing clinical study materials.

We currently have one program to which our research and development costs are attributable. Historically, we have not accumulated and tracked our research and development costs or our personnel and personnel-related costs on a program-by-program basis. Our employee and infrastructure resources, and many of our costs were directed to broadly applicable research endeavors. As a result, we cannot state the historical costs incurred for each of our programs on a program-by-program basis.

General and Administrative Expenses

We obtain from and provide to Aerpio services under the terms of administrative services agreements between the two companies. See “Certain Relationships and Related Party Transactions.” General and administrative expenses consist primarily of salaries and related costs for personnel, including stock-based compensation and travel expenses for our employees in executive, operational, finance and human resource functions. Other general and administrative expenses include facility-related costs and professional fees for directors, accounting and legal services, recruiting fees and expenses associated with obtaining and maintaining patents.

We anticipate that our general and administrative expenses will increase in the future as we increase our headcount to support our continued research and development and potential commercialization of our product candidates. We also anticipate increased expenses related to audit, legal, regulatory and tax-related services associated with maintaining compliance with exchange listing and Securities and Exchange Commission requirements, director and officer insurance premiums, and investor relations costs associated with being a public company. Additionally, if and when we believe a regulatory approval of the first product candidate appears likely, we anticipate an increase in payroll and related expenses as a result of our preparation for commercial operations, especially as it relates to the sales and marketing of our product candidates.

Critical Accounting Policies and Significant Judgments and Estimates

Our management’s discussion and analysis of our financial condition and results of operations are based on our financial statements, which have been prepared in accordance with U.S. generally accepted accounting principles. The preparation of these financial statements requires us to make estimates and judgments that affect the reported amounts of assets, liabilities, and expenses and the disclosure of contingent assets and liabilities in our financial statements. On an ongoing basis, we evaluate our estimates and judgments, including those related to accrued expenses and stock-based compensation. We base our estimates on historical experience, known trends and events, and various other factors that are believed to be reasonable under the circumstances, the results of which form the basis for making judgments about the carrying values of assets and liabilities that are not readily apparent from other sources. Actual results may differ from these estimates under different assumptions or conditions.

While our significant accounting policies are described in more detail in the notes to our financial statements appearing elsewhere in this prospectus, we believe the following accounting policies to be most critical to the judgments and estimates used in the preparation of our financial statements.

Accrued Research and Development Expenses

As part of the process of preparing our financial statements, we are required to estimate our accrued expenses. This process involves reviewing open contracts and purchase orders, communicating with our personnel to identify services that have been performed on our behalf and estimating the level of service performed and the associated cost incurred for the service when we have not yet been invoiced or otherwise notified of the actual cost. The majority of our service providers invoice us monthly in arrears for services performed. We make estimates of our accrued expenses as of each balance sheet date in our financial statements based on facts and circumstances known to us at that time. We confirm the accuracy of our estimates with the service providers and make adjustments if necessary. Examples of estimated accrued research and development expenses include expenses for:

- CROs in connection with clinical studies;
- investigative sites in connection with clinical studies;
- vendors in connection with preclinical development activities; and
- vendors related to product manufacturing, development and distribution of clinical supplies.

We base our expenses related to clinical studies on our estimates of the services received and efforts expended pursuant to contracts with multiple CROs that conduct and manage clinical studies on our behalf. The financial terms of these agreements are subject to negotiation, vary from contract to contract and may result in uneven payment flows. The scope of services under these contracts can be modified and the agreements can be cancelled by either party upon written notice. There may be instances in which payments made to our vendors will exceed the level of services provided and result in a prepayment of the clinical expense. Payments under some of these contracts depend on factors such as the successful enrollment of subjects and the completion of clinical study milestones. In accruing service fees, we estimate the time period over which services will be performed and the level of effort to be expended in each period. If the actual timing of the performance of services or the level of effort varies from our estimate, we adjust the accrual or prepaid accordingly.

Although we do not expect our estimates to be materially different from amounts actually incurred, if our estimates of the status and timing of services performed differ from the actual status and timing of services performed we may report amounts that are too high or too low in any particular period. To date, there have been no material differences between our estimates and the amount actually incurred.

Stock-Based Compensation

Stock-Based Awards

We issue stock-based awards to employees and non-employees, generally in the form of stock options and restricted stock. We account for our stock-based awards to employees in accordance with FASB ASC Topic 718, *Compensation—Stock Compensation*, or ASC 718. ASC 718 requires all stock-based payments to employees, including grants of employee stock options and modifications to existing stock options, to be recognized in the consolidated statements of operations and comprehensive loss based on their fair values. We account for stock-based awards to non-employees in accordance with FASB ASC Topic 505-50, *Equity-Based Payments to Non-Employees*, which requires the fair value of the award to be re-measured at fair value as the award vests. We recognize the compensation cost of our stock-based awards, which are currently all subject only to service-based vesting conditions, to employees on a straight-line basis over the vesting period of the award and using an accelerated attribution model for awards to non-employees. Described below is the methodology we have utilized in measuring stock-based compensation expense. Following the consummation of this offering, stock option and restricted stock values will be determined based on the quoted market price of our common stock.

We estimate the fair value of our stock-based awards to employees and non-employees using the Black-Scholes option pricing model, which requires the input of highly subjective assumptions, including (a) the

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expected volatility of our stock, (b) the expected term of the award, (c) the risk-free interest rate, and (d) expected dividends. Due to the lack of a public market for the trading of our common stock and a lack of company specific historical and implied volatility data, we have based our estimate of expected volatility on the historical volatility of a group of similar companies that are publicly traded. For these analyses, we have selected companies with comparable characteristics to ours including enterprise value, risk profiles, position within the industry, and with historical share price information sufficient to meet the expected life of the stock-based awards. We compute the historical volatility data using the daily closing prices for the selected companies' shares during the equivalent period of the calculated expected term of our stock-based awards. We will continue to apply this process until a sufficient amount of historical information regarding the volatility of our own stock price becomes available. We have estimated the expected life of our employee stock options using the "simplified" method, whereby, the expected life equals the average of the vesting term and the original contractual term of the option. The risk-free interest rates for periods within the expected life of the option are based on the U.S. Treasury yield curve in effect during the period the options were granted.

We are also required to estimate forfeitures at the time of grant, and revise those estimates in subsequent periods if actual forfeitures differ from its estimates. We use historical data to estimate pre-vesting option forfeitures and record stock-based compensation expense only for those awards that are expected to vest. To the extent that actual forfeitures differ from our estimates, the difference is recorded as a cumulative adjustment in the period the estimates were revised. Stock-based compensation expense recognized in the financial statements is based on awards that are ultimately expected to vest.

In June 2011, certain of our employees purchased shares of our restricted stock in exchange for promissory notes. Although these notes are 50% recourse to the employees, we have accounted for the promissory notes as nonrecourse in their entirety since the promissory notes are not aligned with a corresponding percentage of the underlying shares. Accordingly, we have accounted for the combination of the promissory note and restricted stock as a grant of an option, as the substance is similar to the grant of an option. The exercise price of this stock option is the principal and interest due on the promissory note. The fair value of the stock option is recognized over the requisite service period (not the term of the promissory note) through a charge to compensation cost. The maturity date of the promissory notes reflects the legal term of the stock option for purposes of valuing the award. These awards are referred to as promissory note options in the tables below.

We have computed the fair value of employee and non-employee stock options at date of grant using the following weighted-average assumptions:

	Year Ended		Nine Months
	December 31,	December 31,	Ended
	2011	2012	September 30,
			2013
Expected volatility	74.00%	73.00%	79.00%
Expected term (in years)—employee options	6.25	6.25	6.25
Expected term (in years)—non employee options	10	10	10
Expected term (in years)—promissory note options	5	5	5
Risk-free interest rate	1.09%	0.95%	1.71%
Expected dividend yield	0.00%	0.00%	0.00%
Expected dividend yield—promissory note options	6.00%	6.00%	3.00%

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The following table presents the grant dates, number of underlying shares and related exercise prices or purchase prices of stock options granted and restricted stock awards issued between January 1, 2011 and September 30, 2013, along with the fair value per share utilized to calculate stock-based compensation expense:

Year of grant	Type of award	Number of shares	Exercise price (options) or purchase price (promissory note options) per share	Retrospective common stock fair value per share as of grant date
2011	Option	24,330	1.50	1.90
2011	Restricted Stock Award	145,404 ⁽¹⁾	1.50	1.90
2012	Option	40,451	1.50	1.00
2012	Restricted Stock Award	30,047	N/A	1.00
2013	Option	427,519	0.82	6.60
2013	Restricted Stock Award	30,763	N/A	1.56

(1) Represents promissory note options, as described above.

Stock-based compensation totaled approximately \$0.1 million for the year ended December 31, 2012 and approximately \$0.4 million for the nine months ended September 30, 2013. As of September 30, 2013, we had approximately \$2.8 million of total unrecognized compensation expense, net of related forfeiture estimates, which is expected to be recognized over a weighted-average remaining vesting period of approximately 2.4 years. We expect the impact of our stock-based compensation expense for stock options and restricted stock granted to employees and non-employees to grow in future periods due to the potential increases in the value of our common stock and headcount.

Fair Value of Stock Options

We have historically granted stock options at exercise prices not less than the fair value of our common stock as of the actual date of grant. As there has been no public market for our common stock to date, the estimated fair value of our common stock has been determined contemporaneously by our board of directors based on valuation estimates provided by management and prepared in accordance with the framework of the American Institute of Certified Public Accountants' Technical Practice Aid, *Valuation of Privately-Held-Company Equity Securities Issued as Compensation*, or AICPA Practice Aid, as well as independent third-party valuations. In conducting the contemporaneous valuations, our management and the valuation firm considered a number of objective and subjective factors, including external market conditions affecting the biotechnology industry sector and the prices at which we sold shares of preferred stock, the superior rights and preferences of securities senior to our common stock at the time of each grant and the likelihood of achieving a liquidity event such as an initial public offering, or IPO.

On November 13, 2013, our board of directors authorized management to pursue this IPO. As a result, in connection with the preparation of our financial statements for the years ended December 31, 2011 and 2012, the nine months ended September 30, 2013 and the period from February 27, 2007 (date of inception) to September 30, 2013, we reexamined the valuation of our common stock. In connection with that reexamination, we prepared retrospective appraisals of the fair value of our common stock for financial reporting purposes as of December 31, 2011, December 31, 2012 and March 31, 2013. We believe that the valuation methodologies used in the retrospective valuations are reasonable and consistent with the AICPA Practice Aid. The fair values of our common stock shown in the table above reflect these retrospective valuations.

December 31, 2011 Retrospective Valuation

For the retrospective valuation at December 31, 2011, we used the back-solve method of the option-pricing method, or OPM, which derives the implied equity value for one type of equity security from a contemporaneous transaction involving another type of equity security. We applied the OPM back-solve method to solve for the

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equity value and corresponding value of common stock based on our Series B preferred stock financing, which was completed on December 23, 2011. We applied a discount for lack of marketability to the value indicated for our common stock. Our estimate of the appropriate discount for lack of marketability took into consideration put option methodologies consistent with the AICPA Practice Aid.

The OPM treats common stock and preferred stock as call options on the total equity value of a company, with exercise prices based on the value thresholds at which the allocation among the various holders of a company's securities changes. Under this method, the common stock has value only if the funds available for distribution to stockholders exceed the value of the liquidation preference at the time of a liquidity event, such as a strategic sale, merger or IPO. The common stock is modeled as a call option on the underlying equity value at a predetermined exercise price. In the model, the exercise price is based on a comparison with the total equity value rather than, as in the case of a regular call option, a comparison with a per share stock price. Thus, common stock is considered to be a call option with a claim on the enterprise at an exercise price equal to the remaining value immediately after the preferred stock is liquidated. The OPM uses the Black-Scholes option pricing model to price the call options. This model defines the securities' fair values as functions of the fair value of a company's equity as of an appraisal date and uses assumptions such as the anticipated timing of a potential liquidity event and the estimated volatility of the equity securities.

The following table summarizes the significant assumptions used in the OPM to determine the fair value of our common stock as of December 31, 2011:

December 31, 2011 retrospective valuation

Key assumptions

Years to liquidity	2.96
Annual volatility	71%
Risk-free interest rate	0.36%
Discount for lack of marketability (DLOM)	29%

Based on these assumptions, we estimated the fair value of our common stock to be \$1.90 as of December 31, 2011.

December 31, 2012 Retrospective Valuation

For the retrospective valuation at December 31, 2012, we used the guideline public company method under the market approach to value our equity. We estimated our equity value based on a multiple of paid-in capital as indicated by a group of guideline public companies. In our selection of guideline public companies, we took into account each candidate's stage of clinical development and the targeted indications for drugs in development. We used the OPM to allocate the value of our equity among our preferred and common stock. We applied a discount for lack of marketability to the value indicated for our common stock. Our estimate of the appropriate discount for lack of marketability took into consideration put option methodologies consistent with the AICPA Practice Aid.

The following table summarizes the significant assumptions used in the OPM to determine the fair value of our common stock as of December 31, 2012:

December 31, 2012 retrospective valuation

Key assumptions

Years to liquidity	1.96
Annual volatility	58%
Risk-free interest rate	0.25%
Discount for lack of marketability (DLOM)	19%

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Based on these assumptions, we estimated the fair value of our common stock to be \$1.00 as of December 31, 2012.

The estimated per share fair value of our common stock calculated in our December 31, 2012 retrospective valuation of \$1.00 per share decreased from the December 31, 2011 retrospective valuation of \$1.90 per share primarily due to the following factors:

- We did not establish an industry partnership, which would have provided a non-dilutive source of capital.
- Our year-end cash and short-term investments balance decreased from \$6.4 million to \$1.6 million.
- We raised capital by issuing Series X preferred shares, which had a liquidation preference equal to twice the amount invested.
- As a percentage of the common-equivalent shares outstanding, including our convertible preferred shares, our common shares and restricted shares decreased from 18% to 9%.

March 31, 2013 Retrospective Valuation

For the retrospective valuation at March 31, 2013, we used the hybrid method to value our common stock. The hybrid method is a hybrid between the probability-weighted expected returns method and the OPM. We considered an IPO scenario, in which our preferred shares convert to common stock, and a second scenario, in which equity value is allocated using the OPM. We used the guideline public company method under the market approach to value our equity. We estimated our equity value based on a multiple of paid-in capital as indicated by a group of guideline public companies. In addition, we estimated the value of our equity securities in association with an IPO. We considered the enterprise values of guideline public companies and the pricing of IPOs completed by clinical stage drug development companies in the year preceding our appraisal date. For each of the IPO companies, we considered the increase, or step-up, in per share value from the preferred financing preceding the IPO to the common stock value in the IPO. We also considered the equity value of each IPO company, not including the proceeds of the IPO.

The following table summarizes the significant assumptions used in the hybrid method to determine the fair value of our common stock as of March 31, 2013:

<u>March 31, 2013 retrospective valuation</u>	<u>IPO</u>	<u>OPM</u>
Key assumptions		
Probability weighting	5%	95%
Years to liquidity	1.71	1.71
Weighted-average cost of capital	20%	
Annual volatility		59%
Risk-free interest rate		0.22%
Discount for lack of marketability (DLOM)	18%	18%
Estimated per share present value of non-marketable common stock (before probability weighting)	\$9.75	\$ 1.00

Based on these assumptions, we estimated the fair value of our common stock to be \$1.56 as of March 31, 2013.

The estimated per share fair value of our common stock calculated in our March 31, 2013 retrospective valuation of \$1.56 per share increased from the December 31, 2012 valuation of \$1.00 per share primarily due to the following factors:

- Litigation to protect our intellectual property rights in Europe was decided in our favor.

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- We made progress toward completing a Series C preferred stock financing, enhancing our prospects for securing the capital needed for clinical trials prior to an IPO.
- We raised additional capital by issuing Series X preferred shares, and the terms of the Series X shares were revised to the benefit of the common stockholders.

September 30, 2013 Valuation

For the contemporaneous valuation at September 30, 2013, we used the probability-weighted expected returns method (PWERM). Under PWERM, the values of the various equity securities are estimated based upon an analysis of future values for the enterprise, assuming various future outcomes. Share value is based upon the probability-weighted present value of expected future investment returns, considering each of the possible future outcomes available to the enterprise, as well as the rights of each share class.

We considered three scenarios: an IPO, a sale of the Company and a liquidation of the Company's assets. For the IPO scenario, we considered the enterprise values of guideline public companies and the pricing of IPOs completed by clinical stage drug development companies in the year preceding our appraisal date. For each of the IPO companies, we considered the increase, or step-up, in per share value from the preferred financing preceding the IPO to the common stock value in the IPO. We also considered the equity value of each IPO company, not including the proceeds of the IPO. For the sale scenario, we assumed a market participant acquisition premium (MPAP) to the IPO value. Our estimate of the MPAP took into account the premiums observed in eight acquisitions completed in 2013 of publicly-traded clinical stage drug development companies. For the liquidation scenario, we considered a value equal to the amount invested in our Series C preferred stock.

The following table summarizes the significant assumptions used in the PWERM to determine the fair value of our common stock as of September 30, 2013:

	<u>IPO</u>	<u>Sale</u>	<u>Liquidation</u>
Probability	33%	25%	42%
Years to Liquidity	0.48	1.25	1.75
Weighted-average cost of capital	20%	20%	20%
Discount for lack of marketability (DLOM)	12%	20%	NA

Based on these assumptions, we estimated the fair value of our common stock to be \$6.60 as of September 30, 2013.

The estimated per share fair value of our common stock calculated in our September 30, 2013 valuation of \$6.60 per share increased from the March 31, 2013 valuation of \$1.56 per share primarily due to the following factors:

- We completed our Series C preferred stock financing.
- We completed our Phase 2a dose-ranging study of AKB-6548 in patients with stage 3 and 4 CKD.
- Capital market conditions for biotechnology companies improved, as evidenced by an increase in the number of IPOs and their IPO valuations.
- We estimated that the probability of the Company completing an IPO increased.

There are significant judgments and estimates inherent in the determination of these valuations. These judgments and estimates include assumptions regarding our future performance, including the successful enrollment and completion of our clinical studies as well as the determination of the appropriate valuation methods. If we had made different assumptions, our stock-based compensation expense could have been different. The foregoing valuation methodologies are not the only methodologies available and they will not be

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used to value our common stock once this offering is complete. We cannot make assurances as to any particular valuation for our common stock. Accordingly, we caution you not to place undue reliance on the foregoing valuation methodologies as an indicator of future stock prices.

The Company has recognized the following compensation cost related to employee and non-employee based stock option activity:

<u>Year Ended</u>	<u>R&D</u>	<u>General and Administrative</u>	<u>Total</u>
2013 (9 Months)	\$ 63,684	\$ 374,368	\$438,052
2012	52,768	69,573	122,341
2011	175,418	132,011	307,429

Emerging Growth Company Status

The JOBS Act, permits an “emerging growth company” such as us to take advantage of an extended transition period to comply with new or revised accounting standards applicable to public companies. We are choosing to “opt out” of this provision and, as a result, we will comply with new or revised accounting standards as required when they are adopted. This decision to opt out of the extended transition period under the JOBS Act is irrevocable.

Recently Adopted Accounting Pronouncements

In February 2013, the FASB issued guidance to provide information about the amounts reclassified out of accumulated other comprehensive income, or AOCI, by component. An entity is required to present, either on the face of the financial statements or in the notes, significant amounts reclassified out of AOCI by the respective line items of net income, but only if the amount reclassified is required to be reclassified in its entirety in the same reporting period. For amounts that are not required to be reclassified in their entirety to net income, an entity is required to cross-reference to other disclosures that provide additional details about those amounts. On January 1, 2013, we adopted this standard, which had no impact on our financial position or results of operations.

In June 2011, the FASB issued an amendment to the accounting guidance for presentation of comprehensive income. Under the amended guidance, a company may present the total of comprehensive income, the components of net income, and the components of other comprehensive income either in a single continuous statement of comprehensive income or in two separate but consecutive statements. In either case, a company is required to present each component of net income along with total net income, each component of other comprehensive income along with a total for other comprehensive income, and a total amount for comprehensive income. The amendment is effective for fiscal years ending, and interim periods within those years, beginning after December 15, 2011, and is applied retrospectively. The adoption of this amendment has not had a material impact.

In May 2011, the FASB issued amended guidance on fair value measurements. This newly issued accounting standard clarifies the application of certain existing fair value measurement guidance and expands the disclosures for fair value measurements that are estimated using significant unobservable (Level 3) inputs. This accounting standard was effective on a prospective basis for annual and interim reporting periods beginning on or after December 15, 2011. The adoption of this standard has not had a material impact on our financial position or results of operations.

Results of Operations**Comparison of the Nine Months Ended September 30, 2012 (unaudited) and 2013**

	Nine Months Ended September 30,		Increase (Decrease)
	2012 (unaudited)	2013 (in thousands)	
Revenue	\$ —	\$ —	\$ —
Expenses:			
Research and development	5,065	7,591	2,526
General and administrative	1,594	2,141	547
Total expenses	6,659	9,732	3,073
Loss from operations	(6,659)	(9,732)	3,073
Other income, net	866	2,513	1,647
Net loss	\$ (5,793)	\$ (7,219)	\$ 1,426

Research and Development Expenses. Research and development expenses were \$7.6 million for the nine months ended September 30, 2013, compared to \$5.1 million for the nine months ended September 30, 2012. The increase was primarily due to an increase in AKB-6548 clinical trial costs of approximately \$1.7 million due to the initiation of our Phase 2b study in July 2013 and its continued enrollment, and increased patent and regulatory costs of approximately \$0.8 million.

General and Administrative Expenses. General and administrative expenses were \$2.1 million for the nine months ended September 30, 2013, compared to \$1.6 million for the nine months ended September 30, 2012. The increase of \$0.5 million was due to offsetting increases and decrease in all general and administrative costs.

Other Income, Net. Other income, net was \$2.5 million for the nine months ended September 30, 2013 compared to \$0.9 million for the nine months ended September 30, 2012. Other income, net for the nine months ended September 30, 2013 included \$0.8 million in reimbursements from Aerpio for Akebia employee related costs and a \$2.4 million gain on the extinguishment of debt, partially offset by net interest expense of \$0.7 million. Other income, net for the nine months ended September 30, 2012 included \$1.6 million in reimbursements from Aerpio for Akebia employee-related costs, partially offset by net interest expense of \$0.8 million. The decrease in reimbursements from Aerpio for Akebia employee related costs is principally the result of reduced time spent by Akebia employees on Aerpio related activities. Under the terms of administrative services agreements entered into upon disposition of Aerpio by Akebia in 2011, the Company and Aerpio obtain from and provide to each other certain services.

The \$2.4 million gain on extinguishment of debt recognized in the nine months ended September 30, 2013 is the result the March 2013 modification of the 2012 Series X preferred stock. The terms of the 2012 Series X preferred stock were modified to (i) remove the redemption feature and (ii) add a conversion feature. We have accounted for the amendment to the 2012 Series X preferred stock as an extinguishment of the prior security, which was classified as a liability and the issuance of a new preferred stock due to the significance of the modifications to the substantive contractual terms of the preferred stock and the associated fundamental changes to the nature of the preferred stock. The gain is the excess of the book value over the fair value of the revised 2012 Series X preferred stock at the date of the modification. The fair value of the 2012 Series X preferred stock was determined using a hybrid method, in which one scenario assumed the conversion of preferred shares to common stock in an IPO and a second scenario allocated value to the preferred shares using the OPM.

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Comparison of the Years Ended December 31, 2011 and 2012

	Year ended December 31,		Increase (Decrease)
	2011	2012	
Revenue	\$ —	\$ —	\$ —
Expenses:		(in thousands)	
Research and development	12,976	5,632	(7,344)
General and administrative	2,567	2,891	324
Total expenses	15,543	8,523	(7,020)
Loss from operations	(15,543)	(8,523)	(7,020)
Other income, net	246	327	81
Net loss	<u>\$ (15,297)</u>	<u>\$ (8,196)</u>	<u>\$ (7,101)</u>

Research and Development Expenses. Research and development expenses were \$5.6 million for the year ended December 31, 2012, compared to \$13.0 million for the year ended December 31, 2011, a decrease of \$7.3 million. Of the \$7.3 million decrease, approximately \$3.8 million is a result of expenses related to AKB-9778 and AKB-4924, which were no longer Akebia programs in 2012 as a result of the 2011 divestiture of Aerpio. The remainder of the decrease relates to our AKB-6548 program in the following areas: \$1.2 million related to drug substance and drug product costs as the Phase 2b trial did not start until July 2013; \$1.4 million related to clinical trial costs related to studies that were performed in 2011 with nominal costs incurred in 2012; and \$0.7 million of toxicology related expenses due to the completion of the Phase 2a trial.

General and Administrative Expenses. General and administrative expenses were \$2.9 million for the year ended December 31, 2012, compared to \$2.6 million for the year ended December 31, 2011. The increase of \$0.3 million was due to offsetting increases and decrease in all general and administrative costs.

Other Income, Net. Other income, net, was \$0.3 million for the year ended December 31, 2012, compared to \$0.2 million for the year ended December 31, 2011, an increase of approximately \$0.1 million. The increase is primarily related to reimbursements received from Aerpio of \$2.0 million for services which we provided to Aerpio pursuant to the administrative service agreement, partially offset by interest expense of \$1.6 million.

Liquidity and Capital Resources

We have incurred losses and cumulative negative cash flows from operations since our inception in February 2007, and as of September 30, 2013, we had an accumulated deficit of \$119.2 million. We anticipate that we will continue to incur losses for at least the next several years. We expect that our research and development and general and administrative expenses will continue to increase and, as a result, we will need additional capital to fund our operations, which we may raise through a combination of equity offerings, debt financings, other third-party funding, marketing and distribution arrangements and other collaborations, strategic alliances and licensing arrangements.

We have funded our operations principally from the sale of common stock, preferred stock and convertible notes. As of September 30, 2013, we had cash and cash equivalents of approximately \$24.6 million. Cash in excess of immediate requirements is invested in accordance with our investment policy, primarily with a view to liquidity and capital preservation. Currently, our funds are held in money market funds.

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Cash Flows

The following table sets forth the primary sources and uses of cash for each of the periods set forth below:

	Year ended December 31,		Nine months ended September 30,	
	2011	2012	2012 (unaudited)	2013
(in thousands)				
Net cash provided by (used in):				
Operating activities	\$ (14,121)	\$ (7,211)	\$ (5,238)	\$ (6,424)
Investing activities	(732)	1,366	1,366	(13,160)
Financing activities	17,722	2,475	2,475	42,546
Net (decrease) increase in cash and cash equivalents	\$ 2,869	\$ (3,370)	\$ (1,397)	\$ 22,962

Operating Activities. The net cash used in operating activities was \$6.4 million for the nine months ended September 30, 2013, and consisted primarily of a net loss of \$7.2 million adjusted for non-cash items including gain on extinguishment of debt of \$2.4 million, amortization of debt discount of \$0.8 million and stock-based compensation expense of \$0.4 million and a net increase in operating assets and liabilities of \$2.0 million. The significant items in the change in operating assets and liabilities includes an increase in accounts payable of \$2.1 million and a decrease in prepaid expenses and other current assets and accounts receivable of \$0.1 million.

The net cash used in operating activities was \$5.2 million for the nine months ended September 30, 2012, and consisted primarily of a net loss of \$5.8 million adjusted for non-cash items including amortization of debt discount of \$0.8 million and stock-based compensation expense of \$0.1 million and a net decrease in operating assets and liabilities of \$0.4 million. The significant items in the change in operating assets and liabilities include a decrease in accounts payable and accrued expense of \$0.6 million partially offset by an increase in prepaid expenses and other current assets of \$0.3 million.

The net cash used in operating activities was \$7.2 million for the year ended December 31, 2012, and consisted primarily of a net loss of \$8.2 million adjusted for non-cash items including amortization of debt issue costs and debt discount of \$1.7 million and stock-based compensation expense of \$0.1 million and a net decrease in operating assets and liabilities of \$0.8 million. The significant items in the change in operating assets and liabilities include a decrease in accounts payable and accrued expenses of \$1.0 million, offset by an increase in prepaid expenses and current assets of \$0.2 million.

The net cash used in operating activities was \$14.1 million for the year ended December 31, 2011, and consisted primarily of a net loss of \$15.3 million adjusted for non-cash items including stock-based compensation expense of \$0.3 million and a net increase in operating assets and liabilities of \$0.8 million. The significant items in the change in operating assets and liabilities include increases in accounts payable and accrued expenses of \$1.5 million, offset by a decrease in prepaid expenses and other current assets of \$0.6 million.

Investing Activities. Net cash provided by (used in) investing activities consisted of purchases of fixed assets, purchases of marketable securities, and proceeds from the sale of marketable securities. Net cash used in investing activities for the nine months ended September 30, 2013 was \$13.2 million and consisted primarily of purchases of short-term investments. Net cash provided by investing activities for the nine months ended September 30, 2012 was \$1.4 million and consisted completely of proceeds from the maturities of short-term investments. Net cash provided by investing activities for the year ended December 31, 2012 was \$1.4 million and consisted completely of proceeds from maturities of short-term investments. Net cash used in investing activities for the year ended December 31, 2011 was \$0.7 million and was comprised primarily of purchases of short-term investments of \$2.4 million, offset by proceeds from maturities of short-term investments of \$1.7 million.

Financing Activities. Net cash provided by financing activities for the nine months ended September 30, 2013 was \$42.5 million and consisted primarily of \$43.7 million of proceeds from the issuance of preferred stock

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partially offset by stock issue costs of \$1.2 million. Net cash provided by investing activities for the nine months ended September 30, 2012 is the result of the sale of 25,000 shares of our Series X preferred stock for net proceeds of \$2.5 million. Net cash provided by financing activities for the year ended December 31, 2012 is the result of the sale of 25,000 shares of our Series X preferred stock for net proceeds of \$2.5 million. Net cash provided by financing activities for the year ended December 31, 2011 is the result of the issuance and sale of 1.3 million shares of our Series B preferred stock for net proceeds of \$17.7 million.

Operating Capital Requirements

To date, we have not generated any revenue from product sales. We do not know when, or if, we will generate any revenue from product sales. We do not expect to generate significant revenue from product sales unless and until we obtain regulatory approval of and commercialize one of our current or future product candidates. We anticipate that we will continue to generate losses for the foreseeable future, and we expect the losses to increase as we continue the development of, and seek regulatory approvals for, our product candidates, and begin to commercialize any approved products. We are subject to all of the risks incident in the development and commercialization of novel therapeutics, and we may encounter unforeseen expenses, difficulties, complications, delays and other unknown factors that may adversely affect our business. Upon the closing of this offering, we expect to incur additional costs associated with operating as a public company. We anticipate that we will need substantial additional funding in connection with our continuing operations.

We believe that the net proceeds from this offering and our existing cash and cash equivalents will be sufficient to fund our projected operating requirements into . However, we may require additional capital for the further development of our existing product candidates and may also need to raise additional funds sooner to pursue other development activities related to additional product candidates.

If and until we can generate a sufficient amount of revenue from our products, we expect to finance future cash needs through public or private equity or debt offerings. Additional capital may not be available on reasonable terms, if at all. If we are unable to raise additional capital in sufficient amounts or on terms acceptable to us, we may have to significantly delay, scale back or discontinue the development or commercialization of one or more of our product candidates. If we raise additional funds through the issuance of additional debt or equity securities, it could result in dilution to our existing stockholders, increased fixed payment obligations and these securities may have rights senior to those of our common stock. If we incur indebtedness, we could become subject to covenants that would restrict our operations and potentially impair our competitiveness, such as limitations on our ability to incur additional debt, limitations on our ability to acquire, sell or license intellectual property rights and other operating restrictions that could adversely impact our ability to conduct our business. Any of these events could significantly harm our business, financial condition and prospects.

Our forecast of the period of time through which our financial resources will be adequate to support our operations is a forward-looking statement and involves risks and uncertainties, and actual results could vary as a result of a number of factors. We have based this estimate on assumptions that may prove to be wrong, and we could utilize our available capital resources sooner than we currently expect. Our future funding requirements, both near and long-term, will depend on many factors, including, but not limited to:

- the rate of progress, results and cost of completing our Phase 2b clinical trial of AKB-6548 and our operating costs incurred as we conduct these trials and through our end of Phase 2 meeting with the FDA, and equivalent meetings with the EMA and other regulatory authorities;
- assuming positive results from our current Phase 2b trial, the scope, size, rate of progress, results and costs of initiating and completing our Phase 3 development program of AKB-6548;
- assuming favorable clinical results, the cost, timing and outcome of our efforts to obtain marketing approval for AKB-6548 in the United States and in other jurisdictions, including to fund the preparation and filing of regulatory submissions for AKB-6548 with the FDA, the EMA and other regulatory authorities;

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- the scope, progress, results and costs of preclinical development, laboratory testing and clinical trials for AKB-6899 and any other product candidates that we may develop or acquire;
- the timing of, and the costs involved in, obtaining regulatory approvals for AKB-6899 if clinical trials are successful and the outcome of regulatory review of AKB-6899;
- the cost and timing of future commercialization activities for our products, if any of our product candidates are approved for marketing, including product manufacturing, marketing, sales and distribution costs;
- the revenue, if any, received from commercial sales of our product candidates for which we receive marketing approval;
- the cost of having our product candidates manufactured for clinical trials in preparation for regulatory approval and in preparation for commercialization;
- our ability to establish and maintain strategic collaborations, licensing or other arrangements and the financial terms of such agreements;
- the costs involved in preparing, filing, prosecuting patent applications, maintaining, defending and enforcing our intellectual property rights, including litigation costs and the outcome of such litigation;
- the timing, receipt, and amount of sales of, or royalties on, our future products, if any;
- the need to implement additional infrastructure and internal systems; and
- the extent to which we acquire or in-license other products or technologies.

If we cannot expand our operations or otherwise capitalize on our business opportunities because we lack sufficient capital, our business, financial condition and results of operations could be materially adversely affected.

Contractual Obligations and Commitments

We contract with various organizations to conduct research and development activities with remaining contract costs to us of approximately \$4.9 million at September 30, 2013, \$0.7 million at December 31, 2012 and \$1.5 million at December 31, 2011. The scope of services under the research and development contracts can be modified and the contracts can be cancelled by either party upon written notice.

As of December 31, 2012, we did not have any material lease obligations as we are provided office space and other services by Aerpio through the administrative services agreement and are charged based on usage. The scope of services under the administrative services agreement can be modified and the agreement can be cancelled by either party upon written notice.

In December 2013, we entered into a three year office lease agreement for 6,837 square feet of space in Cambridge, Massachusetts, commencing on the date that the premises are delivered to us with the landlord work substantially complete. The lease has monthly lease payments of approximately \$31,000 for the first twelve months with annual rent escalation thereafter and provides a rent abatement of approximately \$31,000 for the first full calendar month of the lease term.

Off-Balance Sheet Arrangements

We did not have during the periods presented, and we do not currently have, any off-balance sheet arrangements, as defined in the rules and regulations of the Securities and Exchange Commission.

Quantitative and Qualitative Disclosures About Market Risks

We are exposed to market risk related to changes in interest rates. As of September 30, 2012 and 2013, we had cash and cash equivalents and short-term investments of \$3.6 million and \$37.7 million, respectively, primarily money market mutual funds consisting of U.S. government-backed securities. Our primary exposure to market risk is interest rate sensitivity, which is affected by changes in the general level of U.S. interest rates, particularly because our investments are in short-term securities. Our available for sale securities are subject to interest rate risk and will fall in value if market interest rates increase. Due to the short-term duration of our investment portfolio and the low risk profile of our investments, an immediate 100 basis point change in interest rates would not have a material effect on the fair market value of our portfolio.

Business

Overview

We are a biopharmaceutical company focused on the development of novel proprietary therapeutics based on HIF biology and the commercialization of these products for patients with kidney disease. HIF is the primary regulator of the production of RBCs in the body and a potentially novel mechanism of treating anemia. Our lead product candidate, AKB-6548, is being developed as a once-daily oral therapy that has successfully completed a Phase 2a proof of concept study demonstrating that AKB-6548 safely and predictably raised hemoglobin levels in patients with anemia secondary to CKD not requiring dialysis.

We are conducting a Phase 2b trial for AKB-6548 in patients with anemia secondary to CKD who are not dependent on dialysis and expect data to be available in the fourth quarter of 2014. We have also initiated a development program for patients dependent on dialysis. If the results of our Phase 2b trial are positive, we would expect to initiate Phase 3 trials for anemia secondary to CKD in 2015, and would anticipate submitting an NDA for AKB-6548 in the United States by 2018 if the Phase 3 data are favorable.

We own worldwide rights to our HIF-based product candidates, including AKB-6548. If approved by regulatory authorities, we plan to commercialize AKB-6548 in the United States ourselves and intend to seek one or more collaborators to commercialize the product candidate in additional markets.

Anemia is a serious medical condition in which blood is deficient in RBCs and hemoglobin, both of which are critical in delivering oxygen to tissue. Anemia generally exists when hemoglobin, a protein in RBCs that carries oxygen, is less than 13 g/dL in men or 12 g/dL in women. Untreated anemia is associated with chronic fatigue, increased risk of progression of multiple diseases, and death. Anemia is common in patients with CKD, cancer, heart failure, inflammatory diseases and other critical illnesses, as well as in the elderly.

More than 30 million people in the United States have CKD, with estimates that over 1.8 million of these patients suffer from anemia. Anemia from these indications is currently treated by injectable recombinant protein erythropoiesis stimulating agents, or rESAs—including Epogen, Aranesp, and Procrit—with iron supplementation or an RBC transfusion. Based on the reported revenues of companies that market and sell rESAs, we estimate that global sales of injectable rESAs were \$6.3 billion in 2012, the vast majority of which were for renal indications.

rESAs are designed to stimulate production of RBCs by binding directly to and saturating EPO receptors. While injectable rESAs and transfusions may be effective in raising hemoglobin levels, they carry significant potential side effects and also need to be delivered subcutaneously or intravenously. In particular, injectable rESAs may lead to thrombosis, stroke, myocardial infarction and death, and these risks are described in black box warnings on the prescribing information of all products marketed in this class. These safety concerns, which became evident starting in 2006, have led to a significant reduction in the use of injectable rESAs. Today anemia is either not treated or inadequately treated in the majority of CKD patients, and we believe that a safe, effective, oral therapeutic option will take significant market share and meaningfully grow the market in patients not requiring dialysis.

AKB-6548 works by a differentiated mechanism of action that we believe has the potential to be safer than that of injectable rESAs. This novel mechanism of action is referred to as HIF-PH inhibition. Instead of binding directly to the EPO receptors on cells in the bone marrow, AKB-6548 leads to activation of critical pathways for hemoglobin and RBC production. This approach mimics the physiological adjustment made by the body when exposed to reduced oxygen levels at higher altitudes.

To date, AKB-6548 has been studied in eight clinical trials across four separate patient populations: healthy volunteers and patients with CKD stages 3, 4 and 5 (non-dialysis). Our largest study was a Phase 2a trial in 91

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patients with anemia secondary to CKD, which showed significantly increased hemoglobin levels among subjects taking AKB-6548 compared to baseline in a dose-dependent manner across all treatment arms ($p < 0.0001$). No drug-related serious adverse events were reported, and dosing was well-tolerated. In addition, AKB-6548 was also shown to stabilize the iron supply to the bone marrow while improving hemoglobin production.

Our ongoing Phase 2b trial explores a dosing approach for AKB-6548 to enable subjects with anemia secondary to CKD to appropriately and safely raise hemoglobin levels. As of December 16, 2013, we had enrolled over 50% of our targeted 200 patients in this study at investigational sites in the United States, with data expected in the fourth quarter of 2014. With positive data, we plan to progress to Phase 3 global registration studies for AKB-6548 in patients with anemia secondary to CKD. We anticipate the design of the Phase 3 studies will mirror the Phase 2b study, except that they will be longer and larger in size, positioning us to file for approval in the United States by 2018.

Given the burdens of the current standard of care and costs associated with administering an injectable rESA, we believe AKB-6548 is a promising alternative for the overall cost-effective treatment of anemia. We intend to commercialize AKB-6548 ourselves in the United States for the treatment of anemia in patients with CKD. These patients are primarily treated by approximately 7,000 nephrologists, and we believe we can reach most of this market with a specialty sales force of approximately 125 people. We intend to seek one or more commercial collaborators for the development and commercialization of AKB-6548 outside of the United States. We may also explore opportunities to expand AKB-6548 into additional markets not adequately addressed by injectable rESAs because of safety or dosing delivery issues, including IAA and anemia of congestive heart failure.

We are led by a team of experienced biopharmaceutical executives with a background in developing and commercializing drugs for the treatment of renal and metabolic disorders. John P. Butler, our CEO, was former President of Genzyme Corp.'s renal division which grew to over \$1 billion in annual revenue under his leadership, and is current Chairman of the Board of the American Kidney Fund, the leading patient advocacy organization for kidney disease patients. Earlier in his career, Mr. Butler held sales and marketing positions at Amgen, working on the early commercial launch of injectable rESAs in the renal anemia market. Our executive team also includes Robert Shalwitz, M.D., CMO and co-founder of Akebia. Dr. Shalwitz is an academic pediatric endocrinologist and has extensive industry experience developing novel pharmaceuticals at Abbott Laboratories and Reliant Pharmaceuticals. He has developed extensive knowledge of HIF biology over his career, particularly over the past seven years in leading development at Akebia.

Our Strategy

Our strategy is to develop novel therapeutics for patients based on HIF biology and to commercialize products for patients with kidney disease, beginning with AKB-6548 for patients with anemia secondary to CKD. The key elements of our strategy are to:

- **Complete the development of AKB-6548 for anemia secondary to CKD.** We plan to complete the Phase 2b trial that is currently enrolling in the United States. We intend to initiate a Phase 3 development program in 2015 following our end of Phase 2 meeting with the FDA.
- **Obtain regulatory approval of AKB-6548 for anemia secondary to CKD in the United States, Europe and other markets.** We plan to complete an end of Phase 2 meeting with the FDA and seek scientific advice from the EMA to define the Phase 3 development program necessary to secure regulatory approval to market AKB-6548. We would expect to initiate Phase 3 trials for anemia secondary to CKD in 2015, and would anticipate submitting an NDA for AKB-6548 in the United States by 2018 if the Phase 3 data are favorable.

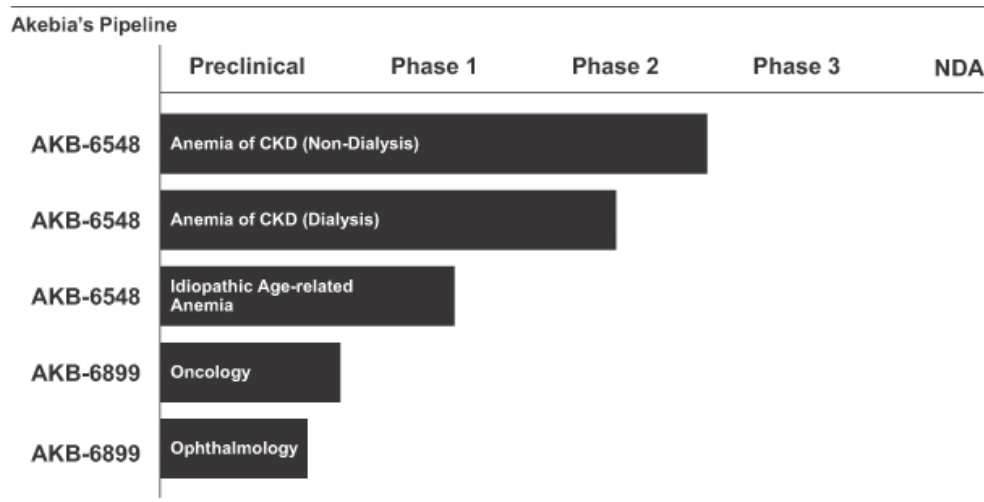
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- **Commercialize AKB-6548 in the United States and other territories.** We will establish a specialty sales and marketing organization to commercialize AKB-6548 in the United States. Outside of the United States, we intend to seek one or more commercial collaborators.
- **Continue to develop AKB-6548 for further indications.** We plan to initiate, in the first half of 2014, a Phase 2 study for AKB-6548 in dialysis patients with anemia, the second indication we intend to pursue. Additionally, we plan to evaluate the product candidate in IAA and other indications.
- **Advance our earlier stage pipeline asset.** We plan to advance AKB-6899, a second HIF-PH inhibitor product candidate, which we believe, based on preclinical testing, has the ability to increase EPO levels while reducing vascular endothelial growth factor, or VEGF, levels. We intend to file an IND application and begin Phase 1 trials to determine its potential use in oncology and ophthalmology.
- **Acquire or in-license additional nephrology products.** If we are able to successfully launch AKB-6548, we will look to leverage our commercial infrastructure with additional products that would be prescribed by nephrologists.

We may enter into strategic collaborations to fully realize all of the elements of our strategy.

Our Product Candidates

The following chart depicts our HIF-based product candidates, their indications and their current development:



Anemia Overview

Anemia is a serious medical condition in which blood is deficient in RBCs and hemoglobin, leading to inadequate oxygen delivery to tissues and cells throughout the body. RBCs are normally formed in the bone marrow from precursor or progenitor cells. EPO, a hormonal factor primarily produced in the kidney and liver, binds to and activates the EPO receptor on these precursor cells. The activation of the EPO receptor stimulates these cells to divide, differentiate into RBCs that contain hemoglobin, and mobilize into circulation. Hemoglobin is an iron-containing protein in RBCs that transports oxygen to, and carbon dioxide from, the tissues of the body.

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Anemia generally exists when hemoglobin is less than 13 g/dL in men and 12 g/dL in women. Anemia has a number of potential causes, including nutritional deficiencies, iron deficiency, bone marrow disease, medications, and abnormalities in EPO production or sensitivity. Common causes of anemia due to inadequate EPO production include CKD, age, heart failure, inflammatory diseases, cancer and other critical illnesses.

Untreated anemia is associated with chronic fatigue, increased risk of progression of multiple diseases, and death. This morbidity and mortality risk has been clearly shown in the CKD population, where in patients age 66 and older, anemic patients with mid-stage CKD (Stage 3) have a 149% increase in cardiovascular events and patients with severe CKD (Stage 4 and 5) have a 24% increase in cardiovascular events versus non-anemic patients in the same group, according to a paper published in 2006 in the peer-reviewed journal *Blood*. Similarly, compared to non-anemic patients, anemia increases the mortality rate by 199% in mid-stage CKD, and 59% in severe CKD. Successful treatment of anemia significantly improves patients' quality of life, especially with respect to vitality, fatigue and physical function. In addition, patients whose anemia has been successfully treated have demonstrated lower mortality rates, less frequent hospitalization, and decreases in cardiovascular morbidity.

Chronic Kidney Disease

CKD, a common cause of anemia, is a condition in which the kidneys are progressively damaged to the point that they cannot properly filter the blood circulating in the body. This damage can cause waste products to build up in the subject's blood and can lead to other health problems, including cardiovascular disease, anemia, and bone disease. CKD patients are classified by the degree of their loss of kidney function as measured by the glomerular filtration rate, or GFR, and albuminuria, the protein levels in urine. As seen in the table below, CKD affects more than 30 million people in the United States. As shown in the table below, the prevalence of anemia is associated with the severity of CKD in this population.

Anemia Prevalence Increases by CKD Stage

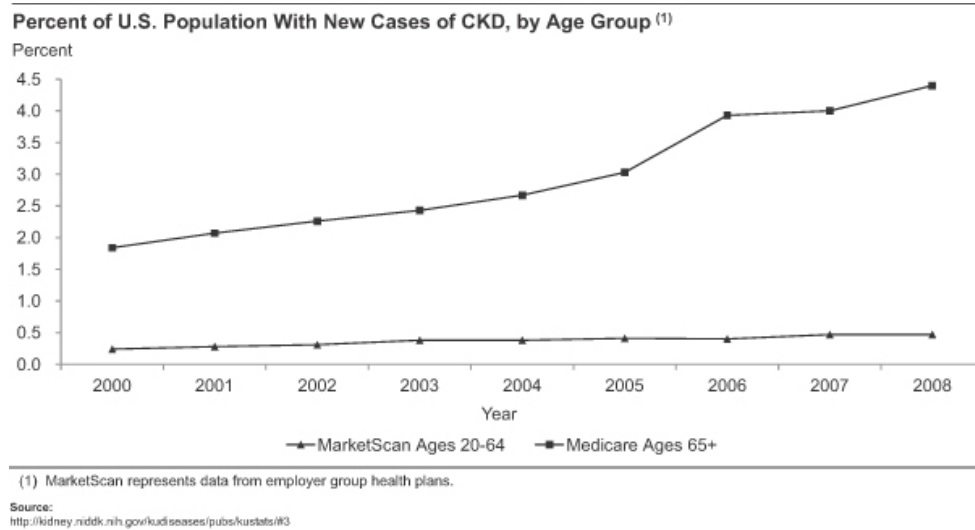
Stage	Prevalence – (diagnosed / undiagnosed)	Kidney Function	Estimated Anemia Prevalence (with CKD diagnosis)	Estimated U.S. Anemia Patients (with CKD diagnosis)
1	~4M	Persistent albuminuria with GFR higher than 90 mL/min/1.73 m2	<1%	~18,000
2	~8M	Persistent albuminuria with GFR of 60 to 89 mL/min/1.73 m2 Mild (GFR: 60 – 89)	1 – 2%	~65,000
3	~17M	GFR of 30 to 59 mL/min/1.73 m2 Moderate (GFR: 30 – 59)	6 - 8	Up to ~605,000
4	~1M	GFR of 15 to 29 mL/min/1.73 m2 Severe (GFR: 15 – 29)	50% to 60	Up to ~500,000
5	~670	Failure (GFR: <15) Includes Dialysis, No Dialysis & Transplants	85%-100%	~650,000

Sources:
 Stages 1-4: JAMA 2007 Coresh et al (Prevalence of CKD in the US), NHANES 1988-94 and 1999-2004.
 Stage 5: USRDS 2013 report (ESRD).

There are many causes of CKD, the most common of which are diabetes and hypertension. The prevalence and incidence of CKD is increasing in all segments of the U.S. population, particularly in patients over 65, as shown below. Risk factors for the development of CKD include underlying disease (hypertension, diabetes and cardiovascular disease), lifestyle factors (tobacco use and inactivity), family history, aging, and prenatal factors (maternal diabetes

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mellitus, low birth weight and small-for-gestational-age status). Beyond the United States, according to a *Lancet* article from May 2013, projected worldwide population changes suggest that the potential number of cases of kidney disease, specifically end-stage, will increase disproportionately in developing countries, such as China and India, where the numbers of elderly people are expanding. This effect will be enhanced further if the trends of increasing hypertension and diabetes prevalence persist, competing causes of death—such as stroke and cardiovascular diseases—are reduced, and access to treatment improves.



The prevalence and severity of anemia in CKD increases as renal function deteriorates. Three variables which may combine to accentuate and accelerate anemia as CKD progresses include:

- Peritubular fibroblasts, a type of cell in the kidney, are designed to sense the amount of oxygen carried by the blood. These cells secrete EPO to adjust the production of RBCs and maintain circulating oxygen levels at normal physiologic levels. As kidney disease progresses, the number of peritubular fibroblasts is reduced and EPO secretion is significantly decreased. This decline in EPO leads to a reduction in RBC production.
- CKD leads to a shorter average life span for RBCs (70 days) as compared to healthy individuals (90 to 120 days), requiring increased RBC production to keep RBC levels consistent with those of a healthy individual.
- The availability of iron to the bone marrow is impaired. Iron is a required component in the formation of hemoglobin, and is essential in the transport of both oxygen and carbon dioxide.

As CKD progresses, the combined effect of decreased RBC production from lower EPO signaling, increased rate of RBC destruction, and reduced iron availability to the bone marrow results in the increased prevalence and severity of anemia.

Current Treatments Leave a Substantial Unmet Need

Injectable rESAs, including epoetin alfa, epoetin beta, and darbepoetin alfa, are currently the standard of care for treating anemia in patients with CKD and must be administered intravenously or subcutaneously with iron supplements. Based on the reported revenues of companies that market and sell rESAs, we estimate that global sales of injectable rESAs were \$6.3 billion in 2012, as compared to an estimated \$12 billion in 2006. Of these 2012

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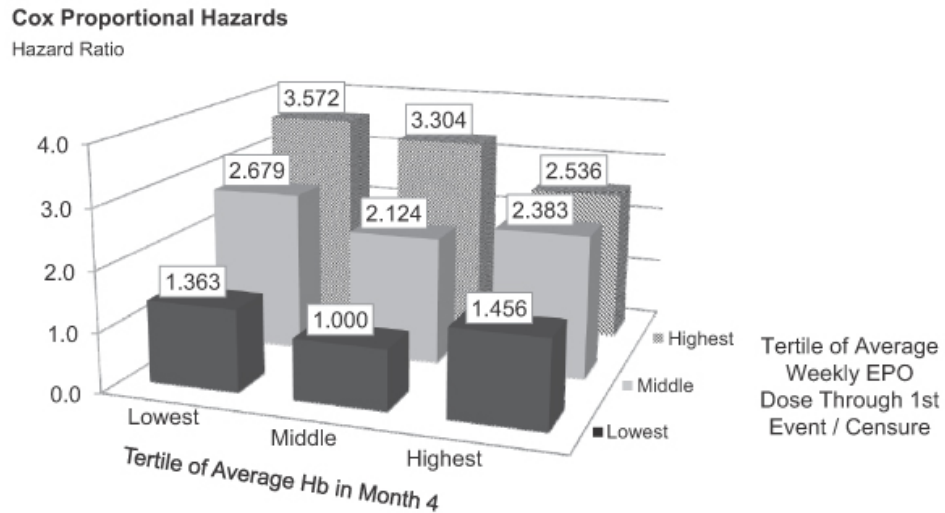
revenues, an estimated \$3.4 billion were generated in the United States, the vast majority of which were for renal indications. In 2006, data on the risks of rESA use among these patients started to become available, forcing physicians to balance serious safety concerns against the efficacy of rESAs. The safety concerns with injectable rESA use include increased risk of cardiovascular disease as well as a potentially increased rate of tumor progression in patients with cancer. We believe that the decline in market revenue since 2007 is a direct result of these increased safety concerns, as well as reimbursement pressures, and that an opportunity exists for a safer, well-tolerated alternative to replace injectable rESAs as the standard of care for anemia secondary to CKD.

As a result of the safety concerns related to rESA use, patients have been forced to live with lower hemoglobin levels, higher rates of transfusions, and more intravenous iron, or IV iron, use. The percentage of dialysis patients in the United States receiving IV iron has increased from 50% in 1999 to 71% during in 2011, which is consistent with the general trend of increasing IV iron. Among U.S. patients receiving IV iron, the mean monthly dose has also increased by 21%. Despite the increased use of IV iron and rate of transfusions, patients are still subject to safety risks related to these alternative treatments to injectable rESAs. The risks of transfusions include the development of antibodies to foreign antigens, transmission of blood-borne pathogens, impairment of venous access in CKD patients (not on dialysis) and iron overload with chronic transfusion. The risks of IV iron include hypersensitivity reactions, such as fatal anaphylactic-type reactions.

Currently, there is no scientific consensus regarding the adverse cardiovascular outcomes associated with the use of injectable rESAs to normalize hemoglobin levels. The results of the four major randomized, controlled clinical trials on the treatment of anemia secondary to CKD with rESAs and adjunctive iron supplementation (Normal Hematocrit Trial/NHCT; CREATE, CHOIR and TREAT) all showed an increased risk of adverse cardiovascular outcomes. These results were surprising at the time and contradicted the extensive body of data from observational studies that showed reduced mortality and improved health outcomes to be associated with higher hemoglobin levels.

A number of critical post-hoc analyses of the randomized controlled trials data have shifted attention to the potential of dose-related toxicity of injectable rESAs in CKD patients as a contributing factor to the reported adverse cardiovascular outcomes, instead of the role of normalized hemoglobin levels. The strongest correlation of adverse outcomes in the post-hoc analyses has been to the level of the injectable rESA dose, not the hemoglobin level achieved. All of the studies analyzed to date demonstrate that both non-dialysis and dialysis-dependent CKD subjects who achieved normal hemoglobin levels with or without minimal doses of injectable rESAs or supplemental iron had better clinical outcomes than subjects assigned to higher hemoglobin targets who failed to reach the assigned level with increasing doses of injectable rESAs and iron. In addition, CKD patients who are able to achieve and maintain normal hemoglobin levels through means other than the use of injectable rESAs (such as hypoxia or iron supplementation) experienced fewer cardiovascular events and reduced morbidity and mortality. Recent studies of injectable rESA use in various preclinical models (including non-human primates) also showed that the frequency of mortality and thrombotic events cannot be explained solely by the achieved higher hemoglobin levels, but is related to the dose, dose frequency, and dose duration of injectable rESAs.

The graphs below highlight these findings. The first chart explores the relative risk of serious cardiovascular adverse events, including death, hospitalization for heart failure, stroke or myocardial infarction based upon the hemoglobin achieved during the study as well as the weekly injectable rESA dose. The data clearly show that the risk of adverse cardiovascular events was greatest in those patients receiving the highest injectable rESA doses, regardless of the hemoglobin level that was achieved.



Source:
McCullough P.A. · Barnhart H.X. · Inrig J.K. · Reddan D. · Sapp S. · Patel U.D. · Singh A.K. · Szczech L.A. · Califf R.M. Am J Nephrol 2013;37:549-558 (DOI:10.1159/000351175); Permission granted by S. Karger AG, Basel.

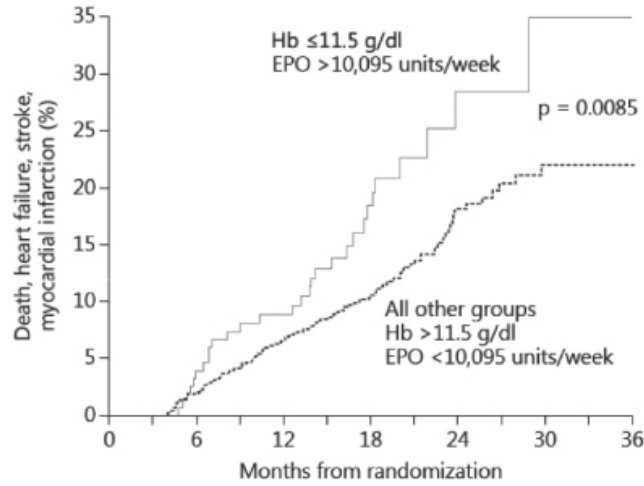
The second graph explores the probability of reaching one of several adverse events (death, stroke, heart failure or myocardial infarction) over time for two different groups:

- patients who achieve the target hemoglobin level with a low injectable rESA dose, and
- patients who do not reach the target hemoglobin level, but receive a high injectable rESA dose in an effort to reach the target level.

This chart is consistent with the previous chart as it shows that patients with high hemoglobin levels on low injectable rESA doses have better outcomes than patients with high injectable rESA doses and low hemoglobin levels. Therefore, high injectable rESA doses, not high hemoglobin levels, appear to be correlated most strongly with adverse outcomes.

Kaplan-Meier Survival Curves

Death, Heart Failure, Stroke, Myocardial Infarction (%)



Source:
McCullough P.A. · Barnhart H.X. · Inrig J.K. · Reddan D. · Sapp S. · Patel U.D. · Singh A.K. ·
Szczzech L.A. · Califf R.M. Am J Nephrol 2013;37:549-558 (DOI:10.1159/000351175);
Permission granted by S. Karger AG, Basel.

The significant safety risks associated with rESAs are outlined in a black-box warning in their prescribing information. This warning arose from numerous events highlighting the safety concerns of injectable rESAs and the responses by the FDA, as highlighted below.

- In 2007, as a result of concerns associated with administering injectable rESAs to target higher hemoglobin levels, the FDA required that revised warnings, including boxed warnings, be added to the labels of marketed injectable rESAs advising physicians to monitor hemoglobin levels and use the lowest dose of injectable rESA, and increase the hemoglobin concentration to the lowest level sufficient to avoid the need for RBC transfusions.
- In November 2007, the FDA found evidence that the use of injectable rESAs to increase hemoglobin to more than 12 g/dL can stimulate progression of some cancers. As a result, injectable rESAs were required to contain black-box labeling for this risk. Following this change in labeling, the use of injectable rESAs in cancer patients has declined significantly.
- In late 2009, Amgen announced the results from the Trial to Reduce Cardiovascular Endpoints with Aranesp Therapy, or TREAT, its large, randomized, double-blind, placebo-controlled Phase 3 study of patients with CKD (not requiring dialysis), anemia and type-2 diabetes. In this study, Aranesp was used

to treat anemia to a target hemoglobin level of 13 g/dL, which was higher than the 10 g/dL - 12 g/dL range previously approved by the FDA in the label. Study results reportedly failed to show benefit compared to the control group with regard to composite of time to all-cause mortality or cardiovascular morbidity (including heart failure, heart attack, stroke, or hospitalization for myocardial ischemia) and composite of time to all-cause mortality or chronic renal replacement therapy. In addition, higher rates of stroke were reported among patients in the 13 g/dL target group compared to the control group. Finally, among a subgroup of patients with a history of cancer at baseline, a statistically significant increase in deaths from cancer was observed in the Aranesp-treated patients compared to placebo-treated patients.

- In January 2010, FDA officials published an editorial in the New England Journal of Medicine noting that a number of randomized trials, including TREAT, had attempted to show that using injectable rESAs to raise hemoglobin concentrations to higher targets improves clinical outcomes but instead suggested the opposite. Accordingly, the article indicated that more conservative hemoglobin targets (well below 12 g/dL), more frequent hemoglobin monitoring, and more cautious dosing should be evaluated.
- In February 2010, the FDA required that injectable rESAs be prescribed and used under a REMS to ensure the safe use of the drugs. As part of the REMS, a medication guide explaining the risks and benefits of injectable rESAs must be provided to all patients receiving injectable rESAs for all indications, and the FDA imposed reporting and monitoring obligations on the manufacturers to ensure compliance.
- In June 2011, the FDA cited increased risks of cardiovascular events as a basis for more conservative dosing guidelines for use of injectable rESAs in CKD patients and announced related changes to injectable rESA labeling. The FDA removed the prior target hemoglobin range of 10-12 g/dL, and recommended that CKD patients initiate treatment when the hemoglobin level is less than 10 g/dL and reduce or interrupt dosing if the hemoglobin level approaches or exceeds 10 g/dL for non-dialysis patients and 11 g/dL for dialysis patients. The FDA also required Amgen to conduct additional clinical trials to explore dosing strategies to minimize hemoglobin variability, rates of change and excursions.

We believe there is now substantial evidence to suggest that EPO level, not hemoglobin, is the cause of the safety issues in the above trials. The collective preclinical and clinical data support a critical re-thinking on the best approach to treating anemia, the appropriate and safe hemoglobin target, and the right time to initiate treatment for these patients.

AKB-6548 as a potential solution

We are developing our lead product candidate, AKB-6548, to be a best in class HIF-PH inhibitor for the treatment of anemia secondary to CKD. We expect AKB-6548 to offer:

- Predictable, meaningful and sustained improvements in hemoglobin levels;
- Once a day therapy delivered orally;
- A dosing regimen that restores the normal diurnal EPO pattern;
- Robust pharmacodynamics and substantially lower peak EPO levels than with injectable rESAs; and
- Reduced administration of IV or oral iron supplementation to patients treated for anemia secondary to CKD.

Novel Mechanism of Action, Which Mimics the Body's Natural Physiologic Response

AKB-6548 is designed to work by a mechanism of action that differs from injectable rESAs. This novel mechanism of action is referred to as a HIF-PH inhibitor. Instead of binding directly to and saturating the EPO receptors in the bone marrow for prolonged periods of time, HIF-PH inhibitors act by simulating the body's natural response to anemia. In this way, AKB-6548 achieves a controlled, adaptive stimulation of the erythropoietic system in the body. This activation of the whole system results in both increased RBC production and improved stabilization of the bone marrow's iron supply, which ensures the proper incorporation of iron into

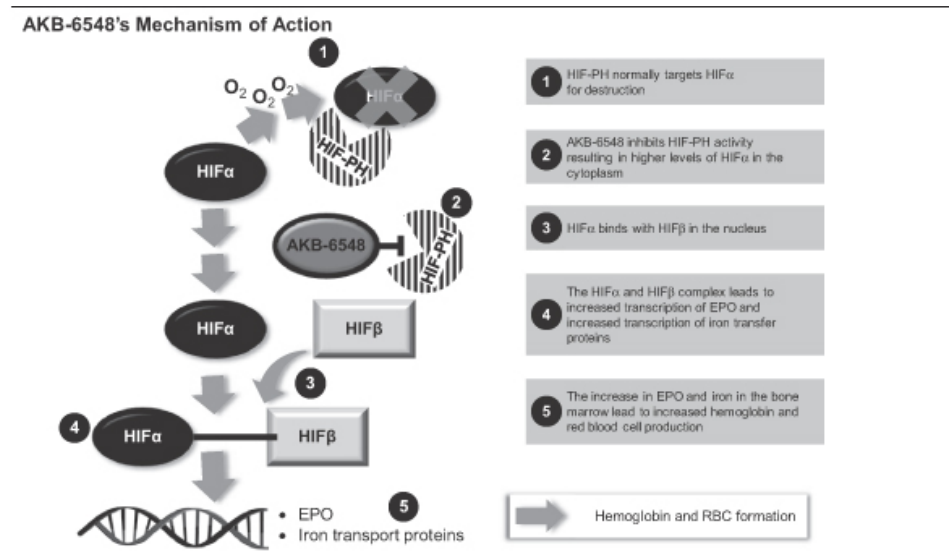
hemoglobin necessary for such RBC production. This adaptive simulation is very similar to the natural response that is induced when a person ascends in altitude. At higher altitudes, low levels of oxygen circulating in the blood stream lead to reduced HIF-PH activity in relevant cells in the kidney and liver. The reduced HIF-PH activity stabilizes and increases levels of HIF α proteins (HIF1 α and HIF2 α) in these cells. For most cells, the stabilization of HIF2 α is greater than that of HIF1 α , ultimately leading to an increase in EPO secretion and a subsequent increase in RBC production.

HIF-PH inhibitors work by blocking the effect of the prolyl-hydroxylase enzymes, which promote the breakdown of HIF α proteins. As the breakdown is inhibited, the level of these HIF α proteins increases in cells. These HIFs are the primary protein mediators that enable the body and all of its individual cells to adapt to changes in levels of oxygen. Both HIF1 α and HIF2 α proteins are consistently produced and their levels in cells are adjusted by the activity of the HIF-PH enzymes, which target the HIF α proteins for degradation. HIF1 α helps cells survive under very low oxygen conditions, whereas HIF2 α helps cells and the body to adapt to modest changes in oxygen, such that would occur with a change in altitude from sea level to up to 7,500 feet.

When HIF α is stabilized, it travels to the nucleus of the cell, where it binds to the protein HIF β . When bound together, they induce the genetic signal for the production of EPO and several other proteins. The HIF-PH inhibitors increase HIF α levels in much the same way that a reduction in oxygen increases HIF α levels by inhibiting the HIF-PH enzymes in the body. With continued stabilization of HIF α (either by staying at higher altitude or by daily dosing of the HIF-PH inhibitor), the level of hemoglobin and RBCs will rise in order to increase the amount of oxygen circulating in the blood. In this way, once-daily dosing of AKB-6548 may have the potential to restore the normal level of EPO for a patient with anemia.

AKB-6548, our lead compound in development, works by inhibiting HIF-PH, leading to stabilization and increased levels of HIF α , and improved production of hemoglobin and RBCs, while maintaining normal levels of EPO in patients. In addition, we believe that AKB-6548's mechanism of action provides for the ability to induce a more prominent HIF2 α response (as naturally occurs with a moderate increase in altitude), and an enhancement in the normal diurnal variation of EPO, which is the normal rise and fall of EPO during the each day.

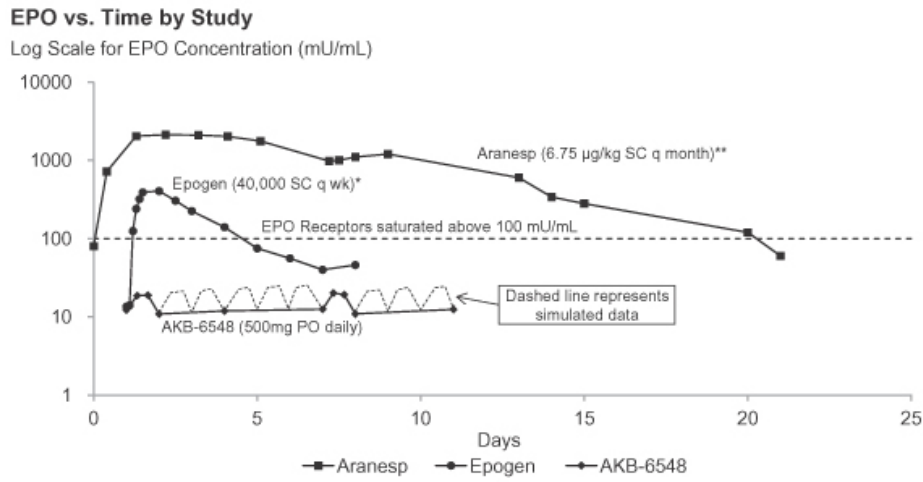
This mechanism of action is illustrated in the graphic below.



Potential Best in Class Profile

We believe AKB-6548 has compelling clinical data demonstrating a best in class profile with several potential safety and efficacy advantages over current injectable rESA therapy in the treatment of anemia secondary to CKD.

- *AKB-6548 significantly increases hemoglobin in anemic CKD patients.* We have successfully completed a Phase 2a trial, in which AKB-6548 significantly increased hemoglobin levels compared to baseline in a dose-dependent manner across all treatment arms ($p < 0.0001$). Further, AKB-6548 provides a physiologic reticulocyte, or newly formed RBC, response, which leads to a more gradual and consistent increase in hemoglobin levels than what is seen with injectable rESA therapies, meaning that these improvements occur without causing patients' hemoglobin to rise to levels that cause concern.
- *AKB-6548 may have the potential to restore the normal diurnal variation of EPO for a patient with anemia in a way that an injectable rESA cannot.* Instead of binding directly to and saturating the EPO receptor for prolonged periods of time as is the case with current injectable rESA treatments, AKB-6548 acts by simulating the body's natural response to hypoxia that is carried out by stabilization of HIF α . We believe the manner in which AKB-6548 works permits a more prominent HIF2 α response (as naturally occurs with a moderate increase in altitude) and there is an enhancement in the normal diurnal variation in EPO, which is the normal rise and fall of EPO during the each day, without continuous elevation of EPO levels. The graph below illustrates the EPO levels that are obtained with AKB-6548 compared with doses of Aranesp and Epogen.



*Source: Arroliga, et. al., Crit Care Med 2009, Vol. 37, No. 4.

**Source: Glaspy, et. al., European Journal of Cancer 41 (2005) 1140-1149. Data based on original Aranesp measurements as adjusted for the higher inherent potency of Aranesp.

- *Oral, once-daily dosing.* Once daily, oral dosing of AKB-6548 offers improved convenience for patients as compared to injectable rESAs. This convenience may increase access to anemia therapy for the largely underserved population of patients with anemia secondary to CKD who are not yet on dialysis and for patients with other forms of anemia, such as IAA. AKB-6548 offers the potential of flexible oral dosing that provides a more gradual and reliable means of titration than that of injectable rESAs.

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- *Ability to stabilize the iron supply to the bone marrow while improving hemoglobin production.* In clinical trials, AKB-6548 has demonstrated a dose-related increase in total iron binding capacity, or TIBC.
- These results indicate that AKB-6548 will stabilize the iron supply to the bone marrow while improving hemoglobin production and should improve EPO responsiveness. As a result, unlike injectable rESAs, which have no effect on iron mobilization, AKB-6548 offers the added potential benefit of reducing the amount of supplemental iron required by anemia patients.
- *Differentiated safety profile.* AKB-6548's novel mechanism of action and dosing profile offer the opportunity to potentially avoid the black box label ascribed to injectable rESAs. In our recently completed Phase 2a study, no drug-related serious adverse events were reported. Dosing was well-tolerated and there was no evidence of undesirable vascular response.

AKB-6548 Clinical Development Overview

Early Clinical Studies (CI-0001 to CI-0004, and CI-0006):

To date, AKB-6548 has been studied in eight clinical trials across four separate patient populations: healthy volunteers and patients with CKD stages 3, 4, and 5 (non-dialysis). These clinical trials consisted of four Phase 2a clinical trials and four Phase 1 clinical trials. The early clinical studies (CI-0001 through CI-0004) for AKB-6548 were designed to demonstrate the efficacy and safety of the compound, starting in healthy male volunteers and progressing to CKD patients with anemia. In healthy males, we demonstrated that AKB-6548 can be dosed daily, and that it induces the desired pharmacodynamics effect, specifically:

- the induction of enhanced diurnal EPO secretion from a single dose;
- an increase in new RBC production by day 5 of dosing; and
- an increase in hemoglobin levels by day 10 of dosing.

Subsequently, we demonstrated a similar induction of a diurnal EPO response in CKD patients. This was followed by a 28 day, dose-titration study to establish the necessary dosing information for increasing hemoglobin levels. Throughout these studies, AKB-6548 was generally well tolerated. There were no serious adverse events, or SAEs, and treatment emergent adverse events, or TEAEs, were limited in number and duration.

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The individual design and summary results of each of our completed clinical trials are highlight below:

Study	Study Design			Subjects Treated		Key Findings
	Subject	Design	Dose, Duration ¹	AKB-6548	Placebo	
Phase 1 CI-0001	Healthy males	Double-blind, placebo-controlled, fasted	80 mg, 160 mg, 300 mg, 600 mg, 900 mg, 1200 mg; single dose	6 (80 mg) 6 (160 mg) 6 (300 mg) 6 (600 mg) 6 (900 mg) 6 (1200 mg)	12 (2 per cohort)	AKB-6548 was well tolerated, and dose responsive increases in EPO levels were demonstrated following a single dose. Half-life of the compound was measured at approximately 4.8 hours. Ten subjects had an AE (seven in the AKB-6548 group and three in the placebo group). No SAEs were reported.
CI-0002	Healthy males	Double-blind, placebo-controlled, fasted	500 mg, 700 mg, 900 mg; 10 days	8 (500 mg) 9 (700 mg) 8 (900 mg)	9 (3 per cohort)	AKB-6548 was well tolerated, and dose responsive increases in reticulocytes and hemoglobin levels were demonstrated. It was also shown that EPO levels returned to baseline by 24 hours following each dose. 26 subjects reported a TEAE. These were evenly distributed across dosing groups. No SAEs were reported.
CI-0006	Healthy males	Randomized, cross-over bioavailability study, fasted	315 mg; single dose of capsule and tablet, with three days between doses	8	0	Both capsules and tablets were well tolerated following a single dose, and shown to be bioequivalent. Six subjects had AEs considered related to study drug. No SAEs were reported.
Phase 2 CI-0003	CKD, Stages 3 & 4	Open-label, fed	500 mg; single dose	22	0	Following a single dose of 500 mg of AKB-6548, the changes in EPO levels followed a similar pattern as that observed in the Phase 1 study at 600 mg in healthy volunteers (CI-0001). In these subjects with CKD, peak levels of EPO were similar to healthy male volunteers, and the half-life was modestly longer at 7.9 hours. Dosing was well tolerated. Five subjects had AEs considered related to study drug. No SAEs were reported.
CI-0004	CKD, Stages 3 & 4	Open-label	Within subject, dose escalation (potential doses of 200 mg, 300 mg, 400 mg, 500 mg, 600 mg, and 700 mg); 28 days of dosing	10	0	In this study, subjects started at 300 mg (CKD 4) or 400 mg (CKD 3). Dose adjustments could be made weekly based on reticulocyte count and hemoglobin data. Dosing was well tolerated. Average hemoglobin levels rose from 9.91 g/dL at baseline to 10.54 g/dL by Day 29. Three subjects had TEAEs considered related to study drug. No SAEs were reported.
CI-0005	CKD, Stages 3, 4 & 5, not on dialysis	Double-blind, placebo-controlled	240 mg, 370 mg, 500 mg, 630 mg; 42 days of dosing	18 (240 mg) 18 (370 mg) 17(500 mg) 19 (630 mg)	19	Dosing was well tolerated. AKB-6548 significantly increased hemoglobin levels in subjects compared to baseline in all dose groups and compared to placebo. The hemoglobin increase occurred without increasing pre-dose EPO levels (prior to daily AKB-6548 dose). Ten subjects had AEs considered related to study drug. There were eight reported SAEs in separated subjects which were all considered unrelated to study drug.
CI-0008	Healthy volunteers	Mass Balance	650 mg; single dose	6	0	Though the final study report is not yet complete, the preliminary data supported earlier findings from human and animal studies. The drug was generally well tolerated during this study.
CI-0009	End-stage renal disease (ESRD)	Pharmacokinetics	450 mg dose four hours prior to start of a dialysis session; 450 mg dose two hours after completion of a different dialysis session	12	0	During the study, dosing of the drug was well tolerated, and there was only one SAE, which was considered unrelated to AKB-6548.

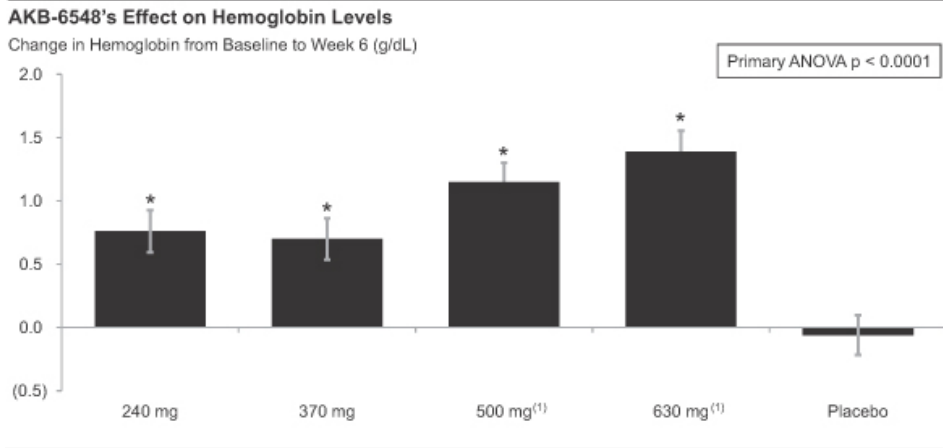
¹ All doses were administered orally, once-daily.

CI-0005: Positive Phase 2a Proof of Concept Trial

CI-0005 was designed to confirm the findings of the early clinical studies and to demonstrate efficacy in CKD patients. In November 2012, we presented at the American Association of Nephrology the results of a randomized, double-blind, placebo controlled trial of AKB-6548 in patients with CKD stages 3, 4 and 5 (not on dialysis) to evaluate the change in hemoglobin levels over 42 days at multiple dose levels. The study enrolled 93 patients with CKD stages 3, 4, or 5 (not on dialysis) who initiated treatment with either placebo or AKB-6548 in the following dose groups: 240 mg, 370 mg, 500 mg, or 630 mg once-daily for 42 days. Depending upon hemoglobin response, patients may have had their initial dose titrated to avoid too rapid of a rise in hemoglobin levels.

At Day 42, AKB-6548 significantly increased hemoglobin levels in a dose-dependent manner compared to baseline in all dose groups. Important findings included:

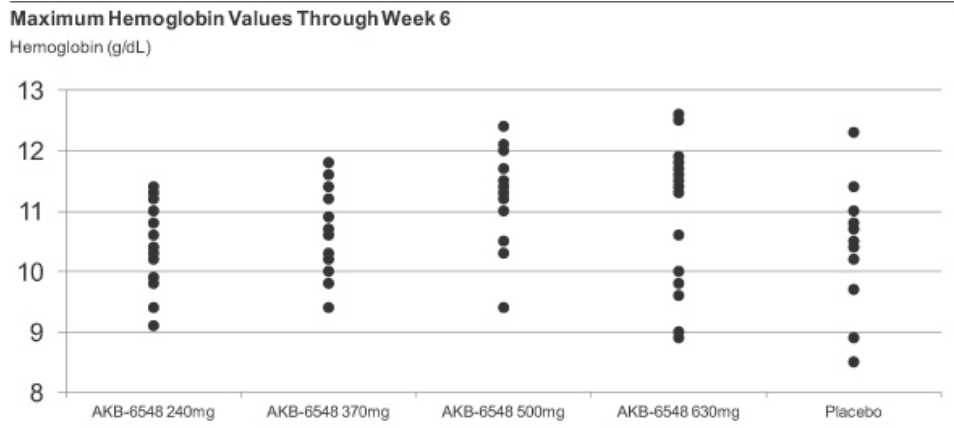
1. AKB-6548 treated patients experienced a statistically significant mean increase in hemoglobin, ranging from 0.7 to 1.4 g/dL by Day 42, while placebo-treated patients experienced a small mean decrease in hemoglobin of 0.1 g/dL. The average baseline hemoglobin level was 9.8 g/dL.



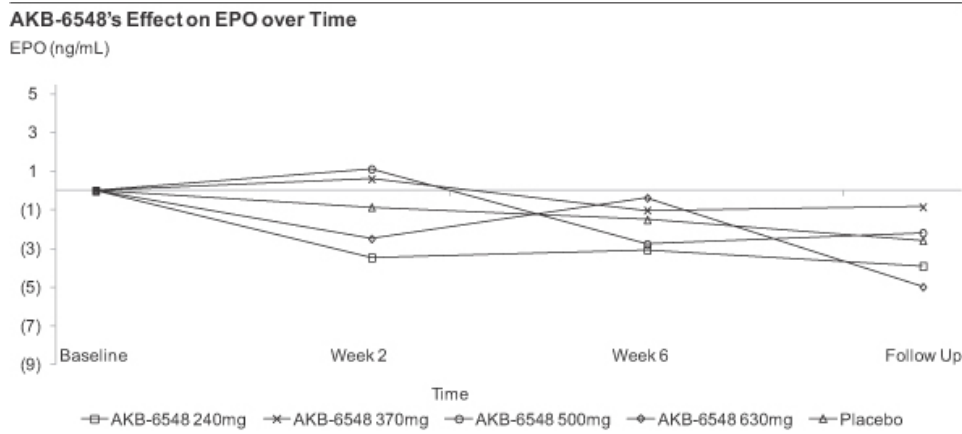
(1) 25% of patients in 630mg and 10% of patients in 500mg had their doses reduced by Week 4.

* Two tailed paired t-test of Hemoglobin: Baseline vs. Week 6 p < 0.01.

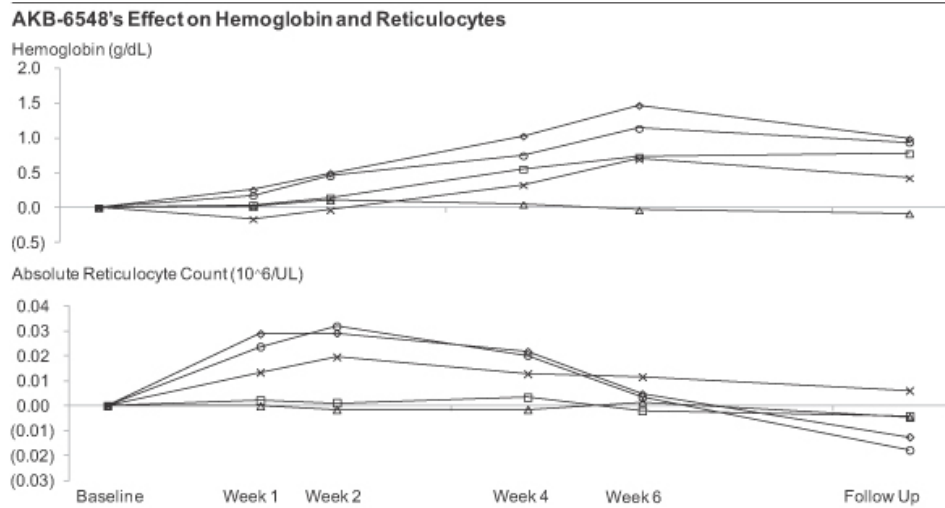
- No patient's measured hemoglobin level exceeded 13 g/dL throughout the study period.



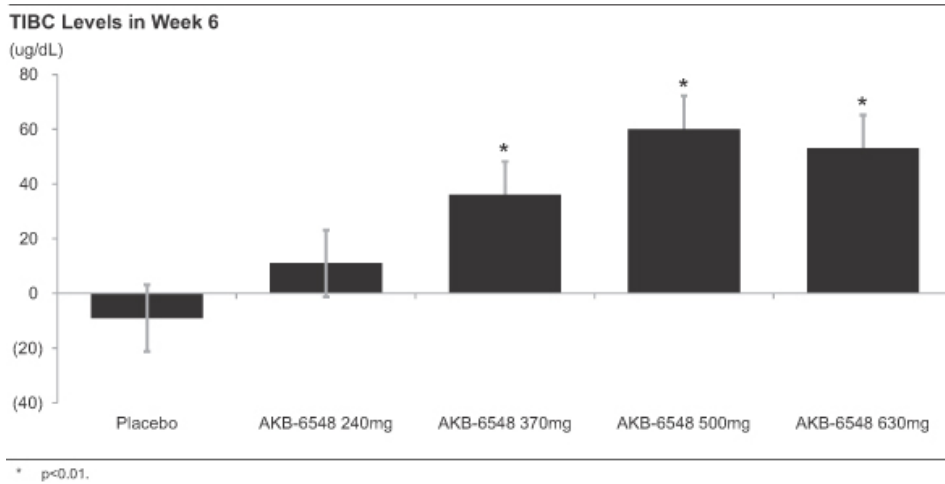
- The dose-dependent increases in hemoglobin occurred even though 26% of patients in the 630 mg dose and 11% of patients in the 500 mg dose decreased their dose, per protocol, as a result of a hemoglobin increase of greater than 1.5 g/dL or more by Day 28.
- The increase in hemoglobin levels occurred without increasing pre-dose EPO levels (prior to daily AKB-6548 dose), demonstrating that AKB-6548 is able to improve RBC production without chronically elevating the body's EPO levels.



5. The increase in hemoglobin levels was preceded by an increase in reticulocytes showing that an increase in hemoglobin levels is a result of a physiologic increase in RBC production.



6. A dose-related increase in TIBC indicated enhanced ability to stabilize the iron supply to the bone marrow while improving hemoglobin production, as shown below with the dose-dependent increase in TIBC.



AKB-6548 was generally well tolerated in the 91 subjects who received study drug. In total, 45 subjects had an AE: 34 (47.2%) in the AKB-6548 groups and 11 (57.9%) in the placebo group. AEs were evenly distributed across the dosing groups with no apparent dose related effect. Ten subjects (13.9%) treated with AKB-6548 and one placebo subject (5.3%) had AEs that were considered study drug related.

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There were eight SAEs in separate subjects which were all considered unrelated to the study drug by the study investigators; seven in the AKB-6548 groups (9.7%) and one in the placebo group (5.3%). These included fluid overload (placebo patient), gastroenteritis, hypoglycemic event, dizziness, triple vessel coronary artery disease with non-ST elevation myocardial infarction, hypertensive crisis, ventricular pacemaker lead replacement, and azotemia (uremia). One subject, who we believe received only three or four doses of study drug, died after being hospitalized for uremia. The subject's death occurred several days into her hospitalization following an in-hospital procedure when she developed sustained ventricular tachycardia and cardiac arrest. The subject's death was not considered to be related to AKB-6548. All other subjects recovered.

VEGF is necessary for the maintenance of healthy kidney function and is regulated by HIF1a. Clinical studies have shown that increased VEGF levels are potentially linked to increased growth of tumors in patients with cancer. AKB-6548 provides for the ability to induce a more prominent HIF2a response, and consistent with this mechanism, no statistically significant change in VEGF levels were observed from baseline for any of the AKB-6548 dose groups.

We also found no statistically significant change in inflammation (C-reactive protein), renal function (Cystatin-C), heart rate, blood pressure and EKG values (including QT assessments).

Ongoing and Planned Clinical Trials

Phase 2b Study (CI-0007)

We are currently enrolling a Phase 2b study of AKB-6548 in subjects with anemia (hemoglobin \leq 10.5 g/dL) secondary to CKD not requiring dialysis. This double-blind, randomized, placebo controlled study will evaluate the efficacy and safety of AKB-6548 in 200 subjects across 62 U.S. sites. The study will enroll patients who have never received rESA therapy, patients previously treated with rESAs, and patients actively treated with rESAs. Patients will initiate treatment with either 450mg of AKB-6548 or placebo once-daily for 20 weeks. The dose of AKB-6548 will be adjusted in accordance with the patient's hemoglobin response. The primary purpose of this study is to demonstrate an adaptive approach to dosing AKB-6548 that will enable subjects to appropriately raise their hemoglobin from baseline without excessive excursions to greater than 13.0 g/dL. Subjects will be extensively evaluated for clinical and laboratory safety, changes in specific biomarkers, and changes in quality-of-life and neuro-cognitive outcomes. We expect that the results for CI-0007 will enable the final design for Phase 3 studies of AKB-6548. It is anticipated that CI-0007 will be fully enrolled by the second quarter of 2014, and we expect that top line results will be announced in the fourth quarter of 2014.

Patients will be assigned in a double-blind fashion in a 2:1 ratio to either AKB-6548 or placebo. After initiating treatment at 450 mg, the dose will be adjusted in accordance with the protocol defined "Dose Adjustment Guidelines and Algorithm." The primary endpoint of our study is the percent of subjects who either (i) achieve or maintain a mean hemoglobin of \geq 11.0 g/dL, or (ii) increase their hemoglobin by \geq 1.2 g/dL over their pre-dose average hemoglobin between screening and baseline. Subjects who receive injectable rESA or transfusion rescue will be counted as failures and subjects receiving transfusion for a non-rescue reason will be removed from the primary analysis. Patients will also be analyzed for safety, including AEs, vital signs, electrocardiograms, and laboratory assay results.

Additional assessments to be conducted during our Phase 2b study include: iron metabolism (changes from baseline in iron, transferrin saturation (TSAT), TIBC, and ferritin); the dose of iron replacement needed to maintain iron levels; actual values and change from baseline in reticulocyte hemoglobin content, HbA1c, and lipids; functional biomarkers; concentration measurements of AKB-6548 and its glucuronide metabolite and measures of neurocognitive functioning and patient reported outcomes.

Studies of AKB-6548 in Dialysis Patients

We plan to initiate a multiple dose, open label Phase 2 study in approximately 60 subjects on dialysis in the first half of 2014. The primary endpoint will compare the change in hemoglobin from baseline for two different doses of AKB-6548 given three times weekly following hemodialysis: 1) 450 mg per dose and 2) 300 mg per dose. The first analysis of change in hemoglobin will be carried out at Week 8, and the second analysis will assess the subsequent change in hemoglobin with dose adjustment starting at Week 8. Key secondary endpoints will include (i) the safety of AKB-6548 in ESRD subjects on dialysis; (ii) the total dose of IV iron therapy for the eight weeks prior to baseline to the first (Weeks 1-8) and second (Weeks 9-16) eight weeks of treatment; and (iii) the effect of dialysis on the pharmacokinetics of AKB-6548.

Projected Phase 3 Clinical Trials

Upon completion of our Phase 2b study, and if we receive positive feedback from the FDA, we intend to initiate our Phase 3 studies. The endpoints, duration, and size of these Phase 3 trials will be based on those used in the Omontys (Peginesatide) approval studies, with modifications to adjust for the shift in focus to CKD. The primary two studies will be double-blind, randomized, and placebo controlled. The anticipated goal of anemia management in these studies will be to raise hemoglobin levels to greater than 10.5 g/dL and include a rescue component for subjects with declining hemoglobin that uses injectable rESAs in accordance with existing guidelines. In this manner, AKB-6548 will be compared to the existing standard of care for both efficacy and safety. The principle requirement for safety will be to demonstrate non-inferiority for cardiovascular safety of AKB-6548 relative to the standard of care provided to the placebo group. We are designing these clinical studies to be applicable for global development with limited protocol differences between geographic regions. The total number of subjects to be enrolled in the Phase 3 studies will be determined upon agreement with FDA, EMA and other regulatory authorities.

Although the exact size and timing cannot be known until final agreement is reached with the FDA, EMA and other regulatory authorities, we estimate that the Phase 3 studies for the indication of anemia secondary to CKD (not including dialysis) will require approximately three studies and include a total of 2,000 subjects. We estimate that the studies will be two years in duration, with an average subject duration on study drug of 1.25 years.

Additional Studies

Prior to initiating the Phase 3 studies, we intend to complete a thorough QT, or TQT, study in accordance with FDA guidance to ensure that AKB-6548 does not affect the cardiac conduction cycle. A lengthened QT interval is a biomarker for certain ventricular arrhythmias and a risk factor for sudden death. To date, AKB-6548 has not shown any tendency to affect the QT interval either in humans or animals. We plan to initiate this study in approximately 50 normal volunteers in the first quarter of 2014.

To test AKB-6548 in a chronic dosing setting, carcinogenicity assessments in two rodent species (rat and mouse) will be pursued. AKB-6548 has been shown to be orally bioavailable and pharmacologically active in both species. The results of a standard battery of tests that evaluate for mutations in cells or animals have indicated that AKB-6548 does not cause mutations that could lead to cancer. However, to satisfy the expected regulatory requirement, two-year carcinogenicity assessments in each of the two rodent species will be conducted. Completion of three-month (mouse; ongoing) and six-month (rat; completed) oral toxicity evaluations will support dose selection for the respective two-year carcinogenicity assessment.

Finally, in order to complete the registration package for drug approval, we are exploring the need to evaluate specific drug interactions with patients taking AKB-6548, as patients with CKD take multiple medications. It is likely we will conduct at least one of these additional clinical studies.

Additional Indications

The two major additional indications for AKB-6548 are anemia associated with aging (also known as IAA) and anemia secondary to congestive heart failure, or CHF. AKB-6548, with its different mechanism of action, offers a completely new approach to these large markets. Both occur in very large segments of the population and are associated with considerable morbidity and mortality. Anemia affects approximately 10% of individuals age 65 and over, and in those individuals age 85 and older, the percentage is greater than 20%, according to a paper published in 2004 in the peer-reviewed journal *Blood*. Of these, approximately one-third are considered to be IAA. Other causes of anemia in this population include CHF, CKD and nutritional deficiencies. Because injectable rESAs are currently associated with increased cardiovascular events, they have not been successful entering either of these markets.

We anticipate studying both indications by using a fixed, low-dose therapeutic approach which would enable a modest increase in hemoglobin, and minimize the requirement for follow-up assessments. Although we will have extensive dosing information from the CKD studies, additional Phase 2 studies will need to be performed to evaluate the required dose level. In addition, the Phase 2 studies will evaluate cardiac performance and other outcomes that will be critical in Phase 3. It is likely that the study in IAA would be undertaken first, as the mechanism of action of AKB-6548 is well supported by the scientific literature in IAA. Specifically, AKB-6548 is expected to stabilize the limited production of HIF2a in older patients. The primary outcome for this study will focus on quality-of-life outcomes, such as the ability of a subject to perform activities of daily living. In addition, the study will need to evaluate standard measures of morbidity and mortality. We expect to initiate the study following the analysis of the Phase 2b study results, particularly the performance of the drug in patients over the age of 70.

AKB-6899

AKB-6899, another HIFa-stabilizing compound, is a very close relative of AKB-6548. In screening AKB-6899 for its HIF-related properties, it was discovered that in cells cultured at low oxygen levels, AKB-6899 significantly inhibited the expression of VEGF and phosphoglycerate kinase, or PGK, mRNA, both of which are associated with the growth of cancerous tumors. In addition, AKB-6899 was found to significantly stimulate the production of soluble vascular endothelial growth factor receptor 1, or sVEGFr1. sVEGFr1 is known to be a potent inhibitor of VEGF signaling by sequestering VEGF and inhibiting its interaction with transmembrane receptors—in so doing, sVEGFr1 can inhibit the growth of certain types of cancer cells. AKB-6899 was also found to stimulate the production of EPO in a manner similar to AKB-6548.

These properties, and others, indicate that AKB-6899 may be an excellent treatment for certain cancers (ovarian, breast, colon, and possibly lung), that could be given in combination with other types of chemotherapy. In addition AKB-6899 may also be a candidate compound for the treatment of chemotherapy-induced anemia and for VEGF-related eye diseases. AKB-6899 has been used effectively in several animal models of cancer, both alone and in combination. In addition, it has been shown to be effective in animal models of colitis.

Manufacturing and Supply

AKB-6548 is a small-molecule drug that is manufactured from readily available commercial starting materials. The manufacturing of AKB-6548 uses standard chemical technologies and equipment. The intended commercial manufacturing route has been successfully scaled up and has produced approximately 40 kg of AKB-6548 drug substance. The drug substance can be readily formulated into compressed tablets using standard USP grade excipients. We have made compressed tablets of varying sizes with no apparent effect on dissolution profile or bioavailability.

The preclinical candidate AKB-6899 has been produced on laboratory scale, but clinical or commercial manufacturing has not been investigated. Based on the similarity of the structure to AKB-6548, similar commercially available starting materials and commercial manufacturing process can be expected.

We have no internal manufacturing capabilities and rely on outside manufacturers to produce all lots of drug substance and drug products. A reputable and well-known U.S.-based contract manufacture facility has been identified to manufacture the required drug substance AKB-6548 for clinical trials and, potentially, commercialization. A high quality U.S.-based manufacturer has been identified to supply drug product for remaining clinical trials. Current tableting methods are amenable for scale up to commercial quantities of drug product. To date, AKB-6548 has been manufactured under strict cGMP regulations and we believe has fully complied with the FDA guidelines for the manufacture of drug substance and drug product used in clinical trials.

Intellectual Property

The proprietary nature of, and protection for, our product candidates and our discovery programs, processes and know-how are important to our business. Our policy is to seek to protect our proprietary position by, among other methods, filing U.S. and foreign patent applications related to our proprietary technology, inventions and improvements that are important to the development and implementation of our business. We also rely on know-how, continuing technological innovation and potential in-licensing opportunities to develop and maintain our proprietary position. Additionally, we may benefit from a variety of statutory frameworks in the United States, Europe and other countries that provide periods of non-patent-based exclusivity for qualifying molecules. See “—Regulatory Matters.”

Our commercial success will depend in part on obtaining and maintaining patent protection of our current and future product candidates, methods of their use and the methods used to develop and manufacture them, as well as successfully defending these patents against third-party challenges. Our ability to stop third parties from making, using, selling, offering to sell or importing our products depends on the extent to which we have rights under valid and enforceable patents that cover these activities. We cannot be sure that patents will be granted with respect to any of our pending patent applications or with respect to any patent applications filed by us in the future, nor can we be sure that any of our existing patents or any patents that may be granted to us in the future will be commercially useful in protecting our product candidates, discovery programs and processes. Even once patents successfully issue, third parties may challenge the validity, enforceability, inventorship, or scope thereof, which may result in such patents being narrowed, invalidated or held unenforceable. For this and more comprehensive risks related to our intellectual property, please see “Risk Factors—Risks Related to Our Intellectual Property.”

Our patent estate, on a worldwide basis, includes 18 allowed applications and issued patents and approximately 39 pending utility and provisional patent applications, with pending and issued claims relating to our current clinical stage candidate AKB-6548 as well as other product candidates, including AKB-6899. We also hold three patents that claim the crystal of a protein-ligand complex of EGLN-1 as well as methods for identifying compounds that bind to EGLN-1.

Individual patents extend for varying periods of time depending on the date of filing of the patent application or the date of patent issuance and the legal term of patents in the countries in which they are obtained. Generally, patents issued from applications filed in the United States are effective for twenty years from the earliest non-provisional filing date. In addition, in certain instances, a patent term can be extended to recapture a portion of the term effectively lost as a result of the FDA regulatory review period, however, the restoration period cannot be longer than five years and the total patent term including the restoration period must not exceed 14 years following FDA approval. The duration of foreign patents varies in accordance with provisions of applicable local law, but typically is also twenty years from the earliest international filing date. Our issued patents and pending applications with respect to our composition of matter, methods of treatment, and pharmaceutical compositions are expected to expire in 2027 or 2028 (depending on eligibility for patent term adjustment) and our pending applications with respect to processes for manufacturing AKB-6548, dosing regimens, formulations, and various other aspects relating to the treatment of anemia using AKB-6548 are expected to expire between 2032 and 2034, exclusive of possible patent term adjustments or extensions; however, the actual protection afforded by a patent varies on a product by product basis, from country to country

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and depends upon many factors, including the type of patent, the scope of its coverage, the availability of extensions of patent term, the availability of legal remedies in a particular country and the validity and enforceability of the patent.

Changes in either the patent laws or interpretations of patent laws in the United States and other countries can diminish our ability to protect our inventions and enforce our intellectual property rights. Accordingly, we cannot predict the breadth or enforceability of claims that may be granted in our patents or in third-party patents. The biotechnology and pharmaceutical industries are characterized by extensive litigation regarding patents and other intellectual property rights. Our ability to maintain and solidify our proprietary position for our drugs and technology will depend on our success in obtaining effective claims and enforcing those claims once granted. We do not know whether any of the patent applications that we may file or license from third parties will result in the issuance of any patents. The issued patents that we own or may receive in the future, may be challenged, invalidated or circumvented, and the rights granted under any issued patents may not provide us with sufficient protection or competitive advantages against competitors with similar technology. Furthermore, our competitors may be able to independently develop and commercialize similar drugs or duplicate our technology, business model or strategy without infringing our patents. Because of the extensive time required for clinical development and regulatory review of a drug we may develop, it is possible that, before any of our drugs can be commercialized, any related patent may expire or remain in force for only a short period following commercialization, thereby reducing any advantage of any such patent. The patent positions for our most advanced programs are summarized below.

AKB-6548 Patent Portfolio

We hold four issued patents and one pending application covering the composition of matter, method of treating anemia, and pharmaceutical compositions of AKB-6548 in the United States, one issued patent in Europe (registered in most countries of the European Patent Convention), and additional patents issued or pending in many other major jurisdictions worldwide, including Japan, China, South Korea, Brazil, Mexico, Russia, Israel and India. The expected expiration date for these composition of matter patents is 2027 plus any extensions or adjustments of term available under national law.

In July of 2011, a third party filed an opposition to our issued European Patent No. 2044005 (the '005 Patent). During the oral proceedings, which took place on April 10, 2013, the Opposition Division of the European Patent Office maintained the '005 Patent on the basis of the third auxiliary request filed during the oral proceedings. This decision resulted in the maintenance of a claim directed to a compound chosen from a group of eight compounds, including AKB-6548, as well as claims to compositions and methods for treating various diseases, including, but not limited to, anemia. Both parties have appealed the decision of the Opposition Division and final resolution of the opposition proceedings will likely take a number of years. We cannot be assured of the breadth of the claims that will remain in the '005 Patent or that the patent will not be revoked in its entirety.

We also hold patents and patent applications directed to processes for manufacturing AKB-6548, dosing regimens, formulations, polymorphs, and various other aspects relating to the treatment of anemia using AKB-6548 that are expected to expire between 2032 and 2034 exclusive of possible patent term extensions.

AKB-6899 Patent Portfolio

We hold two issued patents and one pending application covering the AKB-6899 composition of matter and pharmaceutical compositions in the United States, and additional patents issued or pending in many other major jurisdictions worldwide, including Europe, Japan, China, South Korea, Brazil, Mexico, Russia and India. The expected expiration date for these composition of matter patents is 2027 plus any extensions or adjustments of term available under national law.

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We hold one issued patent that covers the treatment of anemia by administration of AKB-6899, which is expected to expire in 2028. We also hold, either alone or jointly, three pending applications covering various methods, including, but not limited to, the treatment of cancer or chemotherapy-induced anemia by administration of AKB-6899 in the United States. The expected expiration dates for these method of treatment patent applications are expected to be either 2027 or 2032 exclusive of possible patent term extensions or adjustments.

Know-How

In addition to patents, we rely upon unpatented know-how and continuing technological innovation to develop and maintain our competitive position. We seek to protect our proprietary information, in part, using confidentiality agreements with our collaborators, employees and consultants and invention assignment provisions in the confidentiality agreements with our employees. These agreements are designed to protect our proprietary information and, in the case of the invention assignment provisions, to grant us ownership of technologies that are developed by our employees. These agreements may be breached, and we may not have adequate remedies for any breach.

To the extent that our commercial partners, collaborators, employees and consultants use intellectual property owned by others in their work for us, disputes may arise as to the rights in related or resulting know-how and inventions.

Third Party Filings

We are aware of certain U.S. patents issued to FibroGen, directed to, among other things, methods of treating or affecting certain specified conditions in subjects. Such method-of-use patents protect the use of a product for the specified method. This type of patent does not prevent a competitor from making and marketing the product for an indication that is outside the scope of the patented method. We are not aware of any U.S. Patents issued to FibroGen that claim methods of using any of our product candidates for purposes of inhibiting HIF-PHs for the treatment of anemia secondary to CKD.

In addition, we are aware of certain foreign patents owned by FibroGen. For example, in June 2013, the European Patent Office granted European Patent No. 1463823 (the '823 patent) to FibroGen. The '823 patent claims, among other things, the use of a heterocyclic carboxamide compound selected from the group consisting of pyridine carboxamides, quinoline carboxamides, isoquinoline carboxamides, cinnoline carboxamides, and beta-carboline carboxamides that inhibits HIF-PH enzyme activity in the manufacture of a medicament for increasing endogenous EPO in the prevention, pretreatment, or treatment of anemia. On December 5, 2013, we filed an opposition with the European Patent Office to the '823 patent requesting that the '823 patent be revoked in its entirety. While, for the reasons set forth in our opposition, we believe the '823 patent should be revoked in its entirety, the ultimate outcome of the opposition remains uncertain.

Competition

We operate in highly competitive segments of the biotechnology and biopharmaceutical markets. We face competition from many different sources, including commercial pharmaceutical and biotechnology enterprises, academic institutions, government agencies and private and public research institutions. Many of our competitors have significantly greater financial, product development, manufacturing and marketing resources than us. Large pharmaceutical companies have extensive experience in clinical testing and obtaining regulatory approval for drugs. These companies also have significantly greater research capabilities than us. Many universities and private and public research institutes are active in CKD research, some in direct competition with us. Smaller or early-stage companies may also prove to be significant competitors, particularly through collaborative arrangements with large and established companies. The key competitive factors affecting the success of AKB-6548, if approved, are likely to be its efficacy, convenience and safety profile.

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If AKB-6548 is approved and launched commercially, competing drugs will include EPOGEN and potentially Aranesp, which are both marketed by Amgen, Inc., or Amgen, in addition to Procrit and Eprex, which are marketed by Johnson & Johnson. Aranesp, introduced in 2001, has significant market share in the U.S., particularly in the oncology and the non-dialysis markets, although it is approved for treatment in dialysis patients as well. In Europe, Roche has obtained regulatory approval to market, and has launched, a PEGylated rESA called Mircera. Mircera reportedly has greater plasma stability than any of the currently marketed products. PEG is a polymer that increases the time rEPO remains in the circulation and consequently can be dosed less frequently. Mircera has also obtained regulatory approval in the U.S., but as a result of Roche and Amgen's patent infringement litigation, Mircera was found to infringe several U.S. patents owned by Amgen and has been enjoined from being sold in the U.S. until mid-2014 under the terms of a limited license. If Mircera enters the U.S. market, we believe it will be in direct competition with AKB-6548 because of Mircera's ability to be long-acting; therefore, it could potentially limit the market for AKB-6548.

We may also face competition from potential new anemia therapies if we obtain approval for and commercially launch AKB-6548. There are several other HIF product candidates for anemia indications in various stages of development by potential competitors. These candidates are being developed by companies such as FibroGen, AstraZeneca, GlaxoSmithKline and Bayer, all of whom are likely to have greater financial resources than our company. FibroGen, in particular, is ahead of us in the clinical development of its product, FG-4592 (roxadustat). Such HIF compounds under development may have a mechanism of action that is the same or similar to AKB-6548 and promote the production of naturally occurring EPO in patients. Some of these product candidates may enter the market as early as 2015 or 2016. If these product candidates enter the market, they may compete with AKB-6548, if it is approved and marketed.

In addition, certain companies are developing potential new therapies for renal-related diseases that could potentially reduce rESA utilization and thus limit the market for AKB-6548 if it is approved and marketed.

The introduction of biosimilars into the rEPO market in the U.S. will constitute additional competition for AKB-6548 if it is approved and marketed. A biosimilar product is a subsequent version of an existing, branded biologic product. The patent for the existing, branded product must expire in a given market before biosimilars may enter that market. The patents for epoetin alfa, a version of rEPO, expired in 2004 in the European Union, and the remaining patents have expired or will expire in 2012 through 2015 in the U.S. Several biosimilar versions of rEPO are available for sale in the European Union and biosimilar versions of rEPO are currently being studied in clinical trials in the United States.

For example, in January 2012, Hospira, Inc. announced the beginning of its Phase 3 clinical program for its biosimilar rEPO with results anticipated in 2013, and in October 2012, Sandoz announced the beginning of its Phase 3 clinical program for its biosimilar rEPO with results anticipated in 2014. Upon entry into the U.S. market, biosimilars will compete with AKB-6548 if it is approved and marketed, and will likely drive down prices for rEPO, which could also adversely affect our reimbursement.

In the dialysis market, it is typical to compete for and enter into long-term supply agreements with the major operators of dialysis clinics in the U.S. In particular, two of the largest operators of dialysis clinics in the U.S., DaVita Inc., or DaVita, and Fresenius, account for more than half of the rESA sales in the U.S. dialysis market. Both DaVita and Fresenius entered into a long-term supply agreement with Amgen that began in January 2012. We believe that it may be challenging to enter into or expand upon long or short-term supply agreements with DaVita, Fresenius or other operators of dialysis clinics.

Regulatory Matters

The FDA and comparable regulatory authorities in state and local jurisdictions and in other countries impose substantial and burdensome requirements upon companies involved in the clinical development, manufacture, marketing, labeling and packaging storage, distribution, post-approval monitoring and reporting,

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advertising and promotion, pricing and export and import of drugs. The process of obtaining regulatory approvals and the subsequent compliance with appropriate federal, state, local and foreign statutes and regulations require the expenditure of substantial time and financial resources. Moreover, failure to comply with applicable regulatory requirements may result in, among other things, warning letters, clinical holds, civil or criminal penalties, recall or seizure of products, injunction, disbarment, partial or total suspension of production or withdrawal of the product from the market. Any agency or judicial enforcement action could have a material adverse effect on us.

U.S. Government Regulation

In the United States, the FDA regulates drug products under the Federal Food, Drug, and Cosmetic Act, or FFDCFA, and the FDA's implementing regulations. If we fail to comply with applicable FDA or other requirements at any time during the drug development process, clinical testing, the approval process or after approval, we may become subject to administrative or judicial sanctions. These sanctions could include the FDA's refusal to approve pending applications, license suspension or revocation, withdrawal of an approval, warning letters, product recalls, product seizures, total or partial suspension of production or distribution, injunctions, fines, civil penalties or criminal prosecution. Any FDA enforcement action could have a material adverse effect on us. FDA approval is required before any new unapproved drug or dosage form, including a new use of a previously approved drug, can be marketed in the United States.

The process required by the FDA before a drug may be marketed in the United States generally involves:

- completion of extensive nonclinical laboratory tests, nonclinical animal studies and formulation studies performed in accordance with the FDA's current Good Laboratory Practice, or cGMP, regulations;
- submission to the FDA of an IND application which must become effective before human clinical trials in the United States may begin;
- approval by an IRB or ethics committee at each clinical trial site before each trial may be initiated;
- performance of adequate and well-controlled human clinical trials to establish the safety and efficacy of the drug candidate for each proposed indication;
- submission to the FDA of an NDA;
- satisfactory completion of an FDA inspection of the manufacturing facility or facilities at which the drug is produced to assess compliance with current cGMP regulations;
- satisfactory completion of a potential review by an FDA advisory committee, if applicable; and
- FDA review and approval of the NDA prior to any commercial marketing, sale or commercial shipment of the drug in the United States.

The nonclinical and clinical testing and approval process requires substantial time, effort and financial resources, and we cannot be certain that any approvals for our product candidates will be granted on a timely basis, if at all. Nonclinical tests include laboratory evaluation of product chemistry, formulation, stability and toxicity, as well as animal studies to assess the characteristics and potential safety and efficacy of the product.

The results of nonclinical tests, together with manufacturing information, analytical data and a proposed clinical trial protocol and other information, are submitted as part of an IND to the FDA. The central focus of an IND submission is on the general investigational plan and the protocol(s) for human studies. Some nonclinical testing may continue even after the IND is submitted, but an IND must become effective before human clinical trials may begin. The IND automatically becomes effective 30 days after receipt by the FDA, unless the FDA, within the 30-day time period, raises concerns or questions relating to one or more proposed clinical trials and places the clinical trial on a clinical hold, including concerns that human research subjects will be exposed to unreasonable health risks. In such a case, the IND sponsor and the FDA must resolve any outstanding concerns

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before the clinical trial can begin. Accordingly, submission of an IND may or may not result in the FDA allowing clinical trials to commence. A separate submission to an existing IND must also be made for each successive clinical trial conducted during product development.

Clinical trials involve the administration of the investigational drug to human subjects under the supervision of qualified investigators in accordance with cGCPs, which include the requirement that all research subjects provide their informed consent for their participation in any clinical trial. Clinical trials are conducted under protocols detailing, among other things, the objectives of the clinical trial, the parameters to be used in monitoring safety and the effectiveness criteria to be used. Each protocol must be submitted to the FDA as part of the IND. In addition, an independent IRB or ethics committee for each medical center proposing to conduct a clinical trial must also review and approve a plan for any clinical trial before it can begin at that center and the IRB must monitor the clinical trial until it is completed. There are also requirements governing the reporting of ongoing clinical trials and clinical trial results to public registries.

A sponsor who wishes to conduct a clinical trial outside the United States may, but need not, obtain FDA authorization to conduct the clinical trial under an IND. If a foreign clinical trial is not conducted under an IND, the sponsor may submit data from the clinical trial to the FDA in support of an NDA so long as the clinical trial is conducted in compliance with an international guideline for the ethical conduct of clinical research known as the Declaration of Helsinki and/or the laws and regulations of the country or countries in which the clinical trial is performed, whichever provides the greater protection to the participants in the clinical trial.

Clinical Trials

Clinical trials are typically conducted in three or four phases, which may overlap or be combined:

- Phase 1: Clinical trials are initially conducted in a limited population of subjects to test the drug candidate for safety, dose tolerance, absorption, metabolism, distribution and excretion in healthy humans or, on occasion, in patients with severe problems or life threatening diseases to gain an early indication of its effectiveness.
- Phase 2: Clinical trials are generally conducted in a limited patient population to evaluate dosage tolerance and appropriate dosage, identify possible adverse effects and safety risks, and evaluate preliminarily the efficacy of the drug for specific targeted indications in patients with the disease or condition under study.
- Phase 3: Clinical trials are typically conducted when Phase 2 clinical trials demonstrate that a dose range of the product candidate is effective and has an acceptable safety profile. Phase 3 clinical trials are commonly referred to as “pivotal” studies, which typically denotes a study which presents the data that the FDA or other relevant regulatory agency will use to determine whether or not to approve a drug. Phase 3 clinical trials are generally undertaken with large numbers of patients, such as groups of several hundred to several thousand, to further evaluate dosage, to provide substantial evidence of clinical efficacy and to further test for safety in an expanded and diverse patient population at multiple, geographically-dispersed clinical trial sites.
- Phase 4: In some cases, FDA may condition approval of an NDA for a product candidate on the sponsor’s agreement to conduct additional clinical trials after NDA approval. In other cases, a sponsor may voluntarily conduct additional clinical trials post-approval to gain more information about the drug. Such post approval trials are typically referred to as Phase 4 clinical trials.

The FDA, the IRB, or the sponsor may suspend or discontinue a clinical trial at any time on various grounds, including a finding that the subjects are being exposed to an unacceptable health risk. Additionally, some clinical trials are overseen by an independent group of qualified experts organized by the clinical trial sponsor, known as a DSMB or committee. This group provides authorization for whether or not a trial may move forward at designated check points based on access to certain data from the study. A sponsor may also suspend or terminate a clinical trial based on evolving business objectives and/or competitive climate.

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Concurrent with clinical trials, companies usually complete additional animal trials and must also develop additional information about the chemistry and physical characteristics of the drug and finalize a process for manufacturing the drug in commercial quantities in accordance with cGMP requirements. The manufacturing process must be capable of consistently producing quality batches of the drug candidate and, among other things, the manufacturer must develop methods for testing the identity, strength, quality and purity of the final drug product. Additionally, appropriate packaging must be selected and tested and stability studies must be conducted to demonstrate that the drug candidate does not undergo unacceptable deterioration over its shelf life.

New Drug Applications

The results of nonclinical studies and of the clinical trials, together with other detailed information, including extensive manufacturing information and information on the composition of the drug, are submitted to the FDA in the form of an NDA requesting approval to market the drug for one or more specified indications. The FDA reviews an NDA to determine, among other things, whether a drug is safe and effective for its intended use.

Once the NDA submission has been accepted for filing, under the Prescription Drug User Fee Act (PDUFA), the FDA has a goal of responding to standard review NDAs within ten months after the 60-day filing review period, but this timeframe is often extended. The first indication of the FDA's review progress is provided at the mid-cycle review. This typically occurs five months after the NDA is submitted. However, the review process is often significantly extended by FDA requests for additional information or clarification. The FDA may refer the application to an advisory committee for review, evaluation and recommendation as to whether the application should be approved. The FDA is not bound by the recommendation of an advisory committee, but it generally follows such recommendations.

Before approving an application, the FDA will inspect the facility or the facilities at which the finished drug product, and sometimes the active drug ingredient, is manufactured, and will not approve the drug unless cGMP compliance is satisfactory. The FDA may also inspect the sites at which the clinical trials were conducted to assess their compliance, and will not approve the drug unless compliance with GCP requirements is satisfactory.

After the FDA evaluates the NDA and conducts inspections of manufacturing facilities, it may issue an approval letter or a Complete Response Letter. An approval letter authorizes commercial marketing of the drug with specific prescribing information for specific indications. A Complete Response Letter indicates that the review cycle of the application is complete and the application is not ready for approval. A Complete Response Letter may require additional clinical data and/or other significant, expensive, and time-consuming requirements related to clinical trials, preclinical studies and/or manufacturing. Even if such data are submitted, the FDA may ultimately decide that the NDA does not satisfy the criteria for approval. Data from clinical trials are not always conclusive and the FDA may interpret data differently than we interpret data. The FDA could also approve the NDA with a REMS to mitigate risks, which could include medication guides, physician communication plans, or elements to ensure safe use, such as restricted distribution programs, patient registries or other risk minimization tools. The FDA may also condition approval on, among other things, changes to proposed labeling, development of adequate controls and specifications, or a commitment to conduct one or more post-market studies or clinical trials. Such post-market testing may include Phase 4 clinical trials and surveillance to further assess and monitor the product's safety and effectiveness after commercialization.

Once the FDA approves an NDA, or supplement thereto, the FDA may withdraw the approval if ongoing regulatory requirements are not met or if safety problems are identified after the drug reaches the market. Where a withdrawal may not be appropriate, the FDA still may seize existing inventory of such drug or require a recall of any drug already on the market. In addition, the FDA has the authority to prevent or limit further marketing of a drug based on the results of post-market studies or surveillance programs.

After regulatory approval of a drug is obtained, companies are subject to a number of post-approval requirements. For example, there are reporting obligations regarding certain adverse events received and

production problems. Companies are also required to report updated safety and efficacy information and to comply with requirements concerning advertising and promotional labeling. Drugs may be marketed only for the FDA approved indications and in accordance with the provisions of the approved labeling. The FDA closely regulates the post-approval marketing and promotion of drugs, including standards and regulations for direct-to-consumer advertising, off-label promotion, industry-sponsored scientific and educational activities and promotional activities involving the Internet. A company can make only those claims relating to safety and efficacy that are approved by the FDA. Failure to comply with these requirements can result in adverse publicity, warning letters, corrective advertising and potential civil and criminal penalties. Physicians may prescribe legally available drugs for uses that are not described in the product's labeling and that differ from those tested by us and approved by the FDA. Such off-label uses are common across medical specialties. Physicians may believe that such off-label uses are the best treatment for many patients in varied circumstances. The FDA does not regulate the behavior of physicians in their choice of treatments. The FDA does, however, impose stringent restrictions on manufacturers' communications regarding off-label use.

Further, if there are any modifications to the drug, including changes in indications, labeling, or manufacturing processes or facilities, the applicant may be required to submit and obtain FDA approval of a new NDA or NDA supplement, which may require the applicant to develop additional data or conduct additional nonclinical studies and clinical trials. As with new NDAs, the review process is often significantly extended by the FDA requests for additional information or clarification. Also, quality control and manufacturing procedures must continue to conform to cGMP requirements after approval to ensure and preserve the long term stability of the drug product. The FDA periodically inspects manufacturing facilities to assess compliance with cGMP requirements, which imposes extensive procedural, substantive and record keeping requirements. In addition, changes to the manufacturing process are strictly regulated, and, depending on the significance of the change, may require prior FDA approval before being implemented. FDA regulations also require investigation and correction of any deviations from cGMP requirements and impose reporting and documentation requirements upon us and any third-party manufacturers that we may decide to use. Accordingly, manufacturers must continue to expend time, money and effort in the area of production and quality control to maintain compliance with cGMP requirement and other aspects of regulatory compliance.

The testing and approval processes require substantial time, effort and financial resources, and each may take several years to complete. The FDA may not grant approval on a timely basis, or at all. Even if we believe a clinical trial has demonstrated safety and efficacy of one of our drug candidates for the treatment of a disease, the results may not be satisfactory to the FDA. Nonclinical and clinical data may be interpreted by the FDA in different ways, which could delay, limit or prevent regulatory approval. We may encounter difficulties or unanticipated costs in our efforts to secure necessary governmental approvals which could delay or preclude us from marketing drugs. The FDA may limit the indications for use or place other conditions on any approvals that could restrict the commercial application of the drugs.

Europe/Rest of World Government Regulation

In addition to regulations in the United States, we will be subject to a variety of regulations in other jurisdictions governing, among other things, clinical trials and any commercial sales and distribution of our products. Whether or not we obtain FDA approval for a product, we must obtain the requisite approvals from regulatory authorities in foreign countries prior to the commencement of clinical trials or marketing of the product in those countries.

Certain countries outside of the United States have a similar process that requires the submission of a clinical trial application, or CTA, much like the IND prior to the commencement of human clinical trials. In Europe, for example, a CTA must be submitted to each country's national health authority and an independent ethics committee, much like the FDA and IRB, respectively. Once the CTA is approved in accordance with a country's requirements, clinical trial development may proceed.

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The requirements and process governing the conduct of clinical trials, product licensing, pricing and reimbursement vary from country to country. In all cases, the clinical trials are conducted in accordance with cGCP requirements and the applicable regulatory requirements and the ethical principles that have their origin in the Declaration of Helsinki. To obtain regulatory approval of an investigational drug under European Union regulatory systems, we must submit a marketing authorization application. The application used to file the NDA in the United States is similar to that required in Europe, with the exception of, among other things, country-specific document requirements.

For other countries outside of the European Union, such as countries in Eastern Europe, Latin America or Asia, the requirements governing the conduct of clinical trials, product licensing, pricing and reimbursement vary from country to country. In all cases, again, the clinical trials must be conducted in accordance with cGCP requirements and the applicable regulatory requirements and the ethical principles that have their origin in the Declaration of Helsinki.

If we fail to comply with applicable foreign regulatory requirements, we may be subject to, among other things, fines, suspension or withdrawal of regulatory approvals, product recalls, seizure of products, operating restrictions and criminal prosecution.

Special Protocol Assessment

An SPA is a written agreement with the FDA on the details of the design, size, execution and planned analysis for a clinical trial intended to form the primary basis of an effectiveness claim in an NDA. After the clinical trial begins, the agreement may only be changed through a written agreement between the sponsor and the FDA. An SPA is generally binding upon the FDA unless the FDA determines that there are public health concerns unrecognized at the time the SPA agreement was entered into, other new scientific concerns regarding product safety or efficacy arise, or if the sponsor fails to comply with the agreed-upon trial protocol. If the outcome of the clinical trial is successful, the sponsor will ordinarily be able to rely on it as the primary basis for approval with respect to effectiveness.

Fraud and Abuse Laws

In the United States, the research, manufacturing, distribution, sale and promotion of drug products and medical devices are subject to regulation by various federal, state and local authorities in addition to the FDA, including the CMS, other divisions of the U.S. Department of Health and Human Services (e.g., the Office of Inspector General), the U.S. Department of Justice, state Attorneys General, and other state and local government agencies.

These laws include the federal Anti-Kickback Statute, which makes it illegal for any person, including a prescription drug manufacturer (or a party acting on its behalf) to knowingly and willfully solicit, receive, offer, or pay any remuneration that is intended to induce the referral of business, including the purchase, order, or prescription of a particular drug, for which payment may be made under a federal healthcare program, such as Medicare or Medicaid. Violations of this law are punishable by up to five years in prison, criminal fines, administrative civil money penalties, and exclusion from participation in federal healthcare programs. In addition, the PPACA, among other things, amends the intent requirement of the federal Anti-Kickback Statute and criminal healthcare fraud statutes created by HIPAA. A person or entity no longer needs to have actual knowledge of the statute or specific intent to violate it. Moreover, the PPACA provides that the government may assert that a claim including items or services resulting from a violation of the federal anti-kickback statute constitutes a false or fraudulent claim for purposes of the False Claims Act.

Although we would not submit claims directly to payors, drug manufacturers can be held liable under the federal False Claims Act, which prohibits anyone from knowingly presenting, or causing to be presented, for payment to federal programs (including Medicare and Medicaid) claims for items or services, including drugs,

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that are false or fraudulent, claims for items or services not provided as claimed, or claims for medically unnecessary items or services. The government may deem manufacturers to have “caused” the submission of false or fraudulent claims by, for example, providing inaccurate billing or coding information to customers or promoting a product off-label. In addition, our future activities relating to the reporting of wholesaler or estimated retail prices for our products, the reporting of prices used to calculate Medicaid rebate information and other information affecting federal, state, and third-party reimbursement for our products, and the sale and marketing of our products, are subject to scrutiny under this law. Penalties for a False Claims Act violation include three times the actual damages sustained by the government, plus mandatory civil penalties of between \$5,500 and \$11,000 for each separate false claim, the potential for exclusion from participation in federal healthcare programs, and, although the federal False Claims Act is a civil statute, conduct that results in a False Claims Act violation may also implicate various federal criminal statutes. If the government were to allege that we were, or convict us of, violating these false claims laws, we could be subject to a substantial fine and may suffer a decline in our stock price. In addition, private individuals have the ability to bring actions under the federal False Claims Act and certain states have enacted laws modeled after the federal False Claims Act.

In addition to the laws described above, the PPACA also imposed new reporting requirements on drug manufacturers for payments made to physicians and teaching hospitals, as well as ownership and investment interests held by physicians and their immediate family members. Failure to submit required information may result in civil monetary penalties of up to an aggregate of \$150,000 per year (or up to an aggregate of \$1 million per year for “knowing failures”), for all payments, transfers of value or ownership or investment interests that are not timely, accurately and completely reported in an annual submission. Drug manufacturers were required to begin collecting data on August 1, 2013 and will be required to submit reports to CMS by March 31, 2014 (and by the 90th day of each subsequent calendar year).

In addition, many states have adopted laws similar to the federal laws discussed above. Some of these state prohibitions apply to the referral of patients for healthcare services reimbursed by any insurer, not just federal healthcare programs such as Medicare and Medicaid. There has also been a recent trend of increased federal and state regulation of payments made to physicians. Certain states mandate implementation of compliance programs, impose restrictions on drug manufacturers’ marketing practices and/or require the tracking and reporting of gifts, compensation and other remuneration to physicians. Because we intend to commercialize products that could be reimbursed under a federal healthcare program and other governmental healthcare programs, we plan to develop a comprehensive compliance program that establishes internal controls to facilitate adherence to the rules and program requirements to which we will or may become subject. Although compliance programs can mitigate the risk of investigation and prosecution for violations of these laws, the risks cannot be entirely eliminated. Due to the breadth of and ambiguities in these laws, the absence of guidance in the form of regulations or court decisions, and the potential for additional legal or regulatory change in this area, it is possible that our future sales and marketing practices and/or our future relationships with physicians might be challenged under these laws. Any action against us for violation of these laws, even if we successfully defend against it, could cause us to incur significant legal expenses and divert our management’s attention from the operation of our business.

Third-Party Coverage and Reimbursement

Sales of pharmaceutical products depend in significant part on the availability of coverage and adequate reimbursement by third-party payors, such as state and federal governments, including Medicare and Medicaid, and commercial managed care providers. In the United States, no uniform policy of coverage and reimbursement for drug products exists among third-party payors. Accordingly, decisions regarding the extent of coverage and amount of reimbursement to be provided for AKB-6548 will be made on a payor by payor basis. As a result, the coverage determination process is often a time-consuming and costly process that will require us to provide scientific and clinical support for the use of our product candidates to each payor separately, with no assurance that coverage and adequate reimbursement will be obtained. Third-party payors may limit coverage to specific drug products on an approved list, or formulary, which might not include all of the FDA-approved drugs for a particular indication. A decision by a third-party payor not to cover our product candidates could reduce physician utilization of our

products once approved and have a material adverse effect on our future sales, results of operations and financial condition. Moreover, a payor's decision to provide coverage for a drug product does not imply that an adequate reimbursement rate will be approved. Adequate third-party reimbursement may not be available to enable us to maintain price levels sufficient to realize an appropriate return on our investment in product development.

Healthcare Reform

In March 2010, the PPACA was enacted, which includes measures that have or will significantly change the way health care is financed by both governmental and private insurers. Among the provisions of PPACA of greatest importance to the pharmaceutical industry are the following:

- Effective in 2010, PPACA made several changes to the Medicaid Drug Rebate Program, including increasing pharmaceutical manufacturers' rebate liability by raising the minimum basic Medicaid rebate on most branded prescription drugs and biologic agents from 15.1% of average manufacturer price, or AMP, to 23.1% of AMP and adding a new rebate calculation for "line extensions" (i.e., new formulations, such as extended release formulations) of solid oral dosage forms of branded products, as well as potentially impacting their rebate liability by modifying the statutory definition of AMP.
- PPACA also expanded the universe of Medicaid utilization subject to drug rebates by requiring pharmaceutical manufacturers to pay rebates on Medicaid managed care utilization as of 2010 and by expanding the population potentially eligible for Medicaid drug benefits, to be phased-in by 2014. CMS has proposed to expand Medicaid rebate liability to the territories of the United States as well.
- In addition, PPACA provides for the public availability of retail survey prices and certain weighted average AMPs under the Medicaid program. The implementation of this requirement by the CMS may also provide for the public availability of pharmacy acquisition cost data, which could negatively impact our sales.
- Effective in 2010, PPACA expanded the types of entities eligible to receive discounted 340B pricing, although, under the current state of the law, with the exception of children's hospitals, these newly eligible entities will not be eligible to receive discounted 340B pricing on orphan drugs when used for the orphan indication. In addition, as 340B drug pricing is determined based on AMP and Medicaid rebate data, the revisions to the Medicaid rebate formula and AMP definition described above could result in an increase in the required 340B discounts.
- Effective in 2011, PPACA imposed a requirement on manufacturers of branded drugs and biologic agents to provide a 50% discount off the negotiated price of branded drugs dispensed to Medicare Part D patients in the coverage gap (i.e., "donut hole").
- Effective in 2011, PPACA imposed an annual, nondeductible fee on any entity that manufactures or imports certain branded prescription drugs and biologic agents, apportioned among these entities according to their market share in certain government healthcare programs, although this fee would not apply to sales of certain products approved exclusively for orphan indications.
- As of 2010, a new Patient-Centered Outcomes Research Institute was established pursuant to PPACA to oversee, identify priorities in, and conduct comparative clinical effectiveness research, along with funding for such research. The research conducted by the Patient-Centered Outcomes Research Institute may affect the market for certain pharmaceutical products.
- PPACA created the Independent Payment Advisory Board which, beginning in 2014, will have authority to recommend certain changes to the Medicare program to reduce expenditures by the program that could result in reduced payments for prescription drugs. Under certain circumstances, these recommendations will become law unless Congress enacts legislation that will achieve the same or greater Medicare cost savings.
- PPACA established the Center for Medicare and Medicaid Innovation within CMS to test innovative payment and service delivery models to lower Medicare and Medicaid spending, potentially including prescription drug spending. Funding has been allocated to support the mission of the Center for Medicare and Medicaid Innovation from 2011 to 2019.

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Many of the details regarding the implementation of PPACA are yet to be determined, and at this time, it remains unclear the full effect that PPACA would have on our business. In addition, we expect that additional state and federal healthcare reform measures will be adopted in the future. Because we anticipate that a significant proportion of patients eligible for AKB-6548 will be covered by Medicare Part D, any government healthcare reform measures which limit the amounts that federal and state governments will pay for healthcare products and services could result in reduced demand for our products once approved or additional pricing pressures.

Other Regulations

We are also subject to numerous federal, state and local laws relating to such matters as safe working conditions, manufacturing practices, environmental protection, fire hazard control, and disposal of hazardous or potentially hazardous substances. We may incur significant costs to comply with such laws and regulations now or in the future.

Employees

As of December 1, 2013, we had 26 employees, 22 of whom were full-time, eight of whom hold Ph.D. or M.D. degrees, 14 of whom were engaged in research and development activities and 12 of whom were engaged in business development, finance, information systems, facilities, human resources or administrative support. None of our employees are represented by any collective bargaining unit. We believe that we maintain good relations with our employees.

Facilities

Our corporate headquarters are located in Cambridge, Massachusetts. We currently lease approximately 6,837 square feet of office space in Cambridge, Massachusetts under a lease that expires on December 26, 2016. We believe that our existing facilities are adequate to meet our current needs, and that suitable additional alternative spaces will be available in the future on commercially reasonable terms.

Legal Proceedings

We have opposition proceedings pending in the Opposition Division of the European Patent Office. Final resolution of the opposition proceedings will likely take a number of years. For more information, see "Business—Intellectual Property."

We are not currently a party to any other material legal proceedings.

Management

Executive Officers and Directors

Below is a list of the names, ages and positions of the individuals who serve as our executive officers and directors as of December 1, 2013.

<u>Name</u>	<u>Age</u>	<u>Position</u>
John P. Butler	49	President and Chief Executive Officer; Director
Jason A. Amello	45	Senior Vice President, Chief Financial Officer and Treasurer
William Daly	58	Chief Business Officer
Robert Shalwitz, M.D.	59	Chief Medical Officer
Nicole R. Hadas	41	Vice President and General Counsel
Muneer A. Satter	52	Co-Chairman of the Board of Directors
Campbell Murray, M.D.	37	Co-Chairman of the Board of Directors
Jack Nielsen	49	Director
Anupam Dalal, M.D.	42	Director
Giovanni Ferrara	45	Director
Kim Dueholm, Ph.D.	51	Director
Duane Nash, M.D.	42	Director

John P. Butler joined Akebia as director in July 2013 and was appointed as the President and Chief Executive Officer of Akebia in August 2013. Prior to joining Akebia, from 2011 until 2013, Mr. Butler served as the Chief Executive Officer of Inspiration Biopharmaceuticals, Inc., a biopharmaceutical company that filed for protection under Chapter 11 of the U.S. Bankruptcy Code in October 2012 prior to the successful sale of its hemophilia assets to Cangene Corporation and Baxter International in early 2013. From 1997 to 2011, Mr. Butler held various positions at Genzyme Corporation, a biopharmaceutical company, most recently serving as President of the company's rare genetic diseases business. From 2002 until 2010, Mr. Butler led Genzyme's renal division. Prior to his work at Genzyme, Mr. Butler held sales and marketing positions at Amgen and Hoffmann-La Roche. Mr. Butler currently serves as the chairman of board of trustees for the American Kidney Fund and a member of the board of directors of Relypsa, Inc. Mr. Butler received a B.A. in Chemistry from Manhattan College and an M.B.A. degree from Baruch College, City University of New York. We believe that Mr. Butler is qualified to serve on our board of directors due to his industry experience in the biotechnology sector, particularly his experience working in the renal disease market.

Jason A. Amello joined Akebia as Senior Vice President, Chief Financial Officer and Treasurer in 2013. Prior to joining Akebia, Mr. Amello served as Executive Vice President, Chief Financial Officer and Treasurer of ZIOPHARM Oncology, Inc., a biopharmaceutical company, from 2012 to 2013. From 2000 to 2011, Mr. Amello held various positions at Genzyme Corporation, most recently as Senior Vice President, Corporate Controller and Chief Accounting Officer. Earlier in his career, Mr. Amello spent 10 years in the business advisory and assurance practice of Deloitte, serving in various roles of increasing responsibility through senior manager. Mr. Amello holds a B.A. from Boston College and is a Certified Public Accountant in the Commonwealth of Massachusetts.

William Daly joined Akebia as Senior Vice President, Business Development in 2012 and was promoted to Chief Business Officer in 2013. Mr. Daly also currently serves as Chief Business Officer of Aerpio Therapeutics, Inc. Prior to joining Akebia, Mr. Daly served as Senior Vice President, Business Development at Halozyme Therapeutics, a biopharmaceutical company, from 2009 to 2012, where he was the head of business development and alliance management. From 2007 to 2009, Mr. Daly was the Senior Vice President of Business Development at Cougar Biotechnology, a biopharmaceutical company, and then, after the acquisition of Cougar Biotechnology by Johnson & Johnson, co-led the integration of Cougar and Johnson & Johnson and was site head for the Los Angeles facility. Prior to joining Cougar, Mr. Daly held a number of business development and senior executive positions at Allergan, Chiron and Novartis as well as at a number of start-up biotechnology companies. Before moving into industry, Mr. Daly was an investment banker focusing primarily on pharmaceutical and

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biotechnology companies for nine years, most recently served as a Managing Director for Prudential Vector Healthcare Group. Prior to becoming an investment banker, he practiced corporate and securities law for ten years. Mr. Daly holds an A.B. in biochemistry from the University of California at Berkeley and a J.D. and M.B.A. from the University of California at Los Angeles.

Robert Shalwitz, M.D. co-founded Akebia in 2007. Prior to Akebia, Dr. Shalwitz was Vice President of Clinical Development at Reliant Pharmaceuticals, a biopharmaceutical company, from 2005 to 2007. From 1995 to 2005, Dr. Shalwitz was Medical Director at Abbott Labs. Prior to Abbott Labs, Dr. Shalwitz was an academic pediatric endocrinologist for 10 years, and his research at Washington University in St. Louis and at the Children's Hospital of Orange County (CA) focused on glucose and glycogen metabolism. Dr. Shalwitz received a B.G.S. from the University of Michigan and an M.D. from SUNY Buffalo.

Nicole R. Hadas joined Akebia as Vice President and General Counsel in 2013. Prior to Akebia, Ms. Hadas was Vice President and General Counsel at OvaScience, Inc., a biopharmaceutical company, in 2013. From 2011 to 2013, Ms. Hadas served as the Senior Vice President and General Counsel at Inspiration Biopharmaceuticals, Inc., a biopharmaceutical company that filed for protection under Chapter 11 of the U.S. Bankruptcy Code in October 2012, where she managed the successful sale of its hemophilia assets to Cangene Corporation and Baxter International in early 2013. From 2001 to 2011, Ms. Hadas worked at Genzyme Corporation, most recently as Senior Corporate Counsel. Prior to Genzyme, she was an associate at Foley Hoag representing biopharmaceutical companies and healthcare providers in a wide variety of matters. Ms. Hadas received a B.A. from the University of Michigan and a J.D. from Boston College Law School.

Muneer A. Satter has served as a member of our board of directors since 2012. Mr. Satter has been Chairman at Satter Investment Management LLC since 2012. He also manages the Satter Foundation. Prior to Satter Investment Management, Mr. Satter was a partner at Goldman Sachs where he spent 24 years in various roles, most recently as the Global Co-Head of the Principal Debt Group and Global Head of the Mezzanine Group in the Merchant Banking Division. He is Co-Chairman of the Board of Aerpio Therapeutics, Vital Therapies, Inc. and Linq3 Technologies LLC, and Chairman of the Board of Restorsea Holdings, LLC. He also serves as Vice Chairman of Goldman Sachs Foundation and GS Gives, is a director of The Nature Conservancy and World Business Chicago, is on the Board of Advisors of the American Enterprise Institute and is on the Board of Trustees of Northwestern University. Mr. Satter received a B.A. in Economics from Northwestern University, a J.D. from Harvard Law School, and an M.B.A. from Harvard Business School. We believe that Mr. Satter is qualified to serve on our board of directors due to his extensive investment experience.

Campbell Murray, M.D. has served as a member of our board of directors since 2008 and is our Co-Chairman. Dr. Murray has been a Managing Director of Novartis Venture Fund, since 2005. Prior to joining the fund, he worked at the Novartis Institutes for BioMedical Research as the Director of Special Projects. Dr. Murray currently serves on the board of directors of Aerpio Therapeutics, Alios BioPharm, Euthymics Biosciences, Galera Therapeutics, ImaginAb, Neurovance and Tokai Pharmaceuticals. Dr. Murray received a Bachelor of human biology from the University of Auckland Medical School, an M.B.A. from Harvard Business School, an M.P.P. from the John F. Kennedy School of Government, and an M.B.C.L.B. (M.D.) from the University of Auckland Medical School. We believe that Dr. Murray is qualified to serve on our board of directors due to his investment experience in the biotechnology sector.

Jack Nielsen has served as a member of our board of directors since 2013. Mr. Nielsen has worked within the Novo A/S organization and its venture activities since 2001 in several roles, most recently as a Partner based in Copenhagen, Denmark. From 2006 to 2012, Mr. Nielsen worked at Novo Ventures (US) Inc. in San Francisco, where he established the office which provides advisory investment services to Novo A/S. From 1990-2001, he held various positions in the Novo Nordisk business area which in 2000 became Novozymes A/S. Mr. Nielsen currently serves on the board of directors of Alios BioPharma Inc., Apollo Endosurgery Inc., BioClin Therapeutics Inc., ProteinSimple, Reata Pharmaceuticals Inc. and Tobira Therapeutics Inc. Previously, he was a board member of MediQuest Therapeutics Inc., NeoMend Inc. and Protein Forest Inc. Mr. Nielsen received a

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M.Sc. in Chemical Engineering from the Technical University in Denmark, and a Master in Management of Technology from Center for Technology, Economics and Management; Technical University of Denmark. We believe that Mr. Nielsen is qualified to serve on our board of directors due to his experience serving on boards in the biotechnology sector.

Anupam Dalal, M.D. has served as a member of our board of directors since 2008. Dr. Dalal has been a managing director at Kearny Venture Partners since 2008. Prior to working at Kearny Venture Partners, Dr. Dalal was a Principal at Flagship Ventures. Dr. Dalal currently serves on the board of directors of Aerpio Therapeutics. Dr. Dalal has served on the board of Resolvix Pharmaceuticals and Pervasis Therapeutics. Dr. Dalal received a B.A. in Economics from the University of California at Berkeley, an M.B.A. from Harvard Business School, and an M.D. from the University of California, San Francisco. Dr. Dalal was Resident in Surgery at Brigham and Women's Hospital / Harvard Medical School. We believe that Dr. Dalal is qualified to serve on our board of directors due to his investment and board experience in the biotechnology sector.

Giovanni Ferrara has served as a member of our board of directors since 2013. Mr. Ferrara has been a Venture Partner at Novartis Venture Fund since 2011. Prior to joining Novartis, he spent three years as a consultant to west coast venture capital firms and as consulting Chief Business Officer to Sorbent Therapeutics, a biopharmaceutical company. Previously, he was Managing Director and General Partner at Burrill & Company, a venture fund, and began his venture capital career at GeneChem Management, where, in addition to investing, he also held operating positions in portfolio companies, including CEO of Targanta Therapeutics (then Phage Tech, Inc.). Mr. Ferrara received a B.Sc. in Human Genetics and Biology from the University of Toronto, and an M.B.A. and M.Sc. from McGill University. We believe that Mr. Ferrara is qualified to serve on our board of directors due to his management experience in the biotechnology sector.

Kim Dueholm, Ph.D. has served as a member of our board of directors since 2013. Mr. Dueholm has been a partner at Novo A/S, a venture fund, since 2000. Prior to joining Novo A/S, Mr. Dueholm spent five years with Novo Nordisk A/S in positions ranging from Patent Portfolio Analyst to Principal Scientific Analyst, from 1995 to 2000. He currently serves on the board of directors of ObsEva SA and Orphazyme ApS. Previously, he was a board member of Core A/S, F-star GmbH, NeuroKey A/S, Novoxel S.A., Nuevolution A/S and Symphogen A/S. He is also a member of the editorial board of Expert Opinion on Therapeutic Patents. Mr. Dueholm received an M.Sc. in Chemistry and Business Administration from Odense University, and a Ph.D. in organic chemistry from the University of Copenhagen. We believe that Dr. Dueholm is qualified to serve on our board of directors due to his management and director experience in the biotechnology sector.

Duane Nash, M.D. has served as a member of our board of directors since 2013. Dr. Nash has been the Executive Vice President since 2013 and Chief Business Officer since 2012 of Vital Therapies, Inc., a biopharmaceutical company. In 2012 and 2013, he also served as Medical Director. Dr. Nash joined Vital Therapies from Wedbush PacGrow Life Sciences, an investment bank, where he was employed from March 2009 to March 2012 serving most recently as Senior Vice President in Equity Research. Before that he was a research analyst at Pacific Growth Equities, an investment bank, from April 2008 through March 2009, which was subsequently acquired by Wedbush Securities, Inc. Dr. Nash also practiced as an attorney from November 2002 to February 2008, most recently at the law firm of Davis Polk, where he focused on intellectual property litigation and corporate matters. Dr. Nash currently serves on the board of directors of Aerpio Therapeutics Inc. Dr. Nash earned a B.A. in biology from Williams College, an M.D. from Dartmouth Medical School, a J.D. from the University of California, Berkeley, and an M.B.A. from the University of Oxford. Dr. Nash completed his internship in general surgery at the University of California at San Francisco. We believe that Dr. Nash is qualified to serve on our board of directors due to his management experience in the biotechnology sector.

In addition to the individual attributes of each of our directors listed above, we highly value the collective qualifications and experiences of our board members. We believe the collective viewpoints and perspectives of our directors results in a board that is dedicated to advancing the interests of our stockholders.

Board Composition and Election of Directors

Board Composition

Our board of directors is currently comprised of eight members. The members of our board of directors were elected in compliance with the provisions of the voting agreement among us and our major stockholders. The voting agreement will terminate upon the closing of this offering and we will have no further contractual obligations regarding the election of our directors. See “Certain Relationships and Related Party Transactions.” Our directors hold office until their successors have been elected and qualified or until their earlier death, resignation or removal.

Our amended and restated certificate of incorporation and amended and restated bylaws that will become effective upon the closing of this offering provide that the authorized number of directors may be changed only by resolution of our board of directors. Our amended and restated certificate of incorporation and amended and restated bylaws also provide that our directors may be removed only for cause by the affirmative vote of the holders of at least 75% of the votes that all our stockholders would be entitled to cast in an annual election of directors, voting together as a single class, at a meeting of the stockholders called for that purpose, and that any vacancy on our board of directors, including a vacancy resulting from an enlargement of our board of directors, may be filled only by vote of a majority of our directors then in office.

In accordance with the terms of our amended and restated certificate of incorporation that will become effective upon the closing of this offering, our board of directors will be divided into three classes, class I, class II and class III, with members of each class serving staggered three-year terms. Upon the closing of this offering, the members of the classes will be divided as follows:

- the class I directors will be _____ ;
- the class II directors will be _____ ; and
- the class III directors will be _____ .

Upon the expiration of the term of a class of directors, directors in that class will be eligible to be elected for a new three-year term at the annual meeting of stockholders in the year in which their term expires.

We have no formal policy regarding board diversity. Our priority in selection of board members is identification of members who will further the interests of our stockholders through his or her established record of professional accomplishment, the ability to contribute positively to the collaborative culture among board members, knowledge of our business and understanding of the competitive landscape.

Director Independence

Applicable NASDAQ rules require a majority of a listed company’s board of directors to be comprised of independent directors within one year of listing. In addition, the NASDAQ rules require that, subject to specified exceptions, each member of a listed company’s audit, compensation and nominating and corporate governance committees be independent and that audit committee members also satisfy independence criteria set forth in Rule 10A-3 under the Exchange Act. Under applicable NASDAQ rules, a director will only qualify as an “independent director” if, in the opinion of the listed company’s board of directors, that person does not have a relationship that would interfere with the exercise of independent judgment in carrying out the responsibilities of a director. In order to be considered independent for purposes of Rule 10A-3, a member of an audit committee of a listed company may not, other than in his or her capacity as a member of the audit committee, the board of directors, or any other board committee, accept, directly or indirectly, any consulting, advisory, or other compensatory fee from the listed company or any of its subsidiaries or otherwise be an affiliated person of the listed company or any of its subsidiaries.

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In _____, our board of directors undertook a review of the composition of our board of directors and its committees and the independence of each director. Based upon information requested from and provided by each director concerning his or her background, employment and affiliations, including family relationships, our board of directors has determined that _____ are “independent directors” as defined under applicable NASDAQ rules. In making such determination, our board of directors considered the relationships that each such non-employee director has with our company and all other facts and circumstances that our board of directors deemed relevant in determining his or her independence, including the beneficial ownership of our capital stock by each non-employee director. Mr. Butler is not an independent director under these rules because he is an employee of Akebia. Please see the section of this prospectus titled “Certain Relationships and Related Party Transactions.”

There are no family relationships among any of our directors or executive officers.

Board Committees

Our board of directors has three standing committees: the audit committee, the compensation committee and the nominating and corporate governance committee.

Audit Committee

Our audit committee is composed of _____, with _____ serving as chairman of the committee. Our board of directors has determined that each member of the audit committee meets the independence requirements of Rule 10A-3 under the Exchange Act and the applicable listing standards of NASDAQ. Our board of directors has determined that _____ is an “audit committee financial expert” within the meaning of the SEC regulations and applicable listing standards of NASDAQ. Following this offering, the audit committee’s responsibilities will include:

- appointing, approving the compensation of, and assessing the qualifications, performance and independence of our independent registered public accounting firm;
- pre-approving audit and permissible non-audit services, and the terms of such services, to be provided by our independent registered public accounting firm;
- reviewing the internal audit plan with the independent registered public accounting firm and members of management responsible for preparing our financial statements;
- reviewing and discussing with management and the independent registered public accounting firm our annual and quarterly financial statements and related disclosures as well as critical accounting policies and practices used by us;
- reviewing the adequacy of our internal control over financial reporting;
- establishing policies and procedures for the receipt and retention of accounting-related complaints and concerns;
- recommending, based upon the audit committee’s review and discussions with management and the independent registered public accounting firm, whether our audited financial statements shall be included in our Annual Report on Form 10-K;
- monitoring our compliance with legal and regulatory requirements as they relate to our financial statements and accounting matters;
- preparing the audit committee report required by the rules of the SEC to be included in our annual proxy statement;
- viewing related party transactions for potential conflict of interest situations and approving such transactions; and
- reviewing and discussing with management and our independent registered public accounting firm our earnings releases and scripts.

Compensation Committee

Our compensation committee is composed of _____, with _____ serving as chairman of the committee. Our board of directors has determined that each member of the compensation committee is “independent” as defined under the applicable listing standards of NASDAQ. Following this offering, the compensation committee’s responsibilities will include:

- annually reviewing and approving corporate goals and objectives relevant to the compensation of our chief executive officer;
- evaluating the performance of our chief executive officer in light of such corporate goals and objectives and determining and approving the compensation of our chief executive officer;
- reviewing and approving the compensation of our other executive officers;
- appointing, compensating and overseeing the work of any compensation consultant, legal counsel or other advisor retained by the compensation committee;
- conducting the independence assessment outlined in NASDAQ rules with respect to any compensation consultant, legal counsel or other advisor retained by the compensation committee;
- annually reviewing and reassessing the adequacy of the committee charter in its compliance with the listing requirements of NASDAQ;
- reviewing and establishing our overall management compensation, philosophy and policy;
- overseeing and administering our compensation and other compensatory plans;
- reviewing and approving our equity and incentive policies and procedures for the grant of equity-based awards and approving the grant of such equity-based awards;
- reviewing and making recommendations to the board of directors with respect to director compensation; and
- to the extent we no longer qualify as an emerging growth company, reviewing and discussing with management the compensation discussion and analysis to be included in our annual proxy statement or Annual Report on Form 10-K.

Nominating and Corporate Governance Committee

Our nominating and corporate governance committee is composed of _____, with _____ serving as chairman of the committee. Our board of directors has determined that each member of the nominating and corporate governance committee is “independent” as defined under the applicable listing standards of NASDAQ. Following this offering, the nominating and corporate governance committee’s responsibilities will include:

- developing and recommending to the board of directors criteria for board and committee membership;
- establishing procedures for identifying and evaluating board of director candidates, including nominees recommended by stockholders;
- identifying individuals qualified to become members of the board of directors;
- recommending to the board of directors the persons to be nominated for election as directors and to each of the board’s committees;
- developing and recommending to the board of directors a set of corporate governance principles;
- articulating to each director what is expected, including reference to the corporate governance principles and directors’ duties and responsibilities;
- reviewing and recommending to the board of directors practices and policies with respect to directors;

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- reviewing and recommending to the board of directors the functions, duties and compositions of the committees of the board of directors;
- reviewing and assessing the adequacy of the committee charter and submitting any changes to the board of directors for approval;
- considering and reporting to the board of directors any questions of possible conflicts of interest of board of directors members;
- providing for new director orientation and continuing education for existing directors on a periodic basis; and
- overseeing the evaluation of the board of directors and management.

Compensation Committee Interlocks and Insider Participation

None of our executive officers serves as a member of the board of directors or compensation committee, or other committee serving an equivalent function, of any other entity that has one or more of its executive officers serving as a member of our board of directors or compensation committee. None of the members of our compensation committee has ever been employed by us. For a description of transactions between us and members of our compensation committee and affiliates of such members, please see the section of this prospectus titled "Certain Relationships and Related Party Transactions."

Code of Business Conduct and Ethics

Prior to completion of this offering, we will adopt a code of business conduct and ethics that applies to all of our employees, officers and directors, including those officers responsible for financial reporting. Upon the closing of this offering, our code of business conduct and ethics will be available on our website. We intend to disclose amendments to the code, or any waivers of its requirements, on our website as may be required by law or NASDAQ stock market listing standards.

Executive Compensation

This section discusses the material elements of our executive compensation policies and decisions and important factors relevant to an analysis of these policies and decisions. It provides qualitative information regarding the manner and context in which compensation is awarded to and earned by our executive officers named in the “Summary Compensation Table” below and is intended to place in perspective the information presented in the following tables and the corresponding narrative.

Overview

Historically, our executive compensation program has reflected our growth and corporate goals. To date, the compensation of our executive officers has consisted of a combination of base salary, annual cash bonus, long-term equity incentive compensation in the form of restricted stock and stock options and other employee benefits generally available to our employees. Certain of our executive officers are also entitled to certain compensation and benefits upon certain terminations of employment pursuant to their employment agreements, as described below.

Our named executive officers for the year ended December 31, 2012 were as follows:

- Joseph Gardner, Ph.D., our former President and Chief Executive Officer;
- Robert Shalwitz, M.D., our Chief Medical Officer; and
- William Daly, our Senior Vice President, Business Development. Mr. Daly was promoted to Chief Business Officer in 2013.

John P. Butler was hired as our President and Chief Executive Officer in August 2013 and appointed as a member of our board of directors effective July 2013. In connection with Mr. Butler’s appointment, Dr. Gardner resigned as President and Chief Executive Officer and as a member of our board of directors, although he continues to serve as a consultant to the Company.

Elements of Executive Compensation

Base Salaries. Base salaries for our named executive officers are determined annually by our compensation committee, subject to review and approval by our board of directors, based on the scope of each officer’s responsibilities along with his respective experience and contributions to the company during the prior year period. When reviewing base salaries, our compensation committee takes factors into account such as each officer’s experience and individual performance, the company’s performance as a whole, data from surveys of compensation paid by comparable companies, and general industry conditions, but does not assign any specific weighting to any factor.

Annual Cash Bonuses. Our annual cash bonus program promotes and rewards the achievement of key strategic and business goals for the twelve consecutive month period ending each June 30. For the 2012 bonus plan year (the 12-month period beginning on July 1, 2011 and ending on June 30, 2012), the target annual bonus as a percentage of base salary (as determined on the date the bonus is paid) for each of Dr. Gardner, Dr. Shalwitz, and Mr. Daly was 25%, 20% and 20%, respectively. At the beginning of the 2012 bonus plan year, our compensation committee established corporate performance goals, each having a designated weighting, that related to key development, strategic and financial goals of the company. At the end of the 2012 bonus plan year, our compensation committee met and evaluated the performance of the company against the specified performance goals. Based on its evaluation, the compensation committee recommended and the board of directors approved, payment of cash bonuses for the 2012 bonus plan year of: \$36,438 for Dr. Gardner (which represented 53% of his target bonus), \$31,680 for Dr. Shalwitz (which represented 60% of his target bonus), and \$20,625 to Mr. Daly (which represented 75% of his target bonus, as prorated to reflect his commencement of employment in January 2012).

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Equity Awards. Our named executive officers participate in our Akebia Therapeutics, Inc. 2008 Equity Incentive Plan, or the 2008 Equity Incentive Plan. Each of Dr. Gardner and Dr. Shalwitz received a grant of stock options during fiscal 2012 and Mr. Daly received a grant of restricted stock during fiscal 2012. These stock option and restricted stock grants are subject to time-based vesting conditions and generally vest, subject to continued employment, as to 25% of the shares subject to the award after one year and thereafter continue to vest in monthly installments over the following three years. These equity awards serve to align the interests of our named executive officers with our shareholders. They also encourage retention through the use of time-based vesting. For more information regarding the awards granted under the 2008 Equity Incentive Plan, please refer to “Equity Incentive Plans—2008 Equity Incentive Plan” below.

Benefits. Our named executive officers are eligible for benefits, such as participation in our 401(k) plan and basic health and welfare benefit coverage, that are generally available to all of our employees.

Summary Compensation Table

The following table sets forth information regarding compensation awarded to, earned by or paid to each of our named executive officers during the fiscal year ending December 31, 2012.

2012 Summary Compensation Table

<u>Name and Principal Position</u>	<u>Year</u>	<u>Salary (\$)</u>	<u>Bonus (\$)⁽¹⁾</u>	<u>Stock Awards (\$)⁽²⁾</u>	<u>Option Awards (\$)⁽³⁾</u>	<u>All Other Compensation (\$)⁽⁴⁾</u>	<u>Total (\$)</u>
Joseph Gardner, Ph.D. <i>Former President and Chief Executive Officer</i>	2012	275,000	36,438	—	2,882	251	314,571
Robert Shalwitz, M.D. <i>Chief Medical Officer</i>	2012	269,280	31,680	—	6,126	455	307,541
William Daly <i>Senior Vice President, Business Development⁽⁵⁾</i>	2012	259,375	20,625	17,246	—	455	297,701

- (1) The company’s bonus plan covers the twelve consecutive month period from July to June. Amounts represent cash bonuses earned for the 2012 bonus plan period from July 1, 2011 to June 30, 2012.
- (2) The amount reported in the Stock Awards column granted to our named executive officers during 2012 represents the retrospective fair value of the stock awards as of the grant date.
- (3) The amounts reported in the Option Awards column granted to our named executive officers during 2012 represent the retrospective fair value of the stock options as of the grant date as computed in accordance with Accounting Standards Codification, or ASC, Topic 718, not including any estimates of forfeitures. The assumptions used in calculating the grant date fair value of the stock options reported in the Option awards column are set forth in Note 12 to our consolidated financial statements included elsewhere in this prospectus. Note that the amounts reported in this column reflect the accounting cost for these stock options, and do not correspond to the actual economic value that may be received by the named executive officers from the options.
- (4) Amounts represent the dollar value of life insurance premiums paid by the company on behalf of the named executive officers.
- (5) Mr. Daly joined us in January 2012. Mr. Daly’s annual base salary in 2012 was \$275,000. The amounts in the table above reflect his partial year of service in 2012.

2012 Outstanding Equity Awards at Fiscal Year-End

The following table sets forth information concerning outstanding equity awards for each of our named executive officers at December 31, 2012:

Name and Principal Position	Stock Options				Stock Awards ⁽⁸⁾	
	Number of Securities Underlying Unexercised Options (#) Exercisable	Number of Securities Underlying Unexercised Options (#) Unexercisable	Option Exercise Price (\$)	Option Expiration Date	Number of Shares or Units of Stock That Have Not Vested (#)	Market Value of Shares of Stock That Have Not Vested (\$) ⁽⁷⁾
Joseph Gardner, Ph.D.	53,714	—	\$ 1.50	6/1/2018	2,334 ⁽³⁾	2,334
<i>President and Chief Executive Officer</i>	6,771	4,221 ⁽¹⁾	\$ 1.50	7/28/2020	3,468 ⁽⁴⁾	3,468
	3,505	9,991 ⁽²⁾	\$ 1.50	1/12/2022	16,451 ⁽⁵⁾	16,451
Robert Shalwitz, M.D.	38,029	—	\$ 1.50	6/1/2018	2,024 ⁽³⁾	2,024
<i>Chief Medical Officer</i>	10,677	6,656 ⁽¹⁾	\$ 1.50	7/28/2020	3,007 ⁽⁴⁾	3,007
	2,772	7,900 ⁽²⁾	\$ 1.50	1/12/2022	13,079 ⁽⁵⁾	13,079
William Daly	—	—	—	—	30,047 ⁽⁶⁾	30,047
<i>Senior Vice President, Business Development</i>						

- (1) Represents options to purchase shares of our common stock granted on July 28, 2010. The remainder of these options vests in equal monthly installments through July 28, 2014. Vesting of all unvested options shall accelerate in connection with an acquisition event pursuant to the terms of the option agreement.
- (2) Represents options to purchase shares of our common stock granted on January 12, 2012. The remainder of these options vests in equal monthly installments through December 23, 2015. Pursuant to the terms of the award agreement, vesting of all unvested options shall accelerate in connection with an acquisition, in the event the option is not assumed by the acquirer, or in the event the option is assumed by the acquirer and the executive's employment is terminated or materially diminished within the following 12 months.
- (3) Under the terms of the August 31, 2009 restricted stock agreement, the remaining unvested shares will vest in equal monthly installments through August 31, 2013. Vesting of all restricted shares shall accelerate in connection with an acquisition event pursuant to the terms of the restricted stock agreement.
- (4) Under the terms of the June 15, 2011 restricted stock agreement, the remaining unvested shares will vest in equal monthly installments through August 31, 2013. Vesting of all restricted shares shall accelerate in connection with an acquisition event pursuant to the terms of the restricted stock agreement.
- (5) Under the terms of the June 15, 2011 restricted stock agreement, the remaining unvested shares will vest in equal monthly installments through April 6, 2015. Vesting of all restricted shares shall accelerate in connection with an acquisition event pursuant to the terms of the restricted stock agreement.
- (6) Under the terms of the February 21, 2012 restricted stock agreement, the remaining unvested shares will vest as follows: 25% vest on January 22, 2013, with the remainder of the shares vesting in equal monthly installments over the following three years through January 22, 2016. Pursuant to the terms of the award agreement, vesting of all unvested shares shall accelerate in connection with an acquisition, in the event the award is not assumed by the acquirer, or in the event the award is assumed by the acquirer and the executive's employment is terminated or materially diminished within the following 12 months.
- (7) The value of the unvested restricted stock was \$1.00 per share based on an independent, third-party appraisal of our common stock as of December 31, 2012.
- (8) The restricted stock awards for Dr. Gardner and Dr. Shalwitz were purchased using the promissory notes issued by executives to the Company (the "Promissory Notes"). The aggregate balance of the outstanding Promissory Notes at December 31, 2012 was \$140,839 for Dr. Gardner and \$110,692 for Dr. Shalwitz. The Promissory Notes were subsequently amended in 2013 to forgive a

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portion of the principal owed and to reduce the interest rate from 6% to 3% per annum. The Promissory Notes are repayable at the earlier of (a) an initial public offering; (b) the sale of the company or substantially all of its assets; (c) the termination of the employee; or (d) five years from origination and will be extinguished prior to the filing of the registration statement of which this prospectus is a part.

Retention Bonuses

We have established a retention bonus program in which our named executive officers participate. This program provides that if the named executive officers remain employed with us through a "Sale of the Company" (as defined in the Third Amended and Restated Voting Agreement), they will be paid a bonus in the same form and manner as payments made to holders of our Series C Preferred Stock in connection with the "Sale of the Company". Each participant in the program is entitled to a designated percentage of a bonus pool. The size of the bonus pool is based on (i) the percentage of our fully-diluted equity that is represented by vested awards under our 2008 Equity Incentive Plan immediately prior to a Sale of the Company (up to a maximum of 12.5%) multiplied by (ii) fifty percent of the Applicable Accrued Value (as defined in our eighth amended and restated certificate of incorporation) of our Series C Preferred Stock.

Retirement Benefits

We offer a tax-qualified retirement plan, or 401(k) plan, to eligible employees, including our named executive officers. In accordance with this plan, all eligible employees may contribute a percentage of compensation up to a maximum of the statutory limits per year. Company contributions are discretionary and no contributions were made during 2012.

Employment Agreements with Our Named Executive Officers

We have entered into an employment agreement with each of our named executive officers, except for Mr. Daly, with whom we entered into an offer letter at the time of he commenced employment with us. Each of these employment agreements and offer letter provides for "at will" employment, meaning that either we or the named executive officer may terminate our employment relationship at any time without cause.

Dr. Joseph Gardner. On May 2, 2007, we entered into an executive employment agreement with Dr. Gardner for the position of President and CEO; this agreement was subsequently amended on April 6, 2011. Dr. Gardner's base salary during 2012 was \$275,000, which was subject to review by our Board of Directors from time to time during his employment, and at least every 12 months. Dr. Gardner was also eligible to participate in discretionary bonus programs on both a quarterly and annual basis, as determined by our Board of Directors, in its sole discretion; provided; that Dr. Gardner was not entitled to payment of any such bonus unless he remained actively employed through the end of the applicable calendar quarter or year. Dr. Gardner was entitled to four weeks of vacation, as well as holidays and sick leave, and (subject to eligibility criteria under the applicable plan) the right to participate in any profit sharing plan, retirement plan, 401(k) plan, group medical plan, group dental plan, and/or other health or insurance plan maintained by us for our senior executives generally and, if applicable, their family members. Dr. Gardner was also entitled to reimbursement of all reasonable and necessary business and travel expenses incurred in connection with the performance of his duties. Dr. Gardner resigned as President and CEO in September 2013. He continues to serve as a consultant. Although Dr. Gardner was not entitled to any severance under his employment agreement upon his termination of employment, we entered into a separation agreement with him that provided for the accelerated vesting of all his then unvested options and restricted stock. The separation agreement also provided Dr. Gardner with an award of 21,153 shares of unrestricted common stock of the Company, without consideration therefor, and stated that Dr. Gardner would participate in the retention bonuses described above. The separation agreement also contained a release of claims in favor of the Company.

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Dr. Robert Shalwitz. On April 6, 2011, we entered into an executive employment agreement with Dr. Shalwitz for the position of Chief Medical Officer and Vice President. Dr. Shalwitz currently receives a base salary of \$269,280, which is subject to review by our Board of Directors from time to time, and at least every 12 months. Dr. Shalwitz is also eligible to participate in all bonus or similar incentive plans adopted by our Board of Directors including, without limitation, an incentive compensation plan with a yearly performance-based cash bonus of up to 20% of Dr. Shalwitz's base salary. Dr. Shalwitz is entitled to four weeks of vacation, as well as holidays and sick leave, and (subject to eligibility criteria under the applicable plan) the right to participate in any profit sharing plan, retirement plan, 401(k) plan, group medical plan, group dental plan, and/or other health or insurance plan maintained by us for our senior executives generally and, if applicable, their family members. We pay 100% of Dr. Shalwitz's premiums under our medical and dental plans and 50% of the premiums associated with the coverage of his spouses/dependents under those same plans. Dr. Shalwitz is also entitled to reimbursement of all reasonable and necessary business and travel expenses incurred in connection with the performance of his duties.

Mr. William Daly. On January 2, 2012, we entered into an offer letter with Mr. Daly for the position of Senior Vice President of Business Development. Under the offer letter, Mr. Daly currently receives an annual base salary of \$275,000 and is eligible for performance-based cash bonuses of up to a maximum of 20% of base salary per year. Mr. Daly's offer letter also entitled him to an initial grant of restricted stock under our 2008 Equity Incentive Plan, subject to approval by our board of directors. For more information about the restricted stock grant made to Mr. Daly upon the commencement of his employment in 2012, please see the 2012 Outstanding Equity Awards at Fiscal Year-End table above. Mr. Daly's base salary and incentive compensation are subject to review on an annual basis. Mr. Daly is entitled to participate in all benefit plans as may be offered by us from time to time during his employment.

Involuntary Termination of Employment and Change of Control

Pursuant to their employment agreements, Dr. Gardner and Dr. Shalwitz are eligible to receive certain payments and benefits in the event that the executive's employment is terminated by us without "cause" (as defined in the applicable employment agreement), the executive terminates his employment with us for "good reason" (as defined in the applicable employment agreement) or the executive is terminated in connection with or within six months following a "change of control" (as defined in the applicable employment agreement). A termination will be considered to be in connection with a change of control if the executive is not offered employment with compensation and benefit terms (including severance) that are at least materially comparable to those under the executive's employment agreement or if the executive's employment is terminated by the employer without cause or by the executive for good reason within six months following the change of control. The severance payable to Dr. Gardner and Dr. Shalwitz in each of these situations, which is subject to the execution of a release of claims in our favor and continued compliance with a set of restrictive covenants prohibiting certain competitive behaviors by the executive within the one year period following his termination of employment, is equal to six months of salary continuation, the Company's payment of the executive's COBRA premiums for a maximum of six months, and six months of participation in our group insurance benefits (other than health insurance) in which the executive participated immediately prior to termination. As noted above, Dr. Gardner resigned as President and CEO on September 15, 2013. No severance was or is payable to Dr. Gardner under his employment agreement in connection with this termination of employment.

For purposes of Dr. Gardner and Dr. Shalwitz's employment agreements, "cause" means:

(i) the executive's failure to substantially perform his duties under his employment agreement for reasons other than death or disability, which failure, if curable, is not cured to the reasonable satisfaction of the Board of Directors during the fifteen (15) day period following written notice of such failure from us; (ii) the executive's material failure or refusal to comply with reasonable written policies, standards and regulations established by us from time to time which failure, if curable, is not cured to the reasonable satisfaction of the Board of Directors during the fifteen (15) day period following written notice of such

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failure from us; (iii) the commission by the executive of (x) an act of dishonesty or constituting common law fraud, embezzlement or a felony or (y) any tortious act, unlawful act or malfeasance that causes or reasonably would cause (for example, if it became publicly known) material harm to our standing, condition or reputation; or (iv) any material breach by the executive of the provisions of his employment agreement, which breach, if curable, is not cured to the reasonable satisfaction of the Board of Directors during the fifteen (15) day period following written notice of such breach from us. The Board of Directors (excluding the executive if he is at such time a member of the Board of Directors) shall make all determinations relating to termination, including without limitation any determination regarding cause.

For purposes of Dr. Gardner and Dr. Shalwitz's employment agreements, "good reason" means any of the following, without the executive's consent:

(i) a material diminution in the executive's position, duties or responsibilities from those held by or assigned to him as of the effective date of his employment agreement, (ii) a reduction of the executive's base salary, or (iii) a material reduction of the executive's benefits or bonus/incentive compensation opportunities provided to the executive as then in effect, so long as he is the only executive to suffer such a reduction.

For purposes of Dr. Gardner and Dr. Shalwitz's employment agreements "change of control" means any of the following:

a transfer (or license on an exclusive basis) of all or substantially all of our assets or the transfer of ownership of more than a majority of our securities, whether in a single transaction or series of separate transactions, other than in connection with our fundraising activities, including without limitation a transaction in which a portion of our assets are transferred to an acquiror and we do not continue as a going concern during the 6 months thereafter, or our remaining assets are moved following such transfer to an acquiror to a NewCo (regardless of whether the NewCo stockholders are our existing stockholders) and such NewCo does not continue as a going concern during the 6 months after such transfer to such acquiror. If a party obtains an option to close a transaction, the transaction will not be considered as having occurred until such option is exercised and the transaction thereafter closed.

Equity Incentive Plans

All outstanding equity-based awards have been granted under our 2008 Equity Incentive Plan, as described below.

2008 Equity Incentive Plan

Our Board of Directors and shareholders originally approved the 2008 Equity Incentive Plan, effective as of April 4, 2008. The following summary describes the material terms of the 2008 Equity Incentive Plan, as most recently amended effective August 3, 2013. This summary of the 2008 Equity Incentive Plan is not a complete description of all provisions of the 2008 Equity Incentive Plan and is qualified in its entirety by reference to the 2008 Equity Incentive Plan, which will be filed as an exhibit to the registration statement of which this prospectus is a part.

Administration. The 2008 Equity Incentive Plan is administered by our Board of Directors. Our Board of Directors has the authority to, among other things, determine to which of the eligible persons under the plan awards will be granted, determine the type of award to grant, approve forms of award agreements, determine the number of shares subject to, and the terms and conditions of, an award, construe and interpret the plan and awards and establish, amend and revoke rules and regulations for the administration of the plan and awards, correct defects in the plan and awards and generally, to exercise such powers and perform such acts as it deems to be necessary or expedient to make the plan fully effective. Our Board of Directors' determinations under the 2008 Equity Incentive Plan are final and conclusive.

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Eligibility. Our employees, directors, and consultants are eligible to participate in the 2008 Equity Incentive Plan. Eligibility for stock options intended to be incentive stock options, or ISOs, as defined in Section 422 of the Code, is limited to our employees.

Authorized Shares. Subject to adjustment, as described below, as of December 16, 2013, the number of shares of our common stock reserved for future issuance under the 2008 Equity Incentive Plan is 64,408 shares. The shares of our common stock to be issued under the 2008 Equity Incentive Plan may be authorized but unissued shares of our common stock or previously issued shares of our common stock acquired by us. Any shares of our common stock underlying awards that are settled in cash, forfeited, repurchased or otherwise reacquired by us, expired, cancelled or become unexercisable without having been exercised and any shares of our common stock used to satisfy an applicable tax withholding obligation will again be available for issuance under the 2008 Equity Incentive Plan.

Types of Awards. The 2008 Equity Incentive Plan provides for awards of stock options, restricted stock and unrestricted stock.

- *Stock options.* The exercise price of an ISO may not be less than the fair market value (or, in the case of an ISO granted to a ten percent shareholder, 110% of the fair market value) of shares of our common stock on the date of grant. The exercise price of each non-statutory stock option (or NSO) is the exercise price determined by our Board of Directors. The Board of Directors has set the exercise price of all NSOs granted under the 2008 Equity Incentive Plan at the fair market value of shares of our common stock as of the actual date of grant. Our Board of Directors will determine the time or times at which stock options become exercisable and the terms on which such awards remain exercisable.
- *Restricted stock and stock bonuses.* A restricted stock award is an award of shares of our common stock subject to forfeiture restrictions. A stock bonus is not subject to such restrictions. Our Board of Directors will determine the time or times at which any applicable vesting conditions and/or repurchase rights on restricted and unrestricted stock awards will lapse.

Vesting; Termination of Employment or Service. Our Board of Directors has the authority to determine the vesting schedule applicable to each award, and to accelerate the vesting or exercisability of any award. In the case of stock options, our Board of Directors may provide for early exercise of unvested options, with the stock received upon such exercise being subject to vesting. Our Board of Directors will determine the effect of termination of employment or service on an award. Unless otherwise provided in an award agreement, upon a termination of a participant's employment or service, all unvested stock options held by the participant on the date notice of termination is given to the optionee will terminate and all other unvested awards will be forfeited and all vested stock options then held by the participant will remain outstanding for one month following the provision of notice of such termination, or, in the case of death or disability, one year following the date of death or the provision of notice of termination by reason of disability or, in each case, until the applicable expiration date, if earlier. All stock options held by a participant immediately prior to the participant's termination of employment or service will immediately terminate if such termination is for cause, as defined in the 2008 Equity Incentive Plan. Unless otherwise provided by our Board of Directors, a stock bonus or restricted stock award shall cease vesting upon a participant's termination of employment or service (and, if applicable, the right to acquire any stock purchasable under such award will cease).

Non-Transferability of Awards. Awards under the 2008 Equity Incentive Plan may not be transferred other than by will or by the laws of descent and distribution, unless, for awards other than ISOs, otherwise provided in an award agreement (and subject, in the case of restricted stock and unrestricted stock awards to any applicable buy-sell or similar arrangements).

280G cutback. If any payment or right accruing to an individual under the 2008 Equity Incentive Plan would (alone or together with any other payment or right) constitute a "parachute payment" for purposes of Section 280G of the Code, then such payment or right under the 2008 Equity Incentive Plan will be reduced to

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the largest amount or greatest right that will result in no portion of such payment or right being a “parachute payment”. This provision will only apply if the individual would receive less on an after tax basis if he or she did not have his or her payment or right under the 2008 Equity Incentive Plan so reduced.

Certain Transactions; Certain Adjustments. In the event of a merger, consolidation, reorganization, recapitalization, reincorporation, stock dividend, dividend in property other than cash, stock split, liquidating dividend, combination of shares, exchange of shares, change in corporate structure or other transaction not involving the receipt of consideration by the corporation, the Board of Directors will appropriately adjust the maximum number of shares that can be issued under the 2008 Equity Incentive Plan, as well as all outstanding awards, except that no adjustment will be made if it would cause an equity award intended to be an ISO to fail to so qualify.

In the event of a deemed liquidity event (as defined in our certificate of incorporation), our Board of Directors may provide for substitute awards or such alternative consideration, including cash, as it deems equitable in the situation. In the event that our Board of Directors does not provide for such substitution or consideration in connection with a covered transaction, except as otherwise provided in an award agreement, all unexercised options will terminate automatically and, in the case of outstanding unvested restricted stock or stock bonus awards, will be forfeited automatically (in exchange for an amount equal to the original purchase price, if any) upon the consummation of such covered transaction. No additional awards may be made under the 2008 Equity Incentive Plan following a covered transaction.

Amendment; Termination. Our Board of Directors may, in its discretion, amend the 2008 Equity Incentive Plan or suspend or terminate the 2008 Equity Incentive Plan at any time, except that our Board of Directors may not reduce any outstanding award without the participant’s written consent. Shareholder approval will be required for any amendment to the 2008 Equity Incentive Plan to the extent such approval is required by law. Unless earlier terminated by our Board of Directors, the 2008 Equity Incentive Plan will terminate by its terms on April 3, 2018.

2012 Director Compensation

The following table sets forth a summary of the compensation we paid to our non-employee directors during 2012. Other than as set forth in the table below, we did not pay any compensation, make any equity awards or non-equity awards to, or pay any other compensation to any of the other non-employee members of our board of directors in 2012. Mr. Gardner, our former President and Chief Executive Officer, received no compensation for his service as a director, and, consequently, is not included in this table. The compensation received by Mr. Gardner as an employee during 2012 is presented in “Summary Compensation Table” above.

<u>Name</u>	<u>All Other Compensation (\$)⁽¹⁾</u>
Anupam Dalal	5,238
Campbell Murray	—
John Rice	863
Paul Weiss	6,000

(1) Amounts represent reimbursement of travel and expenses in connection with the individual’s service as a director.

Certain Relationships and Related Party Transactions

Since January 1, 2010, we have engaged in the following transactions with our directors and executive officers and holders of more than 5% of our voting securities and affiliates of our directors, executive officers and such 5% stockholders. We believe that all of the transactions described below were made on terms no less favorable to us than could have been obtained from unaffiliated third parties.

Preferred Stock Financings

Series A Preferred Stock Financing

In June 2010, we issued and sold an aggregate of 125,000 shares of our Series A preferred stock at a purchase price of \$40.00 per share for an aggregate purchase price of \$5 million. The following table sets forth the number of shares of our Series A preferred stock that we issued to our directors, executive officers and 5% stockholders at the time of such issuance and their affiliates, in connection with this transaction and the aggregate cash purchase price paid by these related parties:

<u>Investor</u>	<u>Shares of Series A Preferred Stock</u>	<u>Purchase Price (\$)</u>
Triathlon Medical Ventures	12,453	498,120
Novartis Bioventures Ltd.	61,250	2,450,000
Venture Investors Early Stage Fund IV	32,747	1,309,889
Kearny Venture Partners, L.P. and affiliates ⁽¹⁾	6,806	272,226
Joseph Gardner ⁽²⁾	2,834	113,345
Ian Howes ⁽³⁾	1,250	50,000

- (1) Consists of 3,335 shares purchased by Kearny Venture Partners, L.P., 68 shares purchased by Kearny Venture Partners Entrepreneurs Fund, L.P., and 3,403 shares purchased by Thomas Weisel Healthcare Venture.
- (2) Consists of 625 shares purchased by Joseph Gardner and 2,209 shares purchased by the Gardner Family Trust. Dr. Gardner was our former President and Chief Executive Officer.
- (3) Consists of 1,250 shares purchased by Ian A.W. Howes, IRA, Sterling Trust Custodian. Mr. Howes was our former Chief Financial Officer.

Series B Preferred Stock Financing

In April 2011 and December 2011, we issued and sold an aggregate of 1,287,525 shares of our Series B preferred stock at a purchase price of \$14.00 per share for an aggregate purchase price of \$18,025,341. As part of this financing, various trusts and other entities affiliated with Muneer A. Satter collectively purchased 260,873 shares of our Series B preferred stock and immediately following this purchase became a beneficial owner of more than 5% of our voting securities. Furthermore, as part of this financing, AgeChem Venture Fund L.P. purchased 173,915 shares of our Series B preferred stock and immediately following this purchase became a beneficial owner of more than 5% of our voting securities.

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The following table sets forth the number of shares of our Series B preferred stock that we issued to our directors, executive officers and 5% stockholders at the time of such issuance and their affiliates, in connection with this transaction and the aggregate cash purchase price paid by these related parties:

<u>Investor</u>	<u>Shares of Series B Preferred Stock</u>	<u>Purchase Price (\$)</u>
Triathlon Medical Ventures	124,502	1,743,024
Novartis Bioventures Ltd.	347,831	4,869,630
Venture Investors Early Stage Fund IV	173,915	2,434,815
Kearny Venture Partners, L.P. and affiliates ⁽¹⁾	88,478	1,238,692
Joseph Gardner ⁽²⁾	18,882	264,354
Robert Shalwitz	2,070	28,986
Ian Howes ⁽³⁾	5,797	81,161

- (1) Consists of 43,355 shares purchased by Kearny Venture Partners, L.P., 884 shares purchased by Kearny Venture Partners Entrepreneurs Fund, L.P., and 44,239 shares purchased by Thomas Weisel Healthcare Venture.
- (2) Consists of 11,594 shares purchased by Joseph Gardner and 7,288 shares purchased by the Gardner Family Trust.
- (3) Consists of 5,797 shares purchased by Ian A.W. Howes, IRA, Sterling Trust Custodian.

Series X Preferred Stock Financing

In July 2012 and March 2013, we issued and sold an aggregate of 50,000 shares of our Series X preferred stock, at a purchase price of \$100.00 per share, for an aggregate purchase price of \$5,000,002.

The following table sets forth the number of shares of our Series X preferred stock that we issued to our directors, executive officers and 5% stockholders at the time of such issuance and their affiliates, in connection with this transaction and the aggregate cash purchase price paid by these related parties:

<u>Investor</u>	<u>Shares of Series X Preferred Stock</u>	<u>Purchase Price (\$)</u>
Triathlon Medical Ventures	6,576	657,576
Novartis Bioventures Ltd.	15,211	1,521,064
Venture Investors Early Stage Fund IV	8,253	825,348
Kearny Venture Partners, L.P. and affiliates ⁽¹⁾	4,490	449,004
Trusts and Other Entities Affiliated with Muneer A. Satter	3,504	350,394
Joseph Gardner ⁽²⁾	2,042	204,165
Ian Howes ⁽³⁾	406	40,631
AgeChem Venture Fund L.P.	2,240	224,026

- (1) Consists of 220 shares purchased by Kearny Venture Partners, L.P., 45 shares purchased by Kearny Venture Partners Entrepreneurs Fund, L.P., and 2,245 shares purchased by Thomas Weisel Healthcare Venture.
- (2) Consists of 1,694 shares purchased by Joseph Gardner and 348 shares purchased by the Gardner Family Trust.
- (3) Consists of 406 shares purchased by Ian A.W. Howes, IRA, Sterling Trust Custodian.

Series C Preferred Stock Conversion

In May 2013, we issued an aggregate of 357,143 shares of Series C preferred stock at an exchange rate of 7.14286 shares of Series C preferred stock for every share of Series X preferred stock.

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The following table sets forth the number of shares of our Series C preferred stock that we issued to our directors, executive officers and 5% stockholders at the time of such issuance and their affiliates, in exchange for their shares of Series X preferred stock:

<u>Investor</u>	<u>Shares of Series C Preferred Stock</u>
Triathlon Medical Ventures	6,576
Novartis Bioventures Ltd.	15,211
Venture Investors Early Stage Fund IV	8,253
Kearny Venture Partners, L.P. and affiliates ⁽¹⁾	4,490
Trusts and Other Entities Affiliated with Muneer A. Satter	3,504
Joseph Gardner ⁽²⁾	2,042
Ian Howes ⁽³⁾	406
AgeChem Venture Fund L.P.	2,240

- (1) Consists of 2,200 shares held by Kearny Venture Partners, L.P., 45 shares held by Kearny Venture Partners Entrepreneurs Fund, L.P., and 2,245 shares held by Thomas Weisel Healthcare Venture.
- (2) Consists of 1,694 shares held by Joseph Gardner and 348 shares held by the Gardner Family Trust.
- (3) Consists of 406 shares held by Ian A.W. Howes, IRA, Sterling Trust Custodian.

Series C Preferred Stock Financing

In May 2013, we issued and sold an aggregate of 2,945,742 shares of our Series C preferred stock, at a purchase price of \$14.00 per share, for an aggregate purchase price of \$41,240,388. As part of this financing, Novo A/S purchased 714,285 shares of our Series C preferred stock and immediately following this purchase became a beneficial owner of more than 5% of our voting securities.

The following table sets forth the number of shares of our Series C preferred stock that we issued to our directors, executive officers and 5% stockholders at the time of such issuance and their affiliates, in connection with this transaction and the aggregate cash purchase price paid by these related parties:

<u>Investor</u>	<u>Shares of Series C Preferred Stock</u>	<u>Purchase Price (\$)</u>
Triathlon Medical Ventures	71,428	999,992
Novartis Bioventures Ltd.	600,000	8,400,000
Venture Investors Early Stage Fund IV	142,858	2,000,012
Kearny Venture Partners, L.P. and affiliates ⁽¹⁾	357,143	5,000,002
Trusts and Other Entities Affiliated with Muneer A. Satter	471,425	6,599,950
Robert Shalwitz	2,500	35,000
Joseph Gardner	14,285	199,990
Ian Howes	7,142	99,988

- (1) Consists of 292,733 shares purchased by Kearny Venture Partners, L.P., 5,970 shares purchased by Kearny Venture Partners Entrepreneurs Fund, L.P., and 58,440 shares purchased by Thomas Weisel Healthcare Venture.

Indemnification Agreements

Prior to the completion of this offering, we expect to enter into indemnification agreements with each of our directors and executive officers. These agreements will require us to indemnify these individuals and, in certain cases, affiliates of such individuals, to the fullest extent permissible under Delaware law against liabilities that may arise by reason of their service to us or at our direction, and to advance expenses incurred as a result of any proceeding against them as to which they could be indemnified.

Employment Agreements

See the “Executive Compensation—Employment Agreements with Our Named Executive Officers” section of this prospectus for a further discussion of these agreements.

Investors’ Rights Agreement

In connection with our Series C preferred stock financing, on May 10, 2013, we entered into the Third Amended and Restated Investors’ Rights Agreement, or the investors’ rights agreement, with the holders of all of our then-outstanding shares of preferred stock including certain of our executive officers and entities with which certain of our directors are affiliated. The agreement provides that these holders have the right to demand that we file a registration statement with respect to the common stock issued upon conversion of the preferred stock. These holders may also request that shares of common stock held by them be included in certain registration statements that we are otherwise filing. See “Description of Capital Stock — Registration Rights.”

Right of First Refusal and Co-Sale Agreement

In connection with our Series C preferred stock financing, on May 10, 2013, we entered into an amendment to the Second Amended and Restated Right of First Refusal and Co-Sale Agreement with the holders of all of our then-outstanding shares of preferred stock including certain of our executive officers and entities with which certain of our directors are affiliated. Pursuant to the terms of this agreement, in the event of a proposed sale of shares of our common or preferred stock, the seller is required to first offer such shares to the company and to the other investors, subject to certain conditions and restrictions. This agreement will terminate upon the completion of this offering.

Voting Agreement

In connection with our Series C preferred stock financing on May 10, 2013, we entered into the Third Amended and Restated Voting Agreement with the holders of all of our then-outstanding shares of preferred stock including certain of our executive officers and entities with which certain of our directors are affiliated, with respect to the election of directors and certain other matters. All of our current directors were elected pursuant to the terms of this agreement. This agreement will terminate upon the completion of this offering.

Services Agreement

In connection with the spin out of our programs focused on the treatment of diabetic eye disease and inflammatory bowel disease into Aerpio, we entered into administrative services agreements with Aerpio on December 22, 2011, as amended and restated on August 27, 2012, and on November 1, 2012.

Under the terms of the administrative services agreements, starting in 2012, Akebia and Aerpio have obtained from and provided to each other certain services, including consulting services, access to facilities and equipment. Aerpio reimbursed Akebia for employee costs in the amount of \$2.0 million for the year ended December 31, 2012 and \$0.8 million for the nine months ended September 30, 2013. Aerpio paid us for facility-related charges in the amount of \$0.2 million for the year ended December 31, 2012 and \$0.2 million for the nine months ended September 30, 2013. As of October 31, 2013, the amounts due from Aerpio to us total \$98,502, and the amounts due from us to Aerpio total \$35,094.

Promissory Notes

We issued promissory notes to Joseph Gardner, our former President and Chief Executive Officer, in the aggregate amount of \$140,839. Dr. Gardner used these promissory notes to purchase restricted stock awards, as described in “Executive Compensation—2012 Outstanding Equity Awards at Fiscal Year-End.” As of

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December 1, 2013, the current balance of the outstanding promissory notes was \$112,831. The promissory notes are repayable at the earlier of (a) an initial public offering; (b) the sale of the company or substantially all of its assets; (c) the termination of the employee; or (d) five years from origination and will be extinguished prior to the filing of the registration statement of which this prospectus is a part.

Related Person Transactions Policy

Prior to completion of the offering, we will adopt a related person transaction approval policy that will govern the review of related person transactions following the closing of this offering. Pursuant to this policy, if we want to enter into a transaction with a related person or an affiliate of a related person, our Chief Financial Officer will review the proposed transaction to determine, based on applicable NASDAQ and SEC rules, if such transaction requires pre-approval by the audit committee and/or board of directors. If pre-approval is required, such matters will be reviewed at the next regular or special audit committee and/or board of directors meeting. We may not enter into a related person transaction unless our Chief Financial Officer has either specifically confirmed in writing that no further reviews are necessary or that all requisite corporate reviews have been obtained.

Principal Stockholders

The following table sets forth information relating to the beneficial ownership of our common stock as of November 30, 2013 by: each person, or group of affiliated persons, known by us to beneficially own more than 5% of our outstanding shares of common stock; each of our directors; each of our named executive officers; and all directors and executive officers as a group.

The number of shares beneficially owned by each entity, person, director or executive officer is determined in accordance with the rules of the SEC, and the information is not necessarily indicative of beneficial ownership for any other purpose. Under such rules, beneficial ownership includes any shares over which the individual has sole or shared voting power or investment power as well as any shares that the individual has the right to acquire within 60 days of November 30, 2013 through the exercise of any stock options or other rights. Except as otherwise indicated, and subject to applicable community property laws, the persons named in the table have sole voting and investment power with respect to all shares of common stock held by that person.

The percentage of shares beneficially owned is computed on the basis of 7,260,530 shares of our common stock outstanding as of November 30, 2013, which reflects the assumed conversion of all of our outstanding shares of preferred stock into an aggregate of 6,835,341 shares of common stock. Shares of our common stock that a person has the right to acquire within 60 days of November 30, 2013 are deemed outstanding for purposes of computing the percentage ownership of the person holding such rights, but are not deemed outstanding for purposes of computing the percentage ownership of any other person, except with respect to the percentage ownership of all directors and executive officers as a group. Unless otherwise indicated below, the address for each beneficial owner listed is c/o Akebia Therapeutics, Inc., 245 First Street, Suite 1100 Cambridge, MA 02142.

<u>Name and address of beneficial owner</u>	<u>Percentage of shares beneficially owned</u>	
	<u>Number of shares beneficially owned**</u>	<u>Before offering</u>
5% or greater stockholders:		
Triathlon Medical Ventures ⁽¹⁾	634,256	8.7%
Novartis Bioventures Ltd. ⁽²⁾	1,822,996	25.1%
Venture Investors Early Stage Fund IV ⁽³⁾	823,265	11.3%
Kearny Venture Partners, L.P. and related funds ⁽⁴⁾	755,437	10.4%
Novo A/S ⁽⁵⁾	743,286	10.2%
Trusts and Other Entities Affiliated with Muneer A. Satter ⁽⁶⁾	809,630	11.1%
Directors and named executive officers:		
Joseph H. Gardner ⁽⁷⁾	296,579	4.0%
John P. Butler	0	*
William Daly ⁽⁸⁾	74,910	1.0%
Robert Shalwitz, M.D. ⁽⁹⁾	137,596	1.8%
Muneer A. Satter ⁽⁶⁾	809,630	11.1%
Campbell Murray, M.D. ⁽²⁾	1,822,996	25.1%
Jack Nielsen	0	*
Anupam Dalal, M.D.	0	*
Giovanni Ferrara ⁽²⁾	1,822,996	25.1%
Kim Dueholm	0	*
Duane Nash	0	*
All executive officers and directors as a group (12 persons)	2,845,132	38.4%

* Represents beneficial ownership of less than one percent of our outstanding common stock.

** Fractional shares have been rounded down to the nearest whole number.

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- (1) Consists of 20,000 shares of common stock, 365,987 shares of common stock issuable upon conversion of Series A preferred stock, 124,501 shares of common stock issuable upon conversion of Series B preferred stock and 123,768 shares of common stock issuable upon conversion of Series C preferred stock held by Triathlon Medical Ventures Fund. Its general partner, Triathlon Medical Ventures LLC, has sole voting and investment control over the shares owned by Triathlon Medical Ventures Fund. The members of Triathlon Medical Ventures LLC, John Rice, Carrie Bates, Suzette Dutch and Dennis Costello, have sole voting and investment power for Triathlon Medical Ventures LLC with respect to its voting power in its capacity as the general partner for the shares held by Triathlon Medical Ventures Fund.
- (2) Consists of 734,374 shares of common stock issuable upon conversion of Series A preferred stock, 347,830 shares issuable upon conversion of Series B preferred stock and 740,791 shares of common stock issuable upon conversion of Series C preferred stock held by Novartis Bioventures Ltd, a Bermuda corporation. The board of directors of Novartis Bioventures Ltd. has sole voting and investment control and power over such shares. None of the members of its board of directors has individual voting or investment power with respect to such shares and each disclaims beneficial ownership of such shares. Mr. Campbell Murray and Mr. Giovanni Ferrara, two members of our Board of Directors (of which Mr. Murray is co-Chairman), are also employees of a corporation that is affiliated with Novartis Bioventures Ltd. They also disclaim beneficial ownership of shares held by Novartis Bioventures Ltd., except to the extent of their pecuniary interest arising as a result of their employment by that affiliate. Novartis Bioventures Ltd is an indirectly-owned subsidiary of Novartis AG.
- (3) Consists of 438,384 shares of common stock issuable upon conversion of Series A preferred stock, 173,915 shares of common stock issuable upon conversion of Series B preferred stock and 210,965 shares of common stock issuable upon conversion of Series C preferred stock. Venture Investors Early Stage Fund IV Limited Partnership is a Delaware Limited Partnership. Its General Partner, VIESF IV GP LLC, has sole voting and investment control over the shares owned by Venture Investors Early Stage Fund IV Limited Partnership. The members of VIESF IV GP LLC, John Neis, Paul M. Weiss, Scott Button, George Arida, James R. Adox, Loren G. Peterson, and Venture Investors Southeast LLC (of which Roger H. Ganser is the sole member), have sole voting and investment power for VIESF IV GP LLC with respect to its voting power in its capacity as General Partner for the shares held by Venture Investors Early Stage Fund IV Limited Partnership. None of the members of VIESF IV GP LLC has individual voting or investment power with respect to such shares and each disclaims beneficial ownership of such shares except to the extent of any pecuniary interest therein. The address of Venture Investors Early Stage Fund IV Limited Partnership is 505 South Rosa Road, Suite 201, Madison, Wisconsin, 53719.
- (4) Consists of (i) 127,446 shares of common stock issuable upon the conversion of Series A preferred stock, 43,354 shares of common stock issuable upon conversion of Series B preferred stock and 322,439 shares of common stock issuable upon conversion of Series C preferred stock that are held directly by Kearny Venture Partners, L.P. (“KVP”), (ii) 2,599 shares of common stock issuable upon the conversion of Series A preferred stock, 884 shares of common stock issuable upon conversion of Series B preferred stock and 6,575 shares of common stock issuable upon conversion of Series C preferred stock held by Kearny Venture Partners Entrepreneurs’ Fund, L.P. (“KVPE”), and (iii) 130,045 shares of common stock issuable upon conversion of Series A preferred stock, 44,239 shares of common stock issuable upon conversion of Series B preferred stock and 77,854 shares of common stock issuable upon conversion of Series C preferred stock that are held directly by Thomas Weisel Healthcare Venture Partners, L.P. (“TWHVP”). Each of KVP, KVPE and TWHVP is a Delaware limited partnership. The general partner of both KVP and KVPE is Kearny Venture Associates, L.L.C. (“KVA”). KVA has the sole voting and investment control over the shares owned by KVP and KVPE, and the Managing Members of KVA share in the voting and investment control over such shares controlled by KVA. The Managing Members of KVA are Caley Castelein, Richard Spalding and James Shapiro. None of the Managing Members of KVA has individual voting or investment power with respect to such shares and each disclaims beneficial

ownership of such shares except to the extent of any pecuniary interest therein. The address of KVA is 88 Kearny Street, San Francisco, CA 94108. The general partner of TWHVP is Thomas Weisel Healthcare Venture Partners LLC (“TWP GP”). TWP GP has the sole voting and investment control over the shares owned by TWHVP, and the investment committee of TWP GP has sole voting and investment control over the shares controlled by TWP GP. The investment committee of TWP GP consists of Richard Spalding and James Shapiro, neither of whom has individual voting or investment power with respect to such shares and each disclaims beneficial ownership of such shares except to the extent of any pecuniary interest therein. The address of TWP GP is One Montgomery St., San Francisco, CA 94104.

- (5) Consists of 743,286 shares of common stock issuable upon conversion of Series C preferred stock. Novo A/S is a Danish limited liability company. The board of directors of Novo A/S has sole voting and investment control over the shares owned by Novo A/S. The board of directors of Novo A/S, which consists of Sten Scheibye, Göran Ando, Jørgen Boe, Jeppe Christiansen, Steen Risgaard and Per Wold Olsen, has sole voting and investment power with respect to the shares held by Novo A/S. None of the members of the board of directors of Novo A/S has individual voting or investment power with respect to such shares and each disclaims beneficial ownership of such shares except to the extent of any pecuniary interest therein. Mr. Nielsen and Mr. Dueholm, two members of our board of directors, are employed as Partners of Novo A/S. Mr. Nielsen and Mr. Dueholm disclaim beneficial ownership of shares held by Novo A/S, except to the extent of their pecuniary interest arising as a result of their employment with Novo A/S. The address of Novo A/S is Tuborg Havnevej 19, 2900 Hellerup, Denmark.
- (6) Consists of 260,870 shares of common stock issuable upon conversion of Series B preferred stock and 548,759 shares of common stock issuable upon conversion of Series C preferred stock held by Muneer A. Satter Revocable Trust and various other trusts and other entities for which Mr. Satter serves as trustee, investment advisor or manager and, in such capacity, has sole voting and dispositive control over all such shares.
- (7) Joseph Gardner is the Company’s former Chief Executive Officer. This number consists of (i) 48,551 shares of common stock, 78,203 shares of common stock issuable upon the exercise of stock options, 75,220 shares of restricted stock, 25,825 shares of common stock issuable upon conversion of Series A preferred stock, 11,594 shares of common stock issuable upon conversion of Series B preferred stock and 27,579 shares of common stock issuable upon conversion of Series C preferred stock held by Joseph Gardner and (ii) 19,719 shares of common stock issuable upon conversion of Series A preferred stock, 7,288 shares of common stock issuable upon conversion of Series B preferred stock and 2,597 shares of common stock issuable upon conversion of Series C preferred stock held by the Gardner Family Trust.
- (8) Consists of 60,810 shares of restricted stock and 14,100 shares of common stock issuable upon conversion of Series C preferred stock.
- (9) Consists of (i) 4,000 shares of common stock, 59,159 shares of common stock issuable upon the exercise of stock options, 57,604 shares of restricted stock, 2,427 shares of common stock issuable upon conversion of Series A preferred stock, 2,070 shares of common stock issuable upon conversion of Series B preferred stock and 2,613 shares of common stock issuable upon conversion of Series C preferred stock held by Robert Shalwitz and (ii) 9,723 shares of common stock issuable upon conversion of Series A preferred stock held by Fred Shalwitz Trust.

Description of Capital Stock

General

The following description of our capital stock is intended as a summary only and is qualified in its entirety by reference to our ninth amended and restated certificate of incorporation and amended and restated bylaws that will be in effect at the closing of this offering, which will be filed as exhibits to the registration statement of which this prospectus is a part, and to the applicable provisions of the Delaware General Corporation Law. We refer in this section to our ninth amended and restated certificate of incorporation as our certificate of incorporation, and we refer to our amended and restated bylaws as our bylaws. The description of our capital stock reflects changes to our capital structure that will occur upon the closing of this offering.

Upon the closing of this offering, our authorized capital stock will consist of _____ shares of our common stock, par value \$0.0001 per share, and _____ shares of our preferred stock, par value \$0.0001 per share, all of which preferred stock will be undesignated.

As of September 30, 2013, we had issued and outstanding:

- 424,044 shares of our common stock, which includes 244,513 shares of restricted stock;
- 5,324,948 shares of our preferred stock that are convertible into 6,790,149 shares of our common stock; and
- options to purchase a total of 702,625 shares of our common stock with a weighted-average exercise price of \$1.08 per share.

As of September 30, 2013, we had 44 stockholders of record.

Common Stock

Dividend Rights. Subject to preferences that may apply to shares of preferred stock outstanding at the time, holders of outstanding shares of common stock will be entitled to receive dividends out of assets legally available at the times and in the amounts as the board of directors may from time to time determine.

Conversion or Redemption Rights. Our common stock will be neither convertible nor redeemable.

Liquidation Rights. Upon our liquidation, dissolution or winding up, the holders of our common stock will be entitled to receive pro rata our assets which are legally available for distribution, after payment of all debts and other liabilities and the satisfaction of any liquidation preference granted to the holders of any then-outstanding shares of preferred stock.

Rights and Preferences. Holders of common stock have no preemptive, conversion or subscription rights and there are no redemption or sinking fund provisions applicable to the common stock. The rights, preferences and privileges of the holders of common stock are subject to, and may be adversely affected by, the rights of the holders of shares of any series of preferred stock that we may designate in the future.

Preferred Stock

All currently outstanding shares of preferred stock will be converted automatically to common stock upon the completion of this offering.

Following the completion of this offering, our board of directors will have the authority, without further action by our stockholders, to issue up to _____ shares of preferred stock in one or more series, to establish from time to time the number of shares to be included in each such series, to fix the rights, preferences and

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privileges of the shares of each wholly unissued series and any qualifications, limitations or restrictions thereon, and to increase or decrease the number of shares of any such series, but not below the number of shares of such series then outstanding.

Our board of directors may authorize the issuance of preferred stock with voting or conversion rights that could adversely affect the voting power or other rights of the holders of our common stock. The purpose of authorizing our board of directors to issue preferred stock and determine its rights and preferences is to eliminate delays associated with a stockholder vote on specific issuances. The issuance of preferred stock, while providing flexibility in connection with possible acquisitions and other corporate purposes, could, among other things, have the effect of delaying, deferring or preventing a change in control of us and may adversely affect the market price of our common stock and the voting and other rights of the holders of our common stock. It is not possible to state the actual effect of the issuance of any shares of preferred stock on the rights of holders of common stock until the board of directors determines the specific rights attached to that preferred stock.

We have no present plans to issue any shares of preferred stock.

Registration Rights

After our initial public offering, holders of 7,381,221 shares of our common stock issued or issuable (as calculated as of September 30, 2013) will be entitled to certain rights with respect to registration of such shares under the Securities Act. These shares are collectively referred to herein as registrable shares. These rights are provided under the terms of the investors' rights agreement, and include demand registration rights, Form S-3 registration rights and piggyback registration rights. These registration rights are subject to conditions and limitations, including the right, in certain circumstances, of the underwriters of an offering to limit the number of shares included in such registration and our right, in certain circumstances, not to effect a requested S-1 or S-3 registration within 60 days before or 180 days following our estimated date of filing of a registration statement pertaining to an underwritten public offering of securities for the account of an offering of our securities, including this offering.

Demand Registration Rights

Under the terms of the investors' rights agreement, following the six-month anniversary of the completion of this offering, the holders of at least 30% of the registrable shares may require us to file a registration statement on Form S-1 under the Securities Act at our expense with respect to the resale of their registrable shares as soon as practicable, and in any event within 60 days after the date of the request for registration. We are required to effect only two registrations pursuant to this provision of the investors' rights agreement.

Under the terms of the investors' rights agreement, if we are eligible to file a registration statement on Form S-3, the holders of at least 30% of the registrable shares may require us to file a registration statement on Form S-3 at our expense with respect to the resale of their registrable shares as soon as practicable, and in any event within 45 days after the date of the request for registration. We are required to effect only three registrations pursuant to this provision of the investors' rights agreement.

Piggyback Registration Rights

Under the terms of the investors' rights agreement, if we propose to register any of our common stock under the Securities Act in connection with the public offering of such securities solely for cash except for certain excluded registrations, the holders of registrable shares are entitled to notice of such registration and to request that we include registrable shares for resale on such registration statement, subject to our right to terminate or withdraw any registration we initiate prior to its effective date and the right of any underwriter to limit the number of shares included in such registration.

Expenses of Registration

We will pay all expenses relating to any demand, Form S-3 or piggyback registration, other than underwriting discounts, selling commissions and stock transfer taxes applicable to the sale of registrable securities, subject to specified conditions and limitations.

Anti-Takeover Effects of Our Certificate of Incorporation and Our Bylaws

Our certificate of incorporation and bylaws will contain certain provisions that are intended to enhance the likelihood of continuity and stability in the composition of the board of directors and which may have the effect of delaying, deferring or preventing a future takeover or change in control of the company unless such takeover or change in control is approved by the board of directors.

These provisions include:

Classified Board. Our certificate of incorporation will provide that our board of directors will be divided into three classes of directors, with the classes as nearly equal in number as possible. As a result, approximately one-third of our board of directors will be elected each year. The classification of directors will have the effect of making it more difficult for stockholders to change the composition of our board. Our certificate of incorporation will provide that the affirmative vote of holders of at least 85% of the total votes eligible to be cast in the election of directors will be required to amend, alter, change or repeal such classification of directors. Our certificate of incorporation will also provide that, subject to any rights of holders of preferred stock to elect additional directors under specified circumstances, the number of directors will be fixed exclusively pursuant to a resolution adopted by our board of directors. Upon completion of this offering, we expect that our board of directors will have eight members.

Action by Written Consent; Special Meetings of Stockholders. Our certificate of incorporation will provide that stockholder action can be taken only at an annual or special meeting of stockholders and cannot be taken by written consent in lieu of a meeting. Our certificate of incorporation and the bylaws will also provide that, except as otherwise required by law, special meetings of the stockholders can be called only by or at the direction of the board of directors pursuant to a resolution adopted by a majority of the total number of directors. Stockholders will not be permitted to call a special meeting or to require the board of directors to call a special meeting.

Removal of Directors. Our certificate of incorporation will provide that our directors may be removed only for cause by the affirmative vote of at least 75% of the votes that all our stockholders would be entitled to cast in an annual election of directors, voting together as a single class, at a meeting of the stockholders called for that purpose. This requirement of a supermajority vote to remove directors could enable a minority of our stockholders to prevent a change in the composition of our board.

Advance Notice Procedures. Our bylaws will establish an advance notice procedure for stockholder proposals to be brought before an annual meeting of our stockholders, including proposed nominations of persons for election to the board of directors. Stockholders at an annual meeting will only be able to consider proposals or nominations specified in the notice of meeting or brought before the meeting by or at the direction of the board of directors or by a stockholder who was a stockholder of record on the record date for the meeting, who is entitled to vote at the meeting and who has given our Secretary timely written notice, in proper form, of the stockholder's intention to bring that business before the meeting. Although the bylaws will not give the board of directors the power to approve or disapprove stockholder nominations of candidates or proposals regarding other business to be conducted at a special or annual meeting, the bylaws may have the effect of precluding the conduct of certain business at a meeting if the proper procedures are not followed or may discourage or deter a potential acquirer from conducting a solicitation of proxies to elect its own slate of directors or otherwise attempting to obtain control of the company.

Super Majority Approval Requirements. The Delaware General Corporation Law generally provides that the affirmative vote of a majority of the shares entitled to vote on any matter is required to amend a corporation's

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certificate of incorporation or bylaws, unless either a corporation's certificate of incorporation or bylaws requires a greater percentage. A majority vote of our board of directors or the affirmative vote of holders of at least 75% of the total votes of the outstanding shares of capital stock of the Company entitled to vote with respect thereto, voting together as a single class, will be required to amend, alter, change or repeal the bylaws. In addition, the affirmative vote of the holders of at least 75% of the total votes of the outstanding shares of capital stock of the Company entitled to vote with respect thereto, voting together as a single class, will be required to amend, alter, change or repeal, or to adopt any provisions inconsistent with, any of the provisions in our certificate of incorporation relating to amendments to our certificate of incorporation and bylaws and as described under "Action by Written Consent; Special Meetings of Stockholders" and "Removal of Directors" above. This requirement of a supermajority vote to approve amendments to our bylaws and certificate of incorporation could enable a minority of our stockholders to exercise veto power over any such amendments.

Authorized but Unissued Shares. Our authorized but unissued shares of common stock and preferred stock will be available for future issuance without stockholder approval. These additional shares may be utilized for a variety of corporate purposes, including future public offerings to raise additional capital and corporate acquisitions. The existence of authorized but unissued shares of common stock and preferred stock could render more difficult or discourage an attempt to obtain control of a majority of our common stock by means of a proxy contest, tender offer, merger or otherwise.

Exclusive Forum. Our certificate of incorporation will provide that, subject to limited exceptions, the state or federal courts located in the State of Delaware will be the sole and exclusive forum for (i) any derivative action or proceeding brought on our behalf, (ii) any action asserting a claim of breach of a fiduciary duty owed by any of our directors, officers or other employees to us or our stockholders, (iii) any action asserting a claim against us arising pursuant to any provision of the Delaware General Corporation Law, our certificate of incorporation or our bylaws, or (iv) any other action asserting a claim against us that is governed by the internal affairs doctrine. Any person or entity purchasing or otherwise acquiring any interest in shares of our capital stock shall be deemed to have notice of and to have consented to the provisions of our certificate of incorporation described above. Although we believe these provisions benefit us by providing increased consistency in the application of Delaware law for the specified types of actions and proceedings, the provisions may have the effect of discouraging lawsuits against our directors and officers. The enforceability of similar choice of forum provisions in other companies' certificates of incorporation has been challenged in legal proceedings, and it is possible that, in connection with one or more actions or proceedings described above, a court could find the choice of forum provisions contained in our certificate of incorporation to be inapplicable or unenforceable.

Section 203 of the Delaware General Corporation Law

Upon completion of this offering, we will be subject to the provisions of Section 203 of the Delaware General Corporation Law. In general, Section 203 prohibits a publicly-held Delaware corporation from engaging in a "business combination" with an "interested stockholder" for a three-year period following the time that this stockholder becomes an interested stockholder, unless the business combination is approved in a prescribed manner. A "business combination" includes, among other things, a merger, asset or stock sale or other transaction resulting in a financial benefit to the interested stockholder. An "interested stockholder" is a person who, together with affiliates and associates, owns, or did own within three years prior to the determination of interested stockholder status, 15% or more of the corporation's voting stock.

Under Section 203, a business combination between a corporation and an interested stockholder is prohibited unless it satisfies one of the following conditions: before the stockholder became interested, the board of directors approved either the business combination or the transaction which resulted in the stockholder becoming an interested stockholder; upon consummation of the transaction which resulted in the stockholder becoming an interested stockholder, the interested stockholder owned at least 75% of the voting stock of the corporation outstanding at the time the transaction commenced, excluding for purposes of determining the voting stock outstanding, shares owned by persons who are directors and also officers, and employee stock plans, in some instances; or at or after the time the

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stockholder became interested, the business combination was approved by the board of directors of the corporation and authorized at an annual or special meeting of the stockholders by the affirmative vote of at least two-thirds of the outstanding voting stock which is not owned by the interested stockholder.

A Delaware corporation may “opt out” of these provisions with an express provision in its original certificate of incorporation or an express provision in its certificate of incorporation or bylaws resulting from a stockholders’ amendment approved by at least a majority of the outstanding voting shares. We have not opted out of these provisions. As a result, mergers or other takeover or change in control attempts of us may be discouraged or prevented.

Transfer Agent and Registrar

The transfer agent and registrar for our common stock is .

Listing

We intend to apply for listing of our common stock on the NASDAQ Global Market under the symbol AKBA.

Shares Eligible for Future Sale

Prior to this offering, there has been no public market for our common stock. Future sales of our common stock, including shares issued upon the exercise of outstanding options, in the public market after this offering, or the perception that those sales may occur, could cause the prevailing market price for our common stock to fall or impair our ability to raise equity capital at a time and price we deem appropriate. As described below, only a limited number of shares of our common stock will be available for sale in the public market for a period of several months after completion of this offering due to contractual and legal restrictions on resale described below.

Sale of Restricted Shares

Based on the number of shares of our common stock outstanding as of September 30, 2013, upon the closing of this offering and assuming (1) the conversion of our outstanding preferred stock into common stock, (2) no exercise of the underwriters' option to purchase additional shares of common stock, and (3) no exercise of outstanding options, we would have had outstanding an aggregate of approximately _____ shares of common stock. Of these shares, all of the shares of common stock to be sold in this offering, and any shares sold upon exercise of the underwriters' option to purchase additional shares, will be freely tradable in the public market without restriction or further registration under the Securities Act, unless the shares are held by any of our affiliates as such term is defined in Rule 144 of the Securities Act. All remaining shares of common stock held by existing stockholders immediately prior to the completion of this offering will be restricted securities as such term is defined in Rule 144. These restricted securities were issued and sold by us in private transactions and are eligible for public sale only if registered under the Securities Act or if they qualify for an exemption from registration under the Securities Act, including the exemptions provided by Rule 144 or Rule 701, which rules are summarized below.

As a result of the lock-up agreements referred to below and the provisions of Rule 144 and Rule 701 under the Securities Act, the shares of our common stock (excluding the shares sold in this offering) that will be available for sale in the public market are as follows:

<u>Approximate Number of Shares</u>	<u>First Date Available for Sale into Public Market</u>
	180 days after the date of this prospectus upon expiration of the lock-up agreements referred to below, subject in some cases to applicable volume limitations under Rule 144

Lock-up Agreements

In connection with this offering, we, and all of our directors and officers, and the holders of substantially all of our outstanding stock and stock options have agreed that, without the prior written consent of Morgan Stanley & Co. LLC and Credit Suisse Securities (USA) LLC on behalf of the underwriters, we and they will not, during the period ending 180 days after the date of this prospectus (the "restricted period"):

- offer, pledge, sell, contract to sell, sell any option or contract to purchase, purchase any option or contract to sell, grant any option, right or warrant to purchase, lend, or otherwise transfer or dispose of, directly or indirectly, any shares of our common stock or any securities convertible into or exercisable or exchangeable for our common stock; or
- enter into any swap or other arrangement that transfers to another, in whole or in part, any of the economic consequences of ownership of our common stock,

whether any such transaction described above is to be settled by delivery of common stock or such other securities, in cash or otherwise. In addition, we and all of our directors and officers, and the holders of substantially all of our common stock and stock options have agreed that, without the prior written consent of Morgan Stanley & Co. LLC and Credit Suisse Securities (USA) LLC on behalf of the underwriters, during

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the restricted period, no registration statement with the SEC relating to the offering of any shares of common stock or any security convertible into or exercisable or exchangeable for our common stock will be filed.

The restrictions described in the immediately preceding paragraph do not apply to:

- the sale of shares by us to the underwriters;
- the issuance by us of shares of our common stock upon the exercise of an option or warrant or the conversion of a security outstanding on the date of this prospectus of which the underwriters have been advised in writing;
- the establishment of a trading plan pursuant to Rule 10b5-1 under the Exchange Act for the transfer of shares of our common stock; *provided* that such plan does not provide for the transfer of shares of our common stock during the restricted period and to the extent a public announcement or filing under the Exchange Act, if any, is required of or voluntarily made regarding the establishment of such plan, such announcement or filing shall include a statement to the effect that no transfer of common stock may be made under such plan during the restricted period;
- transactions relating to shares of our common stock or other securities acquired in this offering (other than any shares of our common stock directed by us and purchased in this offering by one of our officers or directors) or in open market transactions after the date of the final prospectus;
- transfers of shares of our common stock, or any security convertible into, exercisable or exchangeable for our common stock, as a bona fide gift;
- transfers of shares of our common stock, or any security convertible into, exercisable or exchangeable for our common stock, by will or intestacy;
- the exercise of options to purchase shares of our common stock granted under any existing stock incentive plan or stock purchase plan described in this prospectus, *provided* that any shares of our common stock issued pursuant to such exercise shall be subject to the same restrictions;
- transfers to us for the purpose of satisfying tax withholding obligations upon the vesting of other equity incentive awards granted under any existing stock incentive plan or stock purchase plan described in this prospectus;
- transfers or distributions not involving a disposition for value of shares of our common stock, or any security convertible into, exercisable or exchangeable for our common stock, to any limited or general partners, stockholders or members of a lock-up signatory, or if the lock-up signatory is a corporation, to a wholly-owned subsidiary of such lock-up signatory;
- transfers of shares of our common stock, or any security convertible into, exercisable or exchangeable for our common stock, made by one of the lock-up signatories to (i) any trust, corporation, partnership, limited liability company or other legal entity who, directly or indirectly, controls, is controlled by, or is under common control with such lock-up signatory, (ii) any trust or other legal entity for which a lock-up signatory or the spouse of a lock-up signatory serves as trustee or investment advisor, or (iii) any member of the immediate family of a lock-up signatory, or any trust or other legal entity for the direct or indirect benefit of a lock-up signatory or any member of the immediate family of a lock-up signatory;
- transfers of shares of our common stock, or any securities convertible into, exercisable or exchangeable for our common stock, pursuant to a sale of, or an offer to purchase, 100% of our outstanding common stock, whether pursuant to a merger, tender offer or otherwise, to a third party or group of third parties, *provided* that in the event that such tender offer, merger, or transaction is not completed, our common stock and any security convertible into or exchangeable for our common stock shall remain subject to the same restrictions; or
- the conversion of our outstanding preferred stock into shares of our common stock upon the closing of this offering, *provided* that such shares of our common stock shall remain subject to the same restrictions,

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provided, however, that in the case of any transfer or distribution pursuant to the fifth, sixth, ninth or tenth clauses above, each donee, distributee or transferee shall sign and deliver a lock-up agreement substantially in the form of the lock-up agreements described above; and in the case of any transaction, transfer, exercise or distribution pursuant to the fourth through (and including) the tenth clauses above, no filing under Section 16(a) of the Exchange Act, reporting a reduction in beneficial ownership of shares of our common stock, shall be required or shall be voluntarily made during the restricted period (other than a filing on Form 5 made after the expiration of the restricted period).

Following the lock-up periods set forth in the agreements described above, all of the shares of our common stock that are restricted securities or are held by our affiliates as of the date of this prospectus will be eligible for sale in the public market in compliance with Rule 144 under the Securities Act.

In addition to the restrictions contained in the lock-up agreements described above, we have entered into agreements with certain of our security holders, including our amended and restated investors rights agreement and the standard forms of our option agreements under our equity incentive plans, that contain market stand-off provisions imposing restrictions on the ability of such security holders to offer, sell or transfer our equity securities for a period of 180 days following the date of this prospectus.

Rule 144

Affiliate Resales of Restricted Securities

In general, beginning 90 days after the effective date of the registration statement of which this prospectus is a part, a person who is an affiliate of ours, or who was an affiliate at any time during the 90 days before a sale, who has beneficially owned shares of our common stock for at least six months would be entitled to sell in “broker’s transactions” or certain “riskless principal transactions” or to market makers, a number of shares within any three-month period that does not exceed the greater of:

- 1% of the number of shares of our common stock then outstanding, which will equal approximately shares immediately after this offering; or
- the average weekly trading volume in our common stock on the NASDAQ Global Market during the four calendar weeks preceding the filing of a notice on Form 144 with respect to such sale.

Affiliate resales under Rule 144 are also subject to the availability of current public information about us. In addition, if the number of shares being sold under Rule 144 by an affiliate during any three-month period exceeds 5,000 shares or has an aggregate sale price in excess of \$50,000, the seller must file a notice on Form 144 with the Securities and Exchange Commission and NASDAQ concurrently with either the placing of a sale order with the broker or the execution directly with a market maker.

Non-Affiliate Resales of Restricted Securities

In general, beginning 90 days after the effective date of the registration statement of which this prospectus is a part, a person who is not an affiliate of ours at the time of sale, and has not been an affiliate at any time during the 90 days preceding a sale, and who has beneficially owned shares of our common stock for at least six months but less than a year, is entitled to sell such shares subject only to the availability of current public information about us. If such person has held our shares for at least one year, such person can resell under Rule 144(b)(1) without regard to any Rule 144 restrictions, including the 90-day public company requirement and the current public information requirement.

Non-affiliate resales are not subject to the manner of sale, volume limitation or notice filing provisions of Rule 144.

Rule 701

In general, under Rule 701, any of an issuer's employees, directors, officers, consultants or advisors who purchases shares from the issuer in connection with a qualified compensatory stock or option plan or other written agreement before the effective date of a registration statement under the Securities Act is entitled to sell such shares 90 days after such effective date in reliance on Rule 144. An affiliate of the issuer can resell shares in reliance on Rule 144 without having to comply with the holding period requirement, and non-affiliates of the issuer can resell shares in reliance on Rule 144 without having to comply with the current public information and holding period requirements.

The Securities and Exchange Commission has indicated that Rule 701 will apply to stock options granted by an issuer before it becomes subject to the reporting requirements of the Exchange Act, along with the shares acquired upon exercise of such options, including exercises after an issuer becomes subject to the reporting requirements of the Exchange Act.

Registration Rights

Upon the completion of this offering, the holders of 7,381,221 shares of our common stock issued or issuable (as calculated as of September 30, 2013) will be entitled to specified rights with respect to the registration of the offer and sale of their shares under the Securities Act. Registration of these shares under the Securities Act would result in the shares becoming freely tradable without restriction under the Securities Act immediately upon the effectiveness of the registration statement. See the section of this prospectus titled "Description of Capital Stock—Registration Rights" for additional information.

Equity Incentive Plans

We intend to file with the SEC a registration statement on Form S-8 under the Securities Act covering the shares of common stock that we may issue upon exercise of outstanding options or options or other equity awards to be issued under our Amended and Restated 2008 Equity Incentive Plan and 2014 Equity Incentive Plan. Such registration statement is expected to be filed and become effective as soon as practicable after the completion of this offering. Accordingly, shares registered under such registration statement will be available for sale in the open market following its effective date, subject to Rule 144 restrictions on affiliates and the lock-up agreements described above, if applicable.

**Material United States Federal Income Tax
Considerations for Non-U.S. Holders**

The following is a summary of the material U.S. federal income and estate tax considerations relating to the purchase, ownership and disposition of our common stock by Non-U.S. Holders (defined below). This summary does not purport to be a complete analysis of all the potential tax considerations relevant to Non-U.S. Holders of our common stock. This summary is based upon the Code, the Treasury regulations promulgated or proposed thereunder and administrative and judicial interpretations thereof, all as of the date hereof and all of which are subject to differing interpretations and to change at any time, possibly on a retroactive basis.

This summary assumes that shares of our common stock are held as “capital assets” within the meaning of Section 1221 of the Code (generally, property held for investment). This summary does not purport to deal with all aspects of U.S. federal income and estate taxation that might be relevant to particular Non-U.S. Holders in light of their particular investment circumstances or status, nor does it address specific tax considerations that may be relevant to particular persons (including, for example, financial institutions, broker-dealers, insurance companies, partnerships or other pass-through entities, certain U.S. expatriates, tax-exempt organizations, pension plans, “controlled foreign corporations”, “passive foreign investment companies”, corporations that accumulate earnings to avoid U.S. federal income tax, persons in special situations, such as those who have elected to mark securities to market or those who hold common stock as part of a straddle, hedge, conversion transaction, synthetic security or other integrated investment, or holders subject to the alternative minimum tax or the 3.8% Medicare tax on net investment income). In addition, except as explicitly addressed herein with respect to estate tax, this summary does not address estate and gift tax considerations or considerations under the tax laws of any state, local or non-U.S. jurisdiction.

For purposes of this summary, a “Non-U.S. Holder” means a beneficial owner of common stock that for U.S. federal income tax purposes is not an entity treated as a partnership and is not:

- an individual who is a citizen or resident of the United States;
- a corporation or any other organization taxable as a corporation for U.S. federal income tax purposes, created or organized in or under the laws of the United States, any state thereof or the District of Columbia;
- an estate, the income of which is included in gross income for U.S. federal income tax purposes regardless of its source; or
- a trust if (1) a U.S. court is able to exercise primary supervision over the trust’s administration and one or more U.S. persons have the authority to control all of the trust’s substantial decisions or (2) the trust has a valid election in effect under applicable Treasury regulations to be treated as a U.S. person.

If an entity that is treated as a partnership for U.S. federal income tax purposes holds our common stock, the tax treatment of persons treated as its partners for U.S. federal income tax purposes will generally depend upon the status of the partner and the activities of the partnership. Entities that are treated as partnerships for U.S. federal income tax purposes and persons holding our common stock through an entity treated as a partnership for U.S. federal income tax purposes are urged to consult their own tax advisors.

There can be no assurance that the Internal Revenue Service (“IRS”) will not challenge one or more of the tax consequences described herein, and we have not obtained, nor do we intend to obtain a ruling from the IRS with respect to the U.S. federal income or estate tax consequences to a Non-U.S. Holder of the purchase, ownership or disposition of our common stock.

THIS SUMMARY IS NOT INTENDED TO BE TAX ADVICE. NON-U.S. HOLDERS ARE URGED TO CONSULT THEIR TAX ADVISORS CONCERNING THE U.S. FEDERAL INCOME AND ESTATE TAXATION, STATE, LOCAL AND NON-U.S. TAXATION AND OTHER TAX CONSEQUENCES TO THEM OF THE PURCHASE, OWNERSHIP AND DISPOSITION OF OUR COMMON STOCK.

Distributions on Our Common Stock

As discussed under “Dividend Policy” above, we do not anticipate paying any cash dividends in the foreseeable future. In the event that we do make a distribution of cash or property (other than certain stock distributions) with respect to our common stock (or in the case of certain redemptions that are treated as distributions with respect to our common stock), any such distributions generally will constitute dividends for U.S. federal income tax purposes to the extent of our current and accumulated earnings and profits, if any, as determined under U.S. federal income tax principles. If a distribution exceeds our current and accumulated earnings and profits, the excess will constitute a return of capital and will first reduce the holder’s adjusted tax basis in our common stock, but not below zero. Any remaining excess will be treated as capital gain, subject to the tax treatment described below in “—Gain on Sale, Exchange or Other Taxable Disposition of Our Common Stock”. Any such distribution would also be subject to the discussion below under the sections titled “—Additional Withholding and Reporting Requirements” and “—Backup Withholding and Information Reporting.”

Dividends paid to a Non-U.S. Holder generally will be subject to a 30% U.S. federal withholding tax unless such Non-U.S. Holder provides us or another applicable withholding agent, as the case may be, with the appropriate IRS Form W-8, such as:

- IRS Form W-8BEN (or successor form) certifying, under penalties of perjury, that such holder is not a United States person (as defined under the Code) and is eligible for a reduction in the rate of, or exemption from, withholding under an applicable income tax treaty, or
- IRS Form W-8ECI (or successor form) certifying that a dividend paid on common stock is not subject to withholding tax because it is effectively connected with a trade or business in the United States of the Non-U.S. Holder (in which case such dividend generally will be subject to regular graduated U.S. tax rates as described below).

The certification requirement described above must be provided to us or another applicable withholding agent prior to the payment of dividends and must be updated periodically. The certification also may require a Non-U.S. Holder that claims treaty benefits of a reduction in the rate of, or exemption from, withholding on dividends to provide its U.S. taxpayer identification number. Special certification and other requirements apply in the case of certain Non-U.S. Holders that hold shares of our common stock through intermediaries or are pass-through entities for U.S. federal income tax purposes.

Each Non-U.S. Holder is urged to consult its own tax advisor about the specific methods for satisfying these requirements. A claim for exemption will not be valid if the person receiving the applicable form has actual knowledge or reason to know that the statements on the form are false.

If dividends are effectively connected with a trade or business in the United States of a Non-U.S. Holder (and, if required by an applicable income tax treaty, attributable to a U.S. permanent establishment or fixed base), the Non-U.S. Holder, although exempt from the withholding tax described above (provided that the certifications described above are satisfied), generally will be subject to U.S. federal income tax on such dividends on a net income basis in the same manner as if it were a U.S. person. In addition, if a Non-U.S. Holder is treated as a corporation for U.S. federal income tax purposes, the Non-U.S. Holder may be subject to an additional “branch profits tax” equal to 30% (unless reduced by an applicable income tax treaty) of such effectively connected dividend, as adjusted for certain items.

Non-U.S. Holders that do not timely provide us or another applicable withholding agent with the required certification, but which are eligible for a reduced rate of, or an exemption from, U.S. federal withholding tax, may obtain a refund or credit of any excess amount withheld by timely filing an appropriate claim for refund with the IRS.

Gain on Sale, Exchange or Other Taxable Disposition of Our Common Stock

Subject to the discussion below under the sections titled “—Additional Withholding and Reporting Requirements” and “—Backup Withholding and Information Reporting”, in general, a Non-U.S. Holder will not be subject to U.S. federal income tax or withholding tax on gain realized upon such holder’s sale, exchange or other taxable disposition of shares of our common stock unless (i) such Non-U.S. Holder is an individual who is present in the United States for 183 days or more in the taxable year of disposition, and certain other conditions are met; (ii) we are or have been a “United States real property holding corporation”, as defined in the Code (a “USRPHC”), at any time within the shorter of the five-year period preceding the disposition and the Non-U.S. Holder’s holding period in the shares of our common stock, and certain other requirements are met; or (iii) such gain is effectively connected with the conduct by such Non-U.S. Holder of a trade or business in the United States (and, if required by an applicable income tax treaty, is attributable to a permanent establishment or fixed base maintained by such Non-U.S. Holder in the United States).

If the first exception applies, the Non-U.S. Holder generally will be subject to U.S. federal income tax at a rate of 30% (or at a reduced rate under an applicable income tax treaty) on the amount by which such Non-U.S. Holder’s capital gains allocable to U.S. sources exceed capital losses allocable to U.S. sources during the taxable year of the disposition. If the third exception applies, the Non-U.S. Holder generally will be subject to U.S. federal income tax with respect to such gain on a net income basis in the same manner as if it were a U.S. person, and a Non-U.S. Holder that is a corporation for U.S. federal income tax purposes may also be subject to a branch profits tax with respect to such effectively connected gain, as adjusted for certain items, at a rate of 30% (or at a reduced rate under an applicable income tax treaty).

Regarding the second exception, generally, a corporation is a USRPHC only if the fair market value of its U.S. real property interests (as defined in the Code) equals or exceeds 50% of the sum of the fair market value of its worldwide real property interests plus its other assets used or held for use in a trade or business. Although there can be no assurance in this regard, we believe that we are not, and do not anticipate becoming, a USRPHC. However, because the determination of whether we are a USRPHC depends on the fair market value of our U.S. real property interests relative to the fair market value of our other business assets, there can be no assurance that we have not been a USRPHC in the past and will not become a USRPHC in the future. Even if we became a USRPHC, a Non-U.S. Holder would not be subject to U.S. federal income tax on a sale, exchange or other taxable disposition of our common stock by reason of our status as USRPHC so long as our common stock is regularly traded on an established securities market (within the meaning of the applicable regulations) and such Non-U.S. Holder does not own and is not deemed to own (directly, indirectly or constructively) more than 5% of our outstanding common stock at any time during the shorter of the five year period ending on the date of disposition and such holder’s holding period. However, no assurance can be provided that our common stock will be regularly traded on an established securities market for purposes of the rules described above. Prospective investors are encouraged to consult their own tax advisors regarding the possible consequences to them if we are, or were to become, a USRPHC.

Additional Withholding and Reporting Requirements

Legislation enacted in March 2010 and related guidance (commonly referred to as “FATCA”) will impose, in certain circumstances, U.S. federal withholding at a rate of 30% on payments of (a) dividends on our common stock on or after July 1, 2014, and (b) gross proceeds from the sale or other disposition of our common stock on or after January 1, 2017. In the case of payments made to a “foreign financial institution” as defined under FATCA (including, among other entities, an investment fund), the tax generally will be imposed, subject to certain exceptions, unless such institution (i) enters into (or is otherwise subject to) and complies with an agreement with the U.S. government (a “FATCA Agreement”) or (ii) complies with an applicable intergovernmental agreement between the United States and a foreign jurisdiction (an “IGA”) or any foreign law implementing an applicable IGA, in either case to, among other things, collect and provide to the U.S. or other relevant tax authorities certain information regarding U.S. account holders of such institution. In the case of

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payments made to a foreign entity that is not a foreign financial institution, the tax generally will be imposed, subject to certain exceptions, unless such foreign entity provides the withholding agent with a certification that it does not have any “substantial U.S. owners” (generally, any specified U.S. persons that directly or indirectly owns more than a specified percentage of such entity) or that identifies its substantial U.S. owners. If our common stock is held through a foreign financial institution that enters into (or is otherwise subject to) a FATCA Agreement, such foreign financial institution (or, in certain cases, a person paying amounts to such foreign financial institution) generally will be required, subject to certain exceptions, to apply FATCA withholding on payments of dividends and proceeds described above made to (x) a person (including an individual) that fails to comply with certain information requests or (y) a foreign financial institution that has not entered into a FATCA Agreement and is not otherwise exempt from FATCA pursuant to an IGA.

Prospective investors should consult their own tax advisors regarding the possible impact of these rules on their investment in our common stock, and the entities through which they hold our common stock, including, without limitation, the process and deadlines for meeting the applicable requirements to prevent the imposition of this 30% withholding tax under FATCA.

Backup Withholding and Information Reporting

We must report annually to the IRS and to each Non-U.S. Holder the gross amount of the distributions on our common stock paid to the holder and the tax withheld, if any, with respect to the distributions. Non-U.S. Holders may have to comply with specific certification procedures to establish that the holder is not a United States person (as defined in the Code) in order to avoid backup withholding at the applicable rate, currently 28%, with respect to dividends on our common stock. Dividends paid to Non-U.S. Holders subject to U.S. withholding, as described above under the section titled “—Distributions on Our Common Stock”, generally will be exempt from U.S. backup withholding.

Information reporting and backup withholding will generally apply to the proceeds of a disposition of our common stock by a Non-U.S. Holder effected by or through the U.S. office of any broker, U.S. or foreign, unless the holder certifies, under penalties of perjury, that it is not a United States person (as defined under the Code) and satisfies certain other requirements (and the payor does not have actual knowledge or reason to know that the beneficial owner is a United States person), or otherwise establishes an exemption. Generally, information reporting and backup withholding will not apply to a payment of disposition proceeds to a Non-U.S. Holder where the transaction is effected outside the United States through a non-U.S. office of a broker. However, for information reporting purposes, dispositions effected through a non-U.S. office of a broker with substantial U.S. ownership or operations generally will be treated in a manner similar to dispositions effected through a U.S. office of a broker. Prospective investors should consult their own tax advisors regarding the application of the information reporting and backup withholding rules to them.

Copies of information returns may be made available to the tax authorities of the country in which the Non-U.S. Holder resides or in which the Non-U.S. Holder is incorporated, under the provisions of a specific treaty or agreement.

Backup withholding is not an additional tax. Any amounts withheld under the backup withholding rules from a payment to a Non-U.S. Holder can be refunded or credited against the Non-U.S. Holder’s U.S. federal income tax liability, if any, provided that an appropriate claim is timely filed with the IRS.

Federal Estate Tax

Common stock owned (or treated as owned) by an individual who is not a citizen or a resident of the United States (as defined for U.S. federal estate tax purposes) at the time of death will be included in the individual’s gross estate for U.S. federal estate tax purposes unless an applicable estate or other tax treaty provides otherwise, and therefore, may be subject to U.S. federal estate tax.

Underwriting

Under the terms and subject to the conditions in an underwriting agreement to be dated the date of the final prospectus, the underwriters named below, for whom Morgan Stanley & Co. LLC, Credit Suisse Securities (USA) LLC and UBS Securities LLC are acting as representatives, will severally agree to purchase, and we will agree to sell to them, severally, the number of shares indicated below:

<u>Name</u>	<u>Number of Shares</u>
Morgan Stanley & Co. LLC	
Credit Suisse Securities (USA) LLC	
UBS Securities LLC	
Nomura Securities International, Inc.	
Total	

The underwriters and the representatives are collectively referred to as the “underwriters” and the “representatives,” respectively. The underwriters are offering the shares of common stock subject to their acceptance of the shares from us and subject to prior sale. The underwriting agreement provides that the obligations of the several underwriters to pay for and accept delivery of the shares of common stock offered by this prospectus are subject to the approval of certain legal matters by their counsel and to certain other conditions. The underwriters are obligated to take and pay for all of the shares of common stock offered by this prospectus if any such shares are taken. However, the underwriters are not required to take or pay for the shares covered by the underwriters’ over-allotment option described below.

The underwriters initially propose to offer part of the shares of common stock directly to the public at the offering price listed on the cover page of this prospectus and part to certain dealers at a price that represents a concession not in excess of \$ _____ a share under the public offering price. After the initial offering of the shares of common stock, the offering price and other selling terms may from time to time be varied by the representatives.

We have granted to the underwriters an option, exercisable for 30 days from the date of this prospectus, to purchase up to _____ additional shares of common stock at the public offering price listed on the cover page of this prospectus, less underwriting discounts and commissions. The underwriters may exercise this option solely for the purpose of covering over-allotments, if any, made in connection with the offering of the shares of common stock offered by this prospectus. To the extent the option is exercised, each underwriter will become obligated, subject to certain conditions, to purchase the same percentage of the additional shares of common stock as the number listed next to the underwriter’s name in the preceding table bears to the total number of shares of common stock listed next to the names of all underwriters in the preceding table.

The following table shows the per share and total public offering price, underwriting discounts and commissions, and proceeds before expenses to us. These amounts are shown assuming both no exercise and full exercise of the underwriters’ option to purchase up to an additional _____ shares of common stock.

	<u>Per Share</u>	<u>Total</u>	
		<u>No exercise</u>	<u>Full exercise</u>
Public Offering Price	\$	\$	\$
Underwriting Discount and commissions to be paid by us	\$	\$	\$
Proceeds, before expenses, to us	\$	\$	\$

The estimated offering expenses payable by us, exclusive of the underwriting discounts and commissions, are approximately \$ _____ million. We have agreed to reimburse the underwriters for expenses relating to clearance of this offering with the Financial Industry Regulatory Authority, Inc. and the qualification of our common stock under state securities laws (in an amount not to exceed in the aggregate \$ _____).

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The underwriters have informed us that they do not intend sales to discretionary accounts to exceed 5% of the total number of shares of common stock offered by them.

We intend to apply to have our common stock listed on the Nasdaq Global Market under the trading symbol AKBA.

We, and all of our directors and officers, and the holders of substantially all of our outstanding stock and stock options have agreed that, without the prior written consent of Morgan Stanley & Co. LLC and Credit Suisse Securities (USA) LLC on behalf of the underwriters, we and they will not, during the period ending 180 days after the date of this prospectus (the “restricted period”):

- offer, pledge, sell, contract to sell, sell any option or contract to purchase, purchase any option or contract to sell, grant any option, right or warrant to purchase, lend, or otherwise transfer or dispose of, directly or indirectly, any shares of our common stock or any securities convertible into or exercisable or exchangeable for our common stock; or
- enter into any swap or other arrangement that transfers to another, in whole or in part, any of the economic consequences of ownership of our common stock,

whether any such transaction described above is to be settled by delivery of common stock or such other securities, in cash or otherwise. In addition, we and all of our directors and officers, and the holders of substantially all of our common stock and stock options have agreed that, without the prior written consent of Morgan Stanley & Co. LLC and Credit Suisse Securities (USA) LLC on behalf of the underwriters, during the restricted period, no registration statement with the SEC relating to the offering of any shares of common stock or any security convertible into or exercisable or exchangeable for common stock will be filed.

The restrictions described in the immediately preceding paragraph do not apply to:

- the sale of shares by us to the underwriters;
- the issuance by us of shares of our common stock upon the exercise of an option or warrant or the conversion of a security outstanding on the date of this prospectus of which the Underwriters have been advised in writing;
- the establishment of a trading plan pursuant to Rule 10b5-1 under the Exchange Act for the transfer of shares of our common stock; *provided* that such plan does not provide for the transfer of shares of our common stock during the restricted period and to the extent a public announcement or filing under the Exchange Act, if any, is required of or voluntarily made regarding the establishment of such plan, such announcement or filing shall include a statement to the effect that no transfer of common stock may be made under such plan during the restricted period;
- transactions relating to shares of our common stock or other securities acquired in this offering (other than any shares of our common stock directed by us and purchased in this offering by one of our officers or directors) or in open market transactions after the date of the final prospectus;
- transfers of shares of our common stock, or any security convertible into, exercisable or exchangeable for our common stock, as a bona fide gift;
- transfers of shares of our common stock, or any security convertible into, exercisable or exchangeable for our common stock, by will or intestacy;
- the exercise of options to purchase shares of our common stock granted under any existing stock incentive plan or stock purchase plan described in this prospectus, *provided* that any shares of our common stock issued pursuant to such exercise shall be subject to the same restrictions;
- transfers to us for the purpose of satisfying tax withholding obligations upon the vesting of other equity incentive awards granted under any existing stock incentive plan or stock purchase plan described in this prospectus;

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- transfers or distributions not involving a disposition for value of shares of our common stock, or any security convertible into, exercisable or exchangeable for our common stock, to any limited or general partners, stockholders or members of a lock-up signatory, or if the lock-up signatory is a corporation, to a wholly-owned subsidiary of such lock-up signatory;
- transfers of shares of our common stock, or any security convertible into, exercisable or exchangeable for our common stock, made by one of the lock-up signatories to (i) any trust, corporation, partnership, limited liability company or other legal entity who, directly or indirectly, controls, is controlled by, or is under common control with such lock-up signatory, (ii) any trust or other legal entity for which a lock-up signatory or the spouse of a lock-up signatory serves as trustee or investment advisor, or (iii) any member of the immediate family of a lock-up signatory, or any trust or other legal entity for the direct or indirect benefit of a lock-up signatory or any member of the immediate family of a lock-up signatory;
- transfers of shares of our common stock, or any securities convertible into, exercisable or exchangeable for our common stock, pursuant to a sale of, or an offer to purchase, 100% of our outstanding common stock, whether pursuant to a merger, tender offer or otherwise, to a third party or group of third parties, *provided* that in the event that such tender offer, merger, or transaction is not completed, our common stock and any security convertible into or exchangeable for our common stock shall remain subject to the same restrictions; or
- the conversion of our outstanding preferred stock into shares of our common stock upon the closing of this offering, *provided* that such shares of our common stock shall remain subject to the same restrictions,

provided, however, that in the case of any transfer or distribution pursuant to the fifth, sixth, ninth or tenth clauses above, each donee, distributee or transferee shall sign and deliver a lock-up agreement substantially in the form of the lock-up agreements described above; and in the case of any transaction, transfer, exercise or distribution pursuant to the fourth through (and including) the tenth clauses above, no filing under Section 16(a) of the Exchange Act, reporting a reduction in beneficial ownership of shares of our common stock, shall be required or shall be voluntarily made during the restricted period (other than a filing on Form 5 made after the expiration of the restricted period).

Morgan Stanley & Co. LLC and Credit Suisse Securities (USA) LLC, in their sole discretion, may release the common stock and other securities subject to the lock-up agreements described above in whole or in part at any time.

In order to facilitate the offering of the common stock, the underwriters may engage in transactions that stabilize, maintain or otherwise affect the price of the common stock. Specifically, the underwriters may sell more shares than they are obligated to purchase under the underwriting agreement, creating a short position. A short sale is covered if the short position is no greater than the number of shares available for purchase by the underwriters under the over-allotment option. The underwriters can close out a covered short sale by exercising the over-allotment option or purchasing shares in the open market. In determining the source of shares to close out a covered short sale, the underwriters will consider, among other things, the open market price of shares compared to the price available under the over-allotment option. The underwriters may also sell shares in excess of the over-allotment option, creating a naked short position. The underwriters must close out any naked short position by purchasing shares in the open market. A naked short position is more likely to be created if the underwriters are concerned that there may be downward pressure on the price of the common stock in the open market after pricing that could adversely affect investors who purchase in this offering. As an additional means of facilitating this offering, the underwriters may bid for, and purchase, shares of common stock in the open market to stabilize the price of the common stock. These activities may raise or maintain the market price of the common stock above independent market levels or prevent or retard a decline in the market price of the common stock. The underwriters are not required to engage in these activities and may end any of these activities at any time.

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We and the underwriters have agreed to indemnify each other against certain liabilities, including liabilities under the Securities Act.

A prospectus in electronic format may be made available on websites maintained by one or more underwriters, or selling group members, if any, participating in this offering. The representatives may agree to allocate a number of shares of common stock to underwriters for sale to their online brokerage account holders. Internet distributions will be allocated by the representatives to underwriters that may make Internet distributions on the same basis as other allocations.

Other Relationships

The underwriters and their respective affiliates are full service financial institutions engaged in various activities, which may include securities trading, commercial and investment banking, financial advisory, investment management, principal investment, hedging, financing and brokerage activities. Certain of the underwriters and their respective affiliates have, from time to time, performed, and may in the future perform, various financial advisory and investment banking services for us, for which they received or will receive customary fees and expenses.

In addition, in the ordinary course of their various business activities, the underwriters and their respective affiliates may make or hold a broad array of investments and actively trade debt and equity securities (or related derivative securities) and financial instruments (including bank loans) for their own account and for the accounts of their customers and may at any time hold long and short positions in such securities and instruments. Such investment and securities activities may involve our securities and instruments. The underwriters and their respective affiliates may also make investment recommendations or publish or express independent research views in respect of such securities or instruments and may at any time hold, or recommend to clients that they acquire, long or short positions in such securities and instruments.

Pricing of the Offering

Prior to this offering, there has been no public market for our common stock. The initial public offering price was determined by negotiations between us and the representatives. Among the factors considered in determining the initial public offering price were our future prospects and those of our industry in general, financial and operating information in recent periods, and the price-earnings ratios, price-sales ratios, market prices of securities and certain financial and operating information of companies engaged in activities similar to ours.

Directed Share Program

At our request, the underwriters have reserved % of the shares of common stock to be issued by us and offered by this prospectus for sale, at the initial public offering price, to directors, officers, employees, business associates and related persons of Akebia Therapeutics, Inc. The number of shares of common stock available for sale to the general public will be reduced to the extent these individuals purchase such reserved shares. Any reserved shares that are not so purchased will be offered by the underwriters to the general public on the same basis as the other shares offered by this prospectus.

Selling Restrictions

European Economic Area

In relation to each Member State of the European Economic Area which has implemented the Prospectus Directive (each, a “Relevant Member State”) an offer to the public of any shares of our common stock may not be made in that Relevant Member State, except that an offer to the public in that Relevant Member State of any shares of our common stock may be made at any time under the following exemptions under the Prospectus Directive, if they have been implemented in that Relevant Member State:

- (a) to any legal entity which is a qualified investor as defined in the Prospectus Directive;
- (b) to fewer than 100 or, if the Relevant Member State has implemented the relevant provision of the 2010 PD Amending Directive, 150, natural or legal persons (other than qualified investors as defined in the Prospectus Directive), as permitted under the Prospectus Directive, subject to obtaining the prior consent of the representatives for any such offer; or
- (c) in any other circumstances falling within Article 3(2) of the Prospectus Directive, provided that no such offer of shares of our common stock shall result in a requirement for the publication by us or any underwriter of a prospectus pursuant to Article 3 of the Prospectus Directive.

For the purposes of this provision, the expression an “offer to the public” in relation to any shares of our common stock in any Relevant Member State means the communication in any form and by any means of sufficient information on the terms of the offer and any shares of our common stock to be offered so as to enable an investor to decide to purchase any shares of our common stock, as the same may be varied in that Member State by any measure implementing the Prospectus Directive in that Member State, the expression “Prospectus Directive” means Directive 2003/71/EC (and amendments thereto, including the 2010 PD Amending Directive, to the extent implemented in the Relevant Member State), and includes any relevant implementing measure in the Relevant Member State, and the expression “2010 PD Amending Directive” means Directive 2010/73/EU.

United Kingdom

Each underwriter has represented and agreed that:

- (a) it has only communicated or caused to be communicated and will only communicate or cause to be communicated an invitation or inducement to engage in investment activity (within the meaning of Section 21 of the Financial Services and Markets Act 2000 (“FSMA”)) received by it in connection with the issue or sale of the shares of our common stock in circumstances in which Section 21(1) of the FSMA does not apply to us; and
- (b) it has complied and will comply with all applicable provisions of the FSMA with respect to anything done by it in relation to the shares of our common stock in, from or otherwise involving the United Kingdom.

Switzerland

The Prospectus does not constitute an issue prospectus pursuant to Article 652a or Article 1156 of the Swiss Code of Obligations (“CO”) and the shares will not be listed on the SIX Swiss Exchange. Therefore, the Prospectus may not comply with the disclosure standards of the CO and/or the listing rules (including any prospectus schemes) of the SIX Swiss Exchange. Accordingly, the shares may not be offered to the public in or from Switzerland, but only to a selected and limited circle of investors, which do not subscribe to the shares with a view to distribution.

Legal Matters

The validity of the common stock offered in this prospectus will be passed upon for us by Ropes & Gray LLP, Boston, Massachusetts. Certain legal matters in connection with this offering will be passed upon for the underwriters by Latham & Watkins LLP, Boston, Massachusetts.

Experts

The consolidated financial statements of Akebia Therapeutics, Inc. at September 30, 2013, and December 31, 2012 and 2011, and the statements of operations and comprehensive loss, redeemable convertible preferred stock and stockholders' equity (deficit), and cash flows for the nine-month period ended September 30, 2013, the two years ended December 31, 2012 and 2013, and for the period from February 27, 2007 (inception) through September 30, 2013, appearing in this Prospectus and Registration Statement have been audited by Ernst & Young LLP, independent registered public accounting firm, as set forth in their report thereon appearing elsewhere herein, and are included in reliance upon such report given on the authority of such firm as experts in accounting and auditing.

Where You Can Find More Information

We have filed with the SEC a registration statement on Form S-1 under the Securities Act with respect to the shares of common stock offered hereby. This prospectus, which constitutes a part of the registration statement, does not contain all of the information set forth in the registration statement or the exhibits and schedules filed therewith. For further information with respect to us and the common stock offered hereby, reference is made to the registration statement and the exhibits and schedules filed therewith. Statements contained in this prospectus regarding the contents of any contract or any other document that is filed as an exhibit to the registration statement are not necessarily complete, and each such statement is qualified in all respects by reference to the full text of such contract or other document filed as an exhibit to the registration statement. A copy of the registration statement and the exhibits and schedules filed therewith may be inspected without charge at the public reference room maintained by the SEC, located at 100 F Street N.E., Washington, D.C. 20549, and copies of all or any part of the registration statement may be obtained from such offices upon the payment of the fees prescribed by the SEC. Please call the SEC at 1-800-SEC-0330 for further information about the public reference room. The SEC also maintains a website that contains reports, proxy and information statements and other information regarding registrants that file electronically with the SEC. The address is www.sec.gov.

Upon completion of this offering, we will become subject to the information and periodic reporting requirements of the Exchange Act and, in accordance therewith, will file periodic reports, proxy statements and other information with the SEC. Such periodic reports, proxy statements and other information will be available for inspection and copying at the public reference room and website of the SEC referred to above.

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Akebia Therapeutics, Inc.
(A Development Stage Company)

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Report of Independent Registered Public Accounting Firm

The Board of Directors and Shareholders of
Akebia Therapeutics, Inc.

We have audited the accompanying balance sheets of Akebia Therapeutics, Inc. (a development stage company) as of September 30, 2013, and December 31, 2012 and 2011, and the related statements of operations and comprehensive loss, redeemable convertible preferred stock and stockholders' equity (deficit) and cash flows for the nine-month period ended September 30, 2013, each of the two years in the period ended December 31, 2012 and for the period from February 27, 2007 (inception) through September 30, 2013. These financial statements are the responsibility of the Company's management. Our responsibility is to express an opinion on these financial statements based on our audits.

We conducted our audits in accordance with the standards of the Public Company Accounting Oversight Board (United States). Those standards require that we plan and perform the audit to obtain reasonable assurance about whether the financial statements are free of material misstatement. We were not engaged to perform an audit of the Company's internal control over financial reporting. Our audits included consideration of internal control over financial reporting as a basis for designing audit procedures that are appropriate in the circumstances, but not for the purpose of expressing an opinion on the effectiveness of the Company's internal control over financial reporting. Accordingly, we express no such opinion. An audit also includes examining, on a test basis, evidence supporting the amounts and disclosures in the financial statements, assessing the accounting principles used and significant estimates made by management, and evaluating the overall financial statement presentation. We believe that our audits provide a reasonable basis for our opinion.

In our opinion, the financial statements referred to above present fairly, in all material respects, the financial position of Akebia Therapeutics, Inc. (a development stage company) at September 30, 2013, and December 31, 2012 and 2011, and the results of its operations and its cash flows for the nine-month period ended September 30, 2013, each of the two years in the period ended December 31, 2012, and for the period from February 27, 2007 (inception) through September 30, 2013, in conformity with US generally accepted accounting principles.

/s/ Ernst & Young LLP

Cincinnati, Ohio
December 20, 2013

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Akebia Therapeutics, Inc.
(A Development Stage Company)
Balance Sheets

	December 31,		September 30, 2013	
	2011	2012	Actual	Pro forma (unaudited)
Assets				
Current assets:				
Cash and cash equivalents	\$ 5,011,089	\$ 1,641,038	\$ 24,603,252	\$ 24,603,252
Short-term investments	1,365,943	—	13,113,591	13,113,591
Accounts receivable	82,115	85,633	165,814	165,814
Prepaid expenses and other current assets	752,169	517,202	582,143	582,143
Total current assets	<u>7,211,316</u>	<u>2,243,873</u>	<u>38,464,800</u>	<u>38,464,800</u>
Equipment, net of accumulated depreciation of \$26 at September 30, 2013	—	—	5,588	5,588
Total assets	<u>\$ 7,211,316</u>	<u>\$ 2,243,873</u>	<u>\$ 38,470,388</u>	<u>\$ 38,470,388</u>
Liabilities, redeemable convertible preferred stock and stockholders' (deficit) equity				
Current liabilities:				
Accounts payable	\$ 1,373,056	\$ 417,943	\$ 1,970,476	\$ 1,970,476
Accrued expenses	444,101	351,250	927,933	927,933
2012 Series X preferred stock subject to mandatory redemption	—	4,154,137	—	—
Total current liabilities	<u>1,817,157</u>	<u>4,923,330</u>	<u>2,898,409</u>	<u>2,898,409</u>
Commitments and contingencies (see Note 11)				
Redeemable convertible preferred stock; \$.00001 par value; 2,377,394, 2,427,394 and 5,500,636 shares authorized at December 31, 2011, December 31, 2012 and September 30, 2013, respectively:				
Series A redeemable convertible preferred stock; 734,538 shares issued and outstanding at December 31, 2011, December 31, 2012 and September 30, 2013; no shares issued and outstanding pro forma (unaudited)	34,926,819	37,092,486	38,785,314	—
Series B redeemable convertible preferred stock; 1,287,525 shares issued and outstanding at December 31, 2011, December 31, 2012 and September 30, 2013; no shares issued and outstanding pro forma (unaudited)	18,659,205	19,816,185	20,720,557	—
Series C redeemable convertible preferred stock; no shares issued and outstanding at December 31, 2011 and 2012; 3,302,885 shares issued and outstanding at September 30, 2013; no shares issued and outstanding pro forma (unaudited)	—	—	95,296,787	—
Total redeemable convertible preferred stock	<u>53,586,024</u>	<u>56,908,671</u>	<u>154,802,658</u>	<u>—</u>
Stockholders' (deficit) equity:				
Common stock; \$.00001 par value; 4,466,956, 4,466,956 and 8,400,000 authorized; 321,814, 351,861 and 424,044 shares issued and outstanding at December 31, 2011 and 2012, and September 30, 2013, respectively; 7,214,193 shares issued and outstanding pro forma (unaudited)	3	3	3	71
Additional paid-in capital	—	—	—	93,263,055
Deficit accumulated during the development stage	<u>(48,191,868)</u>	<u>(59,588,131)</u>	<u>(119,230,682)</u>	<u>(57,691,147)</u>
Total stockholders' (deficit) equity	<u>(48,191,865)</u>	<u>(59,588,128)</u>	<u>(119,230,679)</u>	<u>35,571,979</u>
Total liabilities, redeemable convertible preferred stock and stockholders' equity	<u>\$ 7,211,316</u>	<u>\$ 2,243,873</u>	<u>\$ 38,470,388</u>	<u>\$ 38,470,388</u>

See accompanying notes to financial statements.

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Akebia Therapeutics, Inc.
(A Development Stage Company)
Statements of Operations and Comprehensive Loss

	Year Ended December 31,		Nine Months Ended September 30,		February 27,
	2011	2012	2012 (unaudited)	2013	(inception) through September 30, 2013
Revenue	\$ —	\$ —	\$ —	\$ —	\$ —
Operating expenses:					
Research and development	12,976,122	5,631,541	5,065,070	7,591,015	48,557,260
General and administrative	2,566,569	2,891,008	1,593,606	2,140,834	12,258,150
Total operating expenses	15,542,691	8,522,549	6,658,676	9,731,849	60,815,410
Operating loss	(15,542,691)	(8,522,549)	(6,658,676)	(9,731,849)	(60,815,410)
Other income (expense):					
Grant income	221,606	—	—	—	1,481,408
Interest income (expense), net	24,400	(1,644,654)	(771,152)	(722,857)	(4,935,885)
Extinguishment of debt and other liabilities	—	—	—	2,419,766	3,735,499
Reimbursements from Aerpio	—	1,971,246	1,636,868	815,704	2,786,950
Gain on cancellation of preferred stock future tranche rights	—	—	—	—	653,465
Net loss and comprehensive loss	<u>\$ (15,296,685)</u>	<u>\$ (8,195,957)</u>	<u>\$ (5,792,960)</u>	<u>\$ (7,219,236)</u>	<u>\$ (57,093,973)</u>
Reconciliation of net loss to net loss applicable to common stockholders:					
Net loss	\$ (15,296,685)	\$ (8,195,957)	\$ (5,792,960)	\$ (7,219,236)	\$ (57,093,973)
Accretion on preferred stock	(2,970,586)	(3,322,647)	(2,468,546)	(52,861,367)	(63,713,722)
Loss on extinguishment of preferred stock	—	—	—	—	(597,174)
Net loss applicable to common stockholders	<u>\$ (18,267,271)</u>	<u>\$ (11,518,604)</u>	<u>\$ (8,261,506)</u>	<u>\$ (60,080,603)</u>	<u>\$ (121,404,869)</u>
Net loss per share applicable to common stockholders—basic and diluted	<u>\$ (109.36)</u>	<u>\$ (48.68)</u>	<u>\$ (35.80)</u>	<u>\$ (206.32)</u>	<u>\$ (827.64)</u>
Weighted-average number of common shares used in net loss per share applicable to common stockholders'—basic and diluted	167,039	236,633	230,748	291,206	146,688
Pro forma net loss applicable to common stockholders (unaudited)		\$ (8,195,957)		\$ (7,219,236)	
Pro forma net loss per share applicable to common stockholders—basic and diluted (unaudited)		<u>\$ (2.26)</u>		<u>\$ (1.35)</u>	
Pro forma weighted-average number of common shares used in pro forma net loss per share applicable to common stockholders—basic and diluted (unaudited)		<u>3,622,838</u>		<u>5,362,450</u>	

See accompanying notes to financial statements.

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Akebia Therapeutics, Inc.
(A Development Stage Company)
Statements of Redeemable Convertible Preferred Stock and Stockholders' Equity (Deficit)
Period from February 27, 2007 (Inception) through September 30, 2013

	Redeemable Convertible Preferred Stock								Stockholders' Equity (Deficit)						
	Series A		Series B		Series C		2012 Series X		2007 Series X		Common Stock		Additional Paid-In Capital	Deficit Accumulated During the Development Stage	Total Stockholders' Equity (Deficit)
	Convertible Preferred Stock		Redeemable Convertible Preferred Stock		Redeemable Convertible Preferred Stock		Convertible Preferred Stock		Convertible Preferred Stock		Common Stock				
	Number of Shares	Amount	Number of Shares	Amount	Number of Shares	Amount	Number of Shares	Amount	Number of Shares	\$0.00001 Par Value	Number of Shares	\$0.00001 Par Value			
Initial capital contribution	—	—	—	—	—	—	—	—	—	—	40,000	—	\$ 40	—	\$ 40
Issuance of 2007 Series X convertible preferred stock	—	—	—	—	—	—	—	—	—	9,938	—	—	31,800	—	31,800
Issuance costs associated with sale of Series A redeemable convertible preferred stock	—	—	—	—	—	—	—	—	—	—	—	—	(21,219)	—	(21,219)
Net loss	—	—	—	—	—	—	—	—	—	—	—	—	—	(2,701,292)	(2,701,292)
Balance at December 31, 2007	—	—	—	—	—	—	—	—	—	9,938	—	40,000	10,621	(2,701,292)	(2,690,671)
Sale of Series A preferred stock, net of issuance costs of \$90,453	226,500	8,359,507	—	—	—	—	—	—	—	—	—	—	—	—	—
Issuance of Series A preferred stock in settlement of convertible notes	45,228	1,626,386	—	—	—	—	—	—	—	—	—	—	—	—	—
Accretion of preferred stock to redemption value	—	1,515,495	—	—	—	—	—	—	—	—	—	—	(649,509)	(865,986)	(1,515,495)
Loss on extinguishment of 2007 Series X convertible preferred stock	—	—	—	—	—	—	—	—	—	—	—	—	597,174	(597,174)	—
Conversion of 2007 Series X convertible preferred stock into common stock	—	—	—	—	—	—	—	—	—	(9,938)	72,047	1	(1)	—	—
Share-based compensation expense	—	—	—	—	—	—	—	—	—	—	—	—	41,715	—	41,715
Net loss	—	—	—	—	—	—	—	—	—	—	—	—	—	(5,076,316)	(5,076,316)
Balance at December 31, 2008	271,728	11,501,388	—	—	—	—	—	—	—	—	112,047	1	—	(9,240,768)	(9,240,767)
Sale of Series A preferred stock, net of issuance costs of \$44,008	265,609	10,525,461	—	—	—	—	—	—	—	—	—	—	—	—	—
Issuance of Series A preferred stock in settlement of convertible notes	72,201	2,851,929	—	—	—	—	—	—	—	—	—	—	—	—	—
Accretion of preferred stock to redemption value	—	1,226,217	—	—	—	—	—	—	—	—	—	—	(104,737)	(1,121,480)	(1,226,217)
Issuance of restricted common stock	—	—	—	—	—	—	—	—	—	—	31,474	—	—	—	—
Share-based compensation expense	—	—	—	—	—	—	—	—	—	—	—	—	104,737	—	104,737
Net loss	—	—	—	—	—	—	—	—	—	—	—	—	—	(8,506,556)	(8,506,556)
Balance at December 31, 2009	609,538	26,104,995	—	—	—	—	—	—	—	—	143,521	1	—	(18,868,804)	(18,868,803)

Akebia Therapeutics, Inc.
(A Development Stage Company)
Statements of Redeemable Convertible Preferred Stock and Stockholders' Equity (Deficit)
Period from February 27, 2007 (Inception) through September 30, 2013

	Redeemable Convertible Preferred Stock								Stockholders' Equity (Deficit)							
	Series A		Series B				Series C		2012	2007				Deficit Accumulated During the Development Stage	Total Stockholders' Equity (Deficit)	
	Convertible	Preferred Stock	Convertible Preferred Stock		Convertible Preferred Stock		Series X	Series X Convertible Preferred Stock		Common Stock		Additional Paid-In Capital				
	Number of Shares	Amount	Number of Shares	Amount	Number of Shares	Amount	Number of Shares	Amount	Number of Shares	\$0.0001 Par Value	Number of Shares		\$0.0001 Par Value			
Sale of Series A preferred stock, net of issuance costs of \$29,403	125,000	\$ 4,970,597	—	\$ —	—	\$ —	—	\$ —	—	\$ —	—	\$ —	—	\$ —	—	\$ —
Accretion of preferred stock to redemption value	—	1,817,410	—	—	—	—	—	—	—	—	—	—	(169,271)	(1,648,139)	(1,817,410)	
Issuance of restricted common stock	—	—	—	—	—	—	—	—	—	—	10,603	—	—	—	—	—
Share-based compensation expense	—	—	—	—	—	—	—	—	—	—	—	—	169,271	—	—	169,271
Net loss	—	—	—	—	—	—	—	—	—	—	—	—	—	(10,097,931)	(10,097,931)	
Balance at December 31, 2010	734,538	32,893,002	—	—	—	—	—	—	—	—	154,124	1	—	(30,614,874)	(30,614,873)	
Sale of Series B preferred stock, net of issuance costs of \$302,905	—	—	1,287,525	17,722,436	—	—	—	—	—	—	—	—	—	—	—	—
Accretion of preferred stock to redemption value	—	2,033,817	—	936,769	—	—	—	—	—	—	—	—	(690,277)	(2,280,309)	(2,970,586)	
Distribution of subsidiary	—	—	—	—	—	—	—	—	—	—	—	—	382,850	—	382,850	
Issuance of restricted common stock	—	—	—	—	—	—	—	—	—	—	167,690	2	(2)	—	—	
Share-based compensation expense	—	—	—	—	—	—	—	—	—	—	—	—	307,429	—	307,429	
Net loss	—	—	—	—	—	—	—	—	—	—	—	—	—	(15,296,685)	(15,296,685)	
Balance at December 31, 2011	734,538	34,926,819	1,287,525	18,659,205	—	—	—	—	—	—	321,814	3	—	(48,191,868)	(48,191,865)	
Accretion of preferred stock to redemption value	—	2,165,667	—	1,156,980	—	—	—	—	—	—	—	—	(122,341)	(3,200,306)	(3,322,647)	
Issuance of restricted common stock	—	—	—	—	—	—	—	—	—	—	30,047	—	—	—	—	

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Akebia Therapeutics, Inc.
(A Development Stage Company)
Statements of Redeemable Convertible Preferred Stock and Stockholders' Equity (Deficit)
Period from February 27, 2007 (Inception) through September 30, 2013

	Redeemable Convertible Preferred Stock								Stockholders' Equity (Deficit)						
	Series A		Series B		Series C		2012 Series X		2007 Series X		Common Stock		Additional Paid-In Capital	Deficit Accumulated During the Development Stage	Total Stockholders' Equity (Deficit)
	Convertible Preferred Stock	Convertible Preferred Stock	Convertible Preferred Stock	Convertible Preferred Stock	Convertible Preferred Stock	Convertible Preferred Stock	Convertible Preferred Stock	Convertible Preferred Stock	Convertible Preferred Stock	Convertible Preferred Stock	Convertible Preferred Stock				
	Number of Shares	Amount	Number of Shares	Amount	Number of Shares	Amount	Number of Shares	Amount	Number of Shares	\$0.00001 Par Value	Number of Shares	\$0.00001 Par Value			
Share-based compensation expense	—	\$ —	—	\$ —	—	\$ —	—	\$ —	—	\$ —	—	\$ —	\$ 122,341	\$ —	\$ 122,341
Net loss	—	—	—	—	—	—	—	—	—	—	—	—	—	(8,195,957)	(8,195,957)
Balance at December 31, 2012	734,538	37,092,486	1,287,525	19,816,185	—	—	—	—	—	—	351,861	3	—	(59,588,131)	(59,588,128)
Issuance of restricted common stock	—	—	—	—	—	—	—	—	—	—	26,985	—	—	—	—
Issuance of common stock	—	—	—	—	—	—	—	—	—	—	45,198	—	—	—	—
Reclassification of 2012 Series X preferred stock upon modification	—	—	—	—	—	—	25,000	2,486,251	—	—	—	—	—	—	—
Sale of 2012 Series X preferred stock, net of issuance costs of \$42,096	—	—	—	—	—	—	25,000	2,457,904	—	—	—	—	—	—	—
Sale of Series C preferred stock, net of issuance costs of \$1,151,923	—	—	—	—	2,945,742	40,088,465	—	—	—	—	—	—	—	—	—

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Akebia Therapeutics, Inc.
(A Development Stage Company)
Statements of Redeemable Convertible Preferred Stock and Stockholders' Equity (Deficit)
Period from February 27, 2007 (Inception) through September 30, 2013

	Redeemable Convertible Preferred Stock								Stockholders' Equity (Deficit)								
	Series A		Series B				Series C		2012 Series X		2007 Series X Convertible Preferred Stock		Common Stock		Additional Paid-In Capital	Deficit Accumulated During the Development Stage	Total Stockholders' Equity (Deficit)
	Convertible Preferred Stock		Convertible Preferred Stock		Convertible Preferred Stock		Convertible Preferred Stock		Number of Shares	\$0.00001 Par Value	Number of Shares	\$0.00001 Par Value					
	Number of Shares	Amount	Number of Shares	Amount	Number of Shares	Amount	Number of Shares	Amount					Number of Shares	\$0.00001 Par Value			
Conversion of 2012 Series X preferred stock into Series C preferred stock	—	\$ —	—	\$ —	357,143	\$ 4,944,155	(50,000)	\$(4,944,155)	—	\$ —	—	\$ —	\$ —	\$ —	—	\$ —	
Accretion of preferred stock to redemption value	—	1,692,828	—	904,372	—	50,264,167	—	—	—	—	—	—	(438,052)	(52,423,315)	(52,861,367)		
Share-based compensation expense	—	—	—	—	—	—	—	—	—	—	—	—	438,052	—	438,052		
Net loss	—	—	—	—	—	—	—	—	—	—	—	—	—	(7,219,236)	(7,219,236)		
Balance at September 30, 2013	734,538	38,785,314	1,287,525	20,720,557	3,302,885	95,296,787	—	—	—	—	424,044	3	—	(119,230,682)	(119,230,679)		
Conversion of convertible preferred stock into common stock (unaudited)	(734,538)	(38,785,314)	(1,287,525)	(20,720,557)	(3,302,885)	(95,296,787)	—	—	—	—	6,790,149	68	93,263,055	61,539,535	154,802,658		
Pro forma balance at September 30, 2013 (unaudited)	—	\$ —	—	\$ —	—	\$ —	—	\$ —	—	\$ —	7,214,193	\$ 71	\$93,263,055	\$ (57,691,147)	\$ 35,571,979		

See accompanying notes to financial statements.

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Akebia Therapeutics, Inc.
(A Development Stage Company)
Statements of Cash Flows

	Year Ended December 31,		Nine Months Ended September 30,		February 27, 2007 (inception) through September 30, 2013
	2011	2012	2012 (unaudited)	2013	2013
Operating activities:					
Net loss	\$ (15,296,685)	\$ (8,195,957)	\$ (5,792,960)	\$ (7,219,236)	\$ (57,093,973)
Adjustments to reconcile net loss to net cash used in operating activities:					
Gain on extinguishment of debt and other liabilities	—	—	—	(2,419,766)	(3,735,499)
Depreciation	39,155	—	—	26	90,165
Loss on sale of investments	40	—	—	—	—
Amortization of debt issue costs	—	16,613	—	4,153	76,857
Amortization of debt discount and interest expense	—	1,654,136	790,898	751,880	5,159,034
Gain on cancellation of preferred stock future tranche rights	—	—	—	—	(653,465)
Issuance of 2007 Series X preferred stock for Licensing Agreement	—	—	—	—	31,800
Compensation recognized under stock option plan	307,429	122,341	130,883	438,052	1,183,545
Accounts receivable	(45,002)	(3,518)	(69,938)	(80,181)	(165,814)
Prepaid expenses and other current assets	(642,301)	243,511	275,524	(27,858)	(571,056)
Accounts payable and accrued expenses	1,516,264	(1,047,964)	(572,804)	2,129,216	3,474,370
Net cash used in operating activities	(14,121,100)	(7,210,838)	(5,238,397)	(6,423,714)	(52,204,036)
Investing activities:					
Purchase of property and equipment	(61,215)	—	—	(5,614)	(258,674)
Proceeds from maturities of short-term investments	1,721,000	1,365,943	1,365,943	—	11,187,943
Proceeds from sale of short-term investments	50,403	—	—	—	800,234
Purchases of short-term investments	(2,442,255)	—	—	(13,154,827)	(25,143,045)
Net cash (used in) provided by investing activities	(732,067)	1,365,943	1,365,943	(13,160,441)	(13,413,542)
Financing activities:					
Proceeds from issuance of preferred stock	18,025,341	—	—	43,740,388	85,533,938
Stock issue costs	(302,905)	—	—	(1,194,019)	(1,682,007)
Debt issue costs	—	(25,157)	(25,157)	—	(76,857)
Proceeds from issuance of 2012 Series X preferred stock	—	2,500,001	2,500,001	—	2,500,001
Issuance of common stock	—	—	—	—	40
Proceeds from issuance of convertible debt	—	—	—	—	3,945,715
Net cash provided by financing activities	17,722,436	2,474,844	2,474,844	42,546,369	90,220,830
Increase (decrease) in cash and cash equivalents	2,869,269	(3,370,051)	(1,397,610)	22,962,214	24,603,252
Cash and cash equivalents at beginning of period	2,141,820	5,011,089	5,011,089	1,641,038	—
Cash and cash equivalents at end of period	\$ 5,011,089	\$ 1,641,038	\$ 3,613,479	\$ 24,603,252	\$ 24,603,252
Non-cash financing activities:					
Issuance of Series A preferred stock in settlement of convertible notes	\$ —	\$ —	\$ —	\$ —	\$ 5,383,000
Accretion of preferred stock to redemption value	\$ 2,970,586	\$ 3,322,647	\$ 2,468,546	\$ 52,861,367	\$ 63,713,722
Reclassification of 2012 Series X preferred stock from debt to preferred stock	\$ —	\$ —	\$ —	\$ 2,486,251	\$ 2,486,251
Conversion of 2012 Series X preferred stock into Series C preferred stock	\$ —	\$ —	\$ —	\$ 4,944,155	\$ 4,944,155
Loss on extinguishment of 2007 Series X preferred stock	\$ —	\$ —	\$ —	\$ —	\$ 597,174
Book value of assets transferred in distribution of subsidiary	\$ 193,114	\$ —	\$ —	\$ —	\$ 193,114
Book value of liabilities transferred in distribution of subsidiary	\$ (575,964)	\$ —	\$ —	\$ —	\$ (575,964)
Exchange of convertible debt	\$ —	\$ —	\$ —	\$ —	\$ 304,054

See accompanying notes to financial statements.

Akebia Therapeutics, Inc.
(A Development Stage Company)
Notes to Financial Statements
September 30, 2013

1. Nature of Organization and Operations

Akebia Therapeutics, Inc. (the “Company”) is a biopharmaceutical company focused on the development of novel proprietary therapeutics based on hypoxia inducible factor (“HIF”) biology and the commercialization of these products for patients with kidney disease. HIF is the primary regulator of the production of red blood cells in the body and a potentially novel mechanism of treating anemia. The Company’s lead product candidate, AKB-6548, is being developed as a once-daily oral therapy that has successfully completed a Phase 2a proof of concept study demonstrating that AKB-6548 can safely and predictably raise hemoglobin levels in patients with anemia secondary to chronic kidney disease (“CKD”) not requiring dialysis. AKB-6548 is currently being studied in a Phase 2b trial in patients with anemia secondary to CKD, who are not dependent on dialysis, with data expected in the fourth quarter of 2014.

The Company’s operations to date have been limited to organizing and staffing the Company, business planning, raising capital, acquiring and developing its technology, identifying potential product candidates and undertaking preclinical and clinical studies. The Company has not generated any product revenues to date, nor is there any assurance of any future product revenues. The Company’s product candidates are subject to long development cycles and there is no assurance the Company will be able to successfully develop, obtain regulatory approval, for or market its product candidates. Accordingly, the Company is considered to be in the development stage as defined by US generally accepted accounting principles (“US GAAP”).

The Company is subject to a number of risks similar to other life science companies in the development stage, including, but not limited to, the need to obtain adequate additional funding, possible failure of preclinical testing or clinical trials, the need to obtain marketing approval for its product candidates, competitors developing new technological innovations, the need to successfully commercialize and gain market acceptance of any of the Company’s products that are approved and protection of proprietary technology. If the Company does not successfully commercialize any of its products, it will be unable to generate product revenue or achieve profitability. As of December 31, 2012 and September 30, 2013, the Company had a deficit accumulated during the development stage of approximately \$59.6 million and \$119.2 million, respectively.

Unless otherwise indicated, all information in these financial statements gives retrospective effect to the one-hundred-to-one reverse stock split of the Company’s common stock (the “Stock Split”) that was effected on May 10, 2013 (see Note 9).

The Company was incorporated on February 27, 2007, under the laws of the State of Delaware.

2. Summary of Significant Accounting Policies

Basis of Presentation

The financial statements of the Company are prepared in accordance with US GAAP and stated in US dollars.

The accompanying statement of operations and comprehensive loss and statement of cash flows for the nine months ended September 30, 2012 are unaudited. The interim financial statements have been prepared on the same basis as the annual audited financial statements, and in the opinion of management, reflect all adjustments, which include only normal recurring adjustments necessary for the fair statement of the Company’s results of its operations and its cash flows.

Akebia Therapeutics, Inc.
(A Development Stage Company)
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September 30, 2013

The results for the nine months ended September 30, 2013 and 2012 are not necessarily indicative of results to be expected for the year ending December 31, 2013, any other interim periods, or any future year or period.

In December 2011, the Company spun out the Company's programs focused on the treatment of diabetic eye disease and inflammatory bowel disease into Aerpio Therapeutics, Inc ("Aerpio"), as more fully described in Note 3.

Unaudited Pro Forma Presentation

On November 13, 2013, the Company's Board of Directors authorized the management of the Company to file a registration statement with the Securities and Exchange Commission ("SEC") for the Company to sell shares of its common stock (the "Common Stock") to the public. The unaudited pro forma balance sheet and statement of stockholders' (deficit) equity as of September 30, 2013 assumes the automatic conversion of all outstanding convertible preferred stock into shares of Common Stock upon the completion of this proposed offering.

Unaudited pro forma basic and diluted net loss per share was calculated by dividing net loss attributable to common stockholders, excluding the impact of gains (losses) on the extinguishment of preferred stock and accretion of preferred stock, by the pro forma weighted-average number of common shares outstanding. The unaudited pro forma weighted-average number of common shares outstanding was computed after giving effect to the assumed conversion of the redeemable convertible preferred stock (using the if-converted method) as if such conversion had occurred at the beginning of the period presented, or the date of original issuance, if later.

Segment Information

Operating segments are defined as components of an enterprise about which separate discrete information is available for evaluation by the chief operating decision maker, or decision-making group, in deciding how to allocate resources and in assessing performance. The Company views its operations and manages its business in one operating segment, which is the business of developing and commercializing proprietary therapeutics based on HIF biology.

Use of Estimates

The preparation of financial statements in conformity with US GAAP requires management to make estimates and assumptions that affect the reported amounts of assets and liabilities and disclosure of contingent assets and liabilities at the date of the financial statements and the reported amounts of revenues and expenses during the reporting period. Actual results may differ from those estimates. Management considers many factors in selecting appropriate financial accounting policies and controls, and in developing the estimates and assumptions that are used in the preparation of these financial statements. Management must apply significant judgment in this process. In addition, other factors may affect estimates, including: expected business and operational changes, sensitivity and volatility associated with the assumptions used in developing estimates, and whether historical trends are expected to be representative of future trends. The estimation process often may yield a range of potentially reasonable estimates of the ultimate future outcomes, and management must select an amount that falls within that range of reasonable estimates. Estimates are used in the following areas, among others: stock-based compensation expense, fair value of common stock and the Company's other equity instruments, accrued expenses and income taxes.

The Company utilizes significant estimates and assumptions in determining the fair value of its common stock. The Company granted stock options at exercise prices not less than the fair market value of its common stock as determined by the board of directors contemporaneously at the date such grants were made, with input

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Akebia Therapeutics, Inc.
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from management. The fair value of common stock at the grant date was adjusted in connection with the Company's retrospective fair value assessment for financial reporting purposes. The Board of Directors has determined the estimated fair value of the Company's common stock based on a number of objective and subjective factors, including external market conditions affecting the biotechnology industry sector and the prices at which the Company sold shares of preferred stock, the superior rights and preferences of securities senior to the Company's common stock at the time and the likelihood of achieving a liquidity event, such as an initial public offering or sale of the Company.

The Company utilized various valuation methodologies in accordance with the framework of the American Institute of Certified Public Accountants Technical Practice Aid, *Valuation of Privately-Held Company Equity Securities Issued as Compensation*, to estimate the fair value of its common stock. The methodologies included a probability analysis including both a potential public trading scenario and potential sale scenario. In both scenarios, value is estimated using the guideline public company method. The sale scenario includes an adjustment for a market participant acquisition premium. Value is allocated among the preferred and common shares according to the rights associated with each type of security. Valuation methodologies include estimates and assumptions that require the Company's judgment. These estimates include assumptions regarding future performance, including the successful completion of a public offering. Significant changes to the key assumptions used in the valuations could result in different fair values of common stock at each valuation date.

The Company's results can also be affected by economic, political, legislative, regulatory and legal actions. Economic conditions, such as recessionary trends, inflation, interest and monetary exchange rates, government fiscal policies and changes in the prices of research studies, can have a significant effect on operations. While the Company maintains reserves for anticipated liabilities and carries various levels of insurance, the Company could be affected by civil, criminal, regulatory or administrative actions, claims or proceedings.

Cash and Cash Equivalents

Cash and cash equivalents consist of all cash on hand, deposits and funds invested in short-term investments with original maturities of three months or less at the time of purchase. The Company may maintain balances with its banks in excess of federally insured limits.

Investments

Management determines the appropriate classification of securities at the time of purchase and reevaluates such designation as of each balance sheet date. Currently, the Company classifies all investments as trading account assets. Trading account assets are generally available for resale. Trading account assets, consisting principally of corporate and government debt securities, are stated at fair value. Gains and losses, both realized and unrealized, are included in the caption Interest income (expense), net within the statements of operations and comprehensive loss. The Company bases the cost of securities sold upon the specific identification method, and includes interest and dividends on securities in interest income.

Grant Income

Grant income is recognized as earned based on contract work performed. Grant income also includes qualifying therapeutic credits from the US Treasury related to discovery projects.

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Research and Development

Costs incurred in connection with research and development activities are expensed as incurred. Research and development expense consists of (i) employee-related expenses, including salaries, benefits, travel and stock-based compensation expense; (ii) external research and development expenses incurred under arrangements with third parties, such as contract research organizations and consultants; (iii) the cost of acquiring, developing and manufacturing clinical study materials; (iv) facilities and other expenses, which include direct and allocated expenses for rent and maintenance of facilities and laboratory and other supplies; and (v) costs associated with preclinical activities and regulatory operations.

The Company enters into consulting, research and other agreements with commercial firms, researchers, universities and others for the provision of goods and services. Under such agreements, the Company may pay for services on a monthly, quarterly, project or other basis. Such arrangements are generally cancellable upon reasonable notice and payment of costs incurred. Costs are considered incurred based on an evaluation of the progress to completion of specific tasks under each contract using information and data provided to us by the Company's clinical sites and vendors. These costs consist of direct and indirect costs associated with specific projects, as well as fees paid to various entities that perform certain research on behalf of the Company.

Patents

Costs incurred in connection with the application for and issuances of patents are expensed as incurred.

Organizational Costs

All organizational costs and start-up costs are expensed as incurred.

Income Taxes

Income taxes are recorded in accordance with Financial Accounting Standards Board ("FASB") Accounting Standards Codification ("ASC") Topic 740, Income Taxes ("ASC 740"), which provides for deferred taxes using an asset and liability approach. The Company recognizes deferred tax assets and liabilities for the expected future tax consequences of events that have been included in the financial statements or tax returns. Deferred tax assets and liabilities are determined based on the difference between the financial statement and tax bases of assets and liabilities using enacted tax rates in effect for the year in which the differences are expected to reverse. Valuation allowances are provided, if, based upon the weight of available evidence, it is more likely than not that some or all of the deferred tax assets will not be realized.

The Company accounts for uncertain tax positions in accordance with the provisions of ASC 740. When uncertain tax positions exist, the Company recognizes the tax benefit of tax positions to the extent that the benefit will more likely than not be realized. The determination as to whether the tax benefit will more likely than not be realized is based upon the technical merits of the tax position, as well as consideration of the available facts and circumstances. As of December 31, 2011 and 2012, and September 30, 2013, the Company does not have any significant uncertain tax positions.

Stock-Based Compensation

The Company accounts for its stock-based compensation awards in accordance with FASB ASC Topic 718, *Compensation—Stock Compensation* ("ASC 718"). ASC 718 requires all stock-based payments to employees,

Akebia Therapeutics, Inc.
(A Development Stage Company)
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including grants of employee stock options and restricted stock and modifications to existing stock options, to be recognized in the statements of operations and comprehensive loss based on their fair values. All of the Company's stock-based awards are only subject to service based vesting conditions. The Company uses the Black-Scholes option pricing model to determine the fair value of options granted, and an estimate of the Company's Common Stock value to determine the fair value of restricted stock awards.

Compensation expense related to awards to employees is recognized on a straight-line basis based on the grant date fair value over the associated service period of the award, which is generally the vesting term. Awards to non-employees are adjusted through share-based compensation expense as the award vests to reflect the current fair value of such awards, and expensed using an accelerated attribution model.

The Company expenses restricted stock awards based on the fair value of the award on a straight-line basis over the associated service period of the award. Awards of restricted stock to non-employees are adjusted through share-based compensation expense at each reporting period end to reflect the current fair value of such awards, and expensed using an accelerated attribution model.

The Company estimates the fair value of its stock-based awards to employees, non-employees and directors using the Black-Scholes option pricing model, which requires the input of and subjective assumptions, including (a) the expected stock price volatility, (b) the calculation of expected term of the award, (c) the risk-free interest rate and (d) expected dividends.

Due to the lack of a public market for the trading of the Company's common stock and a lack of company-specific historical and implied volatility data, the Company has based its estimate of expected volatility on the historical volatility of a group of similar companies that are publicly traded. The computation of expected volatility is based on the historical volatility of a representative group of companies with similar characteristics to the Company, including stage of product development and life science industry focus. As a result of being a development stage company in a very early stage of product development with no revenues, the representative group of companies has certain similar characteristics to the Company. The Company believes the group selected has sufficient similar economic and industry characteristics, and includes companies that are most representative of the Company. The Company performed a sensitivity analysis to determine the impact a 30% increase or decrease in the volatility rate would have on the fair value of each stock-based award, and determined that such a rate change would be immaterial to the calculation of stock-based compensation.

The Company uses the simplified method as prescribed by the SEC Staff Accounting Bulletin No. 107, *Share-Based Payment*, to calculate the expected term as it does not have sufficient historical exercise data to provide a reasonable basis upon which to estimate the expected term for options granted to employees, and utilizes the contractual term for options granted to non-employees. The expected term is applied to the stock option grant group as a whole, as the Company does not expect substantially different exercise or post-vesting termination behavior among its employee population. The risk-free interest rate is based on a treasury instrument whose term is consistent with the expected life of the stock options.

Consistent with the guidance in FASB ASC Topic 505-50, *Equity-Based Payments to Non-Employees*, the fair value of each non-employee stock option award is estimated at the date of grant using the Black-Scholes option pricing model with assumptions generally consistent with those used for employee stock options, with the exception of expected term, which is over the contractual life.

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Fair Value of Financial Instruments

The Company is required to disclose information on all assets and liabilities reported at fair value that enables an assessment of the inputs used in determining the reported fair values. FASB ASC Topic 820, *Fair Value Measurements and Disclosures* ("ASC 820"), establishes a hierarchy of inputs used in measuring fair value that maximizes the use of observable inputs and minimizes the use of unobservable inputs by requiring that the observable inputs be used when available. Observable inputs are inputs that market participants would use in pricing the asset or liability based on market data obtained from sources independent of the Company. Unobservable inputs are inputs that reflect the Company's assumptions about the inputs that market participants would use in pricing the asset or liability, and are developed based on the best information available in the circumstances. The fair value hierarchy applies only to the valuation inputs used in determining the reported fair value of the investments, and is not a measure of the investment credit quality. The three levels of the fair value hierarchy are described below:

Level 1—Valuations based on unadjusted quoted prices in active markets for identical assets or liabilities that the Company has the ability to access at the measurement date.

Level 2—Valuations based on quoted prices for similar assets or liabilities in markets that are not active, or for which all significant inputs are observable, either directly or indirectly.

Level 3—Valuations that require inputs that reflect the Company's own assumptions that are both significant to the fair value measurement and unobservable.

To the extent that valuation is based on models or inputs that are less observable or unobservable in the market, the determination of fair value requires more judgment. Accordingly, the degree of judgment exercised by the Company in determining fair value is greatest for instruments categorized in Level 3. A financial instrument's level within the fair value hierarchy is based on the lowest level of any input that is significant to the fair value measurement.

Items measured at fair value on a recurring basis include marketable securities (see Note 6). The carrying amounts of accounts payable and accrued expenses approximate their fair values due to their short-term maturities.

Concentrations of Credit Risk and Off-Balance Sheet Risk

Cash and investments are the only financial instruments that potentially subject the Company to concentrations of credit risk. At December 31, 2011, December 31, 2012 and September 30, 2013, all of the Company's cash was deposited in accounts at two principal financial institutions. The Company maintains its cash with a high quality, accredited financial institution and, accordingly, such funds are subject to minimal credit risk. The Company has no significant off-balance sheet concentrations of credit risk, such as foreign currency exchange contracts, option contracts or other hedging arrangements.

Net Loss per Share and Unaudited Pro Forma Net Loss per Share

Basic net loss per share is calculated by dividing net loss attributable to common stockholders by the weighted-average shares outstanding during the period, without consideration for common stock equivalents. Diluted net loss per share is calculated by adjusting weighted-average shares outstanding for the dilutive effect of common stock equivalents outstanding for the period, determined using the treasury-stock method. For purposes of the diluted net loss per share calculation, preferred stock, stock options and unvested restricted stock are considered to be common stock equivalents, but have been excluded from the calculation of diluted net loss per share, as their effect would be anti-dilutive for all periods presented. Therefore, basic and diluted net loss per share were the same for all periods presented.

Akebia Therapeutics, Inc.
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The calculations for the unaudited pro forma basic and diluted net loss applicable to common stockholders per share assume the conversion of all outstanding shares of preferred stock into shares of common stock as if the conversions had occurred at the beginning of the period or the date of issuance, if later (and excludes the gain on extinguishment of preferred stock and the accretion of dividends).

Comprehensive Loss

Comprehensive loss is defined as the change in equity of a business enterprise during a period from transactions and other events and circumstances from non-owner sources. Comprehensive loss was equal to net loss for all periods presented.

Equipment

Equipment is stated at cost, less accumulated depreciation. Equipment is depreciated using the straight-line method over the estimated useful lives of the assets, generally three to seven years. Such costs are periodically reviewed for recoverability when impairment indicators are present. Such indicators include, among other factors, operating losses, unused capacity, market value declines and technological obsolescence. Recorded values of equipment that are not expected to be recovered through undiscounted future net cash flows are written down to current fair value, which generally is determined from estimated discounted future net cash flows (assets held for use) or net realizable value (assets held for sale).

Subsequent Events

The Company evaluates events and transactions occurring subsequent to the date of the financial statements for matters requiring recognition or disclosure in the financial statements. The accompanying financial statements consider events through December 20, 2013, the date on which the financial statements were available to be issued.

Recent Accounting Pronouncements

In February 2013, the FASB issued guidance to provide information about the amounts reclassified out of accumulated other comprehensive income, or AOCI, by component. An entity is required to present, either on the face of the financial statements or in the notes, significant amounts reclassified out of AOCI by the respective line items of net income, but only if the amount reclassified is required to be reclassified in its entirety in the same reporting period. For amounts that are not required to be reclassified in their entirety to net income, an entity is required to cross-reference to other disclosures that provide additional details about those amounts. On January 1, 2013, the Company adopted this standard, which had no impact on the Company's financial position or results of operations.

In June 2011, the FASB issued an amendment to the accounting guidance for presentation of comprehensive income. Under the amended guidance, a company may present the total of comprehensive income, the components of net income and the components of other comprehensive income either in a single continuous statement of comprehensive income or in two separate but consecutive statements. In either case, a company is required to present each component of net income along with total net income, each component of other comprehensive income along with a total for other comprehensive income, and a total amount for comprehensive income. The amendment is effective for fiscal years ending, and interim periods within those years, beginning after December 15, 2011, and is applied retrospectively. The adoption of this amendment has not had a material impact on the Company's financial position or results of operations.

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In May 2011, the FASB issued amended guidance on fair value measurements. This newly issued accounting standard clarifies the application of certain existing fair value measurement guidance, and expands the disclosures for fair value measurements that are estimated using significant unobservable (Level 3) inputs. This accounting standard was effective on a prospective basis for annual and interim reporting periods beginning on or after December 15, 2011. The adoption of this standard has not had a material impact on the Company's financial position or results of operations.

3. Distribution of Aerpio

On December 22, 2011, the Company assigned certain assets and liabilities to a wholly owned subsidiary, Aerpio. The assigned assets and liabilities included all of the Company's fixed assets, the Company's Tie2 activator program, AKB-9778, for diabetic macular edema, the HIF-1 stabilizer program, AKB-4924, for inflammatory bowel disease and contracts, intellectual property, current assets and current liabilities associated with these programs. The Aerpio shares were then distributed to the Company's shareholders as a distribution on the basis of 1 share of Aerpio Series A Preferred Stock for every 35 shares of Akebia Series A Preferred Stock owned, 1 share of Aerpio Series A Preferred Stock for every 100 shares of Akebia Series B Preferred Stock owned, and 1 share of Aerpio Common Stock for every 100 shares of Akebia Common Stock owned.

As of December 22, 2011, the Company assigned the following assets and liabilities to Aerpio at their historical carrying amounts:

Current assets	\$ 30,149
Equipment, net of accumulated depreciation	162,965
Current liabilities	(575,964)
Net gain on distribution of Aerpio, reflected as an increase to additional-paid-in capital	<u>\$ (382,850)</u>

The Company has not presented Aerpio as a discontinued operation in the accompanying financial statements given the significance of on-going cash flows between the Company and Aerpio. Under the terms of administrative services agreements, the Company and Aerpio obtain from and provide to each other certain services beginning in 2012, and as outlined below. These agreements are cancellable upon mutual agreement or a sale of either company.

Below is a summary of the activities included in the statements of operations and comprehensive loss:

Activity	Financial Statement Caption	Year Ended December 31,		Nine Months Ended September 30,		February 27, 2007 (inception) through September 30, 2013
		2011	2012	2012 (unaudited)	2013	
Reimbursement from Aerpio for Akebia employee costs	Reimbursements from Aerpio	\$ —	\$ 1,971,246	\$ 1,636,868	\$ 815,704	\$ 2,786,950
Facility-related charges from Aerpio	General and administrative expenses	\$ —	\$ 177,757	\$ 137,518	\$ 186,518	\$ 364,275

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Below is a summary of the receivables and payables included in the balance sheet related to Aerpio:

Activity	Financial Statement Caption	December 31,		September 30,
		2011	2012	2013
Amounts receivable from Aerpio	Accounts receivable	\$ 24,875	\$ 60,678	\$ 165,814
Amounts payable to Aerpio	Accounts payable	\$ 30,149	\$ 64,294	\$ 70,408

Prior to distribution on December 22, 2011, the direct revenues and expenses attributable to Aerpio related activities are summarized as follows:

	Year Ended December 31, 2011	February 27, 2007 (Inception) through September 30, 2013
Research and development expenses	\$ 5,093,205	\$ 8,889,877
General and administrative expenses	251,340	251,340
Total operating expenses	(5,344,545)	(9,141,217)
Grant income	221,606	834,603
Loss from Aerpio activities	<u>\$ (5,122,939)</u>	<u>\$ (8,306,614)</u>

4. Investments

Investments at fair value consist of the following:

	December 31,		September 30,
	2011	2012	2013
Certificates of deposit	\$ 280,402	\$—	\$ 1,581,667
Government debt securities	1,085,541	—	7,518,299
Corporate debt securities	—	—	4,013,625
	<u>\$ 1,365,943</u>	<u>\$—</u>	<u>\$ 13,113,591</u>

The estimated fair value of the Company's investment balance at September 30, 2013, by contractual maturity, is as follows:

Due in one year or less	\$ 4,995,472
Due after one year through three years	8,118,119
Total investment securities	<u>\$ 13,113,591</u>

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5. Accrued Expenses

Accrued expenses are as follows:

	<u>December 31,</u>		<u>September 30,</u>
	<u>2011</u>	<u>2012</u>	<u>2013</u>
Professional fees	\$ 153,982	\$ 55,420	\$ 375,229
Accrued bonus	179,730	152,293	363,651
Accrued vacation	46,973	35,751	104,347
Other	63,416	107,786	84,706
Total accrued expenses	<u>\$ 444,101</u>	<u>\$ 351,250</u>	<u>\$ 927,933</u>

6. Fair Value of Financial Instruments

The Company utilizes a portfolio management company for the valuation of the majority of securities held. This company is an independent, third-party vendor recognized to be an industry leader with access to market information that obtains or computes fair market values from quoted market prices, pricing for similar securities, recently executed transactions, cash flow models with yield curves and other pricing models. For valuations obtained from the pricing service, the Company performs due diligence to understand how the valuation was calculated or derived, focusing on the valuation technique used and the nature of the inputs.

Based on the fair value hierarchy, the Company classifies its cash equivalents and marketable securities within Level 1 or Level 2. This is because the Company values its cash equivalents and marketable securities using quoted market prices or alternative pricing sources and models utilizing market observable inputs.

Assets measured or disclosed at fair value on a recurring basis as of September 30, 2013 are summarized below:

	<u>Fair Value Measurements Using</u>			<u>Total</u>
	<u>Level 1</u>	<u>Level 2</u>	<u>Level 3</u>	
Assets:				
Cash and cash equivalents	\$ 24,603,252	\$ —	\$ —	\$ 24,603,252
Certificates of deposit	—	1,581,667	—	1,581,667
Government debt securities	—	7,518,299	—	7,518,299
Corporate debt securities	—	4,013,625	—	4,013,625
	<u>\$ 24,603,252</u>	<u>\$ 13,113,591</u>	<u>\$ —</u>	<u>\$ 37,716,843</u>

Assets measured or disclosed at fair value on a recurring basis as of December 31, 2012 are summarized below:

	<u>Fair Value Measurements Using</u>			<u>Total</u>
	<u>Level 1</u>	<u>Level 2</u>	<u>Level 3</u>	
Assets:				
Cash and cash equivalents	\$ 1,641,038	\$ —	\$ —	\$ 1,641,038
	<u>\$ 1,641,038</u>	<u>\$ —</u>	<u>\$ —</u>	<u>\$ 1,641,038</u>

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Assets measured or disclosed at fair value on a recurring basis as of December 31, 2011 are summarized below:

Assets:	Fair Value Measurements Using			Total
	Level 1	Level 2	Level 3	
Cash and cash equivalents	\$ 5,011,089	\$ —	\$ —	\$ 5,011,089
Certificates of deposit	—	280,402	—	280,402
Government debt securities	—	1,085,541	—	1,085,541
	<u>\$ 5,011,089</u>	<u>\$ 1,365,943</u>	<u>\$ —</u>	<u>\$ 6,377,032</u>

The Company had no assets or liabilities measured at fair value on a recurring basis using significant unobservable inputs (Level 3) at September 30, 2013 and December 31, 2012 and 2011.

Investment securities are exposed to various risks such as interest rate, market and credit. Due to the level of risk associated with certain investment securities and the level of uncertainty related to changes in the value of investment securities, it is at least reasonably possible that changes in risks in the near term would result in material changes in the fair value of investments.

7. Convertible Notes

2007 Promissory Notes

In 2007, the Company issued convertible secured promissory notes for aggregate proceeds totaling \$1,350,000 (the "2007 Notes"). The 2007 Notes bore interest at a rate of 12% per annum and were payable at maturity. The 2007 Notes also contained a provision that provided for the automatic settlement with securities of the Company upon completion of a Qualified Financing, as defined in the 2007 Notes, and for the optional settlement with securities of the Company upon the closing of any other financing that did not qualify as a Qualified Financing, as defined in the 2007 Notes, both at 50% of the price per share of the security sold in the financing, among other conversion features. The Company concluded that the settlement of the 2007 Notes in connection with a Qualified Financing was the predominant settlement feature and as a result, accounted for the 2007 Notes as share settled debt. Accordingly, the 2007 Notes were accreted to their redemption value over the expected period to redemption. The 2007 Notes were set to expire during 2007. In anticipation of the sale of Series A Redeemable Convertible Preferred Stock (see Note 8), the maturity dates of the outstanding 2007 Notes were extended to the earlier of the closing of Series A Redeemable Convertible Preferred Stock, or January 31, 2008.

In connection with the issuance of Series A Redeemable Convertible Preferred Stock, in January 2008 (see Note 8), the holders of the 2007 Notes agreed to a modification of the conversion discount from 50% to 20%. On January 23, 2008, all of the outstanding principal and accrued interest thereon of the 2007 Notes were settled through the issuance of 45,228 shares of Series A Redeemable Convertible Preferred Stock (see Note 8). The Company recognized a gain on settlement of the 2007 Notes of \$1,268,180 based on the difference between the aggregate fair value of the Series A Redeemable Convertible Preferred Stock issued upon settlement and the accreted carrying value of the 2007 Notes on January 23, 2008.

2008 Promissory Note

In August 2008, a holder of Series A Redeemable Convertible Preferred Stock advanced \$1,200,000 to the Company in the form of a convertible note (the "2008 Note"). The 2008 Note required, among other things, that

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the Company maintain an amount equal to the outstanding principal balance of the note in either a certificate of deposit, interest bearing account or other cash equivalent, approved by the holder, as collateral for the unpaid principal balance. The 2008 Note bore interest equal to all investment earnings on the cash held as collateral and was payable at maturity. The original maturity of the 2008 Note was June 30, 2009. In anticipation of the July 2009 sale of Series A Redeemable Convertible Preferred Stock (see Note 8), the maturity date was extended to July 15, 2009. The 2008 Note was originally convertible into 3,000,000 shares of Series A Redeemable Convertible Preferred Stock in fulfillment of the investor's commitments under the 2008 Milestone Offering, which was part of the January 23, 2008 Series A Redeemable Convertible Preferred Stock issuance. The 2008 Milestone Offering was cancelled in May 2009, effectively removing the conversion feature included in the 2008 Note. In addition, in May 2009, the principal value of the 2008 Note was reduced to \$895,946 upon the issuance of a new convertible promissory note in the principal amount of \$304,054 (see below).

On July 15, 2009, the principal balance and accrued interest thereon of the 2008 Note of \$916,137 was applied towards the holder's purchase of Series A Redeemable Convertible Preferred Stock in the associated round of financing. As a result, the Company issued 22,904 shares of Series A Redeemable Convertible Preferred Stock in settlement of its obligations under the 2008 Note in July 2009. The gain on the settlement of the 2008 Note was \$11,452, based on the difference between the aggregate fair value of the Series A Redeemable Convertible Preferred Stock issued upon settlement and the carrying value of the 2008 Note at the date of settlement.

2009 Promissory Notes

In May 2009, the Company issued additional convertible secured promissory notes for aggregate proceeds totaling \$1,395,715. These convertible secured promissory notes have the same terms as the \$304,054 of convertible promissory notes issued in exchange for a portion of the 2008 Note, as discussed above (collectively, the "2009 Notes"). The notes bore interest at a rate of 10% per annum and were payable at maturity. The 2009 Notes also contained a provision that provided for the automatic settlement with securities of the Company upon completion of a Qualified Financing, as defined in the 2009 Notes, at 60% of the price per share of the security sold in the financing. In addition, the 2009 Notes were convertible into shares of the Company's Series A Redeemable Convertible Preferred Stock at the option of the holder at \$40.00 per share. The Company concluded that the settlement of the 2009 Notes in connection with a Qualified Financing was the predominant settlement feature and as a result, accounted for the 2009 Notes as share settled debt. Accordingly, the 2009 Notes were accreted to their redemption value over the expected period to redemption. The maturity date of the 2009 Notes was originally December 31, 2009, but was extendable to December 31, 2010, at the option of the Company, with consent of the lender.

On July 15, 2009, all of the outstanding principal and accrued interest thereon of the 2009 Notes were settled through the issuance of 72,201 shares of Series A Redeemable Convertible Preferred Stock (see Note 8). The Company recognized a gain on settlement of the 2009 Notes of \$36,101 based on the difference between the aggregate fair value of the Series A Redeemable Convertible Preferred Stock issued upon settlement and the accreted carrying value of the 2009 Notes on July 15, 2009.

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8. Redeemable Convertible Preferred Stock

Preferred Stock Financings

2007 Series X Convertible Preferred Stock

In September 2007, the Company issued 9,938 shares of 2007 Series X Preferred Stock to The Procter & Gamble Company in consideration for an exclusive worldwide license agreement to certain patents and technology. The value of these shares was charged to research and development expense based on the fair value of the stock at the time of issuance.

The 2007 Series X Preferred Stock shared ratably with the common stock in liquidation, and at issuance was convertible into common stock at a rate equal to 19.9% of the post-conversion common stock equivalents outstanding. In connection with the issuance of Series A Redeemable Convertible Preferred Stock in January 2008, the 2007 Series X Preferred Stock became convertible into common stock at a rate of 7.25 shares for each share outstanding at the option of the holder, and automatically convertible into common stock on May 1, 2008. The modification of the 2007 Series X Preferred Stock has been treated as an extinguishment. On May 1, 2008, all outstanding shares of 2007 Series X Preferred Stock converted into 72,047 shares of common stock.

2012 Series X Preferred Stock

On July 9, 2012, the Company issued 25,000 shares of 2012 Series X Preferred Stock at \$100.00 per share for aggregate proceeds of \$2,500,001. The terms of the 2012 Series X Preferred Stock at issuance provided that the 2012 Series X Preferred Stock were redeemable at a price of \$200.00 per share at the earlier of a Deemed Liquidation Event, as defined, or March 31, 2013. In accordance with the guidance in ASC No. 480, *Distinguishing Liabilities from Equity*, the shares of 2012 Series X Preferred Stock were considered mandatorily redeemable and classified as liabilities upon issuance. The 2012 Series X Preferred Stock were being accreted to the redemption amount over the period from issuance to March 31, 2013.

In March 2013, the Company issued an additional 25,000 shares of 2012 Series X Preferred Stock at \$100.00 per share for aggregate proceeds of \$2,500,001. In addition, the terms of the 2012 Series X were modified to (i) remove the redemption feature, and (ii) add a conversion feature, which provided the 2012 Series X Preferred Stock, unless converted earlier, were convertible upon the earlier of (a) the approval of the holders of at least 60% of the Series X Convertible Preferred Stock, or (b) June 30, 2013 into a number of shares of Series B Redeemable Convertible Preferred Stock determined by dividing \$100.00 per share by the Series X Conversion Price in effect at the time of conversion. The Series X Conversion Price was \$14.00 per share. In addition, the Series X Convertible Preferred Stock was automatically convertible into fully paid non-assessable shares of capital stock of the Company upon the occurrence of a financing which includes (i) the sale of shares of capital stock that are senior or pari passu with the Series B Redeemable Convertible Preferred Stock with gross proceeds to the Company of at least \$10,000,000, or (ii) in the event of a license deal, a financing plus up-front payment from a licensing transaction which results in gross proceeds to the Company of at least \$30,000,000. Each share of Series X Preferred Stock is automatically converted upon closing of the financing into the number of shares of stock issued in the financing equal to the number of shares of Series X Preferred Stock times \$100, divided by the lowest price paid per share by any investor in the financing. Subsequent to the modification in March 2013, as a result of the removal of the mandatory redemption provisions, the shares of 2012 Series X Preferred Stock were reclassified as temporary equity since the 2012 Series X Preferred Stock could be redeemed at the option of the holder upon Deemed Liquidation Events, as defined. The Company has accounted for the amendment to the 2012 Series X Preferred Stock as an extinguishment of the prior security, which was classified as a liability and the issuance of a new preferred stock due to the significance of the modifications to the

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substantive contractual terms of the preferred stock and the associated fundamental changes to the nature of the preferred stock. The Company recorded a gain of \$2,419,766 in the statement of operations and comprehensive loss for the period ended September 30, 2013 based on the excess of the book value over the fair value of the revised 2012 Series X Convertible Preferred Stock at the date of the modification of \$2,486,251. The fair value of the 2012 Series X Convertible Preferred Stock was determined using a hybrid method, in which one scenario assumed the conversion of preferred shares to common stock in an IPO, and a second scenario allocated value to the preferred shares using the option-pricing method.

Upon the closing of the Series C Redeemable Convertible Preferred Stock financing in May 2013, all of the outstanding shares of 2012 Series X Preferred Stock were converted into 357,143 shares of Series C Redeemable Convertible Preferred Stock.

Redeemable Convertible Preferred Stock

On January 23, 2008, the Company issued 226,500 shares of Series A Redeemable Convertible Preferred Stock at \$40.00 per share for aggregate proceeds of \$9,060,000, less issuance costs of \$90,453. In connection with the financing, the Company settled all outstanding 2007 Notes, including accrued interest, with 45,228 shares of Series A Redeemable Convertible Preferred Stock which was recorded at fair value of \$1,626,386 (see Note 7). Additionally, the investors in Series A Redeemable Convertible Preferred Stock agreed to purchase 151,000 additional shares at a price of \$40.00 per share upon the achievement of certain defined milestones (the 2008 Milestone Offering), which the Company concluded should be accounted for as a freestanding financial instrument. The Company allocated \$610,040 of the proceeds to the 2008 Milestone Offering based on the fair value at issuance. The 2008 Milestone Offering was cancelled in May 2009. The reduction of the fair value of the 2008 Milestone Offering has been reflected in net loss for the year ended December 31, 2009.

On July 15, 2009, the Company issued 238,955 shares of Series A Redeemable Convertible Preferred Stock at \$40.00 per share for aggregate proceeds of \$9,558,209, less issuance costs of \$44,008. Additionally, the investors in Series A Redeemable Convertible Preferred Stock agreed to purchase 86,850 additional shares at a price of \$40.00 per share upon the achievement of certain defined milestones (the 2009 Milestone Offering), which the Company concluded should be accounted for as a freestanding financial instrument. The Company allocated \$43,425 of the proceeds to the 2009 Milestone Offerings based on the fair value at issuance. The 2009 Milestone Offering was cancelled in July 2010. The reduction of the fair value of the 2009 Milestone Offering has been reflected in net loss for the year ended December 31, 2010.

In connection with the July 15, 2009 issuance, the Company issued an additional 22,904 shares in settlement of the 2008 Note. In addition, the Company settled all outstanding 2009 Notes including accrued interest with 72,201 shares of Series A Redeemable Convertible Preferred Stock. The shares of Series A Redeemable Convertible Preferred Stock issued in exchange of the 2008 Notes and 2009 Notes, were recorded at fair value at the date of issuance of \$904,685 and \$2,851,929, respectively.

On November 4, 2009, the Company issued 3,750 shares of Series A Redeemable Convertible Preferred Stock at \$40.00 per share for aggregate proceeds of \$150,000. Additionally, the investor in the financing agreed to purchase an additional 1,250 shares at a price of \$40.00 per share as part of the 2009 Milestone Offering. The Company did not allocate any of the proceeds to the additional participant in the 2009 Milestone Offering, since the fair value was immaterial.

On June 7, 2010, the Company issued 125,000 shares of Series A Redeemable Convertible Preferred Stock at \$40.00 per share for aggregate proceeds of \$5,000,000, less issuance costs of \$29,403.

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On April 6, 2011, the Company issued 857,142 shares of Series B Redeemable Convertible Preferred Stock at \$14.00 per share for aggregate proceeds of \$11,999,998, less issuance costs of \$293,905. On April 21, 2011, the Company issued 138,954 shares of Series B Redeemable Convertible Preferred Stock at \$14.00 per share for total aggregate proceeds of \$1,945,343. On December 23, 2011, the Company issued 291,429 shares of Series B Redeemable Convertible Preferred Stock at \$14.00 per share for total proceeds of \$4,080,000, less issuance costs of approximately \$9,000.

On May 10, 2013, the Company issued 2,202,887 shares of Series C Redeemable Convertible Preferred Stock at \$14.00 per share for aggregate proceeds of \$30,840,388. In addition, on May 31, 2013, the Company issued additional 742,855 shares of Series C Redeemable Convertible Preferred Stock at \$14.00 per share for aggregate proceeds of \$10,400,000. The issuance costs related to the issuances of Series C Redeemable Convertible Preferred Stock were \$1,151,923.

Summary of Redeemable Convertible Preferred Stock and 2012 Series X Preferred Stock

As of September 30, 2013, the authorized capital stock of the Company included 5,500,636 shares of preferred stock, par value \$0.00001 per share, of which: (i) 734,538 shares have been designated as Series A redeemable convertible preferred stock (Series A Redeemable Convertible Preferred Stock), (ii) 1,287,525 shares have been designated as Series B redeemable convertible preferred stock (Series B Redeemable Convertible Preferred Stock), (iii) 3,428,572 shares have been designated as Series C redeemable convertible preferred stock (Series C Redeemable Convertible Preferred Stock) and (v) 50,001, shares have been designated as Series X convertible preferred stock (Series X Convertible Preferred Stock). There is no outstanding Series X Convertible Preferred Stock as of September 30, 2013. The Series A Redeemable Convertible Preferred Stock, the Series B Redeemable Convertible Preferred Stock and the Series C Redeemable Convertible Preferred Stock are collectively referred to as the Redeemable Convertible Preferred Stock.

General

The rights, preferences and privileges of the preferred stock are as follows:

Voting

The holders of shares of Redeemable Convertible Preferred Stock are entitled to the number of votes equal to the number of whole shares of common stock into which the shares of the applicable series of Redeemable Convertible Preferred Stock held by such holder are convertible relating to any matter presented to the stockholders of the Company for their action or consideration at any meeting of stockholders of the Company, or by written consents of stockholders in lieu of meetings. Except as provided by law or otherwise, the holders of shares of Redeemable Convertible Preferred Stock vote together with the holders of shares of common stock as a single class.

The affirmative vote or written consent of the holders of shares constituting 50% (the Appropriate Percentage) of the then outstanding shares of Redeemable Convertible Preferred Stock is required in order for the Company to, among other things: (i) liquidate, dissolve or wind-up the business and affairs of the Company or effect any Deemed Liquidation Event, as defined, (ii) amend, alter or repeal any provision of the Company's Certificate of Incorporation or bylaws that adversely affects the powers, preferences or rights of the Redeemable Convertible Preferred Stock, (iii) create or authorize the creation of, or issue or obligate itself to issue shares of, any additional class or series of capital stock or increase the authorized number of shares of any class of Redeemable Convertible Preferred Stock or increase the authorized number of shares of any additional class or series of capital stock,

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(iv) reclassify, alter or amend any security of the Company which is pari passu with or junior to any class of the Redeemable Convertible Preferred Stock with respect to the payment of dividends or rights of redemption if such reclassification, alteration or amendment would render such other security senior to any class of Redeemable Convertible Preferred Stock with respect to any such right, preference or privilege, (v) purchase, redeem, pay or declare any dividend or make any distribution on, any shares of capital stock other than (1) redemptions of or distributions on Redeemable Convertible Preferred Stock as expressly authorized by the certificate of incorporation and (2) dividends or other distributions payable on the Common Stock solely in the form of additional shares of Common Stock, including the conversion of accrued and unpaid dividends on the Series C Redeemable Convertible Preferred Stock into additional shares of Common Stock pursuant to the articles of incorporation or (3) repurchases of stock from former employees, officers, directors, consultants or other persons, who performed services for the Company, in connection with the cessation of such employment or service that are approved by the Board of Directors, (vi) create, or authorize the creation of, or issue, or authorize the issuance of, any debt security, (vii) create or hold capital stock in, any subsidiary that is not wholly owned by the Company or sell, transfer or otherwise dispose of any capital stock of any subsidiary of the Company or permits any subsidiary to sell, lease, transfer or exclusively license or otherwise dispose of all or substantially all of the assets of such subsidiary or (viii) increase or decrease the authorized number of directors constituting the Company's Board of Directors.

Additionally, the affirmative vote by holders of at least 75% of the then-outstanding shares of Series C Redeemable Convertible Preferred Stock is required in order to (1) declare or pay any dividend on Series C Redeemable Convertible Preferred Stock, (2) amend, modify or waive the Company's Certificate of Incorporation in a manner that adversely affects the powers, preferences or rights to the Series C Redeemable Convertible Preferred Stock or (3) increase or decrease the number of authorized shares of Series C Redeemable Convertible Preferred Stock. The affirmative vote by holders of 65% of the then outstanding shares of Series B Redeemable Convertible Preferred Stock is required to (1) amend, modify or waive the Company's Certificate of Incorporation in a manner that adversely affects the powers, preferences or rights of the Series B Redeemable Convertible Preferred Stock or (2) increase or decrease the number of authorized shares of Series B Redeemable Convertible Preferred Stock. The affirmative vote of holders of 65% of the then-outstanding shares of Series A Redeemable Convertible Preferred Stock is required to (1) amend, modify or waive the Company's Certificate of Incorporation in a manner that adversely affects the powers, preferences or rights of the Series A Redeemable Convertible Preferred Stock, or (2) increase or decrease the number of authorized shares of Series B Redeemable Convertible Preferred Stock.

The holders of shares of Redeemable Convertible Preferred Stock are entitled to elect six members of the Company's Board of Directors, which is subject to reduction to not less than four directors under certain circumstances. The holders of shares of Common Stock (including any holders of all shares of Redeemable Convertible Preferred Stock on an as converted basis), are entitled to elect three members of the Company's Board of Directors, which is subject to reduction to two directors under certain circumstances.

Dividends

The holders of shares of Redeemable Convertible Preferred Stock are entitled to receive dividends, at a rate of 6% for the Series A Redeemable Convertible Preferred Stock and the Series B Convertible Preferred Stock and 8% for the Series C Convertible Preferred Stock. Dividends accrue daily (and compound quarterly) whether or not declared and are cumulative. Dividends are payable only if permitted by law and when and if declared by the Board of Directors. The holders of Series X Convertible Preferred Stock are not entitled to dividends.

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Liquidation

In the event of any voluntary or involuntary liquidation, dissolution or winding up of the Company, or upon the occurrence of a Deemed Liquidation Event, as defined, at the election of holders of more than 50% of the shares of Redeemable Convertible Preferred Stock, the holders of shares of Series C Redeemable Convertible Preferred Stock then outstanding are entitled to be paid out, of the assets of the Company available for distribution to stockholders, an amount per share equal to \$28.00, subject to appropriate adjustment, plus an amount equal to 200% of any accrued but unpaid dividends thereon (Series C Liquidation Amount), before any payment is made to the holders of shares of Series B Redeemable Convertible Preferred Stock, holders of shares of Series A Redeemable Convertible Preferred Stock or holders of shares of Common Stock. Next, the holders of shares of Series B Redeemable Convertible Preferred Stock then outstanding are entitled to be paid out of the assets of the Company available for distribution to stockholders, an amount per share equal \$14.00 per share, subject to appropriate adjustment, plus any accrued but unpaid dividends thereon (Series B Liquidation Amount), before any payment is made to the holders of shares of Series A Redeemable Convertible Preferred Stock or holders of shares of Common Stock. Next, the holders of shares of Series A Redeemable Convertible Preferred Stock then outstanding are entitled to be paid, out of the assets of the Company available for distribution to stockholders, an amount per share equal to the greater of \$40.00, subject to appropriate adjustment, plus any accrued but unpaid dividends thereon (Series A Liquidation Amount), before any payment is made to the holders of shares of Common Stock. In the event the assets of the Company available for distribution to stockholders are insufficient to permit payment of the full amount to which each shareholder is entitled, holders of shares of capital stock will share ratably in any distribution of the remaining assets of the Company in proportion to the respective amounts which would otherwise be payable under the circumstances in the order of liquidation preference.

After the payment of all preferential amounts required to be paid to the holders of shares of Redeemable Convertible Preferred Stock, the remaining assets of the Company available for distribution to stockholders will be distributed among the holders of shares of Redeemable Convertible Preferred Stock and Common Stock, pro rata based on the number of shares held by each such holder, treating such securities as if they had been converted to common stock immediately prior to such dissolution, liquidation or winding up of the Company.

Conversion

Each share of Redeemable Convertible Preferred Stock is convertible at the option of the holder, at any time and from time to time, into fully paid and non-assessable shares of Common Stock. Each share of Series C Redeemable Convertible Preferred Stock is convertible into that number of common shares as is determined by dividing the Series C Accrued Value by the Applicable Conversion Price. The Series C Accrued Value is defined as the Applicable Original Purchase Price (\$14.00 per share for the Series C Redeemable Convertible Preferred Stock as of September 30, 2013, subject to adjustment) plus accrued but unpaid dividends. Each share of Series B Redeemable Convertible Preferred Stock and Series A Redeemable Convertible Preferred Stock is convertible into that number of common shares as is determined by dividing the Applicable Original Purchase Price of such share (\$14.00 per share for the Series B Redeemable Convertible Preferred Stock and \$40.00 per share for the Series A Convertible Preferred Stock, as of September 30, 2013, subject to adjustment) by the Applicable Conversion Price (\$14.00 per share for all Redeemable Convertible Preferred Shares, as of September 30, 2013). The Applicable Conversion Price is subject to adjustment in the future upon the occurrence of certain events.

Each share of Redeemable Convertible Preferred Stock is automatically convertible into fully paid and non-assessable shares of common stock upon either: (i) the closing of the sale of shares of the Company's common stock to the public in an underwritten public offering resulting in at least \$40,000,000 of gross proceeds to the

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Company, or (ii) the date and time, or the occurrence of an event, specified by vote or written consent of the holders of shares constituting 65% of the then outstanding shares of either Series C Redeemable Convertible Preferred Stock, Series B Redeemable Convertible Preferred Stock or Series A Redeemable Convertible Preferred Stock, respectively, voting as a separate class.

The Company evaluated each series of its preferred stock and determined that each individual series is considered an equity host under ASC No. 815, *Derivatives and Hedging* (ASC 815). In making this determination, the Company's analysis followed the whole instrument approach, which compares an individual feature against the entire preferred stock instrument which includes that feature. The Company's analysis was based on a consideration of the economic characteristics and risks of each series of preferred stock. More specifically, the Company evaluated all of the stated and implied substantive terms and features, including: (i) whether the preferred stock included redemption features, (ii) how and when any redemption features could be exercised, (iii) whether the holders of preferred stock were entitled to dividends, (iv) the voting rights of the preferred stock and (v) the existence and nature of any conversion rights. As a result of the Company's conclusion that the preferred stock represents an equity host, the conversion feature of all series of preferred stock is considered to be clearly and closely related to the associated preferred stock host instrument. Accordingly, the conversion feature of all series of preferred stock is not considered an embedded derivative that requires bifurcation.

The Company accounts for potentially beneficial conversion features under ASC No. 470-20, *Debt with Conversion and Other Options* (ASC 470-20). At the time of each of the issuances of Redeemable Convertible Preferred Stock and the Series X Convertible Preferred Stock, the Company's Common Stock (or Series B Redeemable Convertible Preferred Stock in the case of the Series X Convertible Preferred Stock) into which each series of the Company's preferred stock is convertible had an estimated fair value less than the effective conversion prices of the convertible preferred stock. Therefore, there was no intrinsic value on the respective commitment dates.

Redemption

The Redeemable Convertible Preferred Stock shall be redeemed by the Company at a price equal to the greater of (i) the Applicable Accrued Value or (ii) the fair market value per share, in three annual installments commencing not more than 60 days following receipt by notice, at any time after July 16, 2015, from holders of more than 50% of the shares of Redeemable Convertible Preferred Stock then outstanding. For the Series C Redeemable Convertible Preferred Stock, the Series B Convertible Preferred Stock and the Series A Convertible Preferred Stock, the Applicable Accrued Value is equal to the Series C Liquidation Amount, the Series B Liquidation Amount and the Series A Liquidation Amount, respectively. In accordance with the guidance in ASC No. 480, *Distinguishing Liabilities from Equity* (ASC 480), shares of Redeemable Convertible Preferred Stock are classified outside of permanent stockholders' deficit.

Extinguishments of Preferred Stock

In connection with the issuance of the Series A Redeemable Convertible Preferred Stock on January 23, 2008, certain rights, preferences and privileges for the outstanding 2007 Series X Convertible Preferred Stock were modified. More specifically, the conversion feature was modified to fix the conversion of each share of 2007 Series X Convertible Preferred Stock into 7.25 shares of common stock. In addition, the conversion feature was modified to include an automatic conversion upon the earlier of (i) a Deemed Liquidation Event, as defined, or (ii) May 1, 2008.

The Company has accounted for the amendment to the rights, preferences and privileges of the 2007 Series X Convertible Preferred Stock as an extinguishment of the old preferred stock and issuance of new

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preferred stock due to the significance of the modifications to the substantive contractual terms of the preferred stock and the associated fundamental changes to the nature of the preferred stock. Accordingly, the Company recorded a loss of \$597,174 within stockholders' deficit equal to the difference between the fair value of the new shares of preferred stock issued and the carrying amount of the old shares of preferred stock extinguished. The loss on extinguishment is reflected as an adjustment to the net loss available to common stockholders in accordance with ASC No. 260, *Earnings per Share* (ASC 260). The fair value of the 2007 Series X Convertible Preferred Stock was determined primarily based on the underlying value of the common stock to be received upon conversion of the 2007 Series X Convertible Preferred Stock.

9. Stockholders' Equity

As of September 30, 2013, the authorized capital stock of the Company included 8,400,000 shares of common stock, par value \$0.00001 per share.

On May 10, 2013, the Company effected a one hundred-for-one reverse stock split. Unless otherwise indicated, all share data and per share amounts in these financial statements have been retroactively adjusted to reflect the reverse stock split.

General

The voting, dividend and liquidation rights of the holders of shares of Common Stock are subject to and qualified by the rights, powers and preferences of the holders of shares of Redeemable Convertible Preferred Stock. The Common Stock has the following characteristics:

Voting

The holders of shares of Common Stock are entitled to one vote for each share of Common Stock held at all meetings of stockholders and written actions in lieu of meetings. Notwithstanding the foregoing, except as otherwise required by law, holders of shares of Common Stock shall not be entitled to vote on any amendment to this Certificate of Incorporation or pursuant to General Corporation Law.

Dividends

The holders of shares of Common Stock are entitled to receive dividends, if and when declared by the Board of Directors. The Company may not declare or pay any cash dividends to the holders of Common Stock unless, in addition to obtaining any necessary consents, dividends are paid on each series of Redeemable Convertible Preferred Stock in accordance with their respective terms. As of September 30, 2013, no dividends have been declared or paid to the holders of Common Stock since the inception of the Company.

Liquidation

After payment to the holders of shares of Redeemable Convertible Preferred Stock of their liquidation preferences, the holders of Common Stock are entitled to share ratably in the Company's assets available for distribution to stockholders, in the event of any voluntary or involuntary liquidation, dissolution or winding up of the Company.

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As of December 31, 2011 and 2012, and September 30, 2013, the Company has reserved the following shares of Common Stock for future issuance based upon the outstanding Redeemable Convertible Preferred Stock and Stock Options:

	December 31, 2012	September 30, 2013
Conversion of Series A Redeemable Convertible Preferred Stock	2,098,680	2,098,680
Conversion of Series B Redeemable Convertible Preferred Stock	1,642,857	1,287,525
Conversion of Series C Redeemable Convertible Preferred Stock	—	3,529,631
Options to purchase common stock	555,529	767,433
Total	<u>4,297,066</u>	<u>7,683,269</u>

10. Income Taxes

There was no current or deferred income tax expense or benefit for the nine-month periods ended September 30, 2013 and 2012, and the years ended December 31, 2012 and 2011, due to the Company's net losses and increases in its deferred tax asset valuation allowance.

The US components of loss before income taxes, and a reconciliation of the statutory federal income tax with the provision for income taxes, follow:

	Years Ended		For the Nine Months Ended	
	December 31, 2011	December 31, 2012	September 30, 2012 (unaudited)	September 30, 2013
Federal tax at statutory rate	34.0%	34.0%	34.0%	34.0%
State and local tax at statutory rate	0.7	0.7	0.7	0.7
Research and development tax credits	3.3	0.0	0.0	6.5
Disqualified interest expense	0.0	(7.0)	(4.7)	(3.6)
Cancellation of debt income	0.0	0.0	0.0	11.6
Distribution of Aerpio	(15.4)	0.0	0.0	0.0
Other	(0.7)	(0.5)	(0.6)	(0.8)
Change in valuation allowance	(21.9)	(27.2)	(29.4)	(48.4)
Effective tax rate	<u>— %</u>	<u>— %</u>	<u>— %</u>	<u>— %</u>

The Company's income tax provision was computed based on the federal statutory rate and the average state statutory rates, net of the related federal benefit.

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Deferred income taxes reflect the net tax effects of temporary differences between the carrying amounts of assets and liabilities for financial reporting purposes and the amounts used for income tax purposes. Significant components of the Company's deferred tax assets and liabilities are as follows:

	December 31, 2011	December 31, 2012	September 30, 2013
Deferred tax assets:			
Net operating loss carry-forwards	\$ 10,785,101	\$ 13,074,606	\$ 16,142,569
Intangible assets	718,485	660,635	617,247
Research and development credit carry-forwards	1,261,955	1,261,955	1,731,573
Accrued expenses	82,183	76,586	76,586
Other	13,827	14,932	16,064
Deferred tax assets	<u>12,861,551</u>	<u>15,088,714</u>	<u>18,584,039</u>
Deferred tax liabilities:			
Accumulated depreciation	<u>—</u>	<u>—</u>	<u>381</u>
Deferred tax liabilities	<u>—</u>	<u>—</u>	<u>381</u>
Less: valuation allowance	(12,861,551)	(15,088,714)	(18,583,658)
Net deferred tax asset	<u>\$ —</u>	<u>\$ —</u>	<u>\$ —</u>

When realization of the deferred tax asset is more likely than not to occur, the benefit related to the deductible temporary differences attributable to operations is recognized as a reduction of income tax expense. Valuation allowances are provided against deferred tax assets when, based on all available evidence, it is considered more likely than not that some portion or all of the recorded deferred tax assets will not be realized in future periods. The Company cannot be certain that future taxable income will be sufficient to realize its deferred tax assets, and accordingly a full valuation allowance has been provided on its deferred tax assets. The Company continues to maintain the underlying tax benefits to offset future taxable income, and to monitor the need for a valuation allowance based on the profitability of its future operations.

At December 31, 2011, December 31, 2012 and September 30, 2013, the Company has approximately \$1,574,000 (after amortization of \$393,500), \$1,443,000 (after amortization of \$524,700) and \$1,345,000 (after amortization of \$623,000), respectively, of start-up expenses capitalized for income tax purposes with amortization available to offset future federal, state and local income tax. Additionally, at December 31, 2011, December 31, 2012 and September 30, 2013, the Company has approximately \$31,079,000, \$37,673,000 and \$46,510,000, respectively, of net operating loss (NOL) carry-forwards, and \$1,261,000, \$1,261,000 and \$1,731,000, respectively, of research and development tax credit carry-forwards. The NOL and research and development tax credit carry-forwards begin to expire in 2027, and will be utilized for tax purposes at such time the Company generates taxable income. The NOL and research and development tax credit carry-forwards may be limited in certain circumstances, including ownership changes.

Under the provisions of the Internal Revenue Code, the net operating loss and tax credit carry-forwards are subject to review and possible adjustment by the Internal Revenue Service and state tax authorities. Net operating loss and tax credit carry-forwards may become subject to an annual limitation in the event of certain cumulative changes in the ownership interest of significant shareholders over a three-year period in excess of 50%, as defined under Sections 382 and 383 of the Internal Revenue Code, respectively, as well as similar state provisions. This could limit the amount of tax attributes that can be utilized annually to offset future taxable income or tax liabilities.

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The amount of the annual limitation is determined based on the value of the Company immediately prior to the ownership change. Subsequent ownership changes may further affect the limitation in future years. The Company has completed several financings since its inception, which may have resulted in a change in control as defined by Sections 382 and 383 of the Internal Revenue Code, or could result in a change in control in the future.

Prior to legislation enacted in January 2013, the research and development tax credit was not in place for the 2012 tax year. Accordingly, the Company has not recorded a deferred tax asset for the 2012 research and development tax credit at December 31, 2012. For applicable years, the Company generated research credits but has not conducted a study to document its qualified activities. Such a study may result in an adjustment to the Company's research and development credit carry-forwards; however, until a study is completed and any adjustment is known, no amounts are being presented as an uncertain tax position. A full valuation allowance has been provided against the Company's research and development credits and, if an adjustment is required, this adjustment would be offset by an adjustment to the deferred tax asset established for the research and development credit carry-forwards and the valuation allowance.

The Company will recognize interest and penalties related to uncertain tax positions in income tax expense. As of December 31, 2011, December 31, 2012 and September 30, 2013, the Company had no accrued uncertain tax positions or associated interest or penalties and no amounts have been recognized in the Company's consolidated statements of operations.

The Company files income tax returns in the US federal jurisdiction, and various state jurisdictions. The Company's 2009-2012 tax years remain open and subject to examination by federal and state taxing authorities.

11. Commitments and Contingencies

The Company contracts with various organizations to conduct research and development activities with remaining contract costs to the Company of \$1,530,149, \$737,030 and \$4,907,529 at December 31, 2011, December 31, 2012 and September 30, 2013, respectively.

12. Stock Based Compensation

In connection with the sale of the Series A and Series B Redeemable Convertible Preferred Stock, the Company approved the creation of an equity incentive plan. There are 64,408 shares of the Company's Common Stock that are reserved for issuance under the plan at September 30, 2013. The plan allows for the grant of incentive stock options and non-qualified stock options to purchase common stock or stock bonuses, or restricted stock awards for management and certain persons performing services for the Company.

Stock Options

The options granted to directors and non-employees vest over periods of between 12 and 48 months. For employees with less than one year's service, options vest in installments of 25% at the one year anniversary and in 36 equal monthly installments beginning in the thirteenth month after the initial Vesting Commencement Date (as defined), subject to the employee's continuous service with the Company. Options granted to other employees vest in 48 equal monthly installments after the initial Vesting Commencement Date (as defined), subject to the employee's continuous service with the Company. The options generally expire ten years after the date of grant. The fair value of these options granted will be recognized as an expense over the requisite service period.

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The fair value of each stock-based award granted to employees is estimated on the grant date using the Black-Scholes option pricing model using the following assumptions:

	<u>December 31,</u>		<u>September 30,</u>
	<u>2011</u>	<u>2012</u>	<u>2013</u>
Risk-free interest rate	1.09%	0.95%	1.71%
Dividend yield	0.00%	0.00%	0.00%
Volatility	74.00%	73.00%	79.00%
Expected term (years)	6.25	6.25	6.25
Weighted-average estimated fair value of options granted during the period	\$ 1.32	\$ 0.57	\$ 6.08

The fair value of each non-employee stock option award is estimated at the date of grant using the Black-Scholes option pricing model, with assumptions generally consistent with those used for employee stock options, with the exception of expected term, which is over the contractual life.

The weighted-average fair values of options granted are as follows:

<u>Period</u>	<u>Weighted-Average Fair Value of Options Granted</u>
2013 (9 Months)	\$ 6.08
2012	\$ 0.57
2011	\$ 1.32
2010	\$ 2.66
2009	\$ 10.45
2008	\$ 7.51

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The following table summarizes the stock option activity for employees and non-employees:

	Shares	Weighted-Average Exercise Price	Weighted-Average Contractual Life (in years)	Aggregate Intrinsic Value
Outstanding, January 1, 2008	—	\$ —		
Granted	103,643	1.50		
Exercised	—	—		
Expired/cancelled	—	—		
Outstanding, December 31, 2008	103,643	\$ 1.50		
Granted	46,149	1.50		
Exercised	—	—		
Expired/cancelled	(900)	1.50		
Outstanding, December 31, 2009	148,892	\$ 1.50		
Granted	66,080	1.50		
Exercised	—	—		
Expired/cancelled	—	—		
Outstanding, December 31, 2010	214,972	\$ 1.50		
Granted	24,330	1.50		
Exercised	—	—		
Expired/cancelled	—	—		
Outstanding, December 31, 2011	239,302	\$ 1.50		
Granted	40,451	1.50		
Exercised	—	—		
Expired/cancelled	—	—		
Outstanding, December 31, 2012	279,753	\$ 1.50		
Granted	427,519	0.82		
Exercised	(4,647)	1.50		
Expired/cancelled	—	—		
Outstanding, September 30, 2013	702,625	\$ 1.08	8.47	\$3,877,000
Options exercisable, September 30, 2013	231,218	\$ 1.50	5.87	\$1,179,000
Expected to vest, September 30, 2013	471,407	\$ 0.88	9.83	\$2,698,000

As of September 30, 2013, there was \$2,818,313 of unrecognized compensation cost related to stock options, which is expected to be recognized over a weighted-average period of 2.4 years.

During 2011, all outstanding stock options granted prior to the issuance of the Series B Preferred Stock were amended to increase the number of option shares from 76,629 to 214,972, and reduce the exercise price per share of each outstanding option from \$4.00 to \$1.50. The increased option shares resulting from the modification were vested in the same proportion as the pre-modification option shares. The fair value of the outstanding options was compared pre- and post-modification, resulting in an incremental fair value of approximately \$93,000 to be recognized as additional compensation cost over the remaining requisite service period.

During November 2012, outstanding stock options granted to certain employees transferred to Aerpio were modified to allow these former employees to continue to vest in their awards. Because the employees were not expected to vest in the original award under its terms, the Company reversed approximately \$62,000 in

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previously recognized compensation cost in November 2012. The Company recognized the fair value of the new modified award over the requisite service period. However, no substantive future services will be provided to the Company. Accordingly, the Company has accounted for the modification as a severance arrangement with no future service requirement, and has immediately expensed the full fair value of the modified award in 2012, resulting in incremental compensation cost of approximately \$10,000.

A single employee was subsequently transferred to Aerpio in September 2013, with a similar modification, resulting in the reversal of compensation expense of approximately \$20,000. The fair value of the new modified award was expensed, resulting in incremental compensation cost of approximately \$5,000.

Restricted Stock

On June 15, 2011, March 26, 2010 and October 15, 2009, certain employees of the Company purchased 145,404, 10,603 and 31,474 shares, respectively of common stock at a purchase price of \$1.50, \$4.00 and \$4.00 per share, respectively, under the terms of a restricted stock award. These shares were purchased in exchange for promissory notes (the "Promissory Notes"). The 2009 and 2010 issuances vest in installments of 25% at the one-year anniversary, and in 36 equal monthly installments beginning in the thirteenth month after the Initial Vesting Commencement Date (as defined), subject to the employee's continuous service with the Company. The 2011 issuances vest ratably over 48 equal monthly installments beginning on the initial Vesting Commencement Date (as defined), subject to the employee's continuous service with the Company. The Company may purchase all of the unvested shares following the employee's termination at the original purchase price. 181,874, 125,438 and 78,568, shares were vested at September 30, 2013, and December 31, 2012 and 2011, respectively. The Promissory Notes accrue interest at the rate of 6% per annum, and are repayable at the earlier of (a) an initial public offering; (b) the sale of the Company or substantially all of its assets; (c) the termination of the employee; or (d) five years from origination. The Promissory Notes are partially collateralized by the assets of the employee.

The Company has accounted for the Promissory Notes as non-recourse in their entirety since the Promissory Notes are not aligned with a corresponding percentage of the underlying shares. Accordingly, the non-recourse notes received by the Company as consideration for the issuance of the restricted stock has been considered a stock option for accounting purposes as the substance is similar to the grant of an option. The exercise price is the principal due on the note. The stated interest rate of the Promissory Notes is reflected as the dividend yield. The fair value of the award is recognized over the requisite service period (not the term of the Promissory Note) through a charge to compensation cost. The maturity date of the Promissory Notes reflects the legal term for purposes of valuing the award.

The fair value of the restricted stock granted to employees in exchange for a Promissory Note is estimated on the grant date using the Black-Scholes option pricing model using the following assumptions:

	<u>December 31,</u>		<u>September 30,</u>
	<u>2011</u>	<u>2012</u>	<u>2013</u>
Risk-free interest rate	1.09%	0.95%	1.71%
Dividend yield	6.00%	6.00%	3.00%
Volatility	74.00%	73.00%	79.00%
Expected term (in years)	5	5	5

On May 9, June 6 and June 15, 2013, the terms of the Promissory Notes were amended to (i) reduce the principal owed, and (ii) reduce the interest rate from 6% per annum to 3% per annum. The fair value of the outstanding awards was compared pre- and post-modification, resulting in an incremental fair value of \$172,448 to be recognized as additional compensation cost over the remaining requisite service period.

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In February 2012 and April 2013, an employee of the Company was granted restricted stock awards of 30,047 and 30,763 shares, respectively, of common stock at a fair value of \$1.00 and \$1.56 per share, respectively. The fair value of the award is based on the estimated fair value of the Company's common stock at the date of grant. The shares vest in installments of 25% at the one-year anniversary, and in 36 equal monthly installments beginning in the thirteenth month after the initial Vesting Commencement Date (as defined), subject to the employee's continuous service with the Company. Total compensation cost of \$11,633 and \$6,886 was recognized during the nine-month period ended September 30, 2013, and year ended December 31, 2012, respectively. As of September 30, 2013, there is \$59,689 of unrecognized compensation cost related to these restricted stock grants, which is expected to be recognized over a weighted-average period of 2.1 years. There were 12,520 shares vested at September 30, 2013.

A summary of the Company's restricted stock activity and related information is as follows:

	<u>Shares</u>	<u>Weighted-Average Grant Date Fair Value</u>
Unvested balance, January 1, 2008	—	\$ —
Granted*	—	—
Vested	—	—
Forfeited	—	—
Unvested balance, December 31, 2008	—	\$ —
Granted*	31,474	7.14
Vested	—	—
Forfeited	—	—
Unvested balance, December 31, 2009	31,474	\$ 7.14
Granted*	10,603	1.63
Vested	(10,060)	7.14
Forfeited	—	—
Unvested balance, December 31, 2010	32,017	\$ 5.32
Granted*	145,404	0.83
Vested	(68,508)	1.61
Forfeited	—	—
Unvested balance, December 31, 2011	108,913	\$ 1.66
Granted**	30,047	1.00
Vested	(46,870)	1.93
Forfeited	—	—
Unvested balance, December 31, 2012	92,090	\$ 1.30
Granted**	30,763	1.56
Vested	(56,435)	1.51
Forfeited	(3,778)	0.83
Unvested balance, September 30, 2013	62,640	\$ 1.32

* Grants of restricted stock awards with corresponding promissory notes; grant date fair value is estimated using the Black-Scholes option pricing model.

** Grants of restricted stock awards; grant date fair value is based on the estimated value of the Company's Common Stock.

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Stock Award

In September 2013, an employee of the Company was granted stock awards, totaling 40,551 shares of Common Stock, at a fair value of \$6.60 per share. The fair value of the award is based on the estimated fair value of the Company's Common Stock at the date of grant. The shares immediately vested, and were not subject to any other restriction. Total compensation cost of \$267,637 was recognized in the nine-month period ended September 30, 2013.

Compensation Expense Summary

The Company has recognized the following compensation cost related to employee and non-employee based stock option activity:

<u>Year Ended</u>	<u>R&D</u>	<u>General and Administrative</u>	<u>Total</u>
2013 (9 Months)	\$ 63,684	\$ 374,368	\$ 438,052
2012	52,768	69,573	122,341
2011	175,418	132,011	307,429
2010	93,489	75,782	169,271
2009	57,234	47,503	104,737
2008	18,388	23,327	41,715
Total	<u>\$ 460,981</u>	<u>\$ 722,564</u>	<u>\$ 1,183,545</u>

13. Employee Retirement Plan

During 2008, the Company established a retirement plan authorized by Section 401(k) of the Internal Revenue Code. In accordance with the Plan, all employees who have attained the age of 21 are eligible to participate in the Plan as of the first Entry Date following their date of employment. Each employee can contribute a percentage of compensation up to a maximum of the statutory limits per year. Company contributions are discretionary, and no contributions were made during 2011, 2012 or 2013.

14. Employee Bonus Plan

During 2008, the Company established a non-calendar year bonus plan for certain employees of the Company based on the achievement of certain milestones. The total amount of eligible bonus is \$239,640 and \$203,057 at December 31, 2011 and 2012, respectively. The Company has accrued an estimate of the bonus of \$179,730 and \$152,293 at December 31, 2011 and 2012, respectively, and \$363,651 at September 30, 2013. These amounts are recorded as a component of accrued expenses in the accompanying balance sheets.

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15. Net Loss per Share

The following table reconciles net loss to net loss applicable to common stockholders:

	<u>Year Ended December 31,</u>		<u>Nine Months Ended</u>		<u>February 27,</u> <u>2007</u> <u>(Inception)</u> <u>through</u> <u>September 30,</u> <u>2013</u>
	<u>2011</u>	<u>2012</u>	<u>2012</u>	<u>2013</u>	
			<u>(unaudited)</u>		
Numerator:					
Net loss	\$ (15,296,685)	\$ (8,195,957)	\$ (5,792,960)	\$ (7,219,236)	\$ (57,093,973)
Accretion on preferred stock	(2,970,586)	(3,322,647)	(2,468,546)	(52,861,367)	(63,713,722)
Loss on extinguishment of preferred stock	—	—	—	—	(597,174)
Net loss applicable to common stockholders	<u>\$ (18,267,271)</u>	<u>\$ (11,518,604)</u>	<u>\$ (8,261,506)</u>	<u>\$ (60,080,603)</u>	<u>\$ (121,404,869)</u>
Denominator:					
Weighted-average common shares—basic and diluted	167,039	236,633	230,748	291,206	146,688
Net loss per share applicable to common stockholders— basic and diluted	<u>\$ (109.36)</u>	<u>\$ (48.68)</u>	<u>\$ (35.80)</u>	<u>\$ (206.32)</u>	<u>\$ (827.64)</u>

The amounts in the table below were excluded from the calculation of diluted net loss per share, prior to the use of the treasury stock method, due to their anti-dilutive effect (in thousands):

	<u>Year Ended December 31,</u>		<u>Nine Months Ended</u>		<u>February 27,</u> <u>2007</u> <u>(Inception)</u> <u>through</u> <u>September 30,</u> <u>2013</u>
	<u>2011</u>	<u>2012</u>	<u>2012</u>	<u>2013</u>	
			<u>(unaudited)</u>		
Preferred stock	3,386,202	3,386,202	3,386,202	6,790,149	6,790,149
Outstanding stock options	239,302	279,753	279,753	702,625	702,625
Unvested restricted stock	108,913	92,090	103,808	62,640	62,640
Total	<u>3,734,417</u>	<u>3,758,045</u>	<u>3,769,763</u>	<u>7,555,414</u>	<u>7,555,414</u>

16. Subsequent Events

In December 2013, the Company entered into a three-year Office Lease Agreement (Lease) for 6,837 square feet of space in Cambridge, Massachusetts, commencing on the date that the premises are delivered to the Company with the landlord work substantially complete. The Company anticipates that the premises shall be available for use on or about December 26, 2013. The Lease has monthly lease payments of approximately \$31,000 for the first twelve months, with annual rent escalation thereafter, and provides a rent abatement of approximately \$31,000 for the first full calendar month of the lease term.



Through and including _____, 2014 (25 days after the commencement of this offering), all dealers that buy, sell or trade shares of our common stock, whether or not participating in this offering, may be required to deliver a prospectus. This delivery is in addition to a dealer's obligation to deliver a prospectus when acting as an underwriter and with respect to their unsold allotments or subscriptions.

Part II
Information Not Required in Prospectus

Item 13. Other Expenses of Issuance and Distribution

The following table sets forth the costs and expenses, other than the underwriting discounts and commissions, payable by the registrant in connection with the sale of common stock being registered. All amounts are estimates except for the SEC registration fee, the Financial Industry Regulatory Authority, or FINRA, filing fee and NASDAQ listing fee.

Item	Amount to be paid
SEC registration fee	\$
FINRA filing fee	650.00
NASDAQ listing fee	*
Printing and engraving expenses	*
Legal fees and expenses	*
Accounting fees and expenses	*
Transfer Agent fees and expenses	*
Miscellaneous expenses	*
Total	\$ *

* To be provided by amendment.

Item 14. Indemnification of Directors and Officers

Section 145 of the General Corporation Law of the State of Delaware provides as follows:

A corporation shall have the power to indemnify any person who was or is a party or is threatened to be made a party to any threatened, pending or completed action, suit or proceeding, whether civil, criminal, administrative or investigative (other than an action by or in the right of the corporation) by reason of the fact that the person is or was a director, officer, employee or agent of the corporation, or is or was serving at the request of the corporation as a director, officer, employee or agent of another corporation, partnership, joint venture, trust or other enterprise, against expenses (including attorneys' fees), judgments, fines and amounts paid in settlement actually and reasonably incurred by him in connection with such action, suit or proceeding if the person acted in good faith and in a manner the person reasonably believed to be in or not opposed to the best interest of the corporation, and, with respect to any criminal action or proceeding, had no reasonable cause to believe his conduct was unlawful. The termination of any action, suit or proceeding by judgment, order, settlement, conviction or upon a plea of nolo contendere or its equivalent shall not, of itself, create a presumption that the person did not act in good faith and in a manner which the person reasonably believed to be in or not opposed to the best interests of the corporation, and, with respect to any criminal action or proceeding, had reasonable cause to believe that his conduct was unlawful.

A corporation shall have the power to indemnify any person who was or is a party or is threatened to be made a party to any threatened, pending or completed action or suit by or in the right of the corporation to procure a judgment in its favor by reason of the fact that the person is or was a director, officer, employee or agent of the corporation, or is or was serving at the request of the corporation as a director, officer, employee or agent of another corporation, partnership, joint venture, trust or other enterprise against expenses (including attorneys' fees) actually and reasonably incurred by him in connection with the defense or settlement of such action or suit if the person acted in good faith and in a manner the person reasonably believed to be in or not opposed to the best interests of the corporation and except that no indemnification shall be made with respect to any claim, issue or matter as to which such person shall have been adjudged to be liable to the corporation unless

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and only to the extent that the Court of Chancery or the court in which such action or suit was brought shall determine upon application that, despite the adjudication of liability but in view of all the circumstances of the case, such person is fairly and reasonably entitled to indemnity for such expenses which the Court of Chancery or such other court shall deem proper.

As permitted by the Delaware General Corporation Law, we have included in our certificate of incorporation a provision to eliminate the personal liability of our directors for monetary damages for breach of their fiduciary duties as directors, subject to certain exceptions. In addition, our certificate of incorporation and bylaws provide that we are required to indemnify our officers and directors under certain circumstances, including those circumstances in which indemnification would otherwise be discretionary, and we are required to advance expenses to our officers and directors as incurred in connection with proceedings against them for which they may be indemnified.

We intend to enter into indemnification agreements with our directors and officers. These agreements will provide broader indemnity rights than those provided under the Delaware General Corporation Law and our certificate of incorporation. The indemnification agreements are not intended to deny or otherwise limit third-party or derivative suits against us or our directors or officers, but to the extent a director or officer were entitled to indemnity or contribution under the indemnification agreement, the financial burden of a third-party suit would be borne by us, and we would not benefit from derivative recoveries against the director or officer. Such recoveries would accrue to our benefit but would be offset by our obligations to the director or officer under the indemnification agreement.

The underwriting agreement provides that the underwriters are obligated, under certain circumstances, to indemnify our directors, officers and controlling persons against certain liabilities, including liabilities under the Securities Act. Reference is made to the form of underwriting agreement filed as Exhibit 1.1 hereto.

We maintain directors' and officers' liability insurance for the benefit of our directors and officers.

Item 15. Recent Sales of Unregistered Securities

In the three years preceding the filing of this registration statement, we have issued the following securities that were not registered under the Securities Act.

(a) Sales of Capital Stock

In June 2010, we issued and sold an aggregate of 125,000 shares of Series A preferred stock at a price per share of \$40.00 for total consideration of \$5 million to 15 investors.

In April 2011 and December 2011, we issued and sold an aggregate of 1,287,524 shares of Series B preferred stock at a price per share of \$14.00 for total consideration of \$18,025,341 to 23 investors.

In July 2012 and March 2013, we issued and sold an aggregate of 50,000 shares of our Series X preferred stock at a price per share of \$100.00 for total consideration of \$5,000,002 to 27 investors.

In May 2013, we issued an aggregate of 357,143 shares of Series C preferred stock at an exchange rate of 7.14286 shares of Series C preferred stock for every share of Series X preferred stock.

In May 2013, we issued and sold an aggregate of 2,945,742 shares of our Series C preferred stock at a price per share of \$14.00 for total consideration of \$41,240,388 to 37 investors.

No underwriters were involved in the foregoing sales and exchanges of securities. The securities issued for cash consideration described in this section (a) of Item 15 were issued to third parties in reliance upon, (i) with

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respect to the Series A preferred stock, Series X preferred stock and the 2,945,742 shares of Series C preferred stock, the exemption from the registration requirements of the Securities Act, as set forth in Section 4(2) under the Securities Act; and (ii) with respect to the shares of Series B preferred stock, pursuant to Regulation D promulgated under the Securities Act, in each case related to transactions by an issuer not involving any public offering. The 357,143 shares of Series C preferred stock issued in exchange for Series X preferred stock described in this section (a) of Item 15 were issued in reliance upon Section 3(a)(9) of the Securities Act. All purchasers of preferred stock described above represented to us in connection with their purchase that they were accredited investors or sophisticated investors with such knowledge and experience in financial matters as to be able to evaluate the merits and risks of an investment in such shares. The purchasers received written disclosures that the securities had not been registered under the Securities Act and that any resale must be made pursuant to a registration statement of an available exemption from such registration.

(b) Grants and Exercises of Stock Options

From January 1, 2010, through December 16, 2013, we granted options to purchase a total of 558,380 shares of our common stock to employees and non-employees, at a weighted-average price of \$0.98 per share. During the same period, we issued 4,647 shares of common stock upon the exercise of options to purchase such shares of common stock at a weighted-average price of \$1.50 per share.

Option grants and issuances of common stock upon exercise of such options were exempt pursuant to Rule 701 and Section 4(2) of the Securities Act. All recipients either received adequate information about us or had access, through employment or other relationships, to such information.

Item 16. Exhibits and financial statement schedules

(a) Exhibits

See the Exhibit Index attached to this Registration Statement, which is incorporated by reference herein.

(b) Financial statement schedules

Schedules not listed above have been omitted because the information required to be set forth therein is not applicable or is shown in the financial statements or notes thereto.

Item 17. Undertakings

The undersigned registrant hereby undertakes to provide to the underwriters at the closing specified in the underwriting agreement certificates in such denominations and registered in such names as required by the underwriters to permit prompt delivery to each purchaser.

Insofar as indemnification for liabilities arising under the Securities Act may be permitted to directors, officers and controlling persons of the registrant pursuant to the foregoing provisions, or otherwise, the registrant has been advised that in the opinion of the Securities and Exchange Commission such indemnification is against public policy as expressed in the Act and is, therefore, unenforceable. In the event that a claim for indemnification against such liabilities (other than the payment by the registrant of expenses incurred or paid by a director, officer or controlling person of the registrant in the successful defense of any action, suit or proceeding) is asserted by such director, officer or controlling person in connection with the securities being registered, the registrant will, unless in the opinion of its counsel the matter has been settled by controlling precedent, submit to a court of appropriate jurisdiction the question whether such indemnification by it is against public policy as expressed in the Act and will be governed by the final adjudication of such issue.

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The undersigned Registrant hereby undertakes:

(1) That for purposes of determining any liability under the Securities Act, the information omitted from the form of prospectus filed as part of this registration statement in reliance upon Rule 430A and contained in a form of prospectus filed by the Registrant pursuant to Rule 424(b)(1) or (4) or 497(h) under the Securities Act shall be deemed to be part of this registration statement as of the time it was declared effective.

(2) That for the purpose of determining any liability under the Securities Act, each post-effective amendment that contains a form of prospectus shall be deemed to be a new registration statement relating to the securities offered therein, and the offering of such securities at that time shall be deemed to be the initial bona fide offering thereof.

SIGNATURES

Pursuant to the requirements of the Securities Act of 1933, as amended, the Registrant has duly caused this Registration Statement on Form S-1 to be signed on its behalf by the undersigned, thereunto duly authorized, in the city of Cambridge, Commonwealth of Massachusetts, on _____, 2013.

AKEBIA THERAPEUTICS, INC.

By: _____
John P. Butler
Chief Executive Officer and President

Signatures and Power of Attorney

We, the undersigned directors and officers of Akebia Therapeutics, Inc. (the "Company"), hereby severally constitute and appoint John P. Butler and Jason A. Amello, and each of them singly, our true and lawful attorneys, with full power to them, and to each of them singly, to sign for us and in our names in the capacities indicated below, the registration statement on Form S-1 filed herewith, and any and all pre-effective and post-effective amendments to said registration statement, and any registration statement filed pursuant to Rule 462(b) under the Securities Act of 1933, as amended, in connection with the registration under the Securities Act of 1933, as amended, of equity securities of the Company, and to file or cause to be filed the same, with all exhibits thereto and other documents in connection therewith, with the Securities and Exchange Commission, granting unto said attorneys, and each of them, full power and authority to do and perform each and every act and thing requisite and necessary to be done in connection therewith, as fully to all intents and purposes as each of us might or could do in person, and hereby ratifying and confirming all that said attorneys, and each of them, or their substitute or substitutes, shall do or cause to be done by virtue of this Power of Attorney.

Pursuant to the requirements of the Securities Act of 1933, as amended, this Registration Statement on Form S-1 has been signed by the following persons in the capacities and on the dates indicated.

<u>Signature</u>	<u>Title</u>	<u>Date</u>
_____ John P. Butler	Chief Executive Officer and President (Principal Executive Officer)	_____, 2013
_____ Jason A. Amello	Senior Vice President, Chief Financial Officer and Treasurer	_____, 2013
_____ Muneer A. Satter	Director	_____, 2013
_____ Campbell Murray, M.D.	Director	_____, 2013
_____ Jack Nielsen	Director	_____, 2013
_____ Anupam Dalal, M.D.	Director	_____, 2013

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Signature	Title	Date
Giovanni Ferrara	Director	, 2013
Kim Dueholm, Ph.D.	Director	, 2013
Duane Nash, M.D.	Director	, 2013

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<u>Exhibit number</u>	<u>Description of Exhibit</u>
1.1*	Form of Underwriting Agreement
3.1*	Form of Amended and Restated Certificate of Incorporation (to be effective upon completion of this offering)
3.2*	Form of Amended and Restated Bylaws (to be effective upon completion of this offering)
3.3	Eighth Amended and Restated Certificate of Incorporation (currently in effect)
3.4	Bylaws of the Registrant, as amended (currently in effect)
4.1*	Form of Common Stock Certificate
4.2	Third Amended and Restated Voting Agreement, dated May 10, 2013
4.3	Amendment No. 1 to the Third Amended and Restated Voting Agreement, dated May 31, 2013
4.4	Third Amended and Restated Investors' Rights Agreement, dated May 10, 2013
4.5	Amendment No. 1 to the Third Amended and Restated Investors' Rights Agreement, dated May 31, 2013
4.6	Second Amended Right of First Refusal and Co-Sale Agreement, dated April 6, 2011
4.7	Amendment Number One to Second Amended and Restated Right of First Refusal and Co-Sale Agreement, dated July 9, 2012
4.8	Amendment Number Two to the Second Amended and Restated Right of First Refusal and Co-Sale Agreement, dated May 10, 2013
5.1*	Opinion of Ropes & Gray LLP
10.1*	Form of Director and Officer Indemnification Agreement
10.2	Office Lease Agreement Between MA-Riverview/245 First Street, L.L.C. and Akebia Therapeutics, Inc., dated December 3, 2013
10.3†	Amended and Restated 2008 Equity Incentive Plan
10.4†	Amendment No. 1 to Amended and Restated 2008 Equity Incentive Plan
10.5†	Executive Employment Agreement with John P. Butler, dated September 16, 2013
10.6†	Executive Employment Agreement with Jason A. Amello, dated September 23, 2013
10.7†	Offer Letter to Nicole R. Hadas, dated November 13, 2013
10.8†	Executive Employment Agreement with Dr. Robert Shalwitz, Dated April 6, 2011
10.9†	Offer Letter to William Daly, Dated January 7, 2012
10.10†	Executive Employment Agreement with Joseph H. Gardner, dated May 2, 2007
10.11†	Amendment No. 1 to Executive Employment Agreement, dated April 6, 2010
10.12†	Consulting Agreement with Joseph H. Gardner, dated September 15, 2013
10.13†	Separation Agreement with Joseph H. Gardner, dated September 15, 2013
10.14†	Amended and Restated Partial Recourse Promissory Note, dated May 9, 2013, with Joseph Gardner
10.15†	Amended and Restated Partial Recourse Promissory Note, dated June 15, 2013, with Joseph Gardner
10.16†	Amended and Restated Partial Recourse Promissory Note, dated May 9, 2013, with Robert Shalwitz
10.17†	Amended and Restated Partial Recourse Promissory Note, dated June 15, 2013, with Robert Shalwitz
23.1*	Consent of Ernst & Young LLP
23.2*	Consent of Ropes & Gray LLP (included in Exhibit 5.1)
24.1	Power of Attorney (included on signature page)

* To be filed by amendment.

† Indicates a management contract or compensatory plan.

**EIGHTH AMENDED AND RESTATED
CERTIFICATE OF INCORPORATION
OF
AKEBIA THERAPEUTICS, INC.**

(Pursuant to Sections 242 and 245 of the
General Corporation Law of the State of Delaware)

Akebia Therapeutics, Inc., a corporation organized and existing under and by virtue of the provisions of the General Corporation Law of the State of Delaware (the "General Corporation Law"),

DOES HEREBY CERTIFY:

1. That the name of this corporation is Akebia Therapeutics, Inc., and that this corporation was originally incorporated pursuant to the General Corporation Law on February 27, 2007. On September 4, 2007, January 23, 2008, July 16, 2009, June 4, 2010, April 6, 2011, July 9, 2012 and March 21, 2013, the corporation's certificate of incorporation was amended and restated in its entirety.

2. That the Board of Directors duly adopted resolutions proposing this eighth amendment and restatement of the Certificate of Incorporation of this corporation, declaring said amendment and restatement to be advisable and in the best interests of this corporation and its stockholders, and authorizing the appropriate officers of this corporation to solicit the consent of the stockholders therefor, which resolution setting forth the proposed amendment and restatement is as follows:

RESOLVED, that the Seventh Amended and Restated Certificate of Incorporation of this corporation be amended and restated in its entirety to read as follows:

FIRST: The name of this corporation is Akebia Therapeutics, Inc. (the "Corporation").

SECOND: The address of the registered office of the Corporation in the State of Delaware is Corporation Trust Center, 1209 Orange Street, in the City of Wilmington, County of New Castle, 19801. The name of its registered agent at such address is The Corporation Trust Company.

THIRD: The nature of the business or purposes to be conducted or promoted is to engage in any lawful act or activity for which corporations may be organized under the General Corporation Law.

FOURTH: Upon the filing and effectiveness (the "Effective Time") of this Eighth Amended and Restated Certificate of Incorporation pursuant to the General Corporation Law, (i) each 100 shares of Common Stock, either issued and outstanding or held by Akebia Therapeutics, Inc. in treasury stock immediately prior to the Effective Time shall, automatically and without any action on the part of the respective holders thereof, be combined and converted

into one (1) share of Common Stock and (ii) each 100 shares of each type of Preferred Stock, either issued and outstanding or held by Akebia Therapeutics, Inc. in treasury stock immediately prior to the Effective Time shall, automatically and without any action on the part of the respective holders thereof, be combined and converted into one (1) share of the applicable type Preferred Stock (collectively, the “Reverse Stock Split”). Fractional shares shall be issued in connection with the Reverse Stock Split. Each certificate that immediately prior to the Effective Time represented shares of Common Stock or Preferred Stock (“Old Certificates”), shall thereafter be deemed to represent that number of shares of Common Stock or Preferred Stock into which the shares of Common Stock or Preferred Stock represented by the Old Certificate shall have been combined.

The total number of shares of all classes of stock which the Corporation shall have authority to issue is (i) 8,400,000 shares of Common Stock, \$0.00001 par value per share (“Common Stock”), and (ii) 5,500,636 shares of Preferred Stock, \$0.00001 par value per share (“Preferred Stock”).

The following is a statement of the designations and the powers, privileges and rights, and the qualifications, limitations or restrictions thereof in respect of each class of capital stock of the Corporation.

COMMON STOCK

1. General. The voting, dividend and liquidation rights of the holders of the Common Stock are subject to and qualified by the rights, powers and preferences of the holders of the Preferred Stock set forth herein.

2. Voting. The holders of the Common Stock are entitled to one vote for each share of Common Stock held at all meetings of stockholders (and written actions in lieu of meetings); provided, however, that, except as otherwise required by law, holders of Common Stock, as such, shall not be entitled to vote on any amendment to this Certificate of Incorporation that relates solely to the terms of one or more outstanding series of Preferred Stock if the holders of such affected series are entitled, either separately or together with the holders of one or more other such series, to vote thereon pursuant to this Certificate of Incorporation or pursuant to the General Corporation Law. There shall be no cumulative voting. The number of authorized shares of Common Stock may be increased or decreased (but not below the number of shares thereof then outstanding) by (in addition to any vote of the holders of one or more series of Preferred Stock that may be required by the terms of this Certificate of Incorporation) the affirmative vote of the holders of shares of capital stock of the Corporation representing a majority of the votes represented by all outstanding shares of capital stock of the Corporation entitled to vote, irrespective of the provisions of Section 242(b)(2) of the General Corporation Law.

PREFERRED STOCK

734,538 shares of the authorized Preferred Stock of the Corporation are hereby designated “Series A Preferred Stock” with the following rights, preferences, powers, privileges, restrictions, qualifications and limitations. 1,287,525 shares of the authorized Preferred Stock of

the Corporation are hereby designated "Series B Preferred Stock" with the following rights, preferences, powers, privileges, restrictions, qualifications and limitations. 3,428,572 shares of the authorized Preferred Stock of the Corporation are hereby designated "Series C Preferred Stock" with the following rights, preferences, powers, privileges, restrictions, qualifications and limitations. The Series A Preferred Stock, Series B Preferred Stock and Series C Preferred Stock are collectively referred to as "Convertible Preferred Stock." 50,001 shares of the authorized Preferred Stock of the Corporation are hereby designated "Series X Preferred Stock" with the following rights, preferences, powers, privileges, restrictions, qualifications and limitations. Unless otherwise indicated, references to "Sections" or "Subsections" in this Article Fourth refer to sections and subsections of this Article Fourth.

1. Dividends.

1.1. Preferred Stock Dividends. The Series X Preferred Stock shall not receive any dividends. From and after the date of the issuance of any shares of Convertible Preferred Stock, dividends at the rate per annum of six percent (6%) (with respect to Series A Preferred Stock and Series B Preferred Stock) and eight percent (8%) (with respect to Series C Preferred Stock) on the Applicable Accrued Value (as defined below) shall accrue on such shares of Convertible Preferred Stock (subject to appropriate adjustment in the event of any stock dividend, stock split, combination or other similar recapitalization with respect to the Convertible Preferred Stock) (the "Accruing Dividends"). For the avoidance of doubt and for purposes of this Section 1.1 of this Article Fourth, Accruing Dividends shall accrue on each share of Convertible Preferred Stock from and after the date of such share's original issuance. Accruing Dividends shall accrue from day to day (and compound quarterly), whether or not declared, and shall be cumulative; provided, however, that except as otherwise set forth in Sections 1, 2, 4 and 6, such Accruing Dividends shall be payable only if permitted by the General Corporation Law and when, as, and if declared by the Board of Directors and the Corporation shall be under no obligation to pay such Accruing Dividends. The Corporation shall not declare, pay or set aside any dividends on shares of any other class or series of capital stock of the Corporation (other than dividends on shares of Common Stock payable in shares of Common Stock) unless (in addition to the obtaining of any consents required elsewhere in the Certificate of Incorporation) the holders of the Convertible Preferred Stock then outstanding shall first receive, or simultaneously receive, a dividend on each outstanding share of Convertible Preferred Stock in an amount at least equal to the sum of (i) the amount of the aggregate Accruing Dividends then accrued on such share of Convertible Preferred Stock and not previously paid and (ii) (A) in the case of a dividend on Common Stock or any class or series that is convertible into Common Stock, that dividend per share of Convertible Preferred Stock as would equal the product of (1) the dividend payable on each share of such class or series determined, if applicable, as if all shares of such class or series had been converted into Common Stock and (2) the number of shares of Common Stock issuable upon conversion of a share of Convertible Preferred Stock, in each case calculated on the record date for determination of holders entitled to receive such dividend or (B) in the case of a dividend on any class or series that is not convertible into Common Stock, at a rate per share of Convertible Preferred Stock determined by (1) dividing the amount of the dividend payable on each share of such class or series of capital stock by the original issuance price of such class or series of capital stock (subject to appropriate adjustment in the event of any stock dividend, stock split, combination or other similar recapitalization with respect to such class or series) and (2) multiplying such fraction by an amount equal to either the

Series A Original Issue Price (as defined below), Series B Original Issue Price (as defined below) or Series C Original Issue Price (as defined below), as applicable; provided, that if the Corporation declares, pays or sets aside, on the same date, a dividend on shares of more than one class or series of capital stock of the Corporation, the dividend payable to the holders of Series B Preferred Stock and Series C Preferred Stock pursuant to this Section 1 shall be calculated based upon the dividend on the class or series of capital stock that would result in the highest Series B Preferred Stock dividend or Series C Preferred Stock dividend, as applicable. The “Applicable Accrued Value” shall mean, (a) with respect to each share of Series X Preferred Stock, \$100.00 (as adjusted for any stock split, combination or other similar recapitalization with respect to the Series X Preferred Stock after May 10, 2013 (the “Eighth Amendment Date”)), (b) with respect to each share of Series C Preferred Stock, the sum (as adjusted for any stock dividend, stock split, combination or other similar recapitalization with respect to the Series C Preferred Stock after the Eighth Amendment Date) of (i) the Applicable Original Purchase Price plus (ii) an amount equal to any dividends on such share of Series C Preferred Stock which have been accrued that have not been paid; provided that, for purposes of Sections 2 and 6, “Applicable Accrued Value” with respect to each share of Series C Preferred Stock shall mean the sum (as adjusted for any stock dividend, stock split, combination or other similar recapitalization with respect to the Series C Preferred Stock after the Eighth Amendment Date) of (x) \$28.00 plus (y) an amount equal to 200% of any dividends on such share of Series C Preferred Stock which have been accrued that have not been paid and (c) with respect to each share of Series A Preferred Stock and Series B Preferred Stock, the sum (as adjusted for any stock dividend, stock split, combination or other similar recapitalization with respect to the Series A Preferred Stock or Series B Preferred Stock after the Eighth Amendment Date) of (i) the Applicable Original Purchase Price plus (ii) an amount equal to any dividends on such share of Series A Preferred Stock or Series B Preferred Stock which have been accrued that have not been paid. The “Applicable Original Purchase Price” shall mean, with respect to each share of Series B Preferred Stock and Series C Preferred Stock, \$14.00 per share, subject to appropriate adjustment in the event of any stock dividend, stock split, combination or other similar recapitalization with respect to the Series B Preferred Stock or Series C Preferred Stock (as applicable) after the Eighth Amendment Date, and with respect to each share of Series A Preferred Stock, \$40.00 per share, subject to appropriate adjustment in the event of any stock dividend, stock split, combination or other similar recapitalization with respect to the Series A Preferred Stock after the Eighth Amendment Date.

1.2. Less than Total Preferred Dividend. Except as otherwise provided herein and subject to obtaining any consents required in this Certificate of Incorporation, if at any time the Corporation pays a dividend on the Convertible Preferred Stock that is less than the total amount of dividends then accrued with respect to the Convertible Preferred Stock, such payment of the Accruing Dividends shall be distributed first to the holders of the Series C Preferred Stock. If such dividend is less than the total amount of dividends then accrued with respect to the Series C Preferred Stock, such payment shall be distributed ratably among the holders of Series C Preferred Stock based upon the aggregate accrued but unpaid dividends on the Series C Preferred Stock held by each holder. Any remainder, after distribution to holders of Series C Preferred Stock of an aggregate amount equal to all accrued dividends thereon, shall be distributed next to the holders of the Series B Preferred Stock. If such remainder is less than the total amount of dividends then accrued with respect to the Series B Preferred Stock, such payment shall be distributed ratably among the holders of Series B Preferred Stock based upon the aggregate accrued but unpaid dividends on the Series B Preferred Stock held by each holder.

Any remainder, after distribution to holders of Series B Preferred Stock of an aggregate amount equal to all accrued dividends thereon, shall be distributed ratably among the holders of Series A Preferred Stock based upon the aggregate accrued but unpaid dividends on the Series A Preferred Stock held by each holder.

2. Liquidation, Dissolution or Winding Up; Certain Mergers, Consolidations and Asset Sales.

2.1. Preferential Payments to Holders of Preferred Stock. In the event of any voluntary or involuntary liquidation, dissolution or winding up of the Corporation, the holders of shares of Preferred Stock then outstanding shall be entitled to be paid out of the assets of the Corporation available for distribution to its stockholders before any payment shall be made to the holders of Common Stock by reason of their ownership thereof, (a) for shares of Series X Preferred Stock, an amount per share equal to the greater of (i) the Applicable Accrued Value, or (ii) the amount a share of Series X Preferred Stock would have received if the Series X Preferred Stock were converted into shares of Series B Preferred Stock and (b) for shares of Convertible Preferred Stock, an amount per share equal to the Applicable Accrued Value. If upon any such liquidation, dissolution or winding up of the Corporation, the assets of the Corporation available for distribution to its stockholders shall be insufficient to pay the holders of shares of Preferred Stock the full amount to which they shall be entitled under this Subsection 2.1, the holders of shares of Series X Preferred Stock shall share ratably in any distribution of the assets available for distribution in proportion to the Applicable Accrued Value of the shares of Series X Preferred Stock held by such holders. Any remainder, after distribution and setting aside the Applicable Accrued Value of the Series X Preferred Stock held by each holder, shall be shared ratably among the holders of shares of Series C Preferred Stock in any distribution of the assets available for distribution in proportion to the Applicable Accrued Value of the Series C Preferred Stock held by each holder. Any remainder, after distribution and setting aside the Applicable Accrued Value for payment to the holders of Series X Preferred Stock and Series C Preferred Stock, shall be distributed ratably among the holders of Series B Preferred Stock based upon the Applicable Accrued Value of the Series B Preferred Stock held by each holder. Any remainder, after distribution and setting aside the Applicable Accrued Value for payment to the holders of Series X Preferred Stock, Series C Preferred Stock and Series B Preferred Stock, shall be distributed ratably among the holders of Series A Preferred Stock based upon the Applicable Accrued Value of the Series A Preferred Stock held by each holder.

2.2. Distribution of Remaining Assets. In the event of any voluntary or involuntary liquidation, dissolution or winding up of the Corporation, after the payment of all preferential amounts required to be paid to the holders of shares of Preferred Stock under Subsection 2.1 the remaining assets of the Corporation available for distribution and setting aside for payment to its stockholders shall be distributed among the holders of the shares of Convertible Preferred Stock and Common Stock, pro rata based on the number of shares held by each such holder, treating for this purpose all such securities as if they had been converted to Common Stock pursuant to the terms of this Certificate of Incorporation immediately prior to such dissolution, liquidation or winding up of the Corporation. The aggregate amount which a holder of a share of Preferred Stock is entitled to receive under Subsections 2.1 and 2.2 is hereinafter referred to as the "Applicable Liquidation Amount."

2.3. Deemed Liquidation Events.

2.3.1. Definition. Each of the following events shall be considered a “Deemed Liquidation Event” unless the holders of more than fifty percent (50%) (the “Appropriate Percentage”) of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Convertible Preferred Stock elect otherwise by written notice sent to the Corporation at least five (5) days prior to the effective date of any such event:

(a) a merger or consolidation in which

- (i) the Corporation is a constituent party or
- (ii) a subsidiary of the Corporation is a constituent party and the Corporation issues shares of its capital stock pursuant to such merger or consolidation,

except any such merger or consolidation involving the Corporation or a subsidiary in which the shares of capital stock of the Corporation outstanding immediately prior to such merger or consolidation continue to represent, or are converted into or exchanged for shares of capital stock that represent, immediately following such merger or consolidation, at least a majority, by voting power, of the capital stock of (1) the surviving or resulting corporation or (2) if the surviving or resulting corporation is a wholly owned subsidiary of another corporation immediately following such merger or consolidation, the parent corporation of such surviving or resulting corporation (provided, that for the purpose of this Subsection 2.3.1, all shares of Common Stock issuable upon exercise of Options (as defined below) outstanding immediately prior to such merger or consolidation or upon conversion of Convertible Securities (as defined below) outstanding immediately prior to such merger or consolidation shall be deemed to be outstanding immediately prior to such merger or consolidation and, if applicable, converted or exchanged in such merger or consolidation on the same terms as the actual outstanding shares of Common Stock are converted or exchanged); or

(b) the sale, lease, transfer, exclusive license or other disposition, in a single transaction or series of related transactions, by the Corporation or any subsidiary of the Corporation of all or substantially all the assets of the Corporation and its subsidiaries taken as a whole, or the sale or disposition (whether by merger or otherwise) of one or more subsidiaries of the Corporation if substantially all of the assets of the Corporation and its subsidiaries taken as a whole are held by such subsidiary or subsidiaries, except where such sale, lease, transfer, exclusive license or other disposition is to a wholly owned subsidiary of the Corporation.

2.3.2. Effecting a Deemed Liquidation Event.

(a) The Corporation shall not have the power to effect a Deemed Liquidation Event referred to in Subsection 2.3.1(a)(i) unless the agreement or plan of merger or consolidation for such transaction (the “Merger Agreement”) provides that the consideration payable to the stockholders of the Corporation shall be allocated among the holders of capital stock of the Corporation in accordance with Subsections 2.1 and 2.2.

(b) In the event of a Deemed Liquidation Event referred to in Subsection 2.3.1(a)(ii) or 2.3.1(b), if the Corporation does not effect a dissolution of the Corporation under the General Corporation Law within 90 days after such Deemed Liquidation Event, then (i) the Corporation shall send a written notice to each holder of Convertible Preferred Stock no later than the 90th day after the Deemed Liquidation Event advising such holders of their right (and the requirements to be met to secure such right) pursuant to the terms of the following clause (ii) to require the redemption of such shares of Convertible Preferred Stock, and (ii) if the holders of the Appropriate Percentage of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Convertible Preferred Stock so request in a written instrument delivered to the Corporation not later than 120 days after such Deemed Liquidation Event, the Corporation shall use the consideration received by the Corporation for such Deemed Liquidation Event (net of any retained liabilities associated with the assets sold or technology licensed, as determined in good faith by the Board of Directors of the Corporation), together with any other assets of the Corporation available for distribution to its stockholders (the "Available Proceeds"), to the extent legally available therefor, on the 150th day after such Deemed Liquidation Event, to redeem all outstanding shares of Preferred Stock at a price per share equal to the Applicable Liquidation Amount. Notwithstanding the foregoing, in the event of a redemption pursuant to the preceding sentence, if the Available Proceeds are not sufficient to redeem all outstanding shares of Preferred Stock, the Corporation shall redeem a pro rata portion of each holder's shares of Series X Preferred Stock to the fullest extent of such Available Proceeds, based on the respective amounts which would otherwise be payable in respect of the shares to be redeemed if the Available Proceeds were sufficient to redeem all such shares, and shall redeem the remaining shares to have been redeemed as soon as practicable after the Corporation has funds legally available therefor. For any remaining Available Proceeds, after redemption of Series X Preferred Stock, the Corporation shall redeem a pro rata portion of each holder's shares of Series C Preferred Stock to the fullest extent of such Available Proceeds, based on the respective amounts which would otherwise be payable in respect of the shares to be redeemed if the Available Proceeds were sufficient to redeem all such shares, and shall redeem the remaining shares to have been redeemed as soon as practicable after the Corporation has funds legally available therefor. For any remaining Available Proceeds, after redemption of Series X Preferred Stock and Series C Preferred Stock, the Corporation shall redeem a pro rata portion of each holder's shares of Series B Preferred Stock to the fullest extent of such Available Proceeds, based on the respective amounts which would otherwise be payable in respect of the shares to be redeemed if the Available Proceeds were sufficient to redeem all such shares, and shall redeem the remaining shares to have been redeemed as soon as practicable after the Corporation has funds legally available therefor. For any remaining Available Proceeds, after redemption of Series X Preferred Stock, Series C Preferred Stock and Series B Preferred Stock, the Corporation shall redeem a pro rata portion of each holder's shares of Series A Preferred Stock to the fullest extent of such Available Proceeds, based on the respective amounts which would otherwise be payable in respect of the shares to be redeemed if the Available Proceeds were sufficient to redeem all such shares, and shall redeem the remaining shares to have been redeemed as soon as practicable after the Corporation has funds legally available therefor. The provisions of Subsections 6.2 through 6.4 shall apply, with such necessary changes in the details thereof as are necessitated by the context, to the redemption of the Preferred Stock pursuant to

this Subsection 2.3.2(b). Prior to the distribution or redemption provided for in this Subsection 2.3.2(b), the Corporation shall not expend or dissipate the consideration received for such Deemed Liquidation Event, except to discharge expenses incurred in connection with such Deemed Liquidation Event.

2.3.3. Amount Deemed Paid or Distributed. The amount deemed paid or distributed to the holders of capital stock of the Corporation upon any such merger, consolidation, sale, transfer, exclusive license, other disposition or redemption shall be the cash or the value of the property, rights or securities paid or distributed to such holders by the Corporation or the acquiring person, firm or other entity. The value of such property, rights or securities shall be determined in good faith by the Board of Directors of the Corporation.

2.3.4. Allocation of Escrow. In the event of a Deemed Liquidation Event pursuant to Subsection 2.3.1(a)(i), if any portion of the consideration payable to the stockholders of the Corporation is placed into escrow and/or is payable to the stockholders of the Corporation subject to contingencies, the Merger Agreement shall provide that (a) the portion of such consideration that is not placed in escrow and not subject to any contingencies (the "Initial Consideration") shall be allocated among the holders of capital stock of the Corporation in accordance with Subsections 2.1 and 2.2 as if the Initial Consideration were the only consideration payable in connection with such Deemed Liquidation Event and (b) any additional consideration which becomes payable to the stockholders of the Corporation upon release from escrow or satisfaction of contingencies shall be allocated among the holders of capital stock of the Corporation in accordance with Subsections 2.1 and 2.2 after taking into account the previous payment of the Initial Consideration as part of the same transaction.

3. Voting.

3.1. General.

3.1.1. Series X Preferred Stock Voting Rights. Any amendment, modification or waiver of this Certificate of Incorporation in a manner that adversely affects the powers, preferences or rights of the Series X Preferred Stock shall require the approval of stockholders holding at least sixty percent (60%) of the outstanding Series X Preferred Stock. On all other matters, except as required by the General Corporation Law, the Series X Preferred Stock shall not have any voting rights.

3.1.2. Convertible Preferred Stock Voting Rights. Any (i) declaration or payment of any accrued and unpaid dividends on the Series C Preferred Stock (but excluding, for the avoidance of doubt, the conversion of accrued and unpaid dividends on the Series C Preferred Stock into additional shares of Common Stock pursuant to Subsection 4.1), (ii) amendment, modification or waiver of this Certificate of Incorporation in a manner that adversely affects the powers, preferences or rights of the Series C Preferred Stock in a manner different from other series of the Corporation's Preferred Stock or (iii) increase or decrease in the number of authorized shares of Series C Preferred Stock shall require the approval of stockholders holding at least seventy-five percent (75%) of the then outstanding shares of Series C Preferred Stock. Any (i) amendment, modification or waiver of this Certificate of Incorporation in a manner that adversely affects the powers, preferences or rights of the Series B

Preferred Stock in a manner different from other series of the Corporation's Preferred Stock or (ii) increase of decrease in the number of authorized shares of Series B Preferred Stock shall require the approval of stockholders holding at least sixty-five percent (65%) of the then outstanding shares of Series B Preferred Stock. Any (i) amendment, modification or waiver of this Certificate of Incorporation in a manner that adversely affects the powers, preferences or rights of the Series A Preferred Stock in a manner different from other series of the Corporation's Preferred Stock or (ii) increase of decrease in the number of authorized shares of Series A Preferred Stock shall require the approval of stockholders holding at least sixty-five percent (65%) of the then outstanding shares of Series A Preferred Stock. On any matter presented to the stockholders of the Corporation for their action or consideration at any meeting of stockholders of the Corporation (or by written consent of stockholders in lieu of meeting), each holder of outstanding shares of Convertible Preferred Stock shall be entitled to cast the number of votes equal to the number of whole shares of Common Stock into which the shares of Convertible Preferred Stock held by such holder are then convertible as of the record date for determining stockholders entitled to vote on such matter. Except as provided by law or by the other provisions of this Certificate of Incorporation, holders of Convertible Preferred Stock shall vote together with the holders of Common Stock as a single class.

3.2. Election of Directors. The holders of record of the shares of Convertible Preferred Stock, exclusively and as a separate class, shall be entitled to elect six (6) directors of the Corporation (which shall be subject to reduction to not less than four (4) directors of the Corporation under certain circumstances as provided in Section 1 of the Voting Agreement) (the "Preferred Directors") and the holders of shares of Common Stock (including, on an as-converted to Common Stock basis, all shares of Convertible Preferred Stock convertible into Common Stock) shall be entitled to elect three (3) directors of the Corporation (which shall be subject to reduction to two (2) directors of the Corporation under certain circumstances as provided in Section 1 of the Voting Agreement). Any director elected as provided in the preceding sentence may be removed without cause by, and only by, the affirmative vote of the holders of the shares of the class or series of capital stock entitled to elect such director or directors, given either at a special meeting of such stockholders duly called for that purpose or pursuant to a written consent of stockholders. If the holders of shares of Convertible Preferred Stock or Common Stock, as the case may be, fail to elect a sufficient number of directors to fill all directorships for which they are entitled to elect directors, voting exclusively and as a separate class, pursuant to the first sentence of this Subsection 3.2, then any directorship not so filled shall remain vacant until such time as the holders of the Convertible Preferred Stock or Common Stock, as the case may be, elect a person to fill such directorship by vote or written consent in lieu of a meeting; and no such directorship may be filled by stockholders of the Corporation other than by the stockholders of the Corporation that are entitled to elect a person to fill such directorship, voting exclusively and as a separate class. The holders of record of the shares of Common Stock and of any other class or series of voting stock (including the Convertible Preferred Stock), exclusively and voting together as a single class, shall be entitled to elect the balance of the total number of directors of the Corporation. At any meeting held for the purpose of electing a director, the presence in person or by proxy of the holders of a majority of the outstanding shares of the class or series entitled to elect such director shall constitute a quorum for the purpose of electing such director. Except as otherwise provided in this Subsection 3.2, a vacancy in any directorship filled by the holders of any class or series shall be filled only by vote or written consent in lieu of a meeting of the holders of such class or series or by any remaining

director or directors elected by the holders of such class or series pursuant to this Subsection 3.2. For purposes hereof, “Voting Agreement” means that certain Third Amended and Restated Voting Agreement, dated on or about the Eighth Amendment Date, by and among the Corporation and certain of the Corporation’s stockholders party thereto (as the same may be further amended, restated, modified, supplemented or waived from time to time).

3.3. Convertible Preferred Stock Protective Provisions. At any time when shares of Convertible Preferred Stock are outstanding, the Corporation shall not, either directly or indirectly by amendment, merger, consolidation or otherwise, do any of the following without (in addition to any other vote or consent required by law or this Certificate of Incorporation) the written consent or affirmative vote of the holders of more than the Appropriate Percentage of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Convertible Preferred Stock, given in writing or by vote at a meeting, consenting or voting (as the case may be) separately as a class:

(a) liquidate, dissolve or wind-up the business and affairs of the Corporation, effect any Deemed Liquidation Event, or consent to any of the foregoing;

(b) amend, alter or repeal any provision of this Certificate of Incorporation or the Bylaws of the Corporation in a manner that adversely affects the powers, preferences or rights of the Series C Preferred Stock, Series B Preferred Stock or Series A Preferred Stock;

(c) create, or authorize the creation of, or issue or obligate itself to issue shares of, any additional class or series of capital stock, or increase the authorized number of shares of Series C Preferred Stock, Series B Preferred Stock or Series A Preferred Stock or increase the authorized number of shares of any additional class or series of capital stock;

(d) (i) reclassify, alter or amend any existing security of the Corporation that is pari passu with the Series C Preferred Stock, Series B Preferred Stock or Series A Preferred Stock in respect of the distribution of assets on the liquidation, dissolution or winding up of the Corporation, the payment of dividends or rights of redemption, if such reclassification, alteration or amendment would render such other security senior to the Series C Preferred Stock, Series B Preferred Stock or Series A Preferred Stock in respect of any such right, preference or privilege, (ii) reclassify, alter or amend any existing security of the Corporation that is junior to the Series C Preferred Stock in respect of the distribution of assets on the liquidation, dissolution or winding up of the Corporation, the payment of dividends or rights of redemption, if such reclassification, alteration or amendment would render such other security senior to or pari passu with the Series C Preferred Stock in respect of any such right, preference or privilege, (iii) reclassify, alter or amend any existing security of the Corporation that is junior to the Series B Preferred Stock in respect of the distribution of assets on the liquidation, dissolution or winding up of the Corporation, the payment of dividends or rights of redemption, if such reclassification, alteration or amendment would render such other security senior to or pari passu with the Series B Preferred Stock in respect of any such right, preference or privilege or (iv) reclassify, alter or amend any existing security of the Corporation that is junior to the Series A Preferred Stock in respect of the distribution of assets on the liquidation,

dissolution or winding up of the Corporation, the payment of dividends or rights of redemption, if such reclassification, alteration or amendment would render such other security senior to or pari passu with the Series A Preferred Stock in respect of any such right, preference or privilege;

(e) purchase or redeem (or permit any subsidiary to purchase or redeem) or pay or declare any dividend or make any distribution on, any shares of capital stock of the Corporation other than (i) redemptions of or dividends or distributions on the Preferred Stock as expressly authorized herein, (ii) dividends or other distributions payable on the Common Stock solely in the form of additional shares of Common Stock and, for the avoidance of doubt, the conversion of accrued and unpaid dividends on the Series C Preferred Stock into additional shares of Common Stock pursuant to Subsection 4.1 or (iii) repurchases of stock from former employees, officers, directors, consultants or other persons who performed services for the Corporation or any subsidiary in connection with the cessation of such employment or service that are approved by the Board of Directors;

(f) create, or authorize the creation of, or issue, or authorize the issuance of any debt security or other funded indebtedness, or permit any subsidiary to take any such action with respect to any debt security or other funded indebtedness;

(g) create, or hold capital stock in, any subsidiary that is not wholly owned (either directly or through one or more other subsidiaries) by the Corporation, or sell, transfer or otherwise dispose of any capital stock of any direct or indirect subsidiary of the Corporation, or permit any direct or indirect subsidiary to sell, lease, transfer, exclusively license or otherwise dispose (in a single transaction or series of related transactions) of all or substantially all of the assets of such subsidiary;

(h) increase or decrease the authorized number of directors constituting the Board of Directors; or

(i) enter into any material transaction with any Affiliate (as defined below) of the Corporation.

In addition to the above provisions of this Section 3.3, at any time when shares of Convertible Preferred Stock are outstanding, the Corporation shall not, either directly or indirectly by amendment, merger, consolidation or otherwise, without (in addition to any other vote or consent required by law or this Certificate of Incorporation) the written consent or affirmative vote of the holders of at least seventy percent (70%) of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Convertible Preferred Stock, given in writing or by vote at a meeting, consenting or voting (as the case may be) separately as a class, authorize, issue or enter into any agreement providing for the issuance (contingent or otherwise) of securities (including any notes or debt securities convertible into or exchangeable for other securities or the reclassification, alteration or amendment of any existing security of the Corporation) senior in any respect to the Series C Preferred Stock.

For purposes of this Section 3.3, "Affiliate" shall mean any person or entity controlling, controlled by or under common control with the Corporation, including, without limitation, any officer, director or stockholder owning greater than five percent (5%) of the Corporation

(assuming conversion of all Convertible Preferred Stock and Common Stock) or any member of the immediate family of such person or any entity controlling, controlled by or under common control with such person or entity.

4. Optional Conversion. The holders of the Convertible Preferred Stock shall have conversion rights as follows (the "Conversion Rights"):

4.1. Right to Convert.

4.1.1. Conversion Ratio. Each share of Convertible Preferred Stock shall be convertible, at the option of the holder thereof, at any time and from time to time, and without the payment of additional consideration by the holder thereof, into such number of fully paid and nonassessable shares of Common Stock as is determined by dividing (a) in the case of Series A Preferred Stock and Series B Preferred Stock, the Applicable Original Purchase Price by the Applicable Conversion Price (as defined below) in effect at the time of conversion and (b) in the case of Series C Preferred Stock, the Applicable Accrued Value of such Series C Preferred Stock as of the time of conversion by the Applicable Conversion Price (as defined below) in effect at the time of conversion. The "Applicable Conversion Price" shall mean \$14.00 for Series A Preferred Stock, Series B Preferred Stock and Series C Preferred Stock, in each case as of the Eighth Amendment Date. Such initial Applicable Conversion Price, and the rate at which shares of Convertible Preferred Stock may be converted into shares of Common Stock, shall be subject to adjustment as provided below.

4.1.2. Termination of Conversion Rights. In the event of a notice of redemption of any shares of Convertible Preferred Stock pursuant to Section 6, the Conversion Rights of the shares designated for redemption shall terminate at the close of business on the last full day preceding the date fixed for redemption, unless the redemption price is not fully paid on such redemption date, in which case the Conversion Rights for such shares shall continue until such price is paid in full. In the event of a liquidation, dissolution or winding up of the Corporation or a Deemed Liquidation Event, the Conversion Rights shall terminate at the close of business on the last full day preceding the date fixed for the payment of any such amounts distributable on such event to the holders of Convertible Preferred Stock.

4.2. Fractional Shares. No fractional shares of Common Stock shall be issued upon conversion of Convertible Preferred Stock unless otherwise approved by the Board of Directors. In lieu of any fractional shares to which the holder would otherwise be entitled, the Corporation shall pay cash equal to such fraction multiplied by the fair market value of a share of Common Stock as determined in good faith by the Board of Directors of the Corporation. Whether or not fractional shares would be issuable upon such conversion shall be determined on the basis of the total number of shares of Convertible Preferred Stock the holder is at the time converting into Common Stock and the aggregate number of shares of Common Stock issuable upon such conversion.

4.3. Mechanics of Conversion.

4.3.1. Notice of Conversion. In order for a holder of Convertible Preferred Stock to voluntarily convert shares of Convertible Preferred Stock into shares

of

Common Stock, such holder shall surrender the certificate or certificates for such shares of Convertible Preferred Stock (or, if such registered holder alleges that such certificate has been lost, stolen or destroyed, a lost certificate affidavit and agreement reasonably acceptable to the Corporation to indemnify the Corporation against any claim that may be made against the Corporation on account of the alleged loss, theft or destruction of such certificate), at the office of the transfer agent for Convertible Preferred Stock (or at the principal office of the Corporation if the Corporation serves as its own transfer agent), together with written notice that such holder elects to convert all or any number of the shares of Convertible Preferred Stock represented by such certificate or certificates and, if applicable, any event on which such conversion is contingent. Such notice shall state such holder's name or the names of the nominees in which such holder wishes the certificate or certificates for shares of Common Stock to be issued. If required by the Corporation, certificates surrendered for conversion shall be endorsed or accompanied by a written instrument or instruments of transfer, in form satisfactory to the Corporation, duly executed by the registered holder or his, her or its attorney duly authorized in writing. The close of business on the date of receipt by the transfer agent (or by the Corporation if the Corporation serves as its own transfer agent) of such certificates (or lost certificate affidavit and agreement) and notice shall be the time of conversion (the "Conversion Time"), and the shares of Common Stock issuable upon conversion of the shares represented by such certificate shall be deemed to be outstanding of record as of such date. The Corporation shall, as soon as practicable after the Conversion Time, (i) issue and deliver to such holder of Convertible Preferred Stock, or to his, her or its nominees, a certificate or certificates for the number of full shares of Common Stock issuable upon such conversion in accordance with the provisions hereof and a certificate for the number (if any) of the shares of Convertible Preferred Stock represented by the surrendered certificate that were not converted into Common Stock, (ii) pay in cash such amount as provided in Subsection 4.2 in lieu of any fraction of a share of Common Stock otherwise issuable upon such conversion and (iii) subject to those approval requirements set forth in Subsection 3.3(e), pay all Accruing Dividends accrued but unpaid on the shares of Series A Preferred Stock or Series B Preferred Stock converted, whether or not declared, together with any other dividends declared but unpaid thereon.

4.3.2. Reservation of Shares. The Corporation shall at all times when the Convertible Preferred Stock shall be outstanding, reserve and keep available out of its authorized but unissued capital stock, for the purpose of effecting the conversion of Convertible Preferred Stock, such number of its duly authorized shares of Common Stock as shall from time to time be sufficient to effect the conversion of all outstanding Convertible Preferred Stock; and if at any time the number of authorized but unissued shares of Common Stock shall not be sufficient to effect the conversion of all then outstanding shares of Convertible Preferred Stock, the Corporation shall take such corporate action as may be necessary to increase its authorized but unissued shares of Common Stock to such number of shares as shall be sufficient for such purposes, including, without limitation, engaging in best efforts to obtain the requisite stockholder approval of any necessary amendment to this Certificate of Incorporation. Before taking any action which would cause an adjustment reducing the Applicable Conversion Price below the then par value of the shares of Common Stock issuable upon conversion of Convertible Preferred Stock, the Corporation will take any corporate action which may, in the opinion of its counsel, be necessary in order that the Corporation may validly and legally issue fully paid and nonassessable shares of Common Stock at such adjusted Applicable Conversion Price.

4.3.3. Effect of Conversion. All shares of Convertible Preferred Stock which shall have been surrendered for conversion as herein provided shall no longer be deemed to be outstanding and all rights with respect to such shares shall immediately cease and terminate at the Conversion Time, except only the right of the holders thereof to receive shares of Common Stock in exchange therefor, to receive payment in lieu of any fraction of a share otherwise issuable upon such conversion as provided in Subsection 4.2 and, in the case of the Series A Preferred Stock and Series B Preferred Stock, to receive payment of any Accruing Dividends not yet paid, whether or not declared, together with any other dividends declared but unpaid thereon, subject to the approval requirements set forth in Subsection 3.3(e). Any shares of Convertible Preferred Stock so converted shall be retired and cancelled and may not be reissued as shares of such series, and the Corporation may thereafter take such appropriate action (without the need for stockholder action) as may be necessary to reduce the authorized number of shares of Convertible Preferred Stock accordingly.

4.3.4. No Further Adjustment. Upon any such conversion, no adjustment to the Applicable Conversion Price shall be made for any Accruing Dividends or for any declared but unpaid dividends on Convertible Preferred Stock surrendered for conversion or on the Common Stock delivered upon conversion.

4.3.5. Taxes. The Corporation shall pay any and all issue and other similar taxes that may be payable in respect of any issuance or delivery of shares of Common Stock upon conversion of shares of Convertible Preferred Stock pursuant to this Section 4. The Corporation shall not, however, be required to pay any tax which may be payable in respect of any transfer involved in the issuance and delivery of shares of Common Stock in a name other than that in which the shares of Convertible Preferred Stock so converted were registered, and no such issuance or delivery shall be made unless and until the person or entity requesting such issuance has paid to the Corporation the amount of any such tax or has established, to the satisfaction of the Corporation, that such tax has been paid.

4.4. Adjustments to Applicable Conversion Price for Diluting Issues.

4.4.1. Special Definitions. For purposes of this Article Fourth, the following definitions shall apply:

(a) "Option" shall mean rights, options or warrants to subscribe for, purchase or otherwise acquire Common Stock or Convertible Securities.

(b) [Intentionally omitted],

(c) "Convertible Securities" shall mean any evidences of indebtedness, shares or other securities directly or indirectly convertible into or exchangeable for Common Stock, but excluding Options.

(d) "Additional Shares of Common Stock" shall mean all shares of Common Stock issued (or, pursuant to Subsection 4.4.3 below, deemed to be issued) by the Corporation after the Eighth Amendment Date, other than (1) the following shares of Common Stock and (2) shares of Common Stock deemed issued pursuant to the following Options and Convertible Securities (clauses (1) and (2), collectively, "Exempted Securities"):

- (i) shares of Common Stock, Options or Convertible Securities issued as a dividend or distribution on Convertible Preferred Stock;
- (ii) shares of Common Stock, Options or Convertible Securities issued by reason of a dividend, stock split, split-up or other distribution on shares of Common Stock that is covered by Subsection 4.5, 4.6, 4.7 or 4.8;
- (iii) up to 969,877 shares of Common Stock or Options issued to employees or directors of, or consultants or advisors to, the Corporation or any of its subsidiaries pursuant to a plan, agreement or arrangement approved by the Board of Directors of the Corporation;
- (iv) shares of Series C Preferred Stock issued pursuant to the Series C Preferred Stock Purchase Agreement, dated on or about the Eighth Amendment Date; or
- (v) shares of Common Stock or Convertible Securities actually issued upon the exercise of Options or shares of Common Stock actually issued upon the conversion or exchange of Convertible Securities, in each case provided such issuance is pursuant to the terms of such Option or Convertible Security.

4.4.2. No Adjustment of Applicable Conversion Price. No adjustment in the Applicable Conversion Price shall be made as the result of the issuance or deemed issuance of Additional Shares of Common Stock if the Corporation receives written notice from the holders of more than the Appropriate Percentage of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Convertible Preferred Stock agreeing that no such adjustment shall be made as the result of the issuance or deemed issuance of such Additional Shares of Common Stock. In addition, and for the avoidance of doubt, no adjustments in the Applicable Conversion Price shall be made as a result of the fact that additional shares of Common Stock are issuable upon conversion of the Series C Preferred Stock in respect of the accrued and unpaid dividends thereon.

4.4.3. Deemed Issue of Additional Shares of Common Stock.

(a) If the Corporation at any time or from time to time after the Eighth Amendment Date shall issue any Options or Convertible Securities (excluding Options or Convertible Securities which are themselves Exempted Securities) or shall fix a record date for the determination of holders of any class of securities entitled to receive any such Options or Convertible Securities, then the maximum number of shares of Common Stock (as set forth in

the instrument relating thereto, assuming the satisfaction of any conditions to exercisability, convertibility or exchangeability but without regard to any provision contained therein for a subsequent adjustment of such number) issuable upon the exercise of such Options or, in the case of Convertible Securities and Options therefor, the conversion or exchange of such Convertible Securities, shall be deemed to be Additional Shares of Common Stock issued as of the time of such issue or, in case such a record date shall have been fixed, as of the close of business on such record date.

(b) If the terms of any Option or Convertible Security, the issuance of which resulted in an adjustment to the Applicable Conversion Price pursuant to the terms of Subsection 4.4.4, are revised as a result of an amendment to such terms or any other adjustment pursuant to the provisions of such Option or Convertible Security (but excluding automatic adjustments to such terms pursuant to anti-dilution or similar provisions of such Option or Convertible Security) to provide for either (1) any increase or decrease in the number of shares of Common Stock issuable upon the exercise, conversion and/or exchange of any such Option or Convertible Security or (2) any increase or decrease in the consideration payable to the Corporation upon such exercise, conversion and/or exchange, then, effective upon such increase or decrease becoming effective, the Applicable Conversion Price computed upon the original issue of such Option or Convertible Security (or upon the occurrence of a record date with respect thereto) shall be readjusted to such Applicable Conversion Price as would have applied had such revised terms been in effect upon the original date of issuance of such Option or Convertible Security. Notwithstanding the foregoing, no readjustment pursuant to this clause (b) shall have the effect of increasing the Applicable Conversion Price to an amount which exceeds the lower of (i) the Applicable Conversion Price in effect immediately prior to the original adjustment made as a result of the issuance of such Option or Convertible Security, or (ii) the Applicable Conversion Price that would have resulted from any issuances of Additional Shares of Common Stock (other than deemed issuances of Additional Shares of Common Stock as a result of the issuance of such Option or Convertible Security) between the original adjustment date and such readjustment date.

(c) If the terms of any Option or Convertible Security (excluding Options or Convertible Securities which are themselves Exempted Securities), the issuance of which did not result in an adjustment to the Applicable Conversion Price pursuant to the terms of Subsection 4.4.4 (either because the consideration per share (determined pursuant to Subsection 4.4.5) of the Additional Shares of Common Stock subject thereto was equal to or greater than the Applicable Conversion Price then in effect, or because such Option or Convertible Security was issued on or before the Eighth Amendment Date), are revised after the Eighth Amendment Date as a result of an amendment to such terms or any other adjustment pursuant to the provisions of such Option or Convertible Security (but excluding automatic adjustments to such terms pursuant to anti-dilution or similar provisions of such Option or Convertible Security) to provide for either (1) any increase in the number of shares of Common Stock issuable upon the exercise, conversion or exchange of any such Option or Convertible Security or (2) any decrease in the consideration payable to the Corporation upon such exercise, conversion or exchange, then such Option or Convertible Security, as so amended or adjusted, and the Additional Shares of Common Stock subject thereto (determined in the manner provided in Subsection 4.4.3(a)) shall be deemed to have been issued effective upon such increase or decrease becoming effective.

(d) Upon the expiration or termination of any unexercised Option or unconverted or unexchanged Convertible Security (or portion thereof) which resulted (either upon its original issuance or upon a revision of its terms) in an adjustment to the Applicable Conversion Price pursuant to the terms of Subsection 4.4.4, the Applicable Conversion Price shall be readjusted to such Applicable Conversion Price as would have applied had such Option or Convertible Security (or portion thereof) never been issued.

(e) If the number of shares of Common Stock issuable upon the exercise, conversion and/or exchange of any Option or Convertible Security, or the consideration payable to the Corporation upon such exercise, conversion and/or exchange, is calculable at the time such Option or Convertible Security is issued or amended but is subject to adjustment based upon subsequent events, any adjustment to the Applicable Conversion Price provided for in this Subsection 4.4.3 shall be effected at the time of such issuance or amendment based on such number of shares or amount of consideration without regard to any provisions for subsequent adjustments (and any subsequent adjustments shall be treated as provided in clauses (b) and (c) of this Subsection 4.4.3). If the number of shares of Common Stock issuable upon the exercise, conversion and/or exchange of any Option or Convertible Security, or the consideration payable to the Corporation upon such exercise, conversion and/or exchange, cannot be calculated at all at the time such Option or Convertible Security is issued or amended, any adjustment to the Applicable Conversion Price that would result under the terms of this Subsection 4.4.3 at the time of such issuance or amendment shall instead be effected at the time such number of shares and/or amount of consideration is first calculable (even if subject to subsequent adjustments), assuming for purposes of calculating such adjustment to the Applicable Conversion Price that such issuance or amendment took place at the time such calculation can first be made.

4.4.4. Adjustment of Applicable Conversion Price Upon Issuance of Additional Shares of Common Stock. In the event the Corporation shall at any time after the Eighth Amendment Date issue Additional Shares of Common Stock (including Additional Shares of Common Stock deemed to be issued pursuant to Subsection 4.4.3), without consideration or for a consideration per share less than the Applicable Conversion Price in effect immediately prior to such issue, then the Applicable Conversion Price shall be reduced, concurrently with such issue, to a price (calculated to the nearest one-hundredth of a cent) determined in accordance with the following formula:

$$CP_2 = CP_1 * (A + B) \div (A + C).$$

For purposes of the foregoing formula, the following definitions shall apply:

- (i) “CP₂” shall mean the Applicable Conversion Price in effect immediately after such issue of Additional Shares of Common Stock
- (ii) “CP₁” shall mean the Applicable Conversion Price in effect immediately prior to such issue of Additional Shares of Common Stock;

- (iii) “A” shall mean the number of shares of Common Stock outstanding immediately prior to such issue of Additional Shares of Common Stock (treating for this purpose as outstanding all shares of Common Stock issuable upon exercise of Options outstanding immediately prior to such issue or upon conversion or exchange of Convertible Securities (including the Convertible Preferred Stock) outstanding (assuming exercise of any outstanding Options therefor) immediately prior to such issue);
- (iv) “B” shall mean the number of shares of Common Stock that would have been issued if such Additional Shares of Common Stock had been issued at a price per share equal to CP_1 (determined by dividing the aggregate consideration received by the Corporation in respect of such issue by CP_1); and
- (v) “C” shall mean the number of such Additional Shares of Common Stock issued in such transaction.

4.4.5. Determination of Consideration. For purposes of this Subsection 4.4, the consideration received by the Corporation for the issue of any Additional Shares of Common Stock shall be computed as follows:

(a) Cash and Property: Such consideration shall:

- (i) insofar as it consists of cash, be computed at the aggregate amount of cash received by the Corporation, excluding amounts paid or payable for accrued interest;
- (ii) insofar as it consists of property other than cash, be computed at the fair market value thereof at the time of such issue, as determined in good faith by the Board of Directors of the Corporation; and
- (iii) in the event Additional Shares of Common Stock are issued together with other shares or securities or other assets of the Corporation for consideration which covers both, be the proportion of such consideration so received, computed as provided in clauses (i) and (ii) above, as determined in good faith by the Board of Directors of the Corporation.

4.4.6. Options and Convertible Securities. The consideration per share received by the Corporation for Additional Shares of Common Stock deemed to have been issued pursuant to Subsection 4.4.3, relating to Options and Convertible Securities, shall be determined by dividing

- (i) the total amount, if any, received or receivable by the Corporation as consideration for the issue of such Options or Convertible Securities, plus the minimum aggregate amount of additional consideration (as set forth in the instruments relating thereto, without regard to any provision contained therein for a subsequent adjustment of such consideration) payable to the Corporation upon the exercise of such Options or the conversion or exchange of such Convertible Securities, or in the case of Options for Convertible Securities, the exercise of such Options for Convertible Securities and the conversion or exchange of such Convertible Securities, by
- (ii) the maximum number of shares of Common Stock (as set forth in the instruments relating thereto, without regard to any provision contained therein for a subsequent adjustment of such number) issuable upon the exercise of such Options or the conversion or exchange of such Convertible Securities, or in the case of Options for Convertible Securities, the exercise of such Options for Convertible Securities and the conversion or exchange of such Convertible Securities.

4.4.7. Multiple Closing Dates. In the event the Corporation shall issue on more than one date Additional Shares of Common Stock that are a part of one transaction or a series of related transactions and that would result in an adjustment to the Applicable Conversion Price pursuant to the terms of Subsection 4.4.4, and such issuance dates occur within a period of no more than ninety (90) days from the first such issuance to the final such issuance, then, upon the final such issuance, the Applicable Conversion Price shall be readjusted to give effect to all such issuances as if they occurred on the date of the first such issuance (and without giving effect to any additional adjustments as a result of any such subsequent issuances within such period).

4.5. Adjustment for Stock Splits and Combinations. If the Corporation shall at any time or from time to time after the Eighth Amendment Date effect a subdivision of the outstanding Common Stock, the Applicable Conversion Price in effect immediately before that subdivision shall be proportionately decreased so that the number of shares of Common Stock issuable on conversion of each share of such series shall be increased in proportion to such increase in the aggregate number of shares of Common Stock outstanding. If the Corporation shall at any time or from time to time after the Eighth Amendment Date combine the outstanding shares of Common Stock, the Applicable Conversion Price in effect immediately before the combination shall be proportionately increased so that the number of shares of Common Stock

issuable on conversion of each share of such series shall be decreased in proportion to such decrease in the aggregate number of shares of Common Stock outstanding. Any adjustment under this subsection shall become effective at the close of business on the date the subdivision or combination becomes effective.

4.6. Adjustment for Certain Dividends and Distributions. In the event the Corporation at any time or from time to time after the Eighth Amendment Date shall make or issue, or fix a record date for the determination of holders of Common Stock entitled to receive, a dividend or other distribution payable on the Common Stock in additional shares of Common Stock, then and in each such event the Applicable Conversion Price in effect immediately before such event shall be decreased as of the time of such issuance or, in the event such a record date shall have been fixed, as of the close of business on such record date, by multiplying the Applicable Conversion Price then in effect by a fraction:

- (1) the numerator of which shall be the total number of shares of Common Stock issued and outstanding immediately prior to the time of such issuance or the close of business on such record date, and
- (2) the denominator of which shall be the total number of shares of Common Stock issued and outstanding immediately prior to the time of such issuance or the close of business on such record date plus the number of shares of Common Stock issuable in payment of such dividend or distribution.

Notwithstanding the foregoing, (a) if such record date shall have been fixed and such dividend is not fully paid or if such distribution is not fully made on the date fixed therefor, the Applicable Conversion Price shall be recomputed accordingly as of the close of business on such record date and thereafter the Applicable Conversion Price shall be adjusted pursuant to this subsection as of the time of actual payment of such dividends or distributions; and (b) no such adjustment shall be made if the holders of Preferred Stock simultaneously receive a dividend or other distribution of shares of Common Stock in a number equal to the number of shares of Common Stock as they would have received if all outstanding shares of Convertible Preferred Stock had been converted into Common Stock on the date of such event.

4.7. Adjustments for Other Dividends and Distributions. In the event the Corporation at any time or from time to time after the Eighth Amendment Date shall make or issue, or fix a record date for the determination of holders of Common Stock entitled to receive, a dividend or other distribution payable in securities of the Corporation (other than a distribution of shares of Common Stock in respect of outstanding shares of Common Stock) or in other property and the provisions of Section 1 do not apply to such dividend or distribution, then in each such event the holders of Convertible Preferred Stock shall receive, simultaneously with the distribution to the holders of Common Stock, a dividend or other distribution of such securities or other property in an amount equal to the amount of such securities or other property as they would have received if all outstanding shares of Convertible Preferred Stock had been converted into Common Stock on the date of such event.

4.8. Adjustment for Merger or Reorganization, etc. Subject to the provisions of Subsection 2.3, if there shall occur any reorganization, recapitalization, reclassification, consolidation or merger involving the Corporation in which Common Stock (but not Preferred Stock) is converted into or exchanged for securities, cash or other property (other than a transaction covered by Subsections 4.4, 4.6 or 4.7), then, following any such reorganization, recapitalization, reclassification, consolidation or merger, each share of Convertible Preferred Stock shall thereafter be convertible in lieu of the Common Stock into which it was convertible prior to such event into the kind and amount of securities, cash or other property which a holder of the number of shares of Common Stock of the Corporation issuable upon conversion of one share of Convertible Preferred Stock immediately prior to such reorganization, recapitalization, reclassification, consolidation or merger would have been entitled to receive pursuant to such transaction; and, in such case, appropriate adjustment (as determined in good faith by the Board of Directors of the Corporation) shall be made in the application of the provisions in this Section 4 with respect to the rights and interests thereafter of the holders of Convertible Preferred Stock, to the end that the provisions set forth in this Section 4 (including provisions with respect to changes in and other adjustments of the Applicable Conversion Price) shall thereafter be applicable, as nearly as reasonably may be, in relation to any securities or other property thereafter deliverable upon the conversion of Convertible Preferred Stock.

4.9. Certificate as to Adjustments. Upon the occurrence of each adjustment or readjustment of the Applicable Conversion Price pursuant to this Section 4, the Corporation at its expense shall, as promptly as reasonably practicable but in any event not later than ten (10) days thereafter, compute such adjustment or readjustment in accordance with the terms hereof and furnish to each holder of Convertible Preferred Stock a certificate setting forth such adjustment or readjustment (including the kind and amount of securities, cash or other property into which the Convertible Preferred Stock is convertible) and showing in detail the facts upon which such adjustment or readjustment is based. The Corporation shall, as promptly as reasonably practicable after the written request at any time of any holder of Convertible Preferred Stock (but in any event not later than ten (10) days thereafter), furnish or cause to be furnished to such holder a certificate setting forth (i) the Applicable Conversion Price then in effect, and (ii) the number of shares of Common Stock and the amount, if any, of other securities, cash or property which then would be received upon the conversion of the holder's Convertible Preferred Stock.

4.10. Notice of Record Date. In the event:

(a) the Corporation shall take a record of the holders of Common Stock (or other capital stock or securities at the time issuable upon conversion of Convertible Preferred Stock) for the purpose of entitling or enabling them to receive any dividend or other distribution, or to receive any right to subscribe for or purchase any shares of capital stock of any class or any other securities, or to receive any other security; or

(b) of any capital reorganization of the Corporation, any reclassification of Common Stock, or any Deemed Liquidation Event; or

(c) of the voluntary or involuntary dissolution, liquidation or winding-up of the Corporation,

then in each such case, the Corporation will send or cause to be sent to the holders of Convertible Preferred Stock a notice specifying, as the case may be, (i) the record date for such dividend, distribution or right, and the amount and character of such dividend, distribution or right, or (ii) the effective date on which such reorganization, reclassification, consolidation, merger, transfer, dissolution, liquidation or winding-up is proposed to take place, and the time, if any is to be fixed, as of which the holders of record of Common Stock (or such other capital stock or securities at the time issuable upon the conversion of Convertible Preferred Stock) shall be entitled to exchange their shares of Common Stock (or such other capital stock or securities) for securities or other property deliverable upon such reorganization, reclassification, consolidation, merger, transfer, dissolution, liquidation or winding-up, and the amount per share and character of such exchange applicable to Convertible Preferred Stock and Common Stock. Such notice shall be sent at least ten (10) days prior to the record date or effective date for the event specified in such notice.

5. Mandatory Conversion.

5.1. Convertible Preferred Stock.

5.1.1. Trigger Events. Upon the closing of the sale of shares of Common Stock to the public at a price of at least \$20.00 per share (subject to appropriate adjustment in the event of any stock dividend, stock split, combination or other similar recapitalization with respect to the Common Stock after the Eighth Amendment Date), in a firm-commitment underwritten public offering pursuant to an effective registration statement under the Securities Act of 1933, as amended, resulting in at least \$40 million of gross proceeds to the Corporation, all outstanding shares of Convertible Preferred Stock shall automatically be converted into shares of Common Stock, at the then effective conversion rate, and such shares may not be reissued by the Corporation. Upon the date and time, or the occurrence of an event, specified by vote or written consent of the holders of at least sixty-five percent (65%) of the then outstanding shares of Series A Preferred Stock, voting as a separate class, all outstanding shares of Series A Preferred Stock shall automatically be converted into shares of Common Stock, at the then effective conversion rate, and such shares may not be reissued by the Corporation. Upon the date and time, or the occurrence of an event, specified by vote or written consent of the holders of at least sixty-five percent (65%) of the then outstanding shares of Series B Preferred Stock, voting as a separate class, all outstanding shares of Series B Preferred Stock shall automatically be converted into shares of Common Stock, at the then effective conversion rate, and such shares may not be reissued by the Corporation. Upon the date and time, or the occurrence of an event, specified by vote or written consent of the holders of at least sixty-five percent (65%) of the then outstanding shares of Series C Preferred Stock, voting as a separate class, all outstanding shares of Series C Preferred Stock shall automatically be converted into shares of Common Stock, at the then effective conversion rate, and such shares may not be reissued by the Corporation. The time of the closing of such underwritten public offering or the date and time specified or the time of the event specified in such vote or written consent otherwise referred to above is referred to in each case herein as the "Mandatory Conversion Time."

5.1.2. Procedural Requirements. All holders of record of shares of Convertible Preferred Stock shall be sent written notice of the Mandatory Conversion Time and the place designated for mandatory conversion of all such shares of Convertible Preferred Stock

(or the applicable shares of Convertible Preferred Stock, as the case may be) pursuant to this Section 5. Such notice need not be sent in advance of the occurrence of the Mandatory Conversion Time. Upon receipt of such notice, each holder of shares of Convertible Preferred Stock (or the applicable shares of Convertible Preferred Stock, as the case may be) shall surrender his, her or its certificate or certificates for such shares (or, if such holder alleges that such certificate has been lost, stolen or destroyed, a lost certificate affidavit and agreement reasonably acceptable to the Corporation to indemnify the Corporation against any claim that may be made against the Corporation on account of the alleged loss, theft or destruction of such certificate) to the Corporation at the place designated in such notice. If so required by the Corporation, certificates surrendered for conversion shall be endorsed or accompanied by written instrument or instruments of transfer, in form satisfactory to the Corporation, duly executed by the registered holder or by his, her or its attorney duly authorized in writing. All rights with respect to the Convertible Preferred Stock converted pursuant to Section 5.1, including the rights, if any, to receive notices and vote (other than as a holder of Common Stock), will terminate at the Mandatory Conversion Time (notwithstanding the failure of the holder or holders thereof to surrender the certificates at or prior to such time), except only the rights of the holders thereof, upon surrender of their certificate or certificates (or lost certificate affidavit and agreement) therefor, to receive the items provided for in the next sentence of this Subsection 5.2. As soon as practicable after the Mandatory Conversion Time and the surrender of the certificate or certificates (or lost certificate affidavit and agreement) for Preferred Stock, the Corporation shall issue and deliver to such holder, or to his, her or its nominees, a certificate or certificates for the number of full shares of Common Stock issuable on such conversion in accordance with the provisions hereof, together with cash as provided in Subsection 4.2 in lieu of any fraction of a share of Common Stock otherwise issuable upon such conversion and the payment of any Accruing Dividends accrued but unpaid on the shares of Series A Preferred Stock or Series B Preferred Stock converted, whether or not declared, together with any other dividends declared but unpaid thereon. Such converted Convertible Preferred Stock shall be retired and cancelled and may not be reissued as shares of such series, and the Corporation may thereafter take such appropriate action (without the need for stockholder action) as may be necessary to reduce the authorized number of shares of Preferred Stock accordingly.

5.2. Series X Preferred Stock.

5.2.1. Conversion into Series B Preferred Stock. Unless earlier converted in accordance with Section 5.2.2 below, upon the earlier of (i) the approval of the holders of at least sixty percent (60%) of the Series X Preferred Stock or (ii) June 30, 2013 (the "Maturity Date"), each outstanding share of Series X Preferred Stock shall be converted into a number of shares of Series B Preferred Stock determined by dividing One Hundred Dollars (\$100.00) by the Series X Conversion Price in effect at the time of conversion. The "Series X Conversion Price" shall mean \$14.00 as of the Eighth Amendment Date.

5.2.2. Conversion upon Qualified Financing. Prior to the Maturity Date, all outstanding shares of Series X Preferred Stock shall automatically be converted into fully paid and non-assessable shares of capital stock of the Corporation upon the occurrence of a Qualified Financing (as defined below). In the event of a Qualified Financing, the type and class of capital stock of the Corporation to be issued to the holders of Series X Preferred Stock upon conversion pursuant to this Section 5.2.2 (and the rights and privileges of the holders thereof)

shall be identical to the type and class of the capital stock issued by the Corporation in connection with such Qualified Financing; provided, that if no capital stock is issued pursuant to the Qualified Financing, the shares of Series X Preferred Stock shall automatically convert into shares of Series B Preferred Stock in accordance with Section 5.2.1 above (as applicable, the "Investor Stock"). Upon conversion of the shares of Series X Preferred Stock in connection with a Qualified Financing, the holder of each share of Series X Preferred Stock shall be entitled to a number of shares of Investor Stock determined by dividing (i) the number of shares of Series X Preferred Stock held by such holder as of the Investor Conversion Date (as defined below) times One Hundred Dollars (\$100.00) by (ii) an amount equal to the lowest price per share of Investor Stock paid by the purchasers of such shares in connection with the Qualified Financing (the "Qualified Financing Conversion Price"). A "Qualified Financing" shall mean and include (x) the sale of shares of capital stock of the Corporation that are senior to or pari passu with the Series B Preferred Stock, in one transaction or a series of related transactions, which sale or sales result in gross proceeds to the Corporation (excluding the conversion of the Series X Preferred Stock) of at least Ten Million Dollars (\$10,000,000) (a "Series C Financing"), or (y) in the event of a license deal, a Series C Financing plus up-front payments from a licensing transaction, which results in gross proceeds to the Corporation of at least Thirty Million Dollars (\$30,000,000) collectively. For the avoidance of doubt, if a Qualified Financing has multiple tranches, all shares of Series X Preferred Stock shall be converted upon the closing of the first tranche.

5.2.3. Adjustment for Stock Splits and Combinations. If the Corporation shall at any time or from time to time after the Eighth Amendment Date effect a subdivision of the outstanding Series B Preferred Stock, the Series X Conversion Price in effect immediately before that subdivision shall be proportionately decreased so that the number of shares of Series B Preferred Stock issuable on conversion of each share of such series shall be increased in proportion to such increase in the aggregate number of shares of Series B Preferred Stock outstanding. If the Corporation shall at any time or from time to time after the Eighth Amendment Date combine the outstanding shares of Series B Preferred Stock, the Series X Conversion Price in effect immediately before the combination shall be proportionately increased so that the number of shares of Series B Preferred Stock issuable on conversion of each share of such series shall be decreased in proportion to such decrease in the aggregate number of shares of Series B Preferred Stock outstanding. Any adjustment under this subsection shall become effective at the close of business on the date the subdivision or combination becomes effective.

5.2.4. Adjustment for Certain Dividends and Distributions. In the event the Corporation at any time or from time to time after the Eighth Amendment Date shall make or issue, or fix a record date for the determination of holders of Series B Preferred Stock entitled to receive, a dividend or other distribution payable on the Series B Preferred Stock in additional shares of Series B Preferred Stock, then and in each such event the Series X Conversion Price in effect immediately before such event shall be decreased as of the time of such issuance or, in the event such a record date shall have been fixed, as of the close of business on such record date, by multiplying the Series X Conversion Price then in effect by a fraction:

- (1) the numerator of which shall be the total number of shares of Series B Preferred Stock issued and outstanding immediately prior to the time of such issuance or the close of business on such record date, and

- (2) the denominator of which shall be the total number of shares of Series B Preferred Stock issued and outstanding immediately prior to the time of such issuance or the close of business on such record date plus the number of shares of Series B Preferred Stock issuable in payment of such dividend or distribution.

Notwithstanding the foregoing, (a) if such record date shall have been fixed and such dividend is not fully paid or if such distribution is not fully made on the date fixed therefor, the Series X Conversion Price shall be recomputed accordingly as of the close of business on such record date and thereafter the Series X Conversion Price shall be adjusted pursuant to this subsection as of the time of actual payment of such dividends or distributions; and (b) no such adjustment shall be made if the holders of Series X Preferred Stock simultaneously receive a dividend or other distribution of shares of Series B Preferred Stock in a number equal to the number of shares of Series B Preferred Stock as they would have received if all outstanding shares of Series X Preferred Stock had been converted into Series B Preferred Stock on the date of such event.

5.2.5. Adjustment for Other Dividends and Distributions. In the event the Corporation at any time or from time to time after the Eighth Amendment Date shall make or issue, or fix a record date for the determination of holders of Series B Preferred Stock entitled to receive, a dividend or other distribution payable in securities of the Corporation (other than a distribution of shares of Series B Preferred Stock in respect of outstanding shares of Series B Preferred Stock) or in other property and the provisions of Section 1 do not apply to such dividend or distribution, then in each such event the holders of Series X Preferred Stock shall receive, simultaneously with the distribution to the holders of Series B Preferred Stock, a dividend or other distribution of such securities or other property in an amount equal to the amount of such securities or other property as they would have received if all outstanding shares of Series X Preferred Stock had been converted into Series B Preferred Stock on the date of such event.

5.2.6. Adjustment for Merger or Reorganization, etc. Subject to the provisions of Subsection 2.3, if there shall occur any reorganization, recapitalization, reclassification, consolidation or merger involving the Corporation in which Series B Preferred Stock (but not Series X Preferred Stock) is converted into or exchanged for securities, cash or other property (other than a transaction covered by Subsections 5.2.4 or 5.2.5), then, following any such reorganization, recapitalization, reclassification, consolidation or merger, each share of Series X Preferred Stock shall thereafter be convertible in lieu of the Series B Preferred Stock into which it was convertible prior to such event into the kind and amount of securities, cash or other property which a holder of the number of shares of Series B Preferred Stock of the Corporation issuable upon conversion of one share of Series X Preferred Stock immediately prior to such reorganization, recapitalization, reclassification, consolidation or merger would have been entitled to receive pursuant to such transaction; and, in such case, appropriate adjustment (as determined in good faith by the Board of Directors of the Corporation) shall be made in the application of the provisions in this Section 5 with respect to the rights and interests

thereafter of the holders of Series X Preferred Stock, to the end that the provisions set forth in this Section 5 (including provisions with respect to changes in and other adjustments of the Series X Conversion Price) shall thereafter be applicable, as nearly as reasonably may be, in relation to any securities or other property thereafter deliverable upon the conversion of Series X Preferred Stock.

5.2.7. Certificate as to Adjustments. Upon the occurrence of each adjustment or readjustment of the Series X Conversion Price pursuant to this Section 5, the Corporation at its expense shall, as promptly as reasonably practicable but in any event not later than ten (10) days thereafter, compute such adjustment or readjustment in accordance with the terms hereof and furnish to each holder of Series X Preferred Stock a certificate setting forth such adjustment or readjustment (including the kind and amount of securities, cash or other property into which the Series X Preferred Stock is convertible) and showing in detail the facts upon which such adjustment or readjustment is based. The Corporation shall, as promptly as reasonably practicable after the written request at any time of any holder of Series X Preferred Stock (but in any event not later than ten (10) days thereafter), furnish or cause to be furnished to such holder a certificate setting forth (i) the Series X Conversion Price then in effect, and (ii) the number of shares of Series B Preferred Stock and the amount, if any, of other securities, cash or property which then would be received upon the conversion of the holder's Series X Preferred Stock.

5.2.8. Procedural Requirements.

(a) Upon the earlier of the Maturity Date or the closing of a Qualified Financing (as applicable, the "Investor Conversion Date") each share of Series X Preferred Stock shall be converted automatically without any further action by the holder.

(b) At least ten (10) days prior to the Maturity Date or a Qualified Financing, as applicable, the Corporation shall cause notice of the Maturity Date or the Qualified Financing, as applicable, to be mailed to the registered holders of Series X Preferred Stock at such holders' address appearing in the records of the Corporation. The Corporation shall, as soon as practicable after the Maturity Date or the Qualified Financing, as applicable, issue and deliver to the holders of such shares of Series X Preferred Stock, or to his or its nominees or respective Affiliates, a certificate or certificates for the number of shares of capital stock to which such holder shall be entitled.

(c) The Corporation shall upon authorization of the sale of shares of its capital stock, or immediately prior to the Maturity Date, for the purpose of effecting the conversion of the shares of Series X Preferred Stock as provided in Section 5.2, authorize a sufficient number of shares of Series B Preferred Stock or Investor Stock, as applicable, to effect the conversion of the outstanding shares of Series X Preferred Stock.

(d) Immediately upon the Investor Conversion Date, each share of Series X Preferred Stock shall no longer be deemed to be outstanding and all rights with respect to the shares of Series X Preferred Stock shall immediately cease and terminate on the Investor Conversion Date, except only the right of the holder to receive the shares of capital stock to which it is entitled as a result of the conversion on the Investor Conversion Date or

assign such right to any of its Affiliates. For purposes of this Section 5.2, “Affiliate” means, with respect to any specified holder, any other person or entity who, directly or indirectly, controls, is controlled by, or is under common control with such holder, including without limitation any general partner, managing member, officer or director of such holder or any venture capital fund now or hereafter existing that is controlled by one or more general partners or managing members of, or shares the same management company with, such holder (including, without limitation, any trust which shares a trustee or investment advisor (or for which an immediate family member acts as trustee or investment advisor) with another holder and any account held for the benefit of the trustee or beneficiary of any such trust).

(e) The Corporation shall pay any and all issue and other taxes that may be payable in respect of any issuance or delivery of shares of capital stock, upon conversion of the Series X Preferred Stock pursuant to Section 5.2. The Corporation shall not, however, be required to pay any tax which may be payable in respect of any transfer involved in the issuance and delivery of shares of capital stock in a name other than that of the registered holder of each share of Series X Preferred Stock, and no such issuance or delivery shall be made unless and until the person or entity requesting such issuance has paid to the Corporation the amount of any such tax or has established, to the satisfaction of the Corporation, that such tax has been paid.

6. Redemption.

6.1. Redemption of Preferred Stock.

6.1.1. Convertible Preferred Stock. Shares of Convertible Preferred Stock shall be redeemed by the Corporation out of funds lawfully available therefor at a price equal to the greater of (a) the Applicable Accrued Value and (b) the Fair Market Value (as defined below) per share (such greater price is the “Convertible Stock Redemption Price”) in three annual installments commencing not more than sixty (60) days after receipt by the Corporation at any time on or after July 16, 2015, from the holders of more than the Appropriate Percentage of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Convertible Preferred Stock, of written notice requesting redemption of all shares of Convertible Preferred Stock (the “Convertible Stock Redemption Notice”). The date of each such installment shall be referred to as a “Convertible Stock Redemption Date.” On each Convertible Stock Redemption Date, the Corporation shall redeem, on a pro rata basis in accordance with the number of shares of Convertible Preferred Stock owned by each holder, that number of outstanding shares of Convertible Preferred Stock determined by dividing (i) the total number of shares of Convertible Preferred Stock outstanding immediately prior to such Convertible Stock Redemption Date by (ii) the number of remaining Convertible Stock Redemption Dates (including the Convertible Stock Redemption Date to which such calculation applies). If the Corporation does not have sufficient funds legally available to redeem on any Convertible Stock Redemption Date all shares of Convertible Preferred Stock to be redeemed on such Convertible Stock Redemption Date, the Corporation shall first redeem a pro rata portion of the Applicable Accrued Value of each holder’s shares of Series C Preferred Stock to the fullest extent possible, up through the Series C Preferred Stock’s Applicable Accrued Value, and only after complete redemption of the Series C Preferred Stock’s Applicable Accrued Value shall then redeem a pro rata portion of each holder’s shares of Series B Preferred Stock to the fullest extent

possible, up through the Series B Preferred Stock's Applicable Accrued Value, and, only after complete redemption of the Series B Preferred Stock's Applicable Accrued Value, shall then redeem a pro rata portion of each holder's shares of Series A Preferred Stock to the fullest extent possible up through the Series A Preferred Stock's Applicable Accrued Value, out of funds legally available therefor, based on the respective amounts which would otherwise be payable in respect of the shares to be redeemed if the legally available funds were sufficient to redeem all such shares, and shall redeem the remaining shares to have been redeemed as soon as practicable after the Corporation has funds legally available therefor. Notwithstanding the other provisions hereof, in the event that the Convertible Stock Redemption Price is determined to be the Applicable Original Issue Price per share (or a multiple thereof in the case of the Series C Preferred Stock), plus any Accruing Dividends accrued but unpaid thereon (or a multiple thereof in the case of the Series C Preferred Stock), whether or not declared, together with any other dividends declared but unpaid thereon, then, in such case and as to Triathlon only, any such Accruing Dividends accrued but unpaid on the shares of Convertible Preferred Stock held by Triathlon, whether or not declared, together with any other dividends declared but unpaid on the shares of Series A Preferred Stock held by Triathlon, shall be paid to Triathlon under this Section 6 only to the extent payable out of retained earnings of the Corporation.

6.1.2. Fair Market Value. For purposes of this Subsection 6.1, "Fair Market Value" shall mean the per share value of the Convertible Preferred Stock determined as of each Convertible Stock Redemption Date without regard to any discount for illiquidity, minority interest or the like. Fair Market Value shall be determined by mutual agreement of the Corporation and the holders of more than the Appropriate Percentage of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Convertible Preferred Stock. If the parties are unable to agree on a Fair Market Value, then Fair Market Value shall be determined by an appraisal prepared by a qualified appraiser acceptable to the Corporation and the holders of more than the Appropriate Percentage of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Convertible Preferred Stock. If the parties are unable to agree on the identity of an appraiser within thirty (30) days after the Corporation's receipt of the Convertible Stock Redemption Notice, then (i) the Corporation and (ii) the holders of more than the Appropriate Percentage of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Convertible Preferred Stock, shall each select an appraiser, and the two appraisers so selected shall choose a third appraiser who shall conduct the appraisal and whose determination shall be binding and final. The costs of any such appraisals pursuant to this Subsection 6.1 shall be borne by the Corporation.

6.2. Redemption Notice. The Corporation shall send written notice of the mandatory redemption (the "Redemption Notice") to each holder of record of Preferred Stock not less than ten (10) days prior to each redemption date applicable to the Series X Preferred Stock and not less than forty (40) days prior to each Convertible Stock Redemption Date. Each Redemption Notice shall state:

(a) the number of shares of Preferred Stock held by the holder that the Corporation shall redeem on the Redemption Date specified in the Redemption Notice;

(b) the Redemption Date and the Redemption Price;

(c) for Convertible Preferred Stock, the date upon which the holder's right to convert such shares terminates (as determined in accordance with Subsection 4.1); and

(d) that the holder is to surrender to the Corporation, in the manner and at the place designated, his, her or its certificate or certificates representing the shares of Preferred Stock to be redeemed.

6.3. Surrender of Certificates; Payment. On or before the applicable Redemption Date, each holder of shares of Preferred Stock to be redeemed on such Redemption Date, unless any holder of Convertible Preferred Stock has exercised his, her or its right to convert such shares as provided in Section 4, shall surrender the certificate or certificates representing such shares (or, if such registered holder alleges that such certificate has been lost, stolen or destroyed, a lost certificate affidavit and agreement reasonably acceptable to the Corporation to indemnify the Corporation against any claim that may be made against the Corporation on account of the alleged loss, theft or destruction of such certificate) to the Corporation, in the manner and at the place designated in the Redemption Notice, and thereupon the Redemption Price for such shares shall be payable to the order of the person whose name appears on such certificate or certificates as the owner thereof. In the event less than all of the shares of Preferred Stock represented by a certificate are redeemed, a new certificate representing the unredeemed shares of Preferred Stock shall promptly be issued to such holder.

6.4. Rights Subsequent to Redemption. If the Redemption Notice shall have been duly given, and if on the applicable Redemption Date the Redemption Price payable upon redemption of the shares of Preferred Stock to be redeemed on such Redemption Date is paid or tendered for payment or deposited with an independent payment agent so as to be available therefor in a timely manner, then notwithstanding that the certificates evidencing any of the shares of Preferred Stock so called for redemption shall not have been surrendered, dividends with respect to such shares of Convertible Preferred Stock shall cease to accrue after such Redemption Date and all rights with respect to such shares shall forthwith after the Redemption Date terminate, except only the right of the holders to receive the Redemption Price without interest upon surrender of their certificate or certificates therefor.

7. Redeemed or Otherwise Acquired Shares. Any shares of Preferred Stock that are redeemed or otherwise acquired by the Corporation or any of its subsidiaries shall be automatically and immediately cancelled and retired and shall not be reissued, sold or transferred. Neither the Corporation nor any of its subsidiaries may exercise any voting or other rights granted to the holders of Preferred Stock following redemption.

8. Waiver. Any of the rights, powers, preferences and other terms of the Series X Preferred Stock set forth herein may be waived on behalf of all holders of Series X Preferred Stock by the affirmative written consent or vote of the holders of at least sixty percent (60%) of the shares of Series X Preferred Stock then outstanding. Any of the rights, powers, preferences and other terms of the Convertible Preferred Stock set forth herein may (in addition and subject to any other vote or consent required by law or this Certificate of Incorporation) be waived on behalf of all holders of Convertible Preferred Stock by the affirmative written consent or vote of the holders of more than the Appropriate Percentage of the shares of Common Stock then

issuable upon conversion of the then outstanding shares of Convertible Preferred Stock. It is understood that the reference in the preceding sentence to “any other vote or consent required by law or this Certificate of Incorporation” would have the effect, for instance, that the waiver or amendment of a provision of this Certificate of Incorporation that by its terms operates by using a percentage of the applicable shares of Common Stock then so issuable that is higher than the Applicable Percentage would require such higher percentage of such applicable shares to be adopted.

9. **Notices.** Any notice required or permitted by the provisions of this Article Fourth to be given to a holder of shares of Preferred Stock shall be mailed, postage prepaid, to the post office address last shown on the records of the Corporation, or given by electronic communication in compliance with the provisions of the General Corporation Law, and shall be deemed sent upon such mailing or electronic transmission.

FIFTH: Subject to any additional vote required by this Certificate of Incorporation or Bylaws, in furtherance and not in limitation of the powers conferred by statute, the Board of Directors is expressly authorized to make, repeal, alter, amend and rescind any or all of the Bylaws of the Corporation.

SIXTH: Subject to any additional vote required by this Certificate of Incorporation, the number of directors of the Corporation shall be determined in the manner set forth in the Bylaws of the Corporation.

SEVENTH: Elections of directors need not be by written ballot unless the Bylaws of the Corporation shall so provide.

EIGHTH: Meetings of stockholders may be held within or without the State of Delaware, as the Bylaws of the Corporation may provide. The books of the Corporation may be kept outside the State of Delaware at such place or places as may be designated from time to time by the Board of Directors or in the Bylaws of the Corporation.

NINTH: To the fullest extent permitted by law, a director of the Corporation shall not be personally liable to the Corporation or its stockholders for monetary damages for breach of fiduciary duty as a director. If the General Corporation Law or any other law of the State of Delaware is amended after approval by the stockholders of this Article Ninth to authorize corporate action further eliminating or limiting the personal liability of directors, then the liability of a director of the Corporation shall be eliminated or limited to the fullest extent permitted by the General Corporation Law as so amended.

Any repeal or modification of the foregoing provisions of this Article Ninth by the stockholders of the Corporation shall not adversely affect any right or protection of a director of the Corporation existing at the time of, or increase the liability of any director of the Corporation with respect to any acts or omissions of such director occurring prior to, such repeal or modification.

TENTH: The following indemnification provisions shall apply to the persons enumerated below:

Right to Indemnification of Directors and Officers. The Corporation shall indemnify and hold harmless, to the fullest extent permitted by applicable law as it presently exists or may hereafter be amended, any person (an “Indemnified Person”) who was or is made or is threatened to be made a party or is otherwise involved in any action, suit or proceeding, whether civil, criminal, administrative or investigative (a “Proceeding”), by reason of the fact that such person, or a person for whom such person is the legal representative, is or was a director or officer of the Corporation or, while a director or officer of the Corporation, is or was serving at the request of the Corporation as a director, officer, employee or agent of another corporation or of a partnership, joint venture, limited liability company, trust, enterprise or nonprofit entity, including service with respect to employee benefit plans, against all liability and loss suffered and expenses (including attorneys’ fees) reasonably incurred by such Indemnified Person in such Proceeding. Notwithstanding the preceding sentence, except as otherwise provided in Section 3 of this Article Tenth, the Corporation shall be required to indemnify an Indemnified Person in connection with a Proceeding (or part thereof) commenced by such Indemnified Person only if the commencement of such Proceeding (or part thereof) by the Indemnified Person was authorized in advance by the Board of Directors.

Prepayment of Expenses of Directors and Officers. The Corporation shall advance payment for the expenses (including attorneys’ fees) incurred by an Indemnified Person in defending any Proceeding in advance of its final disposition, provided, however, that, to the extent required by law, such payment of expenses in advance of the final disposition of the Proceeding shall be made only upon receipt of an undertaking by the Indemnified Person to repay all amounts advanced if it should be ultimately determined that the Indemnified Person is not entitled to be indemnified under this Article Tenth, applicable law or otherwise.

Claims by Directors and Officers. If a claim for indemnification or advancement of expenses under this Article Tenth is not paid in full within thirty (30) days after a written claim therefor by the Indemnified Person has been received by the Corporation, the Indemnified Person may file suit to recover the unpaid amount of such claim and, if successful in whole or in part, shall be entitled to be paid the expense of prosecuting such claim. In any such action the Corporation shall have the burden of proving that the Indemnified Person is not entitled to the requested indemnification or advancement of expenses under this Article Tenth, applicable law or otherwise.

Indemnification of Employees and Agents. The Corporation may indemnify and advance expenses to any person who was or is made or is threatened to be made or is otherwise involved in any Proceeding by reason of the fact that such person, or a person for whom such person is the legal representative, is or was an employee or agent of the Corporation or, while an employee or agent of the Corporation, is or was serving at the request of the Corporation as a director, officer, employee or agent of another corporation or of a partnership, joint venture, limited liability company, trust, enterprise or nonprofit entity, including service with respect to employee benefit plans, against all liability and loss suffered and expenses (including attorney’s fees) reasonably incurred by such person in connection with such Proceeding. The ultimate determination of entitlement to indemnification of persons who are non-director or officer employees or agents under the provisions of this Section 4 of this Article Tenth shall be made in such manner as is determined by the Board of Directors in its sole discretion. Notwithstanding the foregoing sentence, the Corporation shall not be required to indemnify a person in connection with a Proceeding initiated by such person if the Proceeding was not authorized in advance by the Board of Directors.

Advancement of Expenses of Employees and Agents. The Corporation may advance payment for the expenses (including attorney's fees) incurred by an employee or agent in defending any Proceeding in advance of its final disposition on such terms and conditions as may be determined by the Board of Directors.

Non-Exclusivity of Rights. The rights conferred on any person by this Article Tenth shall not be exclusive of any other rights which such person may have or hereafter acquire under any statute, this Certificate of Incorporation, the Bylaws, any agreement, any vote of stockholders or directors who are not Indemnified Persons in the Proceeding or otherwise.

Other Indemnification. The Corporation's obligation, if any, to indemnify or advance payments to any person who was or is serving at its request as a director, officer or employee of another corporation, partnership, limited liability company, joint venture, trust, organization or other enterprise shall be reduced or the Corporation shall be reimbursed, as the case may be, by any amount such person may collect as indemnification from such other corporation, partnership, limited liability company, joint venture, trust, organization or other enterprise.

Insurance. The Board of Directors may, to the full extent permitted by applicable law as it presently exists, or may hereafter be amended from time to time, authorize the Corporation to purchase and maintain insurance: (a) to indemnify the Corporation for any obligation which it incurs as a result of the indemnification of directors, officers and employees under the provisions of this Article Tenth; and (b) to indemnify or insure directors, officers and employees against liability in instances in which they may not otherwise be indemnified by the Corporation under the provisions of this Article Tenth.

Amendment or Repeal. Any repeal or modification of the foregoing provisions of this Article Tenth shall not adversely affect any right or protection hereunder of any person in respect of any act or omission occurring prior to the time of such repeal or modification, nor may any such repeal or modification increase the, or create any, liability of any of the same with respect to any act or omission occurring prior to, or commensurate with, such repeal or modification. The rights provided hereunder shall inure to the benefit of any Indemnified Person and such person's heirs, executors and administrators.

ELEVENTH: The Corporation renounces any interest or expectancy of the Corporation in, or in being offered an opportunity to participate in, any Excluded Opportunity. An "Excluded Opportunity," is any matter, transaction or interest that is presented to, or acquired, created or developed by, or which otherwise comes into the possession of, (i) any director of the Corporation who is not an employee of the Corporation or any of its subsidiaries, or (ii) any holder of Preferred Stock or any partner, member, director, stockholder, employee or agent of any such holder, other than someone who is an employee of the Corporation or any of its subsidiaries (collectively, "Covered Persons"), unless such matter, transaction or interest is presented to, or acquired, created or developed by, or otherwise comes into the possession of, a Covered Person expressly and solely in such Covered Person's capacity as a director of the Corporation.

* * *

3. That the foregoing amendment and restatement was approved by the holders of the requisite number of shares of this corporation in accordance with Section 228 of the General Corporation Law.

4. That this Amended and Restated Certificate of Incorporation, which restates and integrates and further amends the provisions of this corporation's Certificate of Incorporation, has been duly adopted in accordance with Sections 242 and 245 of the General Corporation Law.

[Signature page follows]

IN WITNESS WHEREOF, this Eighth Amended and Restated Certificate of Incorporation has been executed by a duly authorized officer of this corporation on this 10th day of May, 2013.

By: /s/ Joseph Gardner
Joseph Gardner
President and Chief Executive Officer

[Signature Page to Eighth Amended and Restated
Certificate of Incorporation]

AKEBIA THERAPEUTICS, INC.

BYLAWS

Adopted as of April 3, 2007

**Amended pursuant to that certain Joint Action
by Unanimous Written Consent
of the Board of Directors and the Stockholders
of Akebia Therapeutics, Inc.,
Dated as of January 22, 2008:**

RESOLVED FURTHER, that Article II, Section 3 of the Corporation's Bylaws be, and it hereby is, amended by adding to the beginning of the first sentence, "Except as otherwise provided in the certificate of incorporation of the Corporation, as from time to time in effect,";

AKEBIA THERAPEUTICS, INC.

BYLAWS

ARTICLE I.

MEETINGS OF STOCKHOLDERS

Section 1. Time and Place of Meetings. All meetings of the stockholders of Akebia Therapeutics, Inc. (the “**Corporation**”) for the election of directors or for any other purpose shall be held at such time and place, within or without the State of Delaware, as may be designated by the Board of Directors (the “**Board**”), or by the Chairman of the Board, the President or the Secretary in the absence of a designation by the Board, and stated in the notice of the meeting or in a duly executed waiver of notice thereof.

Section 2. Annual Meeting. An annual meeting of the stockholders shall be held at such date and time as shall be designated from time to time by the Board, at which meeting the stockholders shall elect by a plurality vote the directors to succeed those whose terms expire and shall transact such other business as may properly be brought before the meeting.

Section 3. Special Meetings. Special meetings of the stockholders, for any purpose or purposes, unless otherwise prescribed by law or by the Certificate of Incorporation, may be called by the Board, the Chairman of the Board or the President, and shall be called by the President or the Secretary at the request in writing of stockholders owning a majority in interest of the entire capital stock of the Corporation issued and outstanding and entitled to vote. Such request shall be sent to the President and the Secretary and shall state the purpose or purposes of the proposed meeting.

Section 4. Notice of Meetings. Written notice of every meeting of the stockholders, stating the place, date and hour of the meeting and, in the case of a special meeting, the purpose or purposes for which the meeting is called, shall be given not less than ten nor more than sixty days before the date of the meeting to each stockholder entitled to vote at such meeting, except as otherwise provided herein or by law. When a meeting is adjourned to another place, date or time, written notice need not be given of the adjourned meeting if the place, date and time thereof are announced at the meeting at which the adjournment is taken; provided, however, that if the adjournment is for more than thirty days, or if after the adjournment a new record date is fixed for the adjourned meeting, written notice of the place, date and time of the adjourned meeting shall be given in conformity herewith. At any adjourned meeting, any business may be transacted which might have been transacted at the original meeting.

Section 5. Quorum. The holders of a majority of the stock issued and outstanding and entitled to vote thereat, present in person or represented by proxy, shall constitute a quorum at all meetings of the stockholders for the transaction of business except as otherwise provided by law or by the Certificate of Incorporation. If, however, such quorum shall not be present or represented at any meeting of the stockholders, the stockholders entitled to vote thereat, present in person or represented by proxy, shall have power to adjourn the meeting from time to time, without notice other than announcement at the meeting, until a quorum shall be present or represented.

Section 6. Voting. Except as otherwise provided by law or by the Certificate of Incorporation, each stockholder shall be entitled at every meeting of the stockholders to one vote for each share of stock having voting power standing in the name of such stockholder on the books of the Corporation on the record date for the meeting and such votes may be cast either in person or by written proxy. Every proxy must be duly executed and filed with the Secretary of the Corporation. A stockholder may revoke any proxy which is not irrevocable by attending the meeting and voting in person or by filing an instrument in writing revoking the proxy or another duly executed proxy bearing a later date with the Secretary of the Corporation. The vote upon any question brought before a meeting of the stockholders may be by voice vote, unless the holders of a majority of the outstanding shares of all classes of stock entitled to vote thereon present in person or by proxy at such meeting shall so determine. Every vote taken by written ballot shall be counted by one or more inspectors of election appointed by the Board. When a quorum is present at any meeting, the vote of the holders of a majority of the stock that has voting power present in person or represented by proxy shall decide any question properly brought before such meeting, unless the question is one upon which by express provision of law, the Certificate of Incorporation or these Bylaws, a different vote is required, in which case such express provision shall govern and control the decision of such question.

ARTICLE II.

DIRECTORS

Section 1. Powers. The business and affairs of the Corporation shall be managed by or under the direction of its Board, which may exercise all such powers of the Corporation and do all such lawful acts and things as are not by law or by the Certificate of Incorporation directed or required to be exercised or done by the stockholders.

Section 2. Number and Term of Office. Subject to the provisions of the Certificate of Incorporation, the Board shall consist of one or more members, and the number of directors shall be fixed by resolution of the Board or by the stockholders at the annual meeting or a special meeting. The directors shall be elected at the annual meeting of the stockholders, except as provided in Section 3 of this Article, and each director elected shall hold office until his successor is elected and qualified, except as required by law. Any decrease in the authorized number of directors shall not be effective until the expiration of the term of the directors then in office, unless, at the time of such decrease, there shall be vacancies on the Board which are being eliminated by such decrease.

Section 3. Vacancies and New Directorships. Vacancies and newly created directorships resulting from any increase in the authorized number of directors which occur between annual meetings of the stockholders may be filled by a majority of the directors then in office, though less than a quorum, or by a sole remaining director, and the directors so elected shall hold office until the next annual meeting of the stockholders and until their successors are elected and qualified, except as required by law.

Section 4. Regular Meetings. Regular meetings of the Board may be held without notice immediately after the annual meeting of the stockholders and at such other time and place as shall from time to time be determined by the Board.

Section 5. Special Meetings. Special meetings of the Board may be called by the Chairman of the Board or the President on one day's written notice to each director by whom such notice is not waived, given either personally or by mail, facsimile or electronically, and shall be called by the President or the Secretary in like manner and on like notice on the written request of any two directors.

Section 6. Quorum. At all meetings of the Board, a majority of the total number of directors then in office shall constitute a quorum for the transaction of business, and the act of a majority of the directors present at any meeting at which there is a quorum shall be the act of the Board. If a quorum shall not be present at any meeting of the Board, the directors present thereat may adjourn the meeting from time to time to another place, time or date, without notice other than announcement at the meeting, until a quorum shall be present.

Section 7. Written Action. Any action required or permitted to be taken at any meeting of the Board or of any committee thereof may be taken without a meeting if all members of the Board or committee, as the case may be, consent thereto in writing, and the writing or writings are filed with the minutes or proceedings of the Board or Committee.

Section 8. Participation in Meetings by Conference Telephone. Members of the Board, or any committee designated by the Board, may participate in a meeting of the Board, or any such committee, by means of conference telephone or similar communications equipment by means of which all persons participating in the meeting can hear each other, and such participation in a meeting shall constitute presence in person at the meeting.

Section 9. Committees. The Board may, by resolution passed by a majority of the whole Board, designate one or more committees, each committee to consist of one or more of the directors of the Corporation and each to have such lawfully delegable powers and duties as the Board may confer. Each such committee shall serve at the pleasure of the Board. The Board may designate one or more directors as alternate members of any committee, who may replace any absent or disqualified member at any meeting of the committee. Except as otherwise provided by law, any such committee, to the extent provided in the resolution of the Board, shall have and may exercise all the powers and authority of the Board in the management of the business and affairs of the Corporation, and may authorize the seal of the Corporation to be affixed to all papers which may require it. Any committee or committees so designated by the Board shall have such name or names as may be determined from time to time by resolution adopted by the Board. Unless otherwise prescribed by the Board, a majority of the members of the committee shall constitute a quorum for the transaction of business, and the act of a majority of the members present at a meeting at which there is a quorum shall be the act of such committee. Each committee shall prescribe its own rules for calling and holding meetings and its method of procedure, subject to any rules prescribed by the Board, and shall keep a written record of all actions taken by it.

Section 10. Compensation. The Board may establish such compensation for, and reimbursement of the expenses of, directors for attendance at meetings of the Board or committees, or for other services by directors to the Corporation, as the Board may determine.

Section 11. Rules. The Board may adopt such special rules and regulations for the conduct of their meetings and the management of the affairs of the Corporation as they may deem proper, not inconsistent with law or these Bylaws.

ARTICLE III.

NOTICES

Section 1. Generally. Whenever by law or under the provisions of the Certificate of Incorporation or these Bylaws, notice is required to be given to any director or stockholder, it shall not be construed to mean personal notice, but such notice may be given in writing, by mail, addressed to such director or stockholder, at his address as it appears on the records of the Corporation, with postage thereon prepaid, and such notice shall be deemed to be given at the time when the same shall be deposited in the United States mail. Notice to directors may also be given by facsimile, electronic mail or telephone.

Section 2. Waivers. Whenever any notice is required to be given by law or under the provisions of the Certificate of Incorporation or these Bylaws, a waiver thereof in writing, signed by the person or persons entitled to such notice, whether before or after the time of the event for which notice is to be given, shall be deemed equivalent to such notice. Attendance of a person at a meeting shall constitute a waiver of notice of such meeting, except when the person attends a meeting for the express purpose of objecting, at the beginning of the meeting, to the transaction of any business because the meeting is not lawfully called or convened.

ARTICLE IV.

OFFICERS

Section 1. Generally. The officers of the Corporation shall be elected by the Board and shall consist of a President, a Secretary and a Treasurer. The Board may also elect such other officers as the Board deems desirable, including, but not limited to, any or all of the following: a Chairman of the Board, one or more Vice Presidents, a Controller, a General Counsel, and one or more Assistant Secretaries and Assistant Treasurers. Any number of offices may be held by the same person.

Section 2. Compensation. The compensation of all officers and agents of the Corporation who are also directors of the Corporation shall be fixed by the Board. The Board may delegate the power to fix the compensation of other officers and agents of the Corporation to an officer of the Corporation.

Section 3. Succession. The officers of the Corporation shall hold office until their successors are elected and qualified. Any officer elected or appointed by the Board may be removed at any time by the affirmative vote of a majority of the directors. Any vacancy occurring in any office of the Corporation may be filled by the Board.

Section 4. Authority and Duties. Each of the officers of the Corporation shall have such authority and shall perform such duties as are customarily incident to their respective offices, or as may be specified from time to time by the Board in a resolution which is not inconsistent with these Bylaws.

Section 5. Execution of Documents and Action with Respect to Securities of Other Corporations. The President shall have and is hereby given, full power and authority, except as otherwise required by law or directed by the Board, (a) to execute, on behalf of the Corporation, all duly authorized contracts, agreements, deeds, conveyances or other obligations of the Corporation, applications, consents, proxies and other powers of attorney, and other documents and instruments, and (b) to vote and otherwise act on behalf of the Corporation, in person or by proxy, at any meeting of stockholders, members, partners or other equity holders (or with respect to any action of such stockholders, members, partners or other equity holders) of any other corporation, limited liability company, partnership or other entity in which the Corporation may hold securities and otherwise to exercise any and all rights and powers which the Corporation may possess by reason of its ownership of securities. In addition, the President may delegate to other officers, employees and agents of the Corporation the power and authority to take any action which the President is authorized to take under this Section 5, with such limitations as the President may specify; such authority so delegated by the President shall not be re-delegated by the person to whom such execution authority has been delegated.

ARTICLE V.

STOCK

Section 1. Certificates. Certificates representing shares of stock of the Corporation shall be in such form as shall be determined by the Board, subject to applicable legal requirements. Such certificates shall be numbered and their issuance recorded in the books of the Corporation, and such certificate shall exhibit the holder's name and the number of shares and shall be signed by, or in the name of the Corporation by the Chairman of the Board or the President or Vice President and the Secretary or an Assistant Secretary or the Treasurer or an Assistant Treasurer of the Corporation. Any or all of the signatures upon such certificates may be facsimiles, engraved or printed.

Section 2. Transfer. Upon surrender to the Corporation or the transfer agent of the Corporation of a certificate for shares duly endorsed or accompanied by proper evidence of succession, assignment or authority to transfer, it shall be the duty of the Corporation to issue, or to cause its transfer agent to issue, a new certificate to the person entitled thereto, cancel the old certificate and record the transaction upon its books.

Section 3. Lost, Stolen or Destroyed Certificates. The Secretary may direct a new certificate or certificates to be issued in place of any certificate or certificates theretofore issued by the Corporation alleged to have been lost, stolen or destroyed upon the making of an affidavit of that fact, satisfactory to the Secretary, by the person claiming the certificate of stock to be lost, stolen or destroyed. As a condition precedent to the issuance of a new certificate or certificates the Secretary may require the owner of such lost, stolen or destroyed certificate or certificates to give the Corporation a bond in such sum and with such surety or sureties as the Secretary may direct as indemnity against any claims that may be made against the Corporation with respect to the certificate alleged to have been lost, stolen or destroyed or the issuance of the new certificate.

Section 4. Record Date.

(a) In order that the Corporation is able to determine the stockholders entitled to notice of or to vote at any meeting of stockholders or any adjournment thereof, the Board may fix a record date, which record date shall not precede the date upon which the resolution fixing the record date is adopted by the Board and which record date shall not be more than sixty nor less than ten days before the date of such meeting. If no record is fixed by the Board, the record date for determining stockholders entitled to notice of or to vote at a meeting of stockholders shall be at the close of business on the day next preceding the day on which notice is given, or, if notice is waived, at the close of business on the day next preceding the day on which the meeting is held. A determination of stockholders of record entitled to notice of or to vote at a meeting of stockholders shall apply to any adjournment of the meeting; provided, however, that the Board may fix a new record date for the adjourned meeting.

(b) In order that the Corporation may determine the stockholders entitled to consent to corporate action in writing without a meeting, the Board may fix a record date, which record date shall not precede the date upon which the resolution fixing the record date is adopted by the Board, and which date shall not be more than ten days after the date upon which the resolution fixing the record date is adopted by the Board. If no record date has been fixed by the Board, the record date for determining stockholders entitled to consent to corporate action in writing without a meeting, when no prior action by the Board is required, shall be the first date on which a signed written consent setting forth the action taken or proposed to be taken is delivered to the Corporation by delivery to its registered office in Delaware, its principal place of business, or an officer or agent of the Corporation having custody of the book in which proceedings of meetings of stockholders are recorded. Delivery made to a Corporation's registered office shall be by hand or by certified or registered mail, return receipt requested. If no record date has been fixed by the Board and prior action by the Board is required by law, the record date for determining stockholders entitled to consent to corporate action in writing without a meeting shall be at the close of business on the day on which the Board adopts the resolution taking such prior action.

(c) In order that the Corporation may determine the stockholders entitled to receive payment of any dividend or other distribution or allotment of any rights or the stockholders entitled to exercise any rights in respect of any change, conversion or exchange of stock, or for the purpose of any other lawful action, the Board may fix a record date, which record date shall not precede the date upon which the resolution fixing the record date is adopted, and which record date shall be not more than sixty days prior to such action. If no record date is fixed, the record date for determining stockholders for any such purpose shall be at the close of business on the day on which the Board adopts the resolution relating thereto.

ARTICLE VI.

GENERAL PROVISIONS

Section 1. Fiscal Year. The fiscal year of the Corporation shall be fixed from time to time by the Board.

Section 2. Corporate Seal. The Board may adopt a corporate seal and use the same by causing it or a facsimile thereof to be impressed or affixed or reproduced or otherwise.

Section 3. Reliance upon Books, Reports and Records. Each director, each member of a committee designated by the Board, and each officer of the Corporation will, in the performance of his or her duties, be fully protected in relying in good faith upon the records of the Corporation and upon such information, opinions, reports or statements presented to the Corporation by any of the Corporation's officers or employees, or committees of the Board, or by any other person as to matters the director, committee member or officer believes are within such other person's professional or expert competence and who has been selected with reasonable care by or on behalf of the Corporation.

Section 4. Time Periods. In applying any provision of these Bylaws which requires that an act be done or not be done a specified number of days prior to an event or that an act be done during a period of a specified number of days prior to an event, calendar days shall be used, the day of the doing of the act shall be excluded and the day of the event shall be included.

Section 5. Dividends. The Board may from time to time declare and the Corporation may pay dividends upon its outstanding shares of capital stock, in the manner and upon the terms and conditions provided by law and the Certificate of Incorporation.

ARTICLE VII.

AMENDMENTS

Section 1. Amendments. Subject to any additional vote required by the Certificate of Incorporation, these Bylaws may be repealed, altered, amended and rescinded, or new Bylaws may be adopted, by the stockholders or by the Board.

**THIRD AMENDED AND RESTATED
VOTING AGREEMENT**

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Schedule A - Investors

Schedule B - Key Holders

Exhibit A - Adoption Agreement

THIRD AMENDED AND RESTATED VOTING AGREEMENT

THIS THIRD AMENDED AND RESTATED VOTING AGREEMENT is made and entered into as of this 10th day of May, 2013 by and among AKEBIA THERAPEUTICS, INC., a Delaware corporation (the "Company"), each holder of the Company's Series C Preferred Stock, \$0.00001 par value per share ("Series C Preferred Stock"), the Company's Series B Preferred Stock, \$0.00001 par value per share ("Series B Preferred Stock") and Series A Preferred Stock, \$0.00001 par value per share ("Series A Preferred Stock") listed on Schedule A (together with any subsequent investors, or transferees, who become parties hereto as "Investors" pursuant to Sections 6.1 or 6.2 below, the "Investors") and those certain stockholders of the Company listed on Schedule B (together with any subsequent stockholders or option holders, or any transferees, who become parties hereto as "Key Holders" pursuant to Sections 6.1 or 6.2 below, the "Key Holders", and together collectively with the Investors, the "Stockholders").

RECITALS

WHEREAS, certain of the Investors have previously purchased equity securities of the Company;

WHEREAS, the Company and certain of the Investors are parties to the Series C Preferred Stock Purchase Agreement, dated as of the date hereof (as it may be amended, restated, supplemented or otherwise modified from time to time, the "Series C Purchase Agreement");

WHEREAS, the Company, certain of the Investors, the Key Holders and certain other Persons (as defined below) previously entered into a Voting Agreement, dated as of January 23, 2008, as amended and restated as of July 15, 2009 and April 6, 2011 (the "Prior Agreement");

WHEREAS, the Company and certain of the Investors are parties to the Series B Preferred Stock Purchase Agreement, dated as of April 6, 2011;

WHEREAS, the Company and certain Investors entered into Series A Preferred Stock Purchase Agreements dated January 23, 2008 and July 15, 2009 in connection with the purchase of shares of Series A Preferred Stock;

WHEREAS, the Company and its existing Investors desire to induce the Investors to purchase shares of Series C Preferred Stock of the Company, pursuant to the Series C Purchase Agreement, by amending and restating the Prior Agreement to provide the Investors with the rights and privileges as set forth herein; and

WHEREAS, the Eighth Amended and Restated Certificate of Incorporation of the Company (the "Restated Certificate") provides that (a) the holders of record of the shares of the Company's Series A Preferred Stock, Series B Preferred Stock and Series C Preferred Stock (collectively, "Preferred Stock"), exclusively and together as a single class (separate from other classes), voting on a pari passu basis, shall be entitled to elect six (6) directors of the Company (the "Preferred Directors"), and (b) the holders of shares of Common Stock (including, on an as-converted to Common Stock basis, all shares of Preferred Stock convertible into Common Stock) shall be entitled to elect three (3) directors of the Company (the "Common Directors").

For purposes of this Agreement, "Common Stock" means the Common Stock of the Company, par value \$0.00001 per share.

NOW, THEREFORE, the parties agree as follows:

1. Voting Provisions Regarding Board of Directors.

1.1. Size of the Board. Each Stockholder agrees to vote, or cause to be voted, all Shares (as defined below) owned by such Stockholder, or over which such Stockholder has voting control, from time to time and at all times, in whatever manner as shall be necessary to ensure that the size of the Board shall be set and remain at nine (9) directors (subject to reduction for (i) the vacancy described in Section 1.2(e) below should Mr. Nash no longer be a Board member and (ii) any vacancies for Board seats not ultimately filled pursuant to Section 1.2(d), below) and may be increased only with the written consent of holders of more than the Appropriate Percentage (as defined below) of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Preferred Stock. For purposes of this Agreement, the term "Shares" shall mean and include any securities of the Company the holders of which are entitled to vote for members of the Board, including without limitation, all shares of Common Stock and Preferred Stock, by whatever name called, now owned or subsequently acquired by a Stockholder, however acquired, whether through stock splits, stock dividends, reclassifications, recapitalizations, similar events or otherwise. For the purposes of this Agreement, the term "Appropriate Percentage" means fifty percent (50%).

1.2. Board Composition. Each Stockholder agrees to vote, or cause to be voted, all Shares owned by such Stockholder, or over which such Stockholder has voting control, from time to time and at all times, in whatever manner as shall be necessary to ensure that at each annual or special meeting of stockholders at which an election of directors is held or pursuant to any written consent of the stockholders, the following persons shall be elected to the Board:

(a) As a Preferred Director, one person designated by Satter Investment Management, LLC, which individual shall initially be Muneer A. Satter.

(b) As Preferred Directors, two persons designated by Novartis Bioventures Ltd., which individuals shall initially be Campbell Murray and Giovanni Ferrara.

(c) As a Preferred Director, one person designated by Kearny Venture Partners, which individual shall initially be Anupam Dalal.

(d) If an Investor that (together with its Affiliates) does not purchase shares of Series C Preferred Stock at Closing 1 purchases at least \$5,000,000 of Series C Preferred Stock at an Additional Closing (as defined in the Series C Purchase Agreement), such Investor may designate (i) one Preferred Director if such investment is at least \$5,000,000, but less than \$8,400,000 and (ii) two Preferred Directors if such investment is at least \$8,400,000 (each, a "New Investor Director"); provided that the Board seats reserved for such New Investor Directors shall remain vacant unless and until such New Investor Directors are designated pursuant to the terms of this Section 1.2(d).

(e) As Common Directors, two persons, one of whom shall be an independent outside director, to be designated by a majority of the Board members, and the other who shall be Duane Nash; provided that the Board seat reserved for Mr. Nash shall not be filled at any time after the date hereof on which a majority of the Board or Mr. Nash determines that Mr. Nash shall not serve on the Board or his earlier death or disability, and each of the Stockholders shall promptly vote their respective Shares to remove Mr. Nash from the Board upon any such occurrence.

(f) As a Common Director, the Company's Chief Executive Officer, who shall initially be Joseph Gardner (the "CEO Director"), provided that if for any reason the CEO Director shall cease to serve as the Chief Executive Officer of the Company, each of the Stockholders shall promptly vote their respective Shares (i) to remove the former Chief Executive Officer from the Board if such person has not resigned as a member of the Board and (ii) to elect such person's replacement as Chief Executive Officer of the Company as the new CEO Director.

Except with respect to clauses (d) and (e) above, to the extent that any of clauses (a) through (f) above shall not be applicable, any member of the Board who would otherwise have been designated in accordance with the terms thereof shall instead be voted upon by all the stockholders of the Company entitled to vote thereon in accordance with, and pursuant to, the Company's Restated Certificate.

For purposes of this Agreement, an individual, firm, corporation, partnership, association, limited liability company, trust or any other entity (collectively, a "Person") shall be deemed an "Affiliate" of another Person who, directly or indirectly, controls, is controlled by or is under common control with such Person, including, without limitation, any general partner, managing member, officer or director of such Person or any venture capital fund now or hereafter existing that is controlled by one or more general partners or managing members of, or shares the same management company with, such Person, and provided that Satter Investment Management, LLC, Muneer A. Satter and all Persons for which Mr. Satter or any of his immediate family members serves as trustee or investment advisor or any similar capacity (and their respective Affiliates) and any account held for the benefit of any such Person shall be Affiliates of one another, regardless of whether they would otherwise be deemed Affiliates hereunder.

1.3. Failure to Designate a Board Member. In the absence of any designation from the Persons or groups with the right to designate a director as specified above, the director previously designated by them and then serving shall be reelected if still eligible to serve as provided herein.

1.4. Removal of Board Members. Each Stockholder also agrees to vote, or cause to be voted, all Shares owned by such Stockholder, or over which such Stockholder has voting control, from time to time and at all times, in whatever manner as shall be necessary to ensure that:

(a) no director elected pursuant to Sections 1.2 or 1.3 of this Agreement may be removed from office unless (i) such removal is directed or approved by the affirmative vote of the Person entitled under Section 1.2 to designate that director or (ii) the Persons originally entitled to designate or approve such director or occupy such Board seat pursuant to Section 1.2 is no longer so entitled to designate or approve such director or occupy such Board seat;

(b) any vacancies created by the resignation, removal or death of a director elected pursuant to Sections 1.2 or 1.3 shall be filled (or, with respect to Mr. Nash and the New Investor Directors, result in a reduction in the authorized size of the Board as described in Section 1.1 above) pursuant to the provisions of this Section 1; and

(c) upon the request of any party entitled to designate a director as provided in Section 1.2(a) through (f) to remove such director, such director shall be removed.

All Stockholders agree to execute any written consents required to perform the obligations of this Agreement, and the Company agrees at the request of any party entitled to designate directors to call a special meeting of stockholders for the purpose of electing directors.

1.5. No Liability for Election of Recommended Directors. No Stockholder, nor any Affiliate of any Stockholder, shall have any liability as a result of designating a person for election as a director for any act or omission by such designated person in his or her capacity as a director of the Company, nor shall any Stockholder have any liability as a result of voting for any such designee in accordance with the provisions of this Agreement.

1.6. Co-Chairs and Committee Rights. Each Stockholder agrees to vote or cause to be voted all Shares owned by such Stockholder, or over which such Stockholder has voting control, from time to time and at all times, in whatever manner as shall be necessary to cause and the Company agrees to take whatever associated actions are necessary to cause (a) the director elected pursuant to Section 1.2(a) and whichever of the two directors who is elected pursuant to Section 1.2(b) who is thereafter also designated by Novartis Bioventures Ltd. to be elected and maintain the positions of co-Chairmen of the Board (which, for the avoidance of doubt, shall be Board governance positions and not positions as an officer of the Company), and (b) the director elected pursuant to Section 1.2(a) and one of the directors selected by Novartis Bioventures Ltd. pursuant to Section 1.2(b) to have the right to serve on any and all committees of the Board. The Company shall cause management to participate in frequent and regular conference calls with one or both of the co-Chairmen of the Board.

2. Vote to Increase Authorized Common Stock. Each Stockholder agrees to vote or cause to be voted all Shares owned by such Stockholder, or over which such Stockholder has voting control, from time to time and at all times, in whatever manner as shall be necessary to increase the number of authorized shares of Common Stock from time to time to ensure that there will be sufficient shares of Common Stock available for conversion of all of the shares of Preferred Stock outstanding at any given time.

3. Drag-Along Right.

3.1. Definitions. A “Sale of the Company” shall mean either: (a) a transaction or series of related transactions in which a Person, or a group of related Persons, acquires from stockholders of the Company shares representing a majority of the outstanding voting power of the Company (a “Stock Sale”); or (b) a transaction that qualifies as a “Deemed Liquidation Event” as defined in the Restated Certificate.

3.2. Actions to be Taken. In the event that (i) the holders of more than the Appropriate Percentage of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Preferred Stock (the "Selling Investors") and (ii) the Board of Directors approve a Sale of the Company in writing, specifying that this Section 3 shall apply to such transaction, then each Stockholder hereby agrees:

(a) if such transaction requires stockholder approval, with respect to all Shares that such Stockholder owns or over which such Stockholder otherwise exercises voting power, to vote (in person, by proxy or by action by written consent, as applicable) all Shares in favor of, and adopt, such Sale of the Company (together with any related amendment to the Restated Certificate required in order to implement such Sale of the Company) and to vote in opposition to any and all other proposals that could reasonably be expected to delay or impair the ability of the Company to consummate such Sale of the Company;

(b) if such transaction is a Stock Sale, to sell the same proportion of shares of capital stock of the Company beneficially held by such Stockholder as is being sold by the Selling Investors to the Person to whom the Selling Investors propose to sell their Shares, and, except as permitted in Section 3.3 below, on the same terms and conditions as the Selling Investors;

(c) to execute and deliver all related documentation and take such other action in support of the Sale of the Company as shall reasonably be requested by the Company or the Selling Investors in order to carry out the terms and provision of this Section 3, including without limitation executing and delivering instruments of conveyance and transfer, and any purchase agreement, merger agreement, indemnity agreement, escrow agreement, consent, waiver, governmental filing, share certificates duly endorsed for transfer (free and clear of impermissible liens, claims and encumbrances) and any similar or related documents;

(d) not to deposit, and to cause their Affiliates not to deposit, except as provided in this Agreement, any Shares of the Company owned by such party or Affiliate in a voting trust or subject any Shares to any arrangement or agreement with respect to the voting of such Shares, unless specifically requested to do so by the acquiror in connection with the Sale of the Company;

(e) to refrain from exercising any dissenters' rights or rights of appraisal under applicable law at any time with respect to such Sale of the Company; and

(f) if the consideration to be paid in exchange for the Shares pursuant to this Section 3 includes any securities and due receipt thereof by any Stockholder would require under applicable law (x) the registration or qualification of such securities or of any person as a broker or dealer or agent with respect to such securities or (y) the provision to any Stockholder of any information other than such information as a prudent

issuer would generally furnish in an offering made solely to “accredited investors” as defined in Regulation D promulgated under the Securities Act of 1933, as amended, the Company may cause to be paid to any such Stockholder in lieu thereof, against surrender of the Shares which would have otherwise been sold by such Stockholder, an amount in cash equal to the fair value (as determined in good faith by the Company) of the securities which such Stockholder would otherwise receive as of the date of the issuance of such securities in exchange for the Shares.

3.3. Exceptions. Notwithstanding the foregoing, a Stockholder will not be required to comply with Section 3.2 above in connection with any proposed Sale of the Company (the “Proposed Sale”) unless:

(a) [Reserved.]

(b) the Stockholder shall not be liable for the inaccuracy of any representation or warranty made by any other Person in connection with the Proposed Sale, other than the Company (except to the extent that funds may be paid out of an escrow established to cover breach of representations, warranties and covenants of the Company as well as breach by any stockholder of any of identical representations, warranties and covenants provided by all stockholders);

(c) the liability for indemnification, if any, of such Stockholder in the Proposed Sale and for the inaccuracy of any representations and warranties made by the Company in connection with such Proposed Sale, is several and not joint with any other Person (except to the extent that funds may be paid out of an escrow established to cover breach of representations, warranties and covenants of the Company as well as breach by any stockholder of any of identical representations, warranties and covenants provided by all stockholders), and is pro rata in proportion to the amount of consideration paid to such Stockholder in connection with such Proposed Sale (in accordance with the provisions of the Restated Certificate);

(d) liability shall be limited to such Stockholder’s applicable share (determined based on the respective proceeds payable to each Stockholder in connection with such Proposed Sale in accordance with the provisions of the Restated Certificate) of a negotiated aggregate indemnification amount that applies equally to all Stockholders but that in no event exceeds the amount of consideration otherwise payable to such Stockholder in connection with such Proposed Sale, except with respect to claims related to fraud by such Stockholder, the liability for which need not be limited as to such Stockholder;

(e) upon the consummation of the Proposed Sale, (i) each holder of each class or series of the Company’s stock will receive the same form of consideration for their shares of such class or series as is received by other holders in respect of their shares of such same class or series of stock, (ii) each holder of a series of Preferred Stock will receive the same amount of consideration per share of such series of Preferred Stock as is received by other holders in respect of their shares of such same series, (iii) each holder of Common Stock will receive the same amount of consideration per share of

Common Stock as is received by other holders in respect of their shares of Common Stock, and (iv) unless the holders of more than the Appropriate Percentage of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Preferred Stock elect otherwise by written notice given to the Company at least five (5) days prior to the effective date of any such Proposed Sale, the aggregate consideration receivable by all holders of the Preferred Stock and Common Stock shall be allocated among the holders of Preferred Stock and Common Stock on the basis of the relative liquidation preferences to which the holders of each respective series of Preferred Stock and the holders of Common Stock are entitled in a Deemed Liquidation Event (assuming for this purpose that the Proposed Sale is a Deemed Liquidation Event) in accordance with the Company's Certificate of Incorporation in effect immediately prior to the Proposed Sale; and

(f) subject to clause (e) above, requiring the same form of consideration to be available to the holders of any single class or series of capital stock, if any holders of any capital stock of the Company are given an option as to the form and amount of consideration to be received as a result of the Proposed Sale, all holders of such capital stock will be given the same option.

3.4. Restrictions on Sales of Control of the Company. No Stockholder shall be a party to any Stock Sale unless all holders of Preferred Stock are allowed to participate in such transaction and the consideration received pursuant to such transaction is allocated among the parties thereto in the manner specified in the Company's Certificate of Incorporation in effect immediately prior to the Stock Sale (as if such transaction were a Deemed Liquidation Event), unless the holders of more than the Appropriate Percentage of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Preferred Stock elect otherwise by written notice given to the Company at least five (5) days prior to the effective date of any such transaction or series of related transactions.

4. Remedies.

4.1. Covenants of the Company. The Company agrees to use its best efforts, within the requirements of applicable law, to ensure that the rights granted under this Agreement are effective and that the parties enjoy the benefits of this Agreement. Such actions include, without limitation, the use of the Company's best efforts to cause the nomination and election of the directors as provided in this Agreement.

4.2. [Reserved].

4.3. Specific Enforcement. Each party acknowledges and agrees that each party hereto will be irreparably damaged in the event any of the provisions of this Agreement are not performed by the parties in accordance with their specific terms or are otherwise breached. Accordingly, it is agreed that each of the Company and the Stockholders shall be entitled to an injunction to prevent breaches of this Agreement, and to specific enforcement of this Agreement and its terms and provisions in any action instituted in any court of the United States or any state having subject matter jurisdiction.

4.4. Remedies Cumulative. All remedies, either under this Agreement or by law or otherwise afforded to any party, shall be cumulative and not alternative.

5. Term. This Agreement shall be effective as of the date hereof and shall continue in effect until and shall terminate upon the earliest to occur of (a) the consummation of the Company's first underwritten public offering of its Common Stock (other than a registration statement relating either to the sale of securities to employees of the Company pursuant to its stock option, stock purchase or similar plan or an SEC Rule 145 transaction); (b) the consummation of a Sale of the Company and distribution of proceeds to or escrow for the benefit of the Stockholders in accordance with the Restated Certificate, provided that the provisions of Section 3 hereof will continue after the closing of any Sale of the Company to the extent necessary to enforce the provisions of Section 3 with respect to such Sale of the Company; or (c) termination of this Agreement in accordance with Section 6.8 below. The provisions contained in Sections 1.2(a)-(c) shall expire if the Person otherwise entitled to designate or approve such director, and his, her or its Affiliates collectively hold no shares of Preferred Stock.

6. Miscellaneous.

6.1. Additional Parties. In the event that after the date of this Agreement, the Company enters into an agreement with any Person to issue shares of capital stock to such Person, then, the Company shall cause such Person, as a condition precedent to entering into such agreement, to become a party to this Agreement by executing and delivering (i) the Adoption Agreement attached to this Agreement as Exhibit A, or (ii) a counterpart signature page hereto agreeing to be bound by and subject to the terms of this Agreement as a Stockholder and thereafter such person shall be deemed a Stockholder for all purposes under this Agreement.

6.2. Transfers. Each transferee or assignee of any Shares subject to this Agreement shall continue to be subject to the terms hereof, and, as a condition precedent to the Company's recognizing such transfer, each transferee or assignee shall agree in writing to be subject to each of the terms of this Agreement by executing and delivering an Adoption Agreement substantially in the form attached hereto as Exhibit A. Upon the execution and delivery of an Adoption Agreement by any transferee, such transferee shall be deemed to be a party hereto as if such transferee were the transferor and such transferee's signature appeared on the signature pages of this Agreement and shall be deemed to be an Investor and Stockholder, or Key Holder and Stockholder, as applicable. The Company shall not permit the transfer of the Shares subject to this Agreement on its books or issue a new certificate representing any such Shares unless and until such transferee shall have complied with the terms of this Section 6.2. Each certificate representing the Shares subject to this Agreement if issued on or after the date of this Agreement shall be endorsed by the Company with the legend set forth in Section 6.12.

6.3. Successors and Assigns. The terms and conditions of this Agreement shall inure to the benefit of and be binding upon the respective successors and assigns of the parties. Nothing in this Agreement, express or implied, is intended to confer upon any party other than the parties hereto or their respective successors and assigns any rights, remedies, obligations, or liabilities under or by reason of this Agreement, except as expressly provided in this Agreement.

6.4. Governing. This Agreement shall be governed by, and construed in accordance with, the laws of the State of Delaware, regardless of the laws that might otherwise govern under applicable principles of conflicts of law.

6.5. Counterparts; Facsimile or PDE. This Agreement may be executed and delivered by facsimile or PDF signature and in two or more counterparts, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument.

6.6. Titles and Subtitles. The titles and subtitles used in this Agreement are used for convenience only and are not to be considered in construing or interpreting this Agreement.

6.7. Notices. All notices and other communications given or made pursuant to this Agreement shall be in writing and shall be deemed effectively given upon the earlier of actual receipt or: (a) personal delivery to the party to be notified, (b) when sent, if sent by electronic mail or facsimile during normal business hours of the recipient, and if not sent during normal business hours, then on the recipient's next business day, (c) five (5) days after having been sent by registered or certified mail, return receipt requested, postage prepaid, or (d) one (1) business day after the business day of deposit with a nationally recognized overnight courier, freight prepaid, specifying next business day delivery, with written verification of receipt. All communications shall be sent to the respective parties at their address as set forth on Schedule A or Schedule B hereto, or to such email address, facsimile number or address as subsequently modified by written notice given in accordance with this Section 6.7.

If notice is given to the Company, a copy shall also be sent to:

Thompson Hine LLP
312 Walnut Street, 14th Floor
Cincinnati, OH 45202-4089
Attn: David J. Willbrand
david.willbrand@thompsonhine.com

If notice is given to Stockholders, a copy shall also be sent to:

Kirkland & Ellis LLP
300 N. LaSalle St.
Chicago, IL 60654
Attn: Ted H. Zook, P.C.
Roger D. Rhoten
E-mail: ted.zook@kirkland.com
roger.rhoten@kirkland.com

and

Edwards Wildman Palmer LLP
Attn: Albert L. Sokol
111 Huntington Avenue
Boston, MA 02199 USA
asokol@edwardswildman.com

6.8. Consent Required to Amend, Terminate or Waive. This Agreement may be amended or terminated and the observance of any term hereof may be waived (either generally or in a particular instance and either retroactively or prospectively) only by a written instrument executed by (a) the Company; and (b) the holders of more than the Appropriate Percentage of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Preferred Stock. Notwithstanding the foregoing:

(i) this Agreement may not be amended or terminated and the observance of any term of this Agreement may not be waived with respect to any Stockholder without the written consent of such Stockholder unless such amendment, termination or waiver applies to all Stockholders in the same fashion;

(ii) Schedules A and B hereto may be amended by the Company from time to time to add information regarding additional Investors (as defined in the Series B Purchase Agreement) without the consent of the other parties hereto;

(iii) any provision hereof may be waived by the waiving party on such party's own behalf, without the consent of any other party; and

(iv) Section 1.2(a) of this Agreement shall not be amended or waived without the written consent of Satter Investment Management, LLC, Section 1.2(b) of this Agreement shall not be amended or waived without the written consent of Novartis Bioventures Ltd., Section 1.2(c) shall not be amended or waived without the written consent of Kearny Venture Partners, Section 1.2(d) shall not be amended or waived without the written consent of the new Investor (if any) having the right to designate any New Investor Director thereunder, Section 1.2(e) shall not be amended or waived without the written consent of the holders of a majority of the outstanding shares of Common Stock (including, on an as-converted to Common Stock basis, all shares of Preferred Stock convertible into Common Stock) and Section 1.6 shall not be amended or waived without the written consent of Satter Investment Management, LLC and Novartis Bioventures Ltd.

Notwithstanding the foregoing, if any amendment, modification, termination or waiver of this section or any other section or subsection of this Agreement would deprive Satter Investment Management, LLC, Novartis Bioventures Ltd., Kearny Venture Partners or any applicable new Investor (in the case of Section 1.2(d)) of its director designation rights (or of Duane Nash's right to be named to the Board) under Section 1.2, then the written consent of such party or such person (in the case of Mr. Nash) shall be required.

The Company shall give prompt written notice of any amendment, termination or waiver hereunder to any party that did not consent in writing thereto. Any amendment, termination or waiver effected in accordance with this Section 6.8 shall be binding on each party and all of such party's successors and permitted assigns, whether or not any such party, successor or assignee entered into or approved such amendment, termination or waiver.

6.9. Delays or Omissions. No delay or omission to exercise any right, power or remedy accruing to any party under this Agreement, upon any breach or default of any other party under this Agreement, shall impair any such right, power or remedy of such non-breaching or non-defaulting party nor shall it be construed to be a waiver of any such breach or default, or an acquiescence therein, or of or in any similar breach or default thereafter occurring; nor shall any waiver of any single breach or default be deemed a waiver of any other breach or default previously or thereafter occurring. Any waiver, permit, consent or approval of any kind or character on the part of any party of any breach or default under this Agreement, or any waiver on the part of any party of any provisions or conditions of this Agreement, must be in writing and shall be effective only to the extent specifically set forth in such writing. All remedies, either under this Agreement or by law or otherwise afforded to any party, shall be cumulative and not alternative.

6.10. Severability. The invalidity or unenforceability of any provision hereof shall in no way affect the validity or enforceability of any other provision.

6.11. Entire Agreement. This Agreement (including the Exhibits hereto), and the Restated Certificate and the other Transaction Agreements (as defined in the Series C Purchase Agreement) constitute the full and entire understanding and agreement between the parties with respect to the subject matter hereof, and any other written or oral agreement relating to the subject matter hereof existing between the parties is expressly canceled.

6.12. Legend on Share Certificates. Each certificate representing any Shares issued after the date hereof shall be endorsed by the Company with a legend reading substantially as follows:

“THE SHARES EVIDENCED HEREBY ARE SUBJECT TO A VOTING AGREEMENT, AS MAY BE AMENDED FROM TIME TO TIME, (A COPY OF WHICH MAY BE OBTAINED UPON WRITTEN REQUEST FROM THE COMPANY), AND BY ACCEPTING ANY INTEREST IN SUCH SHARES THE PERSON ACCEPTING SUCH INTEREST SHALL BE DEEMED TO AGREE TO AND SHALL BECOME BOUND BY ALL THE PROVISIONS OF THAT VOTING AGREEMENT, INCLUDING CERTAIN RESTRICTIONS ON TRANSFER AND OWNERSHIP SET FORTH THEREIN.”

The Company, by its execution of this Agreement, agrees that it will cause the certificates evidencing the Shares issued after the date hereof to bear the legend required by this Section 6.12 of this Agreement, and it shall supply, free of charge, a copy of this Agreement to any holder of a certificate evidencing Shares upon written request from such holder to the Company at its principal office. The parties to this Agreement do hereby agree that the failure to cause the certificates evidencing the Shares to bear the legend required by this Section 6.12 herein and/or the failure of the Company to supply, free of charge, a copy of this Agreement as provided hereunder shall not affect the validity or enforcement of this Agreement.

6.13. Stock Splits, Stock Dividends, etc. In the event of any issuance of Shares of the Company's voting securities hereafter to any of the Stockholders (including, without limitation, in connection with any stock split, stock dividend, recapitalization, reorganization, or the like), such Shares shall become subject to this Agreement and shall be endorsed with the legend set forth in Section 6.12.

6.14. Manner of Voting. The voting of Shares pursuant to this Agreement may be effected in person, by proxy, by written consent or in any other manner permitted by applicable law.

6.15. Further Assurances. At any time or from time to time after the date hereof, the parties agree to cooperate with each other, and at the request of any other party, to execute and deliver any further instruments or documents and to take all such further action as the other party may reasonably request in order to evidence or effectuate the consummation of the transactions contemplated hereby and to otherwise carry out the intent of the parties hereunder.

6.16. [Reserved]

6.17. [Reserved]

6.18. Aggregation of Stock. All Shares held or acquired by a Stockholder and/or its Affiliates shall be aggregated together for the purpose of determining the availability of any rights under this Agreement, and such Affiliated persons may apportion such rights as among themselves in any manner they deem appropriate.

[Signature Page Follows]

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

AKEBIA THERAPEUTICS, INC.

By: /s/ Joseph H. Gardner

Joseph H. Gardner, Ph.D.

President and Chief Executive Officer

Address:

Suite 420,

9987 Carver Road,

Cincinnati, OH 45242

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

AGECHEM VENTURE FUND L.P.

By: /s/ Louis Lacasse

Name: Louis Lacasse

Title: President

Address:

Attn: Louis Lacasse, President
1 Westmount Square, Suite 800
Montreal, Quebec, Canada
H3Z 2P9

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

ATHENIAN VENTURE PARTNERS III L.P.

By: Athenian III, Ltd
Its: General Partner

By: /s/ Karl O. Elderkin
Name: Karl O. Elderkin
Title: President

AVP OHIO TECHNOLOGY I L.P.

By: AVP Ohio I, Ltd.
Its: General Partner

By: /s/ Karl Elderkin
Name: Karl O. Elderkin
Title: President

Address:

340 West State Street
Unit 29/Suite 137D
Athens OH 45701

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

BLUE CHIP VALIDATION FUND, LTD.

By: Blue Chip Venture Company, LTD
Its: Manager

By: /s/ John McIlwraith
John McIlwraith
Managing Director

Address:

312 Walnut Street
Suite 1120
Cincinnati, OH 45202

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

CINCINNATI CORNERSTONE INVESTORS AKB, LLC

By: /s/ Robert W. Coy, Jr.

Robert W. Coy, Jr.
President

Address:

30 West 3rd Street, 6th Floor
Cincinnati, OH 45202-3559

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

WILLIAM DALY

/s/ William Daly

William Daly

Address:

13 Via Abrazar
San Clemente, CA 92673

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

DIANE H. JANUSZ TRUST

By: /s/ John Janusz

Name: John Janusz

Title: Trustee

JOHN JANUSZ

/s/ John Janusz

John Janusz

Address:

7385 Desert Spring Court

West Chester, OH 45069

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

FAMILY AKEBIA INVESTMENTS LLC

By: /s/ Milton Berlinski

Name: Milton Berlinski

Title: Managing Member

Address:

1185 Park Avenue #11G

New York, NY 10128

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

ALAN FISHMAN

/s/ Alan Fishman

Alan Fishman

Address:

6900 Stonehenge Dr.
Cincinnati, OH 45242-6204

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

FRED SHALWITZ TRUST,
ROBERT SHALWITZ, TRUSTEE

By: /s/ Robert Shalwitz
Name: Robert Shalwitz
Title: Trustee

Address:

2549 Bryden Road
Bexley, OH 43209

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above

GARDNER FAMILY TRUST, JOHN D.
GARDNER TRUSTEE

By: /s/ John D. Gardner
John D. Gardner
Trustee

Address:

111 Pine Court
Bastop, TX 78602

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

GITANA FAMILY TRUST,
ELIZABETH C. ARMITAGE TRUSTEE

By: /s/ Elizabeth C. Armitage
Elizabeth C. Armitage
Trustee

Address:

2207 Upland Place
Cincinnati, OH 45206

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

JOSEPH H. GARDNER

By: /s/ Joseph H. Gardner

Address:

4060 Boomer Road
Cincinnati, OH 45247

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

IAN A. W. HOWES, IRA, STERLING TRUST CUSTODIAN

By: /s/ Ian A. W. Howes

Name: Ian A. W. Howes

Title: Trustee

IAN A. W. HOWES

/s/ Ian A. W. Howes

Ian A. W. Howes

Address:

219 Stratford Drive

Chapel Hill, NC 27516

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

KEARNY VENTURE PARTNERS, L.P.

By: /s/ Anupam Dalal

Name: Kearny Venture Associates, LLC

Title: its General Partner

Address:

Attn: Anupam Dalal

Kearny Venture Associates LLC

88 Kearny Street, Suite 200

San Francisco, CA 94108-5530

KEARNY VENTURE PARTNERS ENTREPRENEURS FUND, L.P.

By: /s/ Anupam Dalal

Name: Kearny Venture Associates, LLC

Title: its General Partner

Address:

Attn: Anupam Dalal

Kearny Venture Associates LLC

88 Kearny Street, Suite 200

San Francisco, CA 94108-5530

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

MCIL WRAITH INVESTMENTS, LLC

By: /s/ John McIlwraith

Name: John McIlwraith

Title: Manager

Address:

Attn: John McIlwraith

7680 Foxgate Lane

Cincinnati, OH 45243

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

MRK INTERNATIONAL, LLC

By: /s/ Richard L. Kiley

Name: Richard L. Kiley

Title: Principal Member

Address:

7800 Tecumseh Trail

Cincinnati, OH 45243

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

NOVARTIS BIOVENTURES LTD.

By: /s/ H.S. Zivi
Name: H. S. Zivi
Title: Deputy Chairman

By: /s/ Rebecca White
Name: Rebecca White
Title: Authorized Signatory

Address:

NOVARTIS BIOVENTURES LTD.
Attn: Henri Simon Zivi
131 Front Street
Hamilton HM 12
Bermuda

But for mail, to:

Novartis BioVentures Ltd.
Attn: Henri Simon Zivi
PO Box HM 2899
Hamilton HM LX Bermuda

And, also send a copy to:

Novartis Venture Fund
Attn: Campbell Murray
Five Cambridge Center, Suite 603
Cambridge, MA 02142

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

KEVIN PETERS

/s/ Kevin Peters

Kevin Peters

Address:

9160 Given Road
Cincinnati, OH 45243

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

QCA FIRST FUND II

By: /s/ John Habbert

Name: John Habbert

Title: Manager

Address:

109 Bentwood Ct.

Cincinnati, OH 45241

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

MUNEER A. SATTER REVOCABLE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

THE SATTER FOUNDATION

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

SATTER FAMILY TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

SATTER CHILDREN'S TRUST I

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

Address:

c/o Satter Investment Management, LLC

676 N. Michigan Avenue, Suite 4000

Chicago, IL 60611

Attn: Muneer A. Satter

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

KRISTEN HAYLER HERTEL REVOCABLE TRUST

By: /s/ Kristen Hayler Hertel

Name: Kristen Hayler Hertel

Title: Trustee

Address:

c/o Satter Investment Management, LLC

676 N. Michigan Avenue, Suite 4000

Chicago, IL 60611

Attn: Muneer A. Satter

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

ANNE-CAROLE WITORT INSURANCE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

ROSE SHEREEN FUQUA INSURANCE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

RABI H. SATTER INSURANCE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

JOHN WOOD TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

Address:

c/o Satter Investment Management, LLC

676 N. Michigan Avenue, Suite 4000

Chicago, IL 60611

Attn: Muneer A. Satter

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

ABDUS SATTER INSURANCE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

GORDON AND BARBARA ANNE HERTEL INSURANCE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

Address:

c/o Satter Investment Management, LLC

676 N. Michigan Avenue, Suite 4000

Chicago, IL 60611

Attn: Muneer A. Satter

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

ROBERT SHALWITZ

By: /s/ Robert Shalwitz

Address:

2549 Bryden Road
Bexley, OH 43209

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

SIGVION FUND I, LP

By: /s/ J.P. Fairbank

J.P. Fairbank
Founding Partner

Address:

806 West Washington Street, Suite 204
Chicago, IL 60607

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

THOMAS WEISEL HEALTHCARE VENTURE PARTNERS, L.P.

By: Thomas Weisel Capital Management LLC
Title: its Managing Member

By: /s/ Anupam Dalal
Name: Anupam Dalal
Title: Managing Director

Address:

88 Kearny Street, 4th Floor
San Francisco, CA 94108

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

TRIATHLON MEDICAL VENTURES FUND, L.P.

By: Triathlon Medical Ventures, LLC
Its: General Partner

By: /s/ John M. Rice
John M. Rice
Managing Partner

Address:

300 E-Business Way
Suite 200
Cincinnati, OH 45241

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

TRI-STATE GROWTH CAPITAL FUND II, L.P.

By: Tri-State Ventures II, LLC
Its: General Partner

By: Fort Washington Investment Advisors, Inc.
Its: Managing Member

By: /s/ Steve Baker
Name: Steve Baker
Title: Managing Director

By: /s/ Maribeth S. Rahe
Name: Maribeth S. Rahe
Title: President and Chief Executive Officer

Address:

303 Broadway, Suite 1200
Cincinnati, OH 45202

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

VENTURE INVESTORS EARLY STAGE FUND IV LIMITED
PARTNERSHIP

By: VIESF IV GP, LLC, its General Partner

By: /s/ Paul M. Weiss

Name: Paul M. Weiss, PhD

Title: Managing Director

Address:

505 South Rosa Road

Madison, WI 53719-1262

Attn: Paul Weiss, Managing Director

Phone: (608) 441-2700

Fax: (608) 441-2727

Email: paul@ventureinvestors.com

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Voting Agreement as of the date first written above.

JOHN H. WYANT

/s/ John H. Wyant

John H. Wyant

Address:

Blue Chip Venture Company
1120 Scripps Center
312 Walnut Street
Cincinnati, OH 45202

SIGNATURE PAGE TO THIRD AMENDED AND RESTATED VOTING AGREEMENT

SCHEDULE A1

INVESTORS

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock</u>	<u>Number of Shares of Series C Preferred Stock</u>
Triathlon Medical Ventures Fund, L.P. Attn: John M. Rice Managing Partner 300 E-Business Way, Suite 200 Cincinnati, OH 45241	20,000.00	128,095.46	124,501.72	118,397.71
Novartis BioVentures Ltd. Attn: Henri Simon Zivi 131 Front Street Hamilton HM 12 Bermuda	0	257,031.16	347,830.73	708,647.43
<i>But for mail, to:</i> Novartis BioVentures Ltd. Attn: Henri Simon Zivi PO Box HM 2899 Hamilton HM LX Bermuda				
<i>And, also send a copy to:</i> Novartis Venture Fund Attn: Campbell Murray Five Cambridge Center, Suite 603 Cambridge, MA 02142				
and Edwards Angell Palmer & Dodge LLP Attn: Al Sokol 111 Huntington Avenue Boston, MA 02199 asokol@eapdlaw.com				

¹ The share amounts set forth in this Schedule A and on Schedule B reflect the effects of the 1-for-100 reverse stock split described in the Restated Certificate.

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock</u>	<u>Number of Shares of Series C Preferred Stock</u>
Venture Investors Early Stage Fund IV Limited Partnership Attn: Paul Weiss Managing Director 505 South Rosa Road Madison, WI 53719-1262	0	153,434.52	173,915.36	201,811.43
Kearny Venture Partners, L.P. Attn: Anupam Dalal Kearny Venture Associates, LLC 88 Kearny Street, Suite 200 San Francisco, CA 94108-5530	0	44,606.15	43,354.71	308,448.29
Kearny Venture Partners Entrepreneurs Fund, L.P. Attn: Anupam Dalal Kearny Venture Associates, LLC 88 Kearny Street, Suite 200 San Francisco, CA 94108-5530	0	909.78	884.26	6,290.57
Thomas Weisel Healthcare Venture Partners, L.P. 88 Kearny Street, 4th Floor San Francisco, CA 94108	0	45,515.99	44,239.03	74,475.86
The Procter & Gamble Company Attn: David Le Neveu Director, Corporate Acquisitions, Divestitures and Equity Ventures 1 Procter & Gamble Plaza Cincinnati, OH 45202	72,047.44	0	8,475.71	0
Athenian Venture Partners III L.P. Attn: Karl O. Elderkin President Athenian III, Ltd. 340 West State Street Unit 29/Suite 137D Athens, OH 45701	0	22,964.77	31,575.85	124,832.79

Schedule A-2

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock</u>	<u>Number of Shares of Series C Preferred Stock</u>
AVP Ohio Technology I L.P. Attn: Karl O. Elderkin President AVP Ohio I, Ltd. 340 West State Street Unit 29/Suite 137D Athens, OH 45701	0	7,654.92	9,004.41	24,194.21
Sigvion Fund I, LP Attn: J. P. Fairbank Founding Partner 738 W. Belden Avenue Chicago, IL 60614	8,000.00	13,660.69	13,277.43	21,402.57
Cincinnati Cornerstone Investors AKB, LLC Attn: Robert W. Coy, Jr. President 30 West 3rd Street, 6th Floor Cincinnati, OH 45202-3559	0	13,122.75	12,754.59	118,391.79
Tri-State Growth Capital Fund II, L.P. Attn: Steve Baker 303 Broadway, Suite 1200 Cincinnati, OH 45202	0	12,804.65	12,445.41	21,488.36
Blue Chip Validation Fund, Ltd. Attn: John McIlwraith Managing Director 1100 Chiquita Center 250 East Fifth Street Cincinnati, OH 45202	0	3,402.09	0	0
QCA First Fund II Attn: John Habbert 1776 Mentor Avenue, MB #302 Cincinnati, OH 45212	0	3,375.00	0	0
Gitana Family Trust, Elizabeth C. Armitage Trustee Attn: Elizabeth C. Armitage Trustee 2207 Upland Place Cincinnati, OH 45206	0	2,765.57	828.17	1,551.57

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock</u>	<u>Number of Shares of Series C Preferred Stock</u>
Robert Shalwitz 2549 Bryden Road Bexley, OH 43209	127,637.41	849.53	2,070.42	2,500.00
Fred Shalwitz Trust, Robert Shalwitz, Trustee Attn: Robert Shalwitz Trustee 2549 Bryden Road Bexley, OH 43209	0	3,403.13	0	0
Joseph H. Gardner 4060 Boomer Road Cincinnati, OH 45247	161,423.79	9,038.87	11,594.35	26,383.21
Gardner Family Trust, John D. Gardner Trustee Attn: John D. Gardner Trustee 111 Pine Court Bastop, TX 78602	0	6,901.99	7,288.07	2,485.00
Ian A. W. Howes, IRA, Sterling Trust Custodian Attn: Ian A. W. Howes Trustee 219 Stratford Drive Chapel Hill, NC 27516	0	5,000.00	5,797.18	7,142.00
Ian A. W. Howes 219 Stratford Drive Chapel Hill, NC 27516	46,925.51	0	0	2,902.21
Kevin Peters 9160 Given Road Cincinnati, OH 45243	46,925.51	0	1,449.29	3,473.00
William Daly 13 Via Abrazar San Clemente, CA 92673	60,810.00	0	0	13,488.86

Schedule A-4

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock</u>	<u>Number of Shares of Series C Preferred Stock</u>
Muneer A. Satter Revocable Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	115,943.58	182,551.50
John Wood Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	3,571.00
The Satter Foundation c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	57,971.78	191,275.86
Muneer A Satter IRA, Millennium Trust Company, Custodian	0	0	0	14,285.00
Satter Children's Trust I c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	57,971.78	66,275.86
Kristen Hayler Hertel Revocable Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	7,142.00
Anne-Carole Witort Insurance Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	7,142.00

Schedule A-5

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock</u>	<u>Number of Shares of Series C Preferred Stock</u>
Rose Shereen Fuqua Insurance Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	7,142.00
Rabi H. Satter Insurance Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	7,142.00
Abdus Satter Insurance Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	3,571.00
Gordon and Barbara Anne Hertel Insurance Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	3,571.00
Satter Family Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	28,985.89	31,351.93
AgeChem Venture Fund L.P. Attn: Louis Lacasse President 1 Westmount Square, Suite 800 Montreal, Quebec, Canada H3Z 2P9	0	0	173,915.35	123,143.86

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock</u>	<u>Number of Shares of Series C Preferred Stock</u>
Diane H. Janusz Trust Attn: John Janusz Trustee 7385 Desert Spring Court West Chester, OH 45069	0	0	1,449.29	139.00
John Janusz 7385 Desert Spring Court West Chester, OH 45069	0	0	0	1,670.79
MRK International, LLC Attn: Richard L. Kiley Principal Member 7800 Tecumseh Trail Cincinnati, OH 45243	0	0	0	396.00
McIlwraith Investments, LLC Attn: John McIlwraith Manager 7680 Foxgate Lane Cincinnati, OH 45243	0	0	0	4,597.00
John H. Wyant Blue Chip Venture Company 1120 Scripps Center 312 Walnut Street Cincinnati, OH 45202	0	0	0	1,039.36
Alan Fishman 6900 Stonehenge Drive Cincinnati, OH 45242-6204	0	0	0	1,428.00
Family Akebia Investments LLC Attn: Milton Berlinski 1185 Park Avenue #11G New York, NY 10128	0	0	0	142,858.00
Novo A/S Novo Ventures Tuborg Havnevej 19 DK - 2900 - Hellerup	0	0	0	714,285.00

Schedule A-7

SCHEDULE B

KEY HOLDERS
Name and Address

The Procter & Gamble Company
One Procter & Gamble Plaza
Cincinnati, OH 45202

Number of Shares Held

7,204,744 shares of Common Stock

Schedule B-1

EXHIBIT A
ADOPTION AGREEMENT

This Adoption Agreement (“Adoption Agreement”) is executed on _____, 20____, by the undersigned (the “Holder”) pursuant to the terms of that certain Voting Agreement dated as of May [], 2013 (the “Agreement”), by and among the Company and certain of its Investors, as such Agreement may be amended or amended and restated hereafter. Capitalized terms used but not defined in this Adoption Agreement shall have the respective meanings ascribed to such terms in the Agreement. By the execution of this Adoption Agreement, the Holder agrees as follows.

1.1 Acknowledgement. Holder acknowledges that Holder is acquiring certain shares of the capital stock of the Company (the “Stock”)[or options, warrants or other rights to purchase such Stock (the “Options”)], for one of the following reasons (Check the correct box):

- as a transferee of Shares from a party in such party’s capacity as an “Investor” bound by the Agreement, and after such transfer, Holder shall be considered an “Investor” and a “Stockholder” for all purposes of the Agreement.
- as a new Investor in accordance with Section 6.1 of the Agreement, in which case Holder will be an “Investor” and a “Stockholder” for all purposes of the Agreement.

1.2 Agreement. Holder hereby (a) agrees that the Stock [Options], and any other shares of capital stock or securities required by the Agreement to be bound thereby, shall be bound by and subject to the terms of the Agreement and (b) adopts the Agreement with the same force and effect as if Holder were originally a party thereto.

1.3 Notice. Any notice required or permitted by the Agreement shall be given to Holder at the address or facsimile number listed below Holder’s signature hereto.

HOLDER: _____

ACCEPTED AND AGREED:

By: _____
Name and Title of Signatory

AKEBIA THERAPEUTICS, INC.

Address: _____

By: _____
Title: _____

Facsimile Number: _____

**AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT**

THIS AMENDMENT NO. 1 TO THE THIRD AMENDED AND RESTATED VOTING AGREEMENT (this "Amendment") is made and effective as of May 31, 2013, by and among Akebia Therapeutics, Inc., a Delaware corporation (the "Company"), and the "Stockholders" as defined in that certain Third Amended and Restated Voting Agreement, dated as of May 10, 2013, by and among the Company and the Stockholders a party thereto (the "Agreement"). Capitalized terms not defined herein have the meanings given them in the Agreement.

RECITALS

WHEREAS, on May 10, 2013, the Agreement was entered into by the Company and the Investors in connection with the transactions and agreements contemplated in that certain Series C Preferred Stock Purchase Agreement by and between the Company and certain parties as listed therein, dated as of May 10, 2013 (the "Series C Stock Purchase Agreement") and the transactions and agreements contemplated therein collectively the "Series C Transaction"; and

WHEREAS, in connection with an Additional Closing (as defined in the Series C Stock Purchase Agreement) to the Series C Transaction dated May 31, 2013, the Company and the Stockholders holding more than the Appropriate Percentage (as defined in the Agreement) of the shares of the Company's Common Stock issuable upon conversion of the outstanding shares of the Company's Preferred Stock (as defined in the Agreement) desire to amend the Agreement in accordance with the terms and conditions more fully set forth in this Amendment.

NOW, THEREFORE, in consideration of the foregoing premises and the mutual covenants hereinafter set forth, and other good and valuable consideration had and received, the parties hereto, upon the terms and subject to the conditions contained herein, hereby agree as follows:

1. Amendments.

(a) Section 1.2(d) is hereby amended by deleting it in its entirety and replacing it with the following: "(d) As Preferred Directors, two persons designated by Novo A/S (each, a "New Investor Director")."

(b) Section 6.8(i) is hereby amended by deleting it in its entirety and replacing it with the following:

(i) this Agreement may not be amended or terminated and the observance of any term of this Agreement may not be waived with respect to any Stockholder without the written consent of such Stockholder unless such amendment, termination or waiver applies to all Stockholders in the same fashion, and provided that (the following provisions of this Section 6.8(i) proviso, the "Liability Proviso") any amendment of Sections 3.3(c) and 3.3(d) hereof, as well as any amendment of this Liability Proviso, shall require, in addition to any other approvals required by this Section 6.8, also the approval of (A) Satter Investment

Management, LLC so long as Satter Investment Management, LLC or any of its Affiliates holds any Preferred Stock or Common Stock issued upon conversion of any of such Preferred Stock, (B) Novartis Bioventures Ltd. so long as Novartis Bioventures Ltd. holds any Preferred Stock or Common Stock issued upon conversion of any of such Preferred Stock, and (C) Novo A/S so long as Novo A/S holds any Preferred Stock or Common Stock issued upon conversion of any of such Preferred Stock;

(c) Section 6.8 (iv) is hereby amended by deleting it in its entirety and replacing it with the following:

(iv) Section 1.2(a) of this Agreement shall not be amended or waived without the written consent of Satter Investment Management, LLC, Section 1.2(b) of this Agreement shall not be amended or waived without the written consent of Novartis Bioventures Ltd., Section 1.2(c) shall not be amended or waived without the written consent of Kearny Venture Partners, Section 1.2(d) shall not be amended or waived without the written consent of Novo A/S, Section 1.2(e) shall not be amended or waived without the written consent of the holders of a majority of the outstanding shares of Common Stock (including, on an as-converted to Common Stock basis, all shares of Preferred Stock convertible into Common Stock) and Section 1.6 shall not be amended or waived without the written consent of Satter Investment Management, LLC and Novartis Bioventures Ltd. Notwithstanding the foregoing, if any amendment, modification, termination or waiver of this section or any other section or subsection of this Agreement would deprive Satter Investment Management, LLC, Novartis Bioventures Ltd., Kearny Venture Partners, or Novo A/S of its director designation rights (or of Duane Nash's right to be named to the Board) under Section 1.2, then the written consent of such party or such person (in the case of Mr. Nash) shall be required.

2. Miscellaneous Amendments. The Agreement is amended hereby so that any reference therein to the Agreement shall be deemed to be a reference to the Agreement as amended by this Amendment.

3. Continuance of Agreement. Except as specifically amended by this Amendment, the Agreement shall remain in full force and effect.

4. Governing Law. The laws of the State of Delaware govern all matters arising out of or relating to this Amendment, including, without limitation, its interpretation, construction, performance, and enforcement, without giving effect to such state's conflicts of law principles or rules of construction concerning the drafter hereof.

5. Counterparts. This Amendment may be executed in two or more counterparts, including by facsimile or PDF signature, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument.

[Signature Page Follows]

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

AKEBIA THERAPEUTICS, INC.

By: /s/ Joseph H. Gardner

Joseph H. Gardner, Ph.D.

President and Chief Executive Officer

Address:

9987 Carver Road, Suite 420

Cincinnati, OH 45242

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

NOVARTIS BIOVENTURES LTD.

By: /s/ Simon Zivi

Name: Simon Zivi

Title: Deputy Chairman

By: /s/ Rebecca White

Name: Rebecca White

Title: Authorized Signatory

Address:

NOVARTIS BIOVENTURES LTD.

Attn: Henri Simon Zivi

131 Front Street

Hamilton HM 12

Bermuda

But for mail, to:

Novartis BioVentures Ltd.

Attn: Henri Simon Zivi

PO Box HM 2899

Hamilton HM LX Bermuda

And, also send a copy to:

Novartis Venture Fund

Attn: Campbell Murray

Five Cambridge Center, Suite 603

Cambridge, MA 02142

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

VENTURE INVESTORS EARLY STAGE FUND IV LIMITED
PARTNERSHIP

By: VIESF IV GP, LLC, its General Partner

By: /s/ Paul M. Weiss

Name: Paul M. Weiss, PhD

Title: Managing Director

Address:

505 South Rosa Road

Madison, WI 53719-1262

Attn: Paul Weiss, Managing Director

Phone: (608) 441-2700

Fax: (608) 441-2727

Email: paul@ventureinvestors.com

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

TRIATHLON MEDICAL VENTURES FUND, L.P.

By: Triathlon Medical Ventures, LLC
Its: General Partner

By: /s/ John M. Rice
John M. Rice
Managing Partner

Address:

300 E-Business Way
Suite 200
Cincinnati, OH 45241

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

KEARNY VENTURE PARTNERS, L.P.

By: /s/ Anupam Dalal

Name: Kearny Venture Associates, LLC

Title: its General Partner

Address:

Attn: Anupam Dalal
Kearny Venture Associates LLC
88 Kearny Street, Suite 1800
San Francisco, CA 94108-5530

KEARNY VENTURE PARTNERS ENTREPRENEURS FUND, L.P.

By: /s/ Anupam Dalal

Name: Kearny Venture Associates, LLC

Title: its General Partner

Address:

Attn: Anupam Dalal
Kearny Venture Associates LLC
88 Kearny Street, Suite 1800
San Francisco, CA 94108-5530

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

THOMAS WEISEL HEALTHCARE VENTURE PARTNERS, L.P.

By: Thomas Weisel Capital Management LLC
Title: its Managing Member

By: /s/ Anupam Dalal
Name: Anupam Dalal
Title: Managing Director

Address:

88 Kearny Street, Suite 1800
San Francisco, CA 94108

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

ATHENIAN VENTURE PARTNERS III L.P.

By: Athenian III, Ltd
Its: General Partner

By: /s/ Karl O. Elderkin
Name: Karl O. Elderkin
Title: President

AVP OHIO TECHNOLOGY I L.P.

By: AVP Ohio I, Ltd.
Its: General Partner

By: /s/ Karl O. Elderkin
Name: Karl O. Elderkin
Title: President

Address:

340 West State Street
Unit 29/Suite 137D
Athens OH 45701

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above,

AGECHEM VENTURE FUND L.P.

By: /s/ Louis Lacasse

Name: Louis Lacasse

Title: President

Address:

Attn: Louis Lacasse, President

1 Westmount Square, Suite 800

Montreal, Quebec, Canada

H3Z 2P9

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

CINCINNATI CORNERSTONE INVESTORS
AKB, LLC

By: /s/ Robert W. Coy, Jr.

Robert W. Coy
President

Address:

30 West 3rd Street, 6th Floor
Cincinnati, OH 45202-3559

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

FAMILY AKEBIA INVESTMENTS LLC

By: /s/ Milton Berlinski

Name: Milton Berlinski

Title: Managing Member

Address:

1185 Park Avenue #11G

New York, NY 10128

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

MUNEER A. SATTER REVOCABLE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

THE SATTER FOUNDATION

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

SATTER FAMILY TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

SATTER CHILDREN'S TRUST I

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

Address:

c/o Satter Investment Management, LLC

676 N. Michigan Avenue, Suite 4000

Chicago, IL 60611

Attn: Muneer A. Satter

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

KRISTEN HAYLER HERTEL REVOCABLE TRUST

By: /s/ Kristen Hayler Hertel

Name: Kristen Hayler Hertel

Title: Trustee

Address:

c/o Satter Investment Management, LLC 676 N. Michigan Avenue,
Suite 4000 Chicago, IL 60611 Attn: Muncer A. Satter

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

ANNE-CAROLE WITORT INSURANCE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

ROSE SHEREEN FUQUA INSURANCE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

RABI H. SATTER INSURANCE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

JOHN WOOD TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

Address:

c/o Satter Investment Management, LLC

676 N. Michigan Avenue, Suite 4000

Chicago, IL 60611

Attn: Muneer A. Satter

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

ABDUS SATTER INSURANCE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

GORDON AND BARBARA ANNE HERTEL INSURANCE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

Address:

c/o Satter Investment Management, LLC

676 N. Michigan Avenue, Suite 4000

Chicago, IL 60611

Attn: Muneer A. Satter

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED VOTING AGREEMENT

**THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT**

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Schedule A - Schedule of Investors

THIRD AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT

THIS THIRD AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT (this "Agreement") is made as of the 10th day of May, 2013 by and among AKEBIA THERAPEUTICS, INC., a Delaware corporation (the "Company"), each of the investors listed on Schedule A hereto, each of which is referred to in this Agreement as an "Investor" and collectively, the "Investors"), and any Additional Investor that becomes a party to this Agreement in accordance with Section 6.9 hereof.

RECITALS

WHEREAS, certain of the Investors have previously purchased equity securities of the Company;

WHEREAS, the Company and certain Investors are parties to the "Series A Preferred Stock Purchase Agreements" dated as of January 23, 2008 and July 15, 2009;

WHEREAS, the Company and certain Investors are parties to the Series B Preferred Stock Purchase Agreement, dated as of April 6, 2011, by and among the Company and certain of the Investors (as it may be amended, restated, supplemented or otherwise modified from time to time, the "Series B Purchase Agreement");

WHEREAS, the Company and certain Investors are parties to the Series C Preferred Stock Purchase Agreement, dated as of the date hereof, by and among the Company and certain of the Investors (as it may be amended, restated, supplemented or otherwise modified from time to time, the "Series C Purchase Agreement");

WHEREAS, the Company, certain of the Investors and certain other Persons (as defined below) previously entered into an Investors' Rights Agreement, dated as of January 23, 2008 as Amended and Restated as of July 15, 2009 and April 6, 2011 (the "Prior Agreement");

WHEREAS, the Company and its existing Investors desire to induce the Investors to purchase shares of Series C Preferred Stock of the Company, pursuant to the Series C Preferred Stock Purchase Agreement by amending and restating the Prior Agreement to provide the Investors with the rights and privileges as set forth herein; and

WHEREAS, in order to induce the Company to enter into the Series C Purchase Agreement and to induce the Investors to invest funds in the Company pursuant to the Series C Purchase Agreement, the Investors and the Company hereby agree that this Agreement shall govern the rights of the Investors to cause the Company to register shares of Common Stock issuable to the Investors, to receive certain information from the Company, and to participate in future equity offerings by the Company, and shall govern certain other matters as set forth in this Agreement;

NOW, THEREFORE, the parties hereby agree as follows:

1. Definitions. For purposes of this Agreement:

“Affiliate” means, with respect to any specified Person, any other Person who, directly or indirectly, controls, is controlled by, or is under common control with such Person, including without limitation any general partner, managing member, officer or director of such Person or any venture capital fund now or hereafter existing that is controlled by one or more general partners or managing members of, or shares the same management company with, such Person, and provided that Satter Investment Management, LLC, Muneer A. Satter and all Persons for which Mr. Satter or any of his Immediate Family Members serves as trustee or investment advisor or any similar capacity (and their respective Affiliates) and any account held for the benefit of any such Person shall be Affiliates of one another, regardless of whether they would otherwise be deemed Affiliates hereunder.

“Appropriate Percentage” means fifty percent (50%).

“Common Stock” means shares of the Company’s common stock, par value \$0.00001 per share.

“Damages” means any loss, damage, or liability (joint or several) to which a party hereto may become subject under the Securities Act, the Exchange Act, or other federal or state law, insofar as such loss, damage, or liability (or any action in respect thereof) arises out of or is based upon (i) any untrue statement or alleged untrue statement of a material fact contained in any registration statement of the Company, including any preliminary prospectus or final prospectus contained therein or any amendments or supplements thereto; (ii) an omission or alleged omission to state therein a material fact required to be stated therein, or necessary to make the statements therein not misleading; or (iii) any violation or alleged violation by the indemnifying party (or any of its agents or Affiliates) of the Securities Act, the Exchange Act, any state securities law, or any rule or regulation promulgated under the Securities Act, the Exchange Act, or any state securities law.

“Derivative Securities” means any securities or rights convertible into, or exercisable or exchangeable for (in each case, directly or indirectly), Common Stock, including options and warrants.

“Exchange Act” means the Securities Exchange Act of 1934, as amended, and the rules and regulations promulgated thereunder.

“Excluded Registration” means (i) a registration relating to the sale of securities to employees of the Company or a subsidiary pursuant to a stock option, stock purchase, or similar plan; (ii) a registration relating to an SEC Rule 145 transaction; (iii) a registration on any form that does not include substantially the same information as would be required to be included in a registration statement covering the sale of the Registrable Securities; or (iv) a registration in which the only Common Stock being registered is Common Stock issuable upon conversion of debt securities that are also being registered.

“Form S-1” means such form under the Securities Act as in effect on the date hereof or any successor registration form under the Securities Act subsequently adopted by the SEC.

“Form S-3” means such form under the Securities Act as in effect on the date hereof or any registration form under the Securities Act subsequently adopted by the SEC that permits incorporation of substantial information by reference to other documents filed by the Company with the SEC.

“GAAP” means generally accepted accounting principles in the United States.

“Holder” means any holder of Registrable Securities who is a party to this Agreement.

“Immediate Family Member” means a child, stepchild, grandchild, parent, stepparent, grandparent, spouse, sibling, mother-in-law, father-in-law, son-in-law, daughter-in-law, brother-in-law, or sister-in-law, including adoptive relationships, of a natural person referred to herein.

“Initiating Holders” means, collectively, Holders who properly initiate a registration request under this Agreement.

“Investor” means the persons named on Schedule A hereto, each person who hereafter becomes a signatory to this Agreement pursuant to Section 6.9 and any one of them, as the context may require; provided, however, that any such person shall cease to be considered an Investor for purposes of this Agreement at any time such person and his, her or its Affiliates collectively hold no shares of Preferred Stock.

“IPO” means the Company’s first underwritten public offering of its Common Stock under the Securities Act.

“Key Employee” means any executive-level employee (including division director and vice president-level positions) as well as any employee who, either alone or in concert with others, develops, invents, programs, or designs any Company Intellectual Property (as defined in the Series C Purchase Agreement).

“Major Investor” means any Investor that, individually or together with such Investor’s Affiliates, holds at least 27,500 shares of Registrable Securities (as adjusted for any stock split, stock dividend, combination, or other recapitalization or reclassification effected after the date hereof).

“New Securities” means, collectively, equity securities of the Company, whether or not currently authorized, as well as rights, options, or warrants to purchase such equity securities, or securities of any type whatsoever that are, or may become, convertible or exchangeable into or exercisable for such equity securities.

“P&G License Agreement” means that certain License Agreement dated as of September 4, 2007 between the Company and The Procter & Gamble Company.

“Person” means any individual, corporation, partnership, trust, limited liability company, association, foundation or other entity.

“Preferred Director” means any director of the Company that the holders of record of the Preferred Stock are entitled to elect pursuant to the Company’s Certificate of Incorporation.

“Preferred Stock” means shares of the Company’s Series A Preferred Stock, Series B Preferred Stock and Series C Preferred Stock.

“Prior Agreement” has the meaning set forth in the Recitals to this Agreement.

“Registrable Securities” means (i) the Common Stock issuable or issued upon conversion of the Preferred Stock; (ii) any Common Stock issued or issuable (directly or indirectly) upon conversion and/or exercise of any other securities of the Company, acquired by the Investors after the date hereof; and (iii) any Common Stock issued as (or issuable upon the conversion or exercise of any warrant, right, or other security that is issued as) a dividend or other distribution with respect to, or in exchange for or in replacement of, the shares referenced in clauses (i) and (ii) above; excluding in all cases, however, any Registrable Securities sold by a Person in a transaction in which the applicable rights under this Agreement are not assigned pursuant to Section 6.1, and excluding for purposes of Section 2 any shares for which registration rights have terminated pursuant to Section 2.13 of this Agreement.

“Registrable Securities then outstanding” means the number of shares determined by adding the number of shares of outstanding Common Stock that are Registrable Securities and the number of shares of Common Stock issuable (directly or indirectly) pursuant to then exercisable and/or convertible securities that are Registrable Securities.

“Restricted Securities” means the securities of the Company required to bear the legend set forth in Section 2.12(b) hereof.

“SEC” means the Securities and Exchange Commission.

“SEC Rule 144” means Rule 144 promulgated by the SEC under the Securities Act.

“SEC Rule 145” means Rule 145 promulgated by the SEC under the Securities Act.

“Securities Act” means the Securities Act of 1933, as amended, and the rules and regulations promulgated thereunder.

“Selling Expenses” means all underwriting discounts, selling commissions, and stock transfer taxes applicable to the sale of Registrable Securities, and fees and disbursements of counsel for any Holder, except for the fees and disbursements of the Selling Holder Counsel borne and paid by the Company as provided in Section 2.6.

“Series A Preferred Stock” means shares of the Company’s Series A Preferred Stock, par value \$0.00001 per share.

“Series B Preferred Stock” means shares of the Company’s Series B Preferred Stock, par value \$0.00001 per share.

“Series C Preferred Stock” means shares of the Company’s Series C Preferred Stock, par value \$0.00001 per share.

“Series C Purchase Agreement” has the meaning set forth in the Recitals to this Agreement.

2. Registration Rights. The Company covenants and agrees as follows:

2.1. Demand Registration.

(a) Form S-1 Demand. If at any time after the earlier of (i) five (5) years after the date hereof or (ii) one hundred eighty (180) days after the effective date of the registration statement for the IPO the Company receives a request from Holders of thirty percent (30%) of the Registrable Securities then outstanding that the Company file a Form S-1 registration statement with respect to Registrable Securities then outstanding, then the Company shall (i) within ten (10) days after the date such request is given, give notice thereof (the “Demand Notice”) to all Holders other than the Initiating Holders; and (ii) as soon as practicable, and in any event within sixty (60) days after the date such request is given by the Initiating Holders, file a Form S-1 registration statement under the Securities Act covering all Registrable Securities that the Initiating Holders requested to be registered and any additional Registrable Securities requested to be included in such registration by any other Holders, as specified by notice given by each such Holder to the Company within ten (10) days of the date the Demand Notice is given, and in each case, subject to the limitations of Sections 2.1(c) and 2.3.

(b) Form S-3 Demand. If at any time when it is eligible to use a Form S-3 registration statement, the Company receives a request from Holders of at least thirty percent (30%) of the Registrable Securities then outstanding that the Company file a Form S-3 registration statement with respect to outstanding Registrable Securities of such Holders, then the Company shall (i) within ten (10) days after the date such request is given, give a Demand Notice to all Holders other than the Initiating Holders; and (ii) as soon as practicable, and in any event within forty-five (45) days after the date such request is given by the Initiating Holders, file a Form S-3 registration statement under the Securities Act covering all Registrable Securities requested to be included in such registration by any other Holders, as specified by notice given by each such Holder to the Company within ten (10) days of the date the Demand Notice is given, and in each case, subject to the limitations of Sections 2.1(c) and 2.3.

(c) Notwithstanding the foregoing obligations, if the Company furnishes to Holders requesting a registration pursuant to this Section 2.1 a certificate signed by the Company’s chief executive officer stating that in the good faith judgment of the Company’s Board of Directors it would be materially detrimental to the Company and its stockholders for such registration statement to either become effective or remain effective for as long as such registration statement otherwise would be required to remain effective, because such action would (i) materially interfere with a significant acquisition, corporate reorganization, or other similar transaction involving the Company; (ii) require premature disclosure of material

information that the Company has a bona fide business purpose for preserving as confidential; or (iii) render the Company unable to comply with requirements under the Securities Act or Exchange Act, then the Company shall have the right to defer taking action with respect to such filing, and any time periods with respect to filing or effectiveness thereof shall be tolled correspondingly, for a period of not more than sixty (60) days after the request of the Initiating Holders is given; provided, however, that the Company may not invoke this right more than once in any twelve (12) month period; and provided, further, that the Company shall not register any securities for its own account or that of any other stockholder during such sixty (60) day period other than an Excluded Registration.

(d) The Company shall not be obligated to effect, or to take any action to effect, any registration pursuant to Section 2.1(a)(i) during the period that is sixty (60) days before the Company's good faith estimate of the date of filing of, and ending on a date that is one hundred eighty (180) days after the effective date of, a Company-initiated registration, provided, that the Company is actively employing in good faith commercially reasonable efforts to cause such registration statement to become effective; (ii) after the Company has effected two registrations pursuant to Section 2.1(a); or (iii) if the Initiating Holders propose to dispose of shares of Registrable Securities that may be immediately registered on Form S-3 pursuant to a request made pursuant to Section 2.1(b). The Company shall not be obligated to effect, or to take any action to effect, any registration pursuant to Section 2.1(b)(i) during the period that is thirty (30) days before the Company's good faith estimate of the date of filing of, and ending on a date that is ninety (90) days after the effective date of, a Company-initiated registration, provided, that the Company is actively employing in good faith commercially reasonable efforts to cause such registration statement to become effective; or (ii) if the Company has effected three registrations pursuant to Section 2.1(b). A registration shall not be counted as "effected" for purposes of this Section 2.1(d), until such time as the applicable registration statement has been declared effective by the SEC, unless the Initiating Holders withdraw their request for such registration, elect not to pay the registration expenses therefor, and forfeit their right to one demand registration statement pursuant to Section 2.6, in which case such withdrawn registration statement shall be counted as "effected" for purposes of this Section 2.1(d).

2.2. Company Registration. If the Company proposes to register (including, for this purpose, a registration effected by the Company for stockholders other than the Holders) any of its Common Stock under the Securities Act in connection with the public offering of such securities solely for cash (other than in an Excluded Registration), the Company shall, at such time, promptly give each Holder notice of such registration. Upon the request of each Holder given within twenty (20) days after such notice is given by the Company, the Company shall, subject to the provisions of Section 2.3, cause to be registered all of the Registrable Securities that each such Holder has requested to be included in such registration. The Company shall have the right to terminate or withdraw any registration initiated by it under this Section 2.2 before the effective date of such registration, whether or not any Holder has elected to include Registrable Securities in such registration. The expenses (other than Selling Expenses) of such withdrawn registration shall be borne by the Company in accordance with Section 2.6.

2.3. Underwriting Requirements.

(a) If, pursuant to Section 2.1, the Initiating Holders intend to distribute the Registrable Securities covered by their request by means of an underwriting, they shall so advise the Company as a part of their request made pursuant to Section 2.1, and the Company shall include such information in the Demand Notice. The underwriter(s) will be selected by the Initiating Holders, subject only to the reasonable approval of the Company. In such event, the right of any Holder to include such Holder's Registrable Securities in such registration shall be conditioned upon such Holder's participation in such underwriting and the inclusion of such Holder's Registrable Securities in the underwriting to the extent provided herein. All Holders proposing to distribute their securities through such underwriting shall (together with the Company as provided in Section 2.4(e)) enter into an underwriting agreement in customary form with the underwriter(s) selected for such underwriting. Notwithstanding any other provision of this Section 2.3, if the managing underwriter(s) advise(s) the Initiating Holders in writing that marketing factors require a limitation on the number of shares to be underwritten, then the Initiating Holders shall so advise all Holders of Registrable Securities that otherwise would be underwritten pursuant hereto, and the number of Registrable Securities that may be included in the underwriting shall be allocated among such Holders of Registrable Securities, including the Initiating Holders, in proportion (as nearly as practicable) to the number of Registrable Securities owned by each Holder or in such other proportion as shall mutually be agreed to by all such selling Holders; provided, however, that the number of Registrable Securities held by the Holders to be included in such underwriting shall not be reduced unless all other securities are first entirely excluded from the underwriting. To facilitate the allocation of shares in accordance with the above provisions, the Company or the underwriters may round the number of shares allocated to any Holder to the nearest 100 shares.

(b) In connection with any offering involving an underwriting of shares of the Company's capital stock pursuant to Section 2.2, the Company shall not be required to include any of the Holders' Registrable Securities in such underwriting unless the Holders accept the terms of the underwriting as agreed upon between the Company and its underwriters, and then only in such quantity as the underwriters in their sole discretion determine will not jeopardize the success of the offering by the Company. If the total number of securities, including Registrable Securities, requested by stockholders to be included in such offering exceeds the number of securities to be sold (other than by the Company) that the underwriters in their reasonable discretion determine is compatible with the success of the offering, then the Company shall be required to include in the offering only that number of such securities, including Registrable Securities, which the underwriters and the Company in their sole discretion determine will not jeopardize the success of the offering. If the underwriters determine that less than all of the Registrable Securities requested to be registered can be included in such offering, then the Registrable Securities that are included in such offering shall be allocated among the selling Holders in proportion (as nearly as practicable to) the number of Registrable Securities owned by each selling Holder or in such other proportions as shall mutually be agreed to by all such selling Holders. To facilitate the allocation of shares in accordance with the above provisions, the Company or the underwriters may round the number of shares allocated to any Holder to the nearest 100 shares. Notwithstanding the foregoing, in no event shall (i) the number of Registrable Securities included in the offering be reduced unless all other securities (other than securities to be sold by the Company) are first entirely excluded from the offering, or (ii) the number of Registrable Securities included in the offering be reduced below thirty percent (30%) of the total number of securities included in such offering, unless such offering is the IPO, in

which case the selling Holders may be excluded further if the underwriters make the determination described above and no other stockholder's securities are included in such offering. For purposes of the provision in this Section 2.3(b) concerning apportionment, for any selling Holder that is a partnership, limited liability company, or corporation, the partners, members, retired partners, retired members, stockholders, and Affiliates of such Holder, or the estates and Immediate Family Members of any such partners, retired partners, members, and retired members and any trusts for the benefit of any of the foregoing Persons, shall be deemed to be a single "selling Holder," and any pro rata reduction with respect to such "selling Holder" shall be based upon the aggregate number of Registrable Securities owned by all Persons included in such "selling Holder," as defined in this sentence.

(c) For purposes of Section 2.1, a registration shall not be counted as "effected" if, as a result of an exercise of the underwriter's cutback provisions in Section 2.3(a), fewer than fifty percent (50%) of the total number of Registrable Securities that Holders have requested to be included in such registration statement are actually included.

2.4. Obligations of the Company. Whenever required under this Section 2 to effect the registration of any Registrable Securities, the Company shall, as expeditiously as reasonably possible:

(a) prepare and file with the SEC a registration statement with respect to such Registrable Securities and use its commercially reasonable efforts to cause such registration statement to become effective and, upon the request of the Holders of a majority of the Registrable Securities registered thereunder, keep such registration statement effective for a period of up to one hundred twenty (120) days or, if earlier, until the distribution contemplated in the registration statement has been completed; provided, however, that (i) such one hundred twenty (120) day period shall be extended for a period of time equal to the period the Holder refrains, at the request of an underwriter of Common Stock (or other securities) of the Company, from selling any securities included in such registration, and (ii) in the case of any registration of Registrable Securities on Form S-3 that are intended to be offered on a continuous or delayed basis, subject to compliance with applicable SEC rules, such one hundred twenty (120) day period shall be extended for up to sixty (60) days, if necessary, to keep the registration statement effective until all such Registrable Securities are sold;

(b) prepare and file with the SEC such amendments and supplements to such registration statement, and the prospectus used in connection with such registration statement, as may be necessary to comply with the Securities Act in order to enable the disposition of all securities covered by such registration statement;

(c) furnish to the selling Holders such numbers of copies of a prospectus, including a preliminary prospectus, as required by the Securities Act, and such other documents as the Holders may reasonably request in order to facilitate their disposition of their Registrable Securities;

(d) use its commercially reasonable efforts to register and qualify the securities covered by such registration statement under such other securities or blue-sky laws of such jurisdictions as shall be reasonably requested by the selling Holders; provided, that the Company shall not be required to qualify to do business or to file a general consent to service of process in any such states or jurisdictions, unless the Company is already subject to service in such jurisdiction and except as may be required by the Securities Act;

(e) in the event of any underwritten public offering, enter into and perform its obligations under an underwriting agreement, in usual and customary form, with the underwriter(s) of such offering;

(f) use its commercially reasonable efforts to cause all such Registrable Securities covered by such registration statement to be listed on a national securities exchange or trading system and each securities exchange and trading system (if any) on which similar securities issued by the Company are then listed;

(g) provide a transfer agent and registrar for all Registrable Securities registered pursuant to this Agreement and provide a CUSIP number for all such Registrable Securities, in each case not later than the effective date of such registration;

(h) promptly make available for inspection by the selling Holders, any managing underwriter(s) participating in any disposition pursuant to such registration statement, and any attorney or accountant or other agent retained by any such underwriter or selected by the selling Holders, all financial and other records, pertinent corporate documents, and properties of the Company, and cause the Company's officers, directors, employees, and independent accountants to supply all information reasonably requested by any such seller, underwriter, attorney, accountant, or agent, in each case, as necessary or advisable to verify the accuracy of the information in such registration statement and to conduct appropriate due diligence in connection therewith;

(i) notify each selling Holder, promptly after the Company receives notice thereof, of the time when such registration statement has been declared effective or a supplement to any prospectus forming a part of such registration statement has been filed; and

(j) after such registration statement becomes effective, notify each selling Holder of any request by the SEC that the Company amend or supplement such registration statement or prospectus.

2.5. Furnish Information. It shall be a condition precedent to the obligations of the Company to take any action pursuant to this Section 2 with respect to the Registrable Securities of any selling Holder that such Holder shall furnish to the Company such information regarding itself, the Registrable Securities held by it, and the intended method of disposition of such securities as is reasonably required to effect the registration of such Holder's Registrable Securities.

2.6. Expenses of Registration. All expenses (other than Selling Expenses) incurred in connection with registrations, filings, or qualifications pursuant to Section 2, including all registration, filing, and qualification fees; printers' and accounting fees; fees and disbursements of counsel for the Company; and the reasonable fees and disbursements, not to exceed \$75,000, of one counsel for the selling Holders ("Selling Holder Counsel"), shall be borne and paid by the Company; provided, however, that the Company shall not be required to

pay for any expenses of any registration proceeding begun pursuant to Section 2.1 if the registration request is subsequently withdrawn at the request of the Holders of a majority of the Registrable Securities to be registered (in which case all selling Holders shall bear such expenses pro rata based upon the number of Registrable Securities that were to be included in the withdrawn registration), unless the Holders of a majority of the Registrable Securities agree to forfeit their right to one registration pursuant to Sections 2.1(a) or 2.1(b), as the case may be; provided, further, that if, at the time of such withdrawal, the Holders shall have learned of a material adverse change in the condition, business, or prospects of the Company from that known to the Holders at the time of their request and have withdrawn the request with reasonable promptness after learning of such information then the Holders shall not be required to pay any of such expenses and shall not forfeit their right to one registration pursuant to Sections 2.1(a) or 2.1(b). All Selling Expenses relating to Registrable Securities registered pursuant to this Section 2 shall be borne and paid by the Holders pro rata on the basis of the number of Registrable Securities registered on their behalf.

2.7. Delay of Registration. No Holder shall have any right to obtain or seek an injunction restraining or otherwise delaying any registration pursuant to this Agreement as the result of any controversy that might arise with respect to the interpretation or implementation of this Section 2.

2.8. Indemnification. If any Registrable Securities are included in a registration statement under this Section 2:

(a) To the extent permitted by law, the Company shall indemnify and hold harmless each selling Holder, and the partners, members, officers, directors, and stockholders of each such Holder; legal counsel and accountants for each such Holder; any underwriter (as defined in the Securities Act) for each such Holder; and each Person, if any, who controls such Holder or underwriter within the meaning of the Securities Act or the Exchange Act, against any Damages, and the Company will pay to each such Holder, underwriter, controlling Person, or other aforementioned Person any legal or other expenses reasonably incurred thereby in connection with investigating or defending any claim or proceeding from which Damages may result, as such expenses are incurred; provided, however, that the indemnity agreement contained in this Section 2.8(a) shall not apply to amounts paid in settlement of any such claim or proceeding if such settlement is effected without the consent of the Company, which consent shall not be unreasonably withheld, nor shall the Company be liable for any Damages to the extent that they arise out of or are based upon actions or omissions made in reliance upon and in conformity with written information furnished by or on behalf of any such Holder, underwriter, controlling Person, or other aforementioned Person expressly for use in connection with such registration.

(b) To the extent permitted by law, each selling Holder, severally and not jointly, shall indemnify and hold harmless the Company, and each of its directors, each of its officers who has signed the registration statement, each Person (if any), who controls the Company within the meaning of the Securities Act, legal counsel and accountants for the Company, any underwriter (as defined in the Securities Act), any other Holder selling securities in such registration statement, and any controlling Person of any such underwriter or other Holder, against any Damages, in each case only to the extent that such Damages arise out of or

are based upon actions or omissions made in reliance upon and in conformity with written information furnished by or on behalf of such selling Holder expressly for use in connection with such registration; and each such selling Holder will pay to the Company and each other aforementioned Person any legal or other expenses reasonably incurred thereby in connection with investigating or defending any claim or proceeding from which Damages may result, as such expenses are incurred; provided, however, that the indemnity agreement contained in this Section 2.8(b), shall not apply to amounts paid in settlement of any such claim or proceeding if such settlement is effected without the consent of the Holder, which consent shall not be unreasonably withheld; and provided, further, that in no event shall the aggregate amounts payable by any Holder by way of indemnity or contribution under Sections 2.8(b) and 2.8(d) exceed the proceeds from the offering received by such Holder (net of any Selling Expenses paid by such Holder), except in the case of fraud or willful misconduct by such Holder.

(c) Promptly after receipt by an indemnified party under this Section 2.8 of notice of the commencement of any action (including any governmental action) for which a party may be entitled to indemnification hereunder, such indemnified party will, if a claim in respect thereof is to be made against any indemnifying party under this Section 2.8, give the indemnifying party notice of the commencement thereof. The indemnifying party shall have the right to participate in such action and, to the extent the indemnifying party so desires, participate jointly with any other indemnifying party to which notice has been given, and to assume the defense thereof with counsel mutually satisfactory to the parties; provided, however, that an indemnified party (together with all other indemnified parties that may be represented without conflict by one counsel) shall have the right to retain one separate counsel, with the fees and expenses to be paid by the indemnifying party, if representation of such indemnified party by the counsel retained by the indemnifying party would be inappropriate due to actual or potential differing interests between such indemnified party and any other party represented by such counsel in such action.

(d) To provide for just and equitable contribution to joint liability under the Securities Act in any case in which either (i) any party otherwise entitled to indemnification hereunder makes a claim for indemnification pursuant to this Section 2.8 but it is judicially determined (by the entry of a final judgment or decree by a court of competent jurisdiction and the expiration of time to appeal or the denial of the last right of appeal) that such indemnification may not be enforced in such case, notwithstanding the fact that this Section 2.8 provides for indemnification in such case, or (ii) contribution under the Securities Act may be required on the part of any party hereto for which indemnification is provided under this Section 2.8, then, and in each such case, such parties will contribute to the aggregate losses, claims, damages, liabilities, or expenses to which they may be subject (after contribution from others) in such proportion as is appropriate to reflect the relative fault of each of the indemnifying party and the indemnified party in connection with the statements, omissions, or other actions that resulted in such loss, claim, damage, liability, or expense, as well as to reflect any other relevant equitable considerations. The relative fault of the indemnifying party and of the indemnified party shall be determined by reference to, among other things, whether the untrue or allegedly untrue statement of a material fact, or the omission or alleged omission of a material fact, relates to information supplied by the indemnifying party or by the indemnified party and the parties' relative intent, knowledge, access to information, and opportunity to correct or prevent such statement or omission; provided, however, that, in any such case, (x) no Holder will be required

to contribute any amount in excess of the public offering price of all such Registrable Securities offered and sold by such Holder pursuant to such registration statement, and (y) no Person guilty of fraudulent misrepresentation (within the meaning of Section 11(f) of the Securities Act) will be entitled to contribution from any Person who was not guilty of such fraudulent misrepresentation; and provided, further, that in no event shall a Holder's liability pursuant to this Section 2.8(d), when combined with the amounts paid or payable by such Holder pursuant to Section 2.8(b), exceed the proceeds from the offering received by such Holder (net of any Selling Expenses) paid by such Holder), except in the case of willful misconduct or fraud by such Holder.

(e) Notwithstanding the foregoing, to the extent that the provisions on indemnification and contribution contained in the underwriting agreement entered into in connection with the underwritten public offering are in conflict with the foregoing provisions, the provisions in the underwriting agreement shall control.

(f) Unless otherwise superseded by an underwriting agreement entered into in connection with the underwritten public offering, the obligations of the Company and Holders under this Section 2.8 shall survive the completion of any offering of Registrable Securities in a registration under this Section 2, and otherwise shall survive the termination of this Agreement.

2.9. Reports Under Exchange Act. With a view to making available to the Holders the benefits of SEC Rule 144 and any other rule or regulation of the SEC that may at any time permit a Holder to sell securities of the Company to the public without registration or pursuant to a registration on Form S-3, the Company shall:

(a) make and keep available adequate current public information, as those terms are understood and defined in SEC Rule 144, at all times after the effective date of the registration statement filed by the Company for the IPO;

(b) use commercially reasonable efforts to file with the SEC in a timely manner all reports and other documents required of the Company under the Securities Act and the Exchange Act (at any time after the Company has become subject to such reporting requirements); and

(c) furnish to any Holder, so long as the Holder owns any Registrable Securities, forthwith upon request (i) to the extent accurate, a written statement by the Company that it has complied with the reporting requirements of SEC Rule 144 (at any time after ninety (90) days after the effective date of the registration statement filed by the Company for the IPO), the Securities Act, and the Exchange Act (at any time after the Company has become subject to such reporting requirements), or that it qualifies as a registrant whose securities may be resold pursuant to Form S-3 (at any time after the Company so qualifies); (ii) a copy of the most recent annual or quarterly report of the Company and such other reports and documents so filed by the Company; and (iii) such other information as may be reasonably requested in availing any Holder of any rule or regulation of the SEC that permits the selling of any such securities without registration (at any time after the Company has become subject to the reporting requirements under the Exchange Act) or pursuant to Form S-3 (at any time after the Company so qualifies to use such form).

2.10. Limitations on Subsequent Registration Rights. From and after the date of this Agreement, the Company shall not, without the prior written consent of the Holders of more than the Appropriate Percentage of the Registrable Securities then outstanding, enter into any agreement with any holder or prospective holder of any securities of the Company that would allow such holder or prospective holder (i) to include such securities in any registration unless, under the terms of such agreement, such holder or prospective holder may include such securities in any such registration only to the extent that the inclusion of such securities will not reduce the number of the Registrable Securities of the Holders that are included or (ii) allow such holder or prospective holder to initiate a demand for registration of any securities held by such holder or prospective holder.

2.11. "Market Stand-off" Agreement. Each Holder hereby agrees that it will not, without the prior written consent of the managing underwriter, during the period commencing on the date of the final prospectus relating to the registration by the Company of shares of its Common Stock or any other equity securities under the Securities Act on a registration statement on Form S-1 or Form S-3, and ending on the date specified by the Company and the managing underwriter (such period not to exceed (x) one hundred eighty (180) days in the case of the IPO, or (y) if requested by the managing underwriter and approved by Holders of more than the Appropriate Percentage of the Registrable Securities, ninety (90) days in the case of any registration other than the IPO, (i) lend; offer; pledge; sell; contract to sell; sell any option or contract to purchase; purchase any option or contract to sell; grant any option, right, or warrant to purchase; or otherwise transfer or dispose of, directly or indirectly, any shares of Common Stock or any securities convertible into or exercisable or exchangeable (directly or indirectly) for Common Stock held immediately before the effective date of the registration statement for such offering or (ii) enter into any swap or other arrangement that transfers to another, in whole or in part, any of the economic consequences of ownership of such securities, whether any such transaction described in clause (i) or (ii) above is to be settled by delivery of Common Stock or other securities, in cash, or otherwise. Notwithstanding clause (i) and (ii) above, each Holder may distribute any or all of its shares of Common Stock or any securities convertible into or exercisable or exchangeable (directly or indirectly) for Common Stock to any of its limited partners. Provided, however, that such limited partners who receive the distribution of any or all shares of Common Stock or any securities convertible into or exercisable or exchangeable (directly or indirectly) for Common Stock shall still be subject to the other provisions of this Section 2.11. The foregoing provisions of this Section 2.11 shall not apply to the sale of any shares to an underwriter pursuant to an underwriting agreement, and shall be applicable to the Holders only if all officers and directors are subject to the same restrictions and the Company uses commercially reasonable efforts to obtain a similar agreement from all stockholders individually owning more than one percent (1%) of the Company's outstanding Common Stock (after giving effect to conversion into Common Stock of all outstanding Preferred Stock). The underwriters in connection with such registration are intended third-party beneficiaries of this Section 2.11 and shall have the right, power, and authority to enforce the provisions hereof as though they were a party hereto. Each Holder further agrees to execute such agreements as may be reasonably requested by the underwriters in connection with such registration that are consistent with this Section 2.11 or that are necessary to give further effect thereto. Any discretionary waiver or termination of the restrictions of any or all of such agreements by the Company or the underwriters shall apply pro rata to all Holders subject to such agreements, based on the number of shares subject to such agreements.

2.12. Restrictions on Transfer.

(a) The Preferred Stock and the Registrable Securities shall not be sold, pledged, or otherwise transferred, and the Company shall not recognize and shall issue stop-transfer instructions to its transfer agent with respect to any such sale, pledge, or transfer, except upon the conditions specified in this Agreement, which conditions are intended to ensure compliance with the provisions of the Securities Act. A transferring Holder will cause any proposed purchaser, pledgee, or transferee of the Preferred Stock and the Registrable Securities held by such Holder to agree to take and hold such securities subject to the provisions and upon the conditions specified in this Agreement.

(b) Each certificate or instrument representing (i) the Preferred Stock, (ii) the Registrable Securities, and (iii) any other securities issued in respect of the securities referenced in clauses (i) and (ii), upon any stock split, stock dividend, recapitalization, merger, consolidation, or similar event, shall (unless otherwise permitted by the provisions of Section 2.12(c)) be stamped or otherwise imprinted with a legend substantially in the following form:

THE SHARES REPRESENTED HEREBY HAVE BEEN ACQUIRED FOR INVESTMENT AND HAVE NOT BEEN REGISTERED UNDER THE SECURITIES ACT OF 1933. SUCH SHARES MAY NOT BE SOLD, PLEDGED, OR TRANSFERRED IN THE ABSENCE OF SUCH REGISTRATION OR A VALID EXEMPTION FROM THE REGISTRATION AND PROSPECTUS DELIVERY REQUIREMENTS OF SAID ACT.

THE SECURITIES REPRESENTED HEREBY MAY BE TRANSFERRED ONLY IN ACCORDANCE WITH THE TERMS OF AN AGREEMENT BETWEEN THE COMPANY AND THE STOCKHOLDER, A COPY OF WHICH IS ON FILE WITH THE SECRETARY OF THE COMPANY.

The Holders consent to the Company making a notation in its records and giving instructions to any transfer agent of the Restricted Securities in order to implement the restrictions on transfer set forth in this Section 2.12.

(c) The holder of each certificate representing Restricted Securities, by acceptance thereof, agrees to comply in all respects with the provisions of this Section 2. Before any proposed sale, pledge, or transfer of any Restricted Securities, unless there is in effect a registration statement under the Securities Act covering the proposed transaction, the Holder thereof shall give notice to the Company of such Holder's intention to effect such sale, pledge, or transfer. Each such notice shall describe the manner and circumstances of the proposed sale, pledge, or transfer in sufficient detail and, if reasonably requested by the Company, shall be accompanied at such Holder's expense by either (i) a written opinion of legal counsel who shall, and whose legal opinion shall, be reasonably satisfactory to the Company, addressed to the Company, to the effect that the proposed transaction may be effected without registration under

the Securities Act; (ii) a “no action” letter from the SEC to the effect that the proposed sale, pledge, or transfer of such Restricted Securities without registration will not result in a recommendation by the staff of the SEC that action be taken with respect thereto; or (iii) any other evidence reasonably satisfactory to counsel to the Company to the effect that the proposed sale, pledge, or transfer of the Restricted Securities may be effected without registration under the Securities Act, whereupon the Holder of such Restricted Securities shall be entitled to sell, pledge, or transfer such Restricted Securities in accordance with the terms of the notice given by the Holder to the Company. The Company will not require such a legal opinion or “no action” letter (x) in any transaction in compliance with SEC Rule 144 or (y) in any transaction in which such Holder distributes Restricted Securities to an Affiliate of such Holder for no consideration; provided, that each transferee agrees in writing to be subject to the terms of this Section 2.12. Each certificate or instrument evidencing the Restricted Securities transferred as above provided shall bear, except if such transfer is made pursuant to SEC Rule 144, the appropriate restrictive legend set forth in Section 2.12(b), except that such certificate shall not bear such restrictive legend if, in the opinion of counsel for such Holder and the Company, such legend is not required in order to establish compliance with any provisions of the Securities Act.

2.13. Termination of Registration Rights. The right of any Holder to request registration or inclusion of Registrable Securities in any registration pursuant to Section 2.1 or 2.2 shall terminate upon the closing of a Deemed Liquidation Event, as such term is defined in the Company’s Certificate of Incorporation.

3. Information and Observer Rights.

3.1. Delivery of Financial Statements. The Company shall deliver to each Major Investor:

(a) as soon as practicable, but in any event within ninety (90) days after the end of each fiscal year of the Company, (i) a balance sheet as of the end of such year, (ii) statements of income and of cash flows for such year, and a comparison between (x) the actual amounts as of and for such fiscal year and (y) the comparable amounts for the prior year and as included in the Budget (as defined in Section 3.1(e)) for such year, with an explanation of any material differences between such amounts and a schedule as to the sources and applications of funds for such year, and (iii) a statement of stockholders’ equity as of the end of such year, all such financial statements audited and certified by independent public accountants of nationally recognized standing selected by the Company;

(b) as soon as practicable, but in any event within thirty (30) days after the end of each of the first three (3) quarters of each fiscal year of the Company, unaudited statements of income and of cash flows for such fiscal quarter, and an unaudited balance sheet as of the end of such fiscal quarter, all prepared in accordance with GAAP (except that such financial statements may (i) be subject to normal year-end audit adjustments and (ii) not contain all notes thereto that may be required in accordance with GAAP);

(c) as soon as practicable, but in any event within thirty (30) days after the end of each month, a statement showing the number of shares of each class and series of capital stock and securities convertible into or exercisable for shares of capital stock outstanding at the end of the period, the Common Stock issuable upon conversion or exercise of any outstanding securities convertible or exercisable for Common Stock and the exchange ratio or exercise price applicable thereto, and the number of shares of issued stock options and stock options not yet issued but reserved for issuance, if any, all in sufficient detail as to permit the Major Investors to calculate their respective percentage equity ownership in the Company, and certified by the chief financial officer or chief executive officer of the Company as being true, complete, and correct;

(d) as soon as practicable, but in any event within thirty (30) days of the end of each month, an unaudited income statement and statement of cash flows for such month, and an unaudited balance sheet as of the end of such month, all prepared in accordance with GAAP (except that such financial statements may (i) be subject to normal year-end audit adjustments and (ii) not contain all notes thereto that may be required in accordance with GAAP);

(e) as soon as practicable, but in any event thirty (30) days before the end of each fiscal year, a budget and business plan for the next fiscal year (collectively, the “Budget”), prepared on a monthly basis, including balance sheets, income statements, and statements of cash flow for such months and, promptly after prepared, any other budgets or revised budgets prepared by the Company;

(f) with respect to the financial statements called for in Sections 3.1(a) and 3.1(b), a management discussion and analysis by the chief executive officer of the Company that includes updates and status of the Company’s material developments and activities; and

(g) such other information relating to the financial condition, business, prospects, or corporate affairs of the Company as any Major Investor may from time to time reasonably request; provided, however, that the Company shall not be obligated under this Section 3.1 to provide information (i) that the Company reasonably determines in good faith to be a trade secret or confidential information (unless covered by an enforceable confidentiality agreement, in form acceptable to the Company) or (ii) the disclosure of which would adversely affect the attorney-client privilege between the Company and its counsel.

If, for any period, the Company has any subsidiary whose accounts are consolidated with those of the Company, then in respect of such period the financial statements delivered pursuant to the foregoing sections shall be the consolidated and consolidating financial statements of the Company and all such consolidated subsidiaries.

Notwithstanding anything else in this Section 3.1 to the contrary, the Company may cease providing the information set forth in this Section 3.1 during the period starting with the date thirty (30) days before the Company’s good-faith estimate of the date of filing of a registration statement if it reasonably concludes it must do so to comply with the SEC rules applicable to such registration statement and related offering; provided, that the Company’s covenants under this Section 3.1 shall be reinstated at such time as the Company is no longer actively employing its commercially reasonable efforts to cause such registration statement to become effective.

3.2. Inspection. The Company shall permit each Major Investor, at such Major Investor's expense, to visit and inspect the Company's properties; examine its books of account and records; and discuss the Company's affairs, finances, and accounts with its officers, during normal business hours of the Company as may be reasonably requested by the Major Investor; provided, however, that the Company shall not be obligated pursuant to this Section 3.2 to provide access to any information that it reasonably and in good faith considers to be a trade secret or confidential information (unless covered by an enforceable confidentiality agreement, in form acceptable to the Company) or the disclosure of which would adversely affect the attorney-client privilege between the Company and its counsel.

3.3. Observer Rights. As long as each of Venture Investors Early Stage Fund IV Limited Partnership, Athenian Venture Partners, AgeChem Venture Fund L.P., Cincinnati Cornerstone Investors AKB, LLC and Family Akebia Investments LLC (a) purchases shares of the Company's Series C Preferred Stock for at least \$1,499,000 in cash (excluding any conversion of Series X Preferred Stock) pursuant to the Series C Purchase Agreement and (b) continues to own not less than fifty percent (50%) of the aggregate number of shares of Preferred Stock that it purchased under the Series A Preferred Stock Purchase Agreements and the Series B Purchase Agreement and that it is purchasing (or acquiring as a result of the conversion of its shares of Series X Preferred Stock) under the Series C Purchase Agreement (or an equivalent number of shares of Common Stock issued upon conversion thereof), and as long as Triathlon Medical Ventures Fund, L.P. continues to own not less than fifty percent (50%) of the aggregate number of shares of Preferred Stock that it purchased under the Series A Preferred Stock Purchase Agreements and the Series B Purchase Agreement and that it is purchasing (or acquiring as a result of the conversion of its shares of Series X Preferred Stock) under the Series C Purchase Agreement (or an equivalent number of shares of Common Stock issued upon conversion thereof), the Company shall invite a representative of each such Person (provided that the representative of Triathlon Medical Ventures Fund, L.P. must be John M. Rice), as applicable, to attend all meetings of its Board of Directors in a nonvoting observer capacity and, in this respect, shall give such representative copies of all notices, minutes, consents, and other materials that it provides to its directors at the same time and in the same manner as provided to such directors; provided, however, that such representative shall agree to hold in confidence and trust and to act in a fiduciary manner with respect to all information so provided; and provided, further, that the Company reserves the right to withhold any information and to exclude such representative from any meeting or portion thereof if access to such information or attendance at such meeting could adversely affect the attorney-client privilege between the Company and its counsel or result in disclosure of trade secrets or a conflict of interest, or if such Investor or its representative is a competitor of the Company.

3.4. Termination of Information and Observer Rights. The covenants set forth in Sections 3.1, 3.2, and 3.3 shall terminate and be of no further force or effect (i) immediately before the consummation of the IPO, (ii) when the Company first becomes subject to the periodic reporting requirements of Section 12(g) or 15(d) of the Exchange Act, or (iii) upon a Deemed Liquidation Event, as such term is defined in the Company's Certificate of Incorporation, whichever event occurs first.

3.5. Confidentiality. Each Investor agrees that such Investor will keep confidential and will not disclose, divulge, or use for any purpose (other than to monitor its investment in the Company) any confidential information obtained from the Company pursuant to the terms of this Agreement (including notice of the Company's intention to file a registration statement), unless such confidential information (a) is known or becomes known to the public in general (other than as a result of a breach of this Section 3.5 by such Investor), (b) is or has been independently developed or conceived by the Investor without use of the Company's confidential information, or (c) is or has been made known or disclosed to the Investor by a third party without a breach of any obligation of confidentiality such third party may have to the Company; provided, however, that an Investor may disclose confidential information (i) to its attorneys, accountants, consultants, and other professionals to the extent necessary to obtain their services in connection with monitoring its investment in the Company; (ii) to any prospective purchaser of any Registrable Securities from such Investor, if such prospective purchaser agrees to be bound by the provisions of this Section 3.5; (iii) to any existing or prospective Affiliate, partner, member, stockholder, or wholly owned subsidiary of such Investor in the ordinary course of business; provided, that such Investor informs such Person that such information is confidential and directs such Person to maintain the confidentiality of such information; or (iv) as may otherwise be required by law; provided, that the Investor promptly notifies the Company of such disclosure and takes reasonable steps to minimize the extent of any such required disclosure.

4. Rights to Future Stock Issuances.

4.1. Right of First Offer. Subject to the terms and conditions of this Section 4.1 and applicable securities laws, if the Company proposes to offer or sell any New Securities, the Company shall first offer such New Securities to each Investor. An Investor shall be entitled to apportion the right of first offer hereby granted to it among itself and its Affiliates in such proportions as it deems appropriate.

(a) The Company shall give notice (the "Offer Notice") to each Investor, stating (i) its bona fide intention to offer such New Securities, (ii) the number of such New Securities to be offered, and (iii) the price and terms, if any, upon which it proposes to offer such New Securities.

(b) By notification to the Company within twenty (20) days after the Offer Notice is given, each Investor may elect to purchase or otherwise acquire, at the price and on the terms specified in the Offer Notice, up to that portion of such New Securities which equals the proportion that the Common Stock then held by such Investor (assuming full conversion and/or exercise, as applicable, of all Preferred Stock and other Derivative Securities then held by such Investor) bears to the total shares of Common Stock of the Company then outstanding (assuming full conversion and/or exercise, as applicable, of all Preferred Stock and other Derivative Securities). At the expiration of such twenty (20) day period, the Company shall promptly notify each Investor that elects to purchase or acquire all the shares available to it (each, a "Fully Exercising Investor") of any other Investor's failure to do likewise. During the ten (10) day period commencing after the Company has given such notice, each Fully Exercising Investor may, by giving notice to the Company, elect to purchase or acquire, in addition to the number of shares specified above, up to that portion of the New Securities for which Investors were entitled to subscribe but that were not subscribed for by the Investors which is equal to the proportion that the Common Stock then held by such Fully Exercising Investor (assuming full conversion and/or exercise, as applicable, of all Preferred Stock and other Derivative Securities

then held by such Fully Exercising Investor) bears to the Common Stock then held by all Fully Exercising Investors who wish to purchase such unsubscribed shares (assuming full conversion and/or exercise, as applicable, of all Preferred Stock and other Derivative Securities then held by all such Fully Exercising Investors). The closing of any sale pursuant to this Section 4.1(b) shall occur within the later of ninety (90) days of the date that the Offer Notice is given and the date of initial sale of New Securities pursuant to Section 4.1(c).

(c) If all New Securities referred to in the Offer Notice are not elected to be purchased or acquired as provided in Section 4.1(b), the Company may, during the ninety (90) day period following the expiration of the periods provided in Section 4.1(b), offer and sell the remaining unsubscribed portion of such New Securities to any Person or Persons at a price not less than, and upon terms no more favorable to the offeree than, those specified in the Offer Notice. If the Company does not enter into an agreement for the sale of the New Securities within such period, or if such agreement is not consummated within thirty (30) days of the execution thereof, the right provided hereunder shall be deemed to be revived and such New Securities shall not be offered unless first reoffered to the Investors in accordance with this Section 4.1.

(d) The right of first offer in this Section 4.1 shall not be applicable to (i) Exempted Securities (as defined in the Company's Certificate of Incorporation); and (ii) shares of Common Stock issued in the IPO.

(e) [Reserved]

(f) Notwithstanding any provision hereof to the contrary, in lieu of complying with the provisions of this Section 4.1, the Company may elect to give notice to the Investors within thirty (30) days after the issuance of New Securities. Such notice shall describe the type, price, and terms of the New Securities. Each Investor shall have twenty (20) days from the date notice is given to elect to purchase up to the number of New Securities that would, if purchased by such Investor, maintain such Investor's percentage-ownership position, calculated as set forth in Section 4.1(b) before giving effect to the issuance of such New Securities. The closing of such sale shall occur within sixty (60) days of the date notice is given to the Investors.

4.2. Termination. The covenants set forth in Section 4.1 shall terminate and be of no further force or effect (i) immediately before the consummation of the IPO, or (ii) upon a Deemed Liquidation Event, as such term is defined in the Company's Certificate of Incorporation, whichever event occurs first.

5. Additional Covenants.

5.1. Insurance. The Company shall use its commercially reasonable efforts to obtain, within ninety (90) days of the date hereof, from financially sound and reputable insurers Directors and Officers liability insurance and term "key-person" insurance on each of Joseph Gardner, Robert Shalwitz and William Daly, each in the amount of \$2,000,000 or such lesser amount determined by the Board of Directors (including a majority of the Preferred Directors) and on terms and conditions satisfactory to the Board of Directors, and will use commercially reasonable efforts to cause such insurance policies to be maintained until such time as the Board

of Directors determines that such insurance should be discontinued. The key-person policy shall name the Company as loss payee, and neither policy shall be cancelable by the Company without prior approval by the Board of Directors and holders of more than the Appropriate Percentage of the shares of Common Stock then issuable upon conversion of the then outstanding shares of Preferred Stock.

5.2. Employee Agreements. The Company will cause (i) each person now or hereafter employed by it or by any subsidiary (or engaged by the Company or any subsidiary as a consultant/independent contractor) with access to confidential information and/or trade secrets to enter into a nondisclosure and proprietary rights assignment agreement and (ii) each Key Employee to enter into a one (1) year noncompetition and nonsolicitation agreement, substantially in the form approved by the Board of Directors. In addition, the Company shall not amend, modify, terminate, waive, or otherwise alter, in whole or in part, any of the above-referenced agreements or any restricted stock agreement between the Company and any employee, without the consent of a majority of the Preferred Directors.

5.3. Qualified Small Business Stock. The Company shall use reasonable best efforts to cause the shares of Preferred Stock, as well as any shares into which such shares are converted, within the meaning of Section 1202(f) of the Internal Revenue Code (the "Code"), to constitute "qualified small business stock" as defined in Section 1202(c) of the Code; provided, however, that such requirement shall not be applicable if the Board of Directors of the Company determines, in its good-faith business judgment, that such qualification would unreasonably defer a sale or liquidation of the Company by virtue of the applicable "minimum holding period" requirements. The Company shall submit to its stockholders (including the Investors) and to the Internal Revenue Service any reports that may be required under Section 1202(d)(1)(C) of the Code and the regulations promulgated thereunder. In addition, within twenty (20) business days after any Investor's written request therefor, the Company shall, at its option, either (i) deliver to such Investor a written statement indicating whether (and what portion of) such Investor's interest in the Company constitutes "qualified small business stock" as defined in Section 1202(c) of the Code or (ii) deliver to such Investor such factual information in the Company's possession as is reasonably necessary to enable such Investor to determine whether (and what portion of) such Investor's interest in the Company constitutes "qualified small business stock" as defined in Section 1202(c) of the Code.

5.4. Matters Requiring Investor Approval. So long as at least twenty percent (20%) of the originally issued Series A Preferred Stock, at least twenty percent (20%) of the originally issued Series B Preferred Stock or at least twenty percent (20%) of the originally issued Series C Preferred Stock remains outstanding, the Company hereby covenants and agrees with each of the Investors that it shall not, without the approval of the Board of Directors, which approval must include the affirmative vote of a majority of the Preferred Directors then in office:

(a) make, or permit any subsidiary to make, any loan or advance in an aggregate amount in excess of \$100,000 to, or own any stock or other securities of, any subsidiary or other corporation, partnership, or other entity unless it is wholly owned by the Company;

(b) make, or permit any subsidiary to make, any loan or advance to any Person, including, without limitation, any employee or director of the Company or any subsidiary, except advances and similar expenditures in the ordinary course of business or under the terms of an employee stock or option plan approved by the Board of Directors;

(c) guarantee, directly or indirectly, or permit any subsidiary to guarantee, directly or indirectly, any indebtedness in an aggregate amount in excess of \$100,000, except for trade accounts of the Company or any subsidiary arising in the ordinary course of business;

(d) make any investment inconsistent with any investment policy approved by the Board of Directors;

(e) incur any aggregate indebtedness in excess of \$250,000, other than trade credit incurred in the ordinary course of business;

(f) otherwise enter into or be a party to any transaction with any director, officer, or employee of the Company or any “associate” (as defined in Rule 12b-2 promulgated under the Exchange Act) of any such Person, except for transactions contemplated by this Agreement, the Series A Purchase Agreements, the Series B Purchase Agreement, and the P&G License Agreement in effect as of the date of this Agreement;

(g) hire, terminate, or change the compensation of the executive officers, including approving any option grants or stock awards to executive officers;

(h) change the principal business of the Company, enter new lines of business, or exit the current line of business;

(i) sell, assign, license, pledge, or encumber material technology or intellectual property, other than licenses granted in the ordinary course of business;

(j) make any capital expenditure in excess of \$100,000; or

(k) adopt the Budget.

5.5. **Board Matters.** Unless otherwise determined by the vote of a majority of the directors then in office, the Board of Directors shall meet at least bi-monthly in accordance with an agreed-upon schedule. The Company shall reimburse the nonemployee directors for all reasonable out-of-pocket travel expenses incurred (consistent with the Company’s travel policy) in connection with attending meetings of the Board of Directors. The Company shall cause to be established, as soon as practicable after such request, and will maintain, an audit and compensation committee, each of which shall consist solely of non-management directors. Each non-employee director shall be entitled in such person’s discretion to be a member of any Board committee.

5.6. Successor Indemnification. If the Company or any of its successors or assignees consolidates with or merges into any other Person and is not the continuing or surviving corporation or entity of such consolidation or merger, then to the extent necessary, proper provision shall be made so that the successors and assignees of the Company assume the obligations of the Company with respect to indemnification of members of the Board of Directors as in effect immediately before such transaction, whether such obligations are contained in the Company's Bylaws, its Certificate of Incorporation, or elsewhere, as the case may be.

5.7. Termination of Covenants. The covenants set forth in this Section 5, except for Section 5.6, shall terminate and be of no further force or effect (i) immediately before the consummation of the IPO, (ii) when the Company first becomes subject to the periodic reporting requirements of Section 12(g) or 15(d) of the Exchange Act, or (iii) upon a Deemed Liquidation Event, as such term is defined in the Company's Certificate of Incorporation, whichever event occurs first.

5.8. Limitation of Liability. The Company shall not enter into any agreement that could reasonably be expected to result in requiring such Investor in any series of the Company's Preferred Stock, without the written consent of such Investor, to participate in any transaction which would expose such Investor to liability (a) in excess of the proceeds actually received by such Investor under such transaction or (b) where the share of such liability of such Investor is higher than its pro rata portion of the proceeds actually received by such Investor under such transaction.

6. Miscellaneous.

6.1. Successors and Assigns. The rights under this Agreement may be assigned (but only with all related obligations) by a Holder to a transferee of Registrable Securities that (i) is an Affiliate of a Holder; (ii) is a Holder's Immediate Family Member or trust for the benefit of an individual Holder or one or more of such Holder's Immediate Family Members; or (iii) after such transfer, holds at least 1,000 shares of Registrable Securities (subject to appropriate adjustment for stock splits, stock dividends, combinations, and other recapitalizations after the date hereof); provided, however, that (x) the Company is, within a reasonable time after such transfer, furnished with written notice of the name and address of such transferee and the Registrable Securities with respect to which such rights are being transferred; and (y) such transferee agrees in a written instrument delivered to the Company to be bound by and subject to the terms and conditions of this Agreement, including the provisions of Section 2.11. For the purposes of determining the number of shares of Registrable Securities held by a transferee, the holdings of a transferee (1) that is an Affiliate or stockholder of a Holder; (2) who is a Holder's Immediate Family Member; or (3) that is a trust for the benefit of an individual Holder or such Holder's Immediate Family Member shall be aggregated together and with those of the transferring Holder; provided, further, that all transferees who would not qualify individually for assignment of rights shall have a single attorney-in-fact for the purpose of exercising any rights, receiving notices, or taking any action under this Agreement. The terms and conditions of this Agreement inure to the benefit of and are binding upon the respective successors and permitted assignees of the parties. Nothing in this Agreement, express or implied, is intended to confer upon any party other than the parties hereto or their respective successors and permitted assignees any rights, remedies, obligations or liabilities under or by reason of this Agreement, except as expressly provided herein.

6.2. Governing Law. This Agreement shall be governed by, and construed in accordance with, the laws of the State of Delaware, regardless of the laws that might otherwise govern under applicable principles of conflicts of law.

6.3. Counterparts; Facsimile or PDE. This Agreement may be executed in two or more counterparts, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument. This Agreement may also be executed and delivered by facsimile or PDF signature and in two or more counterparts, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument.

6.4. Titles and Subtitles. The titles and subtitles used in this Agreement are for convenience only and are not to be considered in construing or interpreting this Agreement.

6.5. Notices. All notices and other communications given or made pursuant to this Agreement shall be in writing and shall be deemed effectively given upon the earlier of actual receipt or: (i) personal delivery to the party to be notified; (ii) when sent, if sent by electronic mail or facsimile during the recipient's normal business hours, and if not sent during normal business hours, then on the recipient's next business day; (iii) five (5) days after having been sent by registered or certified mail, return receipt requested, postage prepaid; or (iv) one (1) business day after the business day of deposit with a nationally recognized overnight courier, freight prepaid, specifying next-day delivery, with written verification of receipt. All communications shall be sent to the respective parties at their addresses as set forth on Schedule A hereto, or to the principal office of the Company and to the attention of the Chief Executive Officer, in the case of the Company, or to such email address, facsimile number, or address as subsequently modified by written notice given in accordance with this Section 6.5.

If notice is given to the Company, a copy shall also be sent to:

Thompson Hine LLP
312 Walnut Street, 14th Floor
Cincinnati, OH 45202-4089
Attn: David J. Willbrand
david.willbrand@thompsonhine.com

If notice is given to the Investors, a copy shall also be sent to:

Kirkland & Ellis LLP
300 N. LaSalle St.
Chicago, IL 60654
Attn: Ted H. Zook, P.C.
Roger D. Rhoten
E-mail: ted.zook@kirkland.com
roger.rhoten@kirkland.com

and

Edwards Wildman Palmer LLP
Attn: Albert L. Sokol
111 Huntington Avenue
Boston, MA 02199 USA
asokol@edwardswildman.com

6.6. Amendments and Waivers. Any term of this Agreement may be amended and the observance of any term of this Agreement may be waived (either generally or in a particular instance, and either retroactively or prospectively) only with the written consent of the Company and the holders of more than the Appropriate Percentage of the Registrable Securities then outstanding; provided, that the Company may in its sole discretion waive compliance with Section 2.12(c) (and the Company's failure to object promptly in writing after notification of a proposed assignment allegedly in violation of Section 2.12(c) shall be deemed to be a waiver); and provided, further, that any provision hereof may be waived by any waiving party on such party's own behalf, without the consent of any other party. Notwithstanding the foregoing, this Agreement may not be amended or terminated and the observance of any term hereof may not be waived with respect to any Investor without the written consent of such Investor, unless such amendment, termination, or waiver applies to all Investors in the same fashion (it being agreed that a waiver of the provisions of Section 4 with respect to a particular transaction shall be deemed to apply to all Investors in the same fashion if such waiver does so by its terms, notwithstanding the fact that certain Investors may nonetheless, by agreement with the Company, purchase securities in such transaction). Notwithstanding the foregoing, the provisions of (a) Section 3.3 may not be amended to terminate a designated right therein to appoint an observer to the Board of Directors without the written consent of the Company and at least fifty percent (50%) of the shares of Common Stock issuable upon conversion of the then outstanding shares of Preferred Stock held by the Persons entitled to appoint such observer, (b) Sections 4.1 and 4.2 may not be waived without the written consent of the holders of more than seventy percent (70%) of the shares of Common Stock issuable upon conversion of the then outstanding shares of Preferred Stock and (c) Section 5.8 may not be amended or waived without the written consent of the Company and the holders of ninety percent (90%) of the shares of Common Stock issuable upon conversion of the then outstanding shares of Preferred Stock. The Company shall give prompt notice of any amendment or termination hereof or waiver hereunder to any party hereto that did not consent in writing to such amendment, termination, or waiver. Any amendment, termination, or waiver effected in accordance with this Section 6.6 shall be binding on all parties hereto, regardless of whether any such party has consented thereto. No waivers of or exceptions to any term, condition, or provision of this Agreement, in any one or more instances, shall be deemed to be or construed as a further or continuing waiver of any such term, condition, or provision.

6.7. Severability. In case any one or more of the provisions contained in this Agreement is for any reason held to be invalid, illegal or unenforceable in any respect, such invalidity, illegality, or unenforceability shall not affect any other provision of this Agreement, and such invalid, illegal, or unenforceable provision shall be reformed and construed so that it will be valid, legal, and enforceable to the maximum extent permitted by law.

6.8. Aggregation of Stock. All shares of Registrable Securities held or acquired by Affiliates shall be aggregated together for the purpose of determining the availability of any rights under this Agreement and such Affiliated persons may apportion such rights as among themselves in any manner they deem appropriate.

6.9. Additional Investors. Notwithstanding anything to the contrary contained herein, if the Company issues additional shares of the Company's Preferred Stock after the date hereof, whether pursuant to the Series C Purchase Agreement or otherwise or if any Investor transfers any shares of the Company's Preferred Stock to any other Person, any purchaser or new holder of such shares of Preferred Stock may become a party to this Agreement by executing and delivering an additional counterpart signature page to this Agreement, and thereafter shall be deemed an "Investor" for all purposes hereunder. No action or consent by the Investors shall be required for such joinder to this Agreement by such additional Investor, so long as such additional Investor has agreed in writing to be bound by all of the obligations as an "Investor" hereunder.

6.10. Entire Agreement. This Agreement (including any Schedules and Exhibits hereto) amends and restates in its entirety the Prior Agreement, constitutes the full and entire understanding and agreement among the parties with respect to the subject matter hereof, and any other written or oral agreement relating to the subject matter hereof existing between the parties is expressly canceled.

6.11. [Reserved].

6.12. Delays or Omissions. No delay or omission to exercise any right, power, or remedy accruing to any party under this Agreement, upon any breach or default of any other party under this Agreement, shall impair any such right, power, or remedy of such nonbreaching or nondefaulting party, nor shall it be construed to be a waiver of or acquiescence to any such breach or default, or to any similar breach or default thereafter occurring, nor shall any waiver of any single breach or default be deemed a waiver of any other breach or default theretofore or thereafter occurring. All remedies, whether under this Agreement or by law or otherwise afforded to any party, shall be cumulative and not alternative.

6.13. Acknowledgment. The Company acknowledges that certain Investors are in the business of venture capital investing and therefore review the business plans and related proprietary information of many enterprises, including enterprises which may have products or services which compete directly or indirectly with those of the Company. Nothing in this Agreement shall preclude or in any way restrict the Investors from investing or participating in any particular enterprise whether or not such enterprise has products or services which compete with those of the Company.

[Remainder of Page Intentionally Left Blank]

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

AKEBIA THERAPEUTICS, INC.

By: /s/ Joseph H. Gardner
Joseph H. Gardner, Ph.D.
President and Chief Executive Officer

Address:
Suite 420,
9987 Carver Road,
Cincinnati, OH 45242

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

AGECHEM VENTURE FUND L.P.

By: /s/ Louis Lacasse

Name: Louise Lacasse

Title: President

Address:

Attn: Louis Lacasse, President

1 Westmount Square, Suite 800

Montreal, Quebec, Canada

H3Z 2P9

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

ATHENIAN VENTURE PARTNERS III L.P.

By: /s/ Karl O. Elderkin
Name: Karl O. Elderkin
Title: President

AVP OHIO TECHNOLOGY I L.P.

By: /s/ Karl O. Elderkin
Name: Karl O. Elderkin
Title: President

Address:

340 West State Street
Unit 29/Suite I37D
Athens OH 45701

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

BLUE CHIP VALIDATION FUND, LTD.

By: /s/ John McIlwraith
John McIlwraith
Managing Director

Address:

312 Walnut Street
Suite 1120
Cincinnati, OH 45202

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

CINCINNATI CORNERSTONE INVESTORS AKB, LLC

By: /s/ Robert W. Coy, Jr.

Robert W. Coy, Jr.
President

Address:

30 West 3rd Street, 6th Floor
Cincinnati, OH 45202-3559

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

WILLIAM DALY

By: /s/ William Daly
William Daly

Address:

13 Via Abrazar
San Clemente, CA 92673

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

DIANE H. JANUSZ TRUST

By: /s/ John Janusz

Name: John Janusz

Title: Trustee

JOHN JANUSZ

By: /s/ John Janusz

John Janusz

Address:

7385 Desert Spring Court
West Chester, OH 45069

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

FAMILY AKEBIA INVESTMENTS LLC

By: /s/ Milton Berlinski

Name: Milton Berlinski

Title: Managing Member

Address:

1185 Park Avenue #11G
New York, NY 10128

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

ALAN FISHMAN

By: /s/ Alan Fishman
Alan Fishman

Address:

6900 Stonehenge Dr.
Cincinnati, OH 45242-6204

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

FRED SHALWITZ TRUST,
ROBERT SHALWITZ, TRUSTEE

By: /s/ Robert Schalwitz
Name: Robert Schalwitz
Title: Trustee

Address:

2549 Bryden Road
Bexley, OH 43209

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

JOSEPH H. GARDNER

By: /s/ Joseph H. Gardner

Address:

4060 Boomer Road
Cincinnati, OH 45247

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

GARDNER FAMILY TRUST. JOHN D. GARDNER TRUSTEE

By: /s/ John D. Gardner

John D. Gardner
Trustee

Address:

111 Pine Court
Bastop, TX 78602

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

GHANA FAMILY TRUST, ELIZABETH C. ARMITAGE TRUSTEE

By: /s/ Elizabeth C. Armitage

Elizabeth C. Armitage
Trustee

Address:

2207 Upland Place
Cincinnati, OH 45206

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

IAN A. W. HOWES, IRA.
STERLING TRUST CUSTODIAN

By: /s/ Ian A. W. Howes
Name: Ian A. W. Howes
Title: Trustee

IAN A. W. HOWES

By: /s/ Ian A. W. Howes
Ian A. W. Howes

Address:

219 Stratford Drive
Chapel Hill, NC 27516

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

KEARNY VENTURE PARTNERS, L.P.

By: /s/ Anupam Dalal

Name: Kearny Venture Associates, LLC

Title: its General Partner

Address:

Attn: Anupam Dalal
Kearny Venture Associates LLC
88 Kearny Street, Suite 200
San Francisco, CA 94108-5530

KEARNY VENTURE PARTNERS ENTREPRENEURS FUND, L.P.

By: /s/ Anupam Dalal

Name: Kearny Venture Associates, LLC

Title: its General Partner

Address:

Attn: Anupam Dalal
Kearny Venture Associates LLC
88 Kearny Street, Suite 200
San Francisco, CA 94108-5530

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

MCILWRAITH INVESTMENTS, LLC

By: /s/ John McIlwraith

Name: John McIlwraith

Title: Manager

Address:

Attn: John McIlwraith

7680 Foxgate Lane

Cincinnati, OH 45243

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

MRK INTERNATIONAL, LLC

By: /s/ Richard L. Kiley

Name: Richard L. Kiley

Title: Principal Member

Address:

7800 Tecumseh Trail
Cincinnati, OH 45243

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

NOVARTIS BIOVENTURES LTD.

By: /s/ H.S. Zivi
Name: H.S. Zivi
Title: Deputy Chairman

By: /s/ Rebecca White
Name: Rebecca White
Title: Authorized Signatory

Address:

NOVARTIS BIOVENTURES LTD.
Attn: Henri Simon Zivi
131 Front Street
Hamilton HM 12
Bermuda

But for mail, to:

Novartis Bio Ventures Ltd.
Attn: Henri Simon Zivi
PO Box HM 2899
Hamilton HM LX Bermuda

And, also send a copy to:

Novartis Venture Fund
Attn: Campbell Murray
Five Cambridge Center, Suite 603
Cambridge, MA 02142

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

KEVIN PETERS

By: /s/ Kevin Peters
Kevin Peters

Address:

9160 Given Road
Cincinnati, OH 45243

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

QCA FIRST FUND II

By: /s/ John Habbert

Name: John Habbert

Title: Manager

Address:

109 Bentwood Ct.
Cincinnati, OH 45241

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

MUNEER A. SATTER REVOCABLE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Trustee

THE SATTER FOUNDATION

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Trustee

SATTER FAMILY TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

SATTER CHILDREN'S TRUST I

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

Address:

c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

KRISTEN HAYLER HERTEL REVOCABLE TRUST

By: /s/ Kristen Hayler Hertel

Name: Kristen Hayler Hertel

Title: Trustee

Address:

c/o Satter Investment Management, LLC

676 N. Michigan Avenue, Suite 4000

Chicago, IL 60611

Attn: Muneer A. Satter

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

ANNE-CAROLE WITORT INSURANCE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Trustee

ROSE SHEREEN FUQUA INSURANCE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

RABI H. SATTER INSURANCE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

JOHN WOOD TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

Address:

c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

ABDUS SATTER INSURANCE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Trustee

GORDON AND BARBARA ANNE HERTEL INSURANCE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Trustee

Address:

c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

ROBERT A. SHALWITZ

By: /s/ Robert A. Shalwitz

Address:

2549 Bryden Road
Bexley, OH 43209

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

SIGVION FUND I, LP

By: /s/ J.P. Fairbank
J.P. Fairbank
Founding partner

Address:

806 West Washington Street, Suite 204
Chicago, IL 60607

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

THOMAS WEISEL HEALTHCARE VENTURE PARTNERS, L.P.

By: Thomas Weisel Capital Management LLC
Title: its Managing Member

By: /s/ Anupam Dalal
Name: Anupam Dalal
Title: Managing Director

Address:

88 Kearny Street, 4th Floor
San Francisco, CA 94108

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

TRIATHLON MEDICAL VENTURES FUND, L.P.

By: Triathlon Medical Ventures, LLC
Title: its General Partner

By: /s/ John M. Rice
John M. Rice
Managing Partner

Address:

300 E-Business Way
Suite 200
Cincinnati, OH 45241

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

TRI-STATE GROWTH CAPITAL FUND II, L.P.

By: Tri-State Ventures II, LLC
Title: General Partner

By: Fort Washington Investment Advisors, Inc.
Title: Managing Member

By: /s/ Steve Baker
Name: Steve Baker
Title: Managing Director

By: /s/ Maribeth S. Rahe
Name: Maribeth S. Rahe
Title: President and Chief Executive Officer

Address:

303 Broadway, Suite 1200
Cincinnati, OH 45202

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

VENTURE INVESTORS EARLY STAGE
FUND IV LIMITED PARTNERSHIP

By: VIESF IV GP, LLC, its General Partner

By: /s/ Paul M. Weiss

Name: Paul M. Weiss, Ph.D.

Title: Managing Director

Address:

505 South Rosa Road

Madison, WI 53719-1262

Attn: Paul Weiss, Managing Director

Phone: (608) 441-2700

Fax: (608) 441-2727

Email: paul@ventureinvestors.com

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Third Amended and Restated Investors' Rights Agreement as of the date first written above.

JOHN H. WYANT

By: /s/ John H. Wyant
John H. Wyant

Address:

Blue Chip Venture Company
1120 Scripps Center
312 Walnut Street
Cincinnati, OH 45202

SIGNATURE PAGE OF THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT

SCHEDULE A**Investors****Name and Address**

Triathlon Medical Ventures Fund, L.P.
Attn: John M. Rice
Managing Partner
300 E-Business Way, Suite 200
Cincinnati, OH 45241

Novartis BioVentures Ltd.
Attn: Henri Simon Zivi
131 Front Street
Hamilton HM 12
Bermuda

But for mail, to:

Novartis BioVentures Ltd.
Attn: Henri Simon Zivi
PO Box HM 2899
Hamilton HM LX Bermuda

And, also send a copy to:

Novartis Venture Fund
Attn: Campbell Murray
Five Cambridge Center, Suite 603
Cambridge, MA 02142

and

Edwards Angell Palmer & Dodge LLP
Attn: Al Sokol
111 Huntington Avenue
Boston, MA 02199
asokol@eapdlaw.com

Venture Investors Early Stage Fund IV Limited Partnership
Attn: Paul Weiss
Managing Director
505 South Rosa Road
Madison, WI 53719-1262

Schedule A-1

Kearny Venture Partners, L.P.
Attn: Anupam Dalal
Kearny Venture Associates, LLC
88 Kearny Street, Suite 200
San Francisco, CA 94108-5530

Kearny Venture Partners Entrepreneurs Fund, L.P.
Attn: Anupam Dalal
Kearny Venture Associates, LLC
88 Kearny Street, Suite 200
San Francisco, CA 94108-5530

Thomas Weisel Healthcare Venture Partners, L.P.
88 Kearny Street, 4th Floor
San Francisco, CA 94108

The Procter & Gamble Company
Attn: David Le Neveu
Director, Corporate Acquisitions, Divestitures and Equity Ventures
1 Procter & Gamble Plaza
Cincinnati, OH 45202

Athenian Venture Partners III L.P.
Attn: Karl O. Elderkin
President, Athenian III, Ltd.
340 West State Street
Unit 29/Suite 137D
Athens, OH 45701

AVP Ohio Technology I L.P.
Attn: Karl O. Elderkin
President
AVP Ohio I, Ltd.
340 West State Street
Unit 29/Suite 137D
Athens, OH 45701

Sigvion Fund I, LP
Attn: J. P. Fairbank
Founding Partner
738 W. Belden Avenue
Chicago, IL 60614

Cincinnati Cornerstone Investors AKB, LLC
Attn: Robert W. Coy, Jr.
President
30 West 3rd Street, 6th Floor
Cincinnati, OH 45202-3559

Tri-State Growth Capital Fund II, L.P.
Attn: Steve Baker
303 Broadway, Suite 1200
Cincinnati, OH 45202

Blue Chip Validation Fund, Ltd.
Attn: John McIlwraith
Managing Director
1100 Chiquita Center
250 East Fifth Street
Cincinnati, OH 45202

QCA First Fund II
Attn: John Habbert
1776 Mentor Avenue, MB #302
Cincinnati, OH 45212

Gitana Family Trust, Elizabeth C. Armitage Trustee
Attn: Elizabeth C. Armitage
Trustee
2207 Upland Place
Cincinnati, OH 45206

Robert Shalwitz
2549 Bryden Road
Bexley, OH 43209

Fred Shalwitz Trust, Robert Shalwitz, Trustee
Attn: Robert Shalwitz
Trustee
2549 Bryden Road
Bexley, OH 43209

Joseph H. Gardner
4060 Boomer Road
Cincinnati, OH 45247

Gardner Family Trust, John D. Gardner Trustee
Attn: John D. Gardner
Trustee
111 Pine Court
Bastop, TX 78602

Ian A. W. Howes, IRA, Sterling Trust Custodian
Attn: Ian A. W. Howes
Trustee
219 Stratford Drive
Chapel Hill, NC 27516

Ian A. W. Howes
219 Stratford Drive
Chapel Hill, NC 27516

Kevin Peters
9160 Given Road
Cincinnati, OH 45243

William Daly
13 Via Abrazar
San Clemente, CA 92673

Muneer A. Satter Revocable Trust
c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

John Wood Trust
c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

The Satter Foundation
c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

Muneer A Satter IRA, Millennium Trust Company, Custodian

Satter Children's Trust I
c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

Kristen Hayler Hertel Revocable Trust
c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

Anne-Carole Witort Insurance Trust
c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

Rose Shereen Fuqua Insurance Trust
c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

Rabi H. Satter Insurance Trust
c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611

Abdus Satter Insurance Trust
c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

Gordon and Barbara Anne Hertel Insurance Trust
c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

Satter Family Trust
c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

AgeChem Venture Fund L.P.
Attn: Louis Lacasse
President
1 Westmount Square, Suite 800
Montreal, Quebec, Canada
H3Z 2P9

Diane H. Janusz Trust
Attn: John Janusz
Trustee
7385 Desert Spring Court
West Chester, OH 45069

John Janusz
7385 Desert Spring Court
West Chester, OH 45069

MRK International, LLC
Attn: Richard L. Kiley
Principal Member
7800 Tecumseh Trail
Cincinnati, OH 45243

McIlwraith Investments, LLC
Attn: John McIlwraith
Manager
7680 Foxgate Lane
Cincinnati, OH 45243

John H. Wyant
Blue Chip Venture Company
1120 Scripps Center
312 Walnut Street
Cincinnati, OH 45202

Alan Fishman
6900 Stonehenge Drive
Cincinnati, OH 45242-6204

Schedule A-6

Family Akebia Investments LLC
1185 Park Avenue #11G
New York, NY 10128

Novo A/S Novo Ventures
Tuborg Havnevej 19
DK - 2900 - Hellerup

Schedule A-7

**AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED
INVESTORS' RIGHTS AGREEMENT**

THIS AMENDMENT NO. 1 TO THE THIRD AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT (this "Amendment") is made and effective as of May 31, 2013, by and among Akebia Therapeutics, Inc., a Delaware corporation (the "Company"), and the "Investors" as defined in that certain Third Amended and Restated Investors' Rights Agreement, dated as of May 10, 2013, by and among the Company and the Investors a party thereto (the "Agreement"). Capitalized terms not defined herein have the meanings given them in the Agreement.

RECITALS

WHEREAS, on May 10, 2013, the Agreement was entered into by the Company and the Investors in connection with the transactions and agreements contemplated in that certain Series C Preferred Stock Purchase Agreement by and between the Company and certain parties as listed therein, dated as of May 10, 2013 (the "Series C Stock Purchase Agreement") and the transactions and agreements contemplated therein collectively the "Series C Transaction"; and

WHEREAS, in connection with an Additional Closing (as defined in the Series C Stock Purchase Agreement) to the Series C Transaction dated May 31, 2013, the Company and the Investors holding more than the Appropriate Percentage (as defined in the Agreement) of the Registrable Securities (as defined in the Agreement) outstanding desire to amend the Agreement in accordance with the terms and conditions more fully set forth in this Amendment.

NOW, THEREFORE, in consideration of the foregoing premises and the mutual covenants hereinafter set forth, and other good and valuable consideration had and received, the parties hereto, upon the terms and subject to the conditions contained herein, hereby agree as follows:

1. Amendment of Section 6.6. Section 6.6 of the Agreement is hereby amended by deleting it in its entirety and replacing it with the following:

Amendments and Waivers. Any term of this Agreement may be amended and the observance of any term of this Agreement may be waived (either generally or in a particular instance, and either retroactively or prospectively) only with the written consent of the Company and the holders of more than the Appropriate Percentage of the Registrable Securities then outstanding; provided, that the Company may in its sole discretion waive compliance with Section 2.12(c) (and the Company's failure to object promptly in writing after notification of a proposed assignment allegedly in violation of Section 2.12(c) shall be deemed to be a waiver); and provided, further, that any provision hereof may be waived by any waiving party on such party's own behalf, without the consent of any other party. Notwithstanding the foregoing, this Agreement may not be amended or terminated and the observance of any term hereof may not be waived with respect to any Investor without the written consent of such Investor, unless such

amendment, termination, or waiver applies to all Investors in the same fashion (it being agreed that a waiver of the provisions of Section 4 with respect to a particular transaction shall be deemed to apply to all Investors in the same fashion if such waiver does so by its terms, notwithstanding the fact that certain Investors may nonetheless, by agreement with the Company, purchase securities in such transaction). Notwithstanding the foregoing, the provisions of (a) Section 2.8(d), commencing with its words “provided, however”, may not be amended or waived without the written consent of (i) the Company and (ii) the holders of ninety percent (90%) of the shares of Common Stock issuable upon conversion of the then outstanding shares of Preferred Stock and (iii) (A) Satter Investment Management, LLC so long as Satter Investment Management, LLC or any of its Affiliates holds any Preferred Stock or Common Stock issued upon conversion of any of such Preferred Stock, (B) Novartis Bioventures Ltd. so long as Novartis Bioventures Ltd. holds any Preferred Stock or Common Stock issued upon conversion of any of such Preferred Stock, and (C) Novo A/S so long as Novo A/S holds any Preferred Stock or Common Stock issued upon conversion of any of such Preferred Stock; (b) Section 3.3 may not be amended to terminate a designated right therein to appoint an observer to the Board of Directors without the written consent of the Company and at least fifty percent (50%) of the shares of Common Stock issuable upon conversion of the then outstanding shares of Preferred Stock held by the Persons entitled to appoint such observer; (c) Sections 4.1 and 4.2 may not be waived without the written consent of the holders of more than seventy percent (70%) of the shares of Common Stock issuable upon conversion of the then outstanding shares of Preferred Stock; and (d) Section 5.8, Section 6.6(a) and this Section 6.6(d) may not be amended or waived without the written consent of (i) the Company, (ii) the holders of ninety percent (90%) of the shares of Common Stock issuable upon conversion of the then outstanding shares of Preferred Stock and (iii) (A) Satter Investment Management, LLC so long as Satter Investment Management, LLC or any of its Affiliates holds any Preferred Stock or Common Stock issued upon conversion of any of such Preferred Stock, (B) Novartis Bioventures Ltd. so long as Novartis Bioventures Ltd. holds any Preferred Stock or Common Stock issued upon conversion of any of such Preferred Stock, and (C) Novo A/S so long as Novo A/S holds any Preferred Stock or Common Stock issued upon conversion of any of such Preferred Stock. The Company shall give prompt notice of any amendment or termination hereof or waiver hereunder to any party hereto that did not consent in writing to such amendment, termination, or waiver. Any amendment, termination, or waiver effected in accordance with this Section 6.6 shall be binding on all parties hereto, regardless of whether any such party has consented thereto. No waivers of or exceptions to any term, condition, or provision of this Agreement, in any one or more instances, shall be deemed to be or construed as a further or continuing waiver of any such term, condition, or provision.

2. Amendment of Section 2.8(d). The last sentence of Section 2.8(d) of the Agreement is hereby amended by deleting the parenthetical which reads “(net of any Selling Expenses) paid by such Holder)” and replacing it with the parenthetical which reads “(net of any Selling Expenses paid by such Holder)”.

3. Miscellaneous Amendments. The Agreement is amended hereby so that any reference therein to the Agreement shall be deemed to be a reference to the Agreement as amended by this Amendment.

4. Continuance of Agreement. Except as specifically amended by this Amendment, the Agreement shall remain in full force and effect.

5. Governing Law. The laws of the State of Delaware govern all matters arising out of or relating to this Amendment, including, without limitation, its interpretation, construction, performance, and enforcement, without giving effect to such state's conflicts of law principles or rules of construction concerning the drafter hereof.

6. Counterparts. This Amendment may be executed in two or more counterparts, including by facsimile or PDF signature, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument.

[Signature Page Follows]

IN WITNESS WHEREOF, the parties have caused this Amendment to be executed as of the date first written above.

AKEBIA THERAPEUTICS, INC.

By: /s/ Joseph H. Gardner
Joseph H. Gardner, Ph.D.
President and Chief Executive Officer

Address:
Suite 420,
9987 Carver Road,
Cincinnati, OH 45242

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have caused this Amendment to be executed as of the date first written above.

NOVARTIS BIO VENTURES LTD.

By: /s/ H.S. Zivi
Name: Simon Zivi
Title: Deputy Chairman

By: /s/ Rebecca White
Name: Rebecca White
Title: Authorized Signatory

Address:

NOVARTIS BIOVENTURES LTD.
Attn: Henri Simon Zivi
131 Front Street
Hamilton HM 12
Bermuda

But for mail, to:

Novartis Bio Ventures Ltd.
Attn: Henri Simon Zivi
PO Box HM 2899
Hamilton HM LX Bermuda

And, also send a copy to:

Novartis Venture Fund
Attn: Campbell Murray
Five Cambridge Center, Suite 603
Cambridge, MA 02142

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have caused this Amendment to be executed as of the date first written above.

VENTURE INVESTORS EARLY STAGE
FUND IV LIMITED PARTNERSHIP

By: VIESF IV GP, LLC, its General Partner

By: /s/ Paul M. Weiss

Name: Paul M. Weiss, Ph.D.

Title: Managing Director

Address:

505 South Rosa Road

Madison, WI 53719-1262

Attn: Paul Weiss, Managing Director

Phone: (608) 441-2700

Fax: (608) 441-2727

Email: paul@ventureinvestors.com

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have caused this Amendment to be executed as of the date first written above.

TRIATHLON MEDICAL VENTURES FUND, L.P.

By: Triathlon Medical Ventures, LLC
Title: its General Partner

By: /s/ John M. Rice
John M. Rice
Managing Partner

Address:

300 E-Business Way
Suite 200
Cincinnati, OH 45241

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have caused this Amendment to be executed as of the date first written above.

KEARNY VENTURE PARTNERS, L.P.

By: /s/ Anupam Dalal

Name: Kearny Venture Associates, LLC

Title: its General Partner

Address:

Attn: Anupam Dalal
Kearny Venture Associates LLC
88 Kearny Street, Suite 1800
San Francisco, CA 94108-5530

KEARNY VENTURE PARTNERS

ENTREPRENEURS FUND, L.P.

By: /s/ Anupam Dalal

Name: Kearny Venture Associates, LLC

Title: its General Partner

Address:

Attn: Anupam Dalal
Kearny Venture Associates LLC
88 Kearny Street, Suite 1800
San Francisco, CA 94108-5530

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
THE THIRD AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT

IN WITNESS WHEREOF, the parties have caused this Amendment to be executed as of the date first written above.

THOMAS WEISEL HEALTHCARE VENTURE PARTNERS, L.P.

By: Thomas Weisel Capital Management LLC
Title: its Managing Member

By: /s/ Anupam Dalal
Name: Anupam Dalal
Title: Managing Director

Address:
88 Kearny Street, Suite 1800
San Francisco, CA 94108

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IN WITNESS WHEREOF, the parties have caused this Amendment to be executed as of the date first written above.

ATHENIAN VENTURE PARTNERS III L.P.

By: /s/ Karl Elderkin
Name: Karl O. Elderkin
Title: President

AVP OHIO TECHNOLOGY I L.P.

By: /s/ Karl Elderkin
Name: Karl O. Elderkin
Title: President

Address:

340 West State Street
Unit 29/Suite I37D
Athens OH 45701

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IN WITNESS WHEREOF, the parties have caused this Amendment to be executed as of the date first written above.

AGECHEM VENTURE FUND L.P.

By: /s/ Louis Lacasse

Name: Louis Lacasse

Title: President

Address:

Attn: Louis Lacasse, President
1 Westmount Square, Suite 800
Montreal, Quebec, Canada
H3Z 2P9

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
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IN WITNESS WHEREOF, the parties have caused this Amendment to be executed as of the date first written above.

CINCINNATI CORNERSTONE INVESTORS
AKB, LLC

By: /s/ Robert W. Coy, Jr.
Robert W. Coy, Jr.
President

Address:

30 West 3rd Street, 6th Floor
Cincinnati, OH 45202-3559

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FAMILY AKEBIA INVESTMENTS LLC

By: /s/ Milton Berlinski

Name: Milton Berlinski

Title: Managing Member

Address:

1185 Park Avenue #11G

New York, NY 10128

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IN WITNESS WHEREOF, the parties have caused this Amendment to be executed as of the date first written above.

MUNEER A. SATTER REVOCABLE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Trustee

THE SATTER FOUNDATION

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Trustee

SATTER FAMILY TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

SATTER CHILDREN'S TRUST I

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

Address:

c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

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IN WITNESS WHEREOF, the parties have caused this Amendment to be executed as of the date first written above.

KRISTEN HAYLER HERTEL REVOCABLE TRUST

By: /s/ Kristen Hayler Hertel

Name: Kristen Hayler Hertel

Title: Trustee

Address:

c/o Satter Investment Management, LLC

676 N. Michigan Avenue, Suite 4000

Chicago, IL 60611

Attn: Muneer A. Satter

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IN WITNESS WHEREOF, the parties have caused this Amendment to be executed as of the date first written above.

ANNE-CAROLE WITORT INSURANCE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Trustee

ROSE SHEREEN FUQUA INSURANCE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

RABI H. SATTER INSURANCE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

JOHN WOOD TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

Address:

c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

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IN WITNESS WHEREOF, the parties have caused this Amendment to be executed as of the date first written above.

ABDUS SATTER INSURANCE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Trustee

GORDON AND BARBARA ANNE HERTEL INSURANCE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Trustee

Address:

c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

SIGNATURE PAGE TO AMENDMENT NO. 1 TO
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**SECOND AMENDED AND RESTATED
RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT**

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**SECOND AMENDED AND RESTATED RIGHT OF
FIRST REFUSAL AND CO-SALE AGREEMENT**

THIS SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT is made as of the 6th day of April, 2011 by and among AKEBIA THERAPEUTICS, INC., a Delaware corporation (the "Company"), the Investors listed on Schedule A and the Major Holders listed on Schedule B.

WHEREAS, each Major Holder is the beneficial owner of the number of shares of Common Stock (as defined below) set forth opposite the name of the Major Holder on Schedule B;

WHEREAS, certain of the Investors have previously purchased equity securities of the Company;

WHEREAS, the Company, certain of the Investors and certain other Persons (as defined below) previously entered into a Right of First Refusal and Co-Sale Agreement, dated as of January 23, 2008, as amended and restated as of July 15, 2009 (the "Prior Agreement");

WHEREAS, the Company and certain Investors entered into Series A Preferred Stock Purchase Agreements, dated January 23, 2008 and July 15, 2009, in connection with the purchase of shares of Series A Preferred Stock of the Company, par value \$0.00001 per share ("Series A Preferred Stock");

WHEREAS, the Company and certain Investors are parties to the Series B Preferred Stock Purchase Agreement, of even date herewith (as it may be amended, restated, supplemented or otherwise modified from time to time, the "Purchase Agreement"), pursuant to which certain Investors have agreed to purchase shares of the Series B Preferred Stock of the Company, par value \$0.00001 per share ("Series B Preferred Stock"); and

WHEREAS, the Major Holders and the Company desire further to induce the Investors to purchase the Series B Preferred Stock;

NOW, THEREFORE, the Company, the Major Holders and the Investors agree as follows:

1. Definitions.

"Affiliate" means, with respect to any specified Investor, any other Investor who directly or indirectly, controls, is controlled by or is under common control with such Investor, including without limitation any general partner, managing member, officer or director of such Investor, or any venture capital fund now or hereafter existing which is controlled by one or more general partners or managing members of, or shares the same management company with, such Investor.

"Appropriate Percentage" means sixty percent (60%) from the date hereof through June 30, 2012 and fifty-five percent (55%) as of and after July 1, 2012.

“Capital Stock” means (a) shares of Common Stock and Preferred Stock (whether now outstanding or hereafter issued in any context), (b) shares of Common Stock issued or issuable upon conversion of Preferred Stock and (c) shares of Common Stock issued or issuable upon exercise or conversion, as applicable, of stock options, warrants or other convertible securities of the Company, in each case now owned or subsequently acquired by any Major Holder, any Investor, or their respective successors or permitted transferees or assigns. Unless the context otherwise requires, for purposes of the number of shares of Capital Stock held by an Investor or Major Holder (or any other calculation based thereon), all shares of Preferred Stock shall be deemed to have been converted into Common Stock at the then-applicable conversion ratio.

“Common Stock” means shares of Common Stock of the Company, \$0.00001 par value per share.

“Company Notice” means written notice from the Company notifying a selling Major Holder that the Company intends to exercise its Right of First Refusal as to some or all of the Transfer Stock with respect to any Proposed Major Holder Transfer.

“Investor Notice” means written notice from an Investor notifying the Company and the selling Major Holder that such Investor intends to exercise its Secondary Refusal Right as to a portion of the Transfer Stock with respect to any Proposed Major Holder Transfer.

“Investors” means the persons named on Schedule A hereto, each person to whom the rights of an Investor are assigned pursuant to Section 6.9 and each person who hereafter becomes a signatory to this Agreement pursuant to Section 6.11 and any one of them, as the context may require; provided, however, that any such person shall cease to be considered an Investor for purposes of this Agreement at any time such person and his, her or its Affiliates collectively hold no shares of Preferred Stock. Notwithstanding the foregoing, for purposes of this Agreement, any reference herein to an “Investor” shall refer to that holder of shares of Preferred Stock in his, her or its capacity as a holder of such shares, and as to such shares only (exclusive of any other shares of Capital Stock which may be owned or beneficially held by such party).

“Major Holders” means the persons named on Schedule B hereto and each person who hereafter becomes a signatory to this Agreement pursuant to Section 6.17 and any one of them, as the context may require. Notwithstanding the foregoing, for purposes of this Agreement, any reference herein to a “Major Holder” shall refer to that holder of shares of Common Stock in his, her or its capacity as a holder of such shares, and as to such shares only (inclusive of any other shares of Capital Stock which may be owned or beneficially held by such party, but exclusive of any shares of Preferred Stock which may be owned or beneficially held by such party).

“Preferred Stock” means all shares of Series A Preferred Stock and Series B Preferred Stock.

“Prior Agreement” has the meaning set forth in the Recitals to this Agreement.

“Proposed Major Holder Transfer” means any assignment, sale, offer to sell, pledge, mortgage, hypothecation, encumbrance, disposition of or any other like transfer or encumbering of any Transfer Stock (or any interest therein)-proposed by any of the Major Holders.

“Proposed Transfer Notice” means written notice from a Major Holder setting forth the terms and conditions of a Proposed Major Holder Transfer.

“Prospective Transferee” means any person to whom a Major Holder proposes to make a Proposed Major Holder Transfer.

“Purchase Agreement” has the meaning set forth in the Recitals to this Agreement.

“Right of Co-Sale” means the right, but not an obligation, of an Investor to participate in a Proposed Major Holder Transfer on the terms and conditions specified in the Proposed Transfer Notice.

“Right of First Refusal” means the right, but not an obligation, of the Company, or its permitted transferees or assigns, to purchase some or all of the Transfer Stock with respect to a Proposed Major Holder Transfer, on the terms and conditions specified in the Proposed Transfer Notice.

“Secondary Notice” means written notice from the Company notifying the Investors that the Company does not intend to exercise its Right of First Refusal as to all shares of Transfer Stock with respect to any Proposed Major Holder Transfer.

“Secondary Refusal Right” means the right, but not an obligation, of each Investor to purchase up to its pro rata portion (based upon the total number of shares of Preferred Stock then held by all Investors, on an as-converted to Common Stock basis) of any Transfer Stock not purchased pursuant to the Right of First Refusal, on the terms and conditions specified in the Proposed Transfer Notice.

“Stockholders” means, collectively, the Investors and the Major Holders.

“Transfer Stock” means shares of Capital Stock, other than and in no event including Preferred Stock, owned by a Major Holder, or issued to a Major Holder after the date hereof (including, without limitation, in connection with any stock split, stock dividend, recapitalization, reorganization, or the like).

“Undersubscription Notice” means written notice from an Investor notifying the Company and the selling Major Holder that such Investor intends to exercise its option to purchase all or any portion of the Transfer Stock not purchased pursuant to the Right of First Refusal or the Secondary Refusal Right.

2. Agreement among the Company, the Investors and the Major Holders.

2.1 Right of First Refusal.

(a) Grant. Subject to the terms of Section 3 below, each Major Holder hereby unconditionally and irrevocably grants to the Company a Right of First Refusal to purchase all or any portion of Transfer Stock that such Major Holder may propose to transfer in a Proposed Major Holder Transfer, at the same price and on the same terms and conditions as those offered to the Prospective Transferee.

(b) Notice. Each Major Holder proposing to make a Proposed Major Holder Transfer must deliver a Proposed Transfer Notice to the Company and each Investor not later than forty-five (45) days prior to the consummation of such Proposed Major Holder Transfer. Such Proposed Transfer Notice shall contain the material terms and conditions (including price and form of consideration) of the Proposed Major Holder Transfer and the identity of the Prospective Transferee. To exercise its Right of First Refusal under this Section 2, the Company must deliver a Company Notice to the selling Major Holder within fifteen (15) days after delivery of the Proposed Transfer Notice. In the event of a conflict between this Agreement and any other agreement that may have been entered into by a Major Holder with the Company that contains a preexisting right of first refusal, the Company and the Major Holder acknowledge and agree that the terms of this Agreement shall control and the preexisting right of first refusal shall be deemed satisfied by compliance with Section 2.1(a) and this Section 2.1(b).

(c) Grant of Secondary Refusal Right to Investors. Subject to the terms of Section 3 below, each Major Holder hereby unconditionally and irrevocably grants to the Investors a Secondary Refusal Right to purchase all or any portion of the Transfer Stock not purchased by the Company pursuant to the Right of First Refusal, as provided in this Section 2.1(c). If the Company does not intend to exercise its Right of Refusal with respect to all Transfer Stock subject to a Proposed Major Holder Transfer, the Company must deliver a Secondary Notice to the selling Major Holder and to each Investor to that effect no later than fifteen (15) days after the selling Major Holder delivers the Proposed Transfer Notice to the Company. To exercise its Secondary Refusal Right, an Investor must deliver an Investor Notice to the selling Major Holder and the Company within ten (10) days after the Company's deadline for its delivery of the Secondary Notice as provided in the preceding sentence.

(d) Undersubscription of Transfer Stock. If options to purchase have been exercised by the Company and the Investors with respect to some but not all of the Transfer Stock by the end of the 10-day period specified in the last sentence of Section 2.1(c) (the "Investor Notice Period"), then the Company shall, immediately after the expiration of the Investor Notice Period, send written notice (the "Company Undersubscription Notice") to those Investors who fully exercised their Secondary Refusal Right within the Investor Notice Period (the "Exercising Investors"). Each Exercising Investor shall, subject to the provisions of this Section 2.1(d), have an additional option to purchase all or any part of the balance of any such remaining unsubscribed shares of Transfer Stock on the terms and conditions set forth in the Proposed Transfer Notice. To exercise such option, an Exercising Investor must deliver an Undersubscription Notice to the selling Major Holder and the Company within ten (10) days after the expiration of the Investor Notice Period. In the event there are two or more such Exercising Investors that choose to exercise the last-mentioned option for a total number of remaining shares in excess of the number available, the remaining shares available for purchase under this Section 2.1(d) shall be allocated to such Exercising Investors pro rata based on the number of shares of Transfer Stock such Exercising Investors have elected to purchase pursuant to the Secondary Refusal Right (without giving effect to any shares of Transfer Stock that any such Exercising Investor has elected to purchase pursuant to the Company Undersubscription Notice). If the options to purchase the remaining shares are exercised in full by the Exercising Investors, the Company shall immediately notify all of the Exercising Investors and the selling Major Holder of that fact.

(e) Forfeiture of Rights. Notwithstanding the foregoing, if the total number of shares of Transfer Stock that the Company and the Investors have agreed to purchase in the Company Notice, Investor Notices and Undersubscription Notices is less than the total number of shares of Transfer Stock, then the Company and the Investors shall be deemed to have forfeited any right to purchase such Transfer Stock, and the selling Major Holder shall be free to sell all, but not less than all, of the Transfer Stock to the Prospective Transferee on terms and conditions substantially similar to (and in no event more favorable than) the terms and conditions set forth in the Proposed Transfer Notice, it being understood and agreed that (i) any such sale or transfer shall be subject to the other terms and restrictions of this Agreement, including without limitation the terms and restrictions set forth in Sections 2.2 and 6.9(b); (ii) any future Proposed Major Holder Transfer shall remain subject to the terms and conditions of this Agreement, including this Section 2; and (iii) such sale shall be consummated within forty-five (45) days after receipt of the Proposed Transfer Notice by the Company and, if such sale is not consummated within such forty-five (45) day period, such sale shall again become subject to the Right of First Refusal and Secondary Refusal Right on the terms set forth herein.

(f) Consideration: Closing. If the consideration proposed to be paid for the Transfer Stock is in property, services or other non-cash consideration, the fair market value of the consideration shall be as determined in good faith by the Company's Board of Directors and as set forth in the Company Notice. If the Company or any Investor cannot, for any reason, pay for the Transfer Stock in the same form of non-cash consideration, then the Company or such Investor may pay the cash value equivalent thereof, as determined in good faith by the Board of Directors and as set forth in the Company Notice. The closing of the purchase of Transfer Stock by the Company and the Investors shall take place, and all payments from the Company and the Investors shall have been delivered to the selling Major Holder, by the later of (i) the date specified in the Proposed Transfer Notice as the intended date of the Proposed Major Holder Transfer and (ii) forty-five (45) days after delivery of the Proposed Transfer Notice.

2.2 Right of Co-Sale.

(a) Exercise of Right. Subject to the terms of Section 3, if any Transfer Stock that is the subject of a Proposed Major Holder Transfer is not purchased by the Company and/or the Investors pursuant to Section 2.1 above, and thereafter is to be sold to a Prospective Transferee, then each Investor may elect to exercise its Right of Co-Sale and participate on a pro rata basis in the Proposed Major Holder Transfer as set forth in Section 2.2(b) below and otherwise on the same terms and conditions specified in the Proposed Transfer Notice (provided that if an Investor wishes to sell Preferred Stock, then the price set forth in the Proposed Transfer Notice shall be appropriately adjusted based on the conversion ratio of the Preferred Stock into Common Stock). Each Investor who desires to exercise its Right of Co-Sale must give the selling Major Holder written notice to that effect within fifteen (15) days after the deadline for delivery of the Secondary Notice described above, and upon giving such notice, such Investor shall be deemed to have effectively exercised the Right of Co-Sale.

(b) Shares Includable. Each Investor who timely exercises such Investor's Right of Co-Sale by delivering the written notice provided for above in Section 2.2(a), may include in the Proposed Major Holder Transfer all or any part of such Investor's Capital Stock equal to the product obtained by multiplying (i) the aggregate number of shares of

Transfer Stock subject to the Proposed Major Holder Transfer (excluding shares purchased by the Company or the Investors pursuant to the Right of First Refusal or the Secondary Refusal Right) by (ii) a fraction, the numerator of which is the number of shares of Preferred Stock owned by such Investor immediately before consummation of the Proposed Major Holder Transfer (on an as-converted to Common Stock basis) and the denominator of which is the total number of shares of Preferred Stock owned, in the aggregate, by all Investors immediately prior to the consummation of the Proposed Major Holder Transfer (on an as-converted to Common Stock basis), plus the number of shares of Transfer Stock held by the selling Major Holder. To the extent one or more of the Investors exercise such right of participation in accordance with the terms and conditions set forth herein, the number of shares of Transfer Stock that the selling Major Holder may sell in the Proposed Major Holder Transfer shall be correspondingly reduced.

(c) Delivery of Certificates. Each Investor shall effect its participation in the Proposed Major Holder Transfer by delivering to the transferring Major Holder, no later than fifteen (15) days after such Investor's exercise of the Right of Co-Sale, one or more stock certificates, properly endorsed for transfer to the Prospective Transferee, representing:

(i) the number of shares of Common Stock that such Investor elects to include in the Proposed Major Holder Transfer; or

(ii) the number of shares of Preferred Stock that is at such time convertible into the number of shares of Common Stock that such Investor elects to include in the Proposed Major Holder Transfer; provided, however, that if the Prospective Transferee objects to the delivery of convertible Preferred Stock in lieu of Common Stock, such Investor shall first convert the Preferred Stock into Common Stock and deliver Common Stock as provided above. The Company agrees to make any such conversion concurrent with, and contingent upon, the actual transfer of such shares to the Prospective Transferee.

(d) Purchase Agreement. The parties hereby agree that the terms and conditions of any sale pursuant to this Section 2.2 will be memorialized in, and governed by, a written purchase and sale agreement with customary terms and provisions for such a transaction and the parties further covenant and agree to enter into such an agreement as a condition precedent to any sale or other transfer pursuant to this Section 2.2.

(e) Deliveries. Each stock certificate an Investor delivers to the selling Major Holder pursuant to Section 2.2(c) above will be transferred to the Prospective Transferee against payment therefor in consummation of the sale of the Transfer Stock pursuant to the terms and conditions specified in the Proposed Transfer Notice and the purchase and sale agreement, and the selling Major Holder shall concurrently therewith remit or direct payment to each Investor the portion of the sale proceeds to which such Investor is entitled by reason of its participation in such sale. If any Prospective Transferee refuses to purchase securities subject to the Right of Co-Sale from any Investor exercising its Right of Co-Sale hereunder, no Major Holder may sell any Transfer Stock to such Prospective Transferee or Transferees unless and until, simultaneously with such sale, such Major Holder purchases all securities subject to the Right of Co-Sale from such Investor on the same terms and conditions (including the proposed purchase price) as set forth in the Proposed Transfer Notice.

(f) Additional Compliance. If any Proposed Major Holder Transfer is not consummated within sixty (60) days after receipt of the Proposed Transfer Notice by the Company, the Major Holder proposing the Proposed Major Holder Transfer may not sell any Transfer Stock unless he or she first complies in full with each provision of this Section 2. The exercise or election not to exercise any right by any Investor hereunder shall not adversely affect its right to participate in any other sales of Transfer Stock subject to this Section 2.2.

2.3 Effect of Failure to Comply.

(a) Transfer Void: Equitable Relief. Any Proposed Major Holder Transfer not made in compliance with the requirements of this Agreement shall be null and void ab initio, shall not be recorded on the books of the Company or its transfer agent and shall not be recognized by the Company. Each party hereto acknowledges and agrees that any breach of this Agreement would result in substantial harm to the other parties hereto for which monetary damages alone could not adequately compensate. Therefore, the parties hereto unconditionally and irrevocably agree that any non-breaching party hereto shall be entitled to seek protective orders, injunctive relief and other remedies available at law or in equity (including, without limitation, seeking specific performance or the rescission of purchases, sales and other transfers of Transfer Stock not made in strict compliance with this Agreement).

(b) Violation of First Refusal Right. If any Major Holder becomes obligated to sell any Transfer Stock to the Company or any Investor under this Agreement and fails to deliver such Transfer Stock in accordance with the terms of this Agreement, the Company and/or such Investor may, at its option, in addition to all other remedies it may have, send to such Major Holder the purchase price for such Transfer Stock as is herein specified and transfer to the name of the Company or such Investor (or request that the Company effect such transfer in the name of an Investor) on the Company's books the certificate or certificates representing the Transfer Stock to be sold.

(c) Violation of Co-Sale Right. If any Major Holder purports to sell any Transfer Stock in contravention of the Right of Co-Sale (a "Prohibited Transfer"), each Investor who desires to exercise its Right of Co-Sale under Section 2.2 may, in addition to such remedies as may be available by law, in equity or hereunder, require such Major Holder to purchase from such Investor the type and number of shares of Capital Stock that such Investor would have been entitled to sell to the Prospective Transferee under Section 2.2 had the Prohibited Transfer been effected pursuant to and in compliance with the terms of Section 2.2. The sale will be made on the same terms and subject to the same conditions as would have applied had the Major Holder not made the Prohibited Transfer, except that the sale (including, without limitation, the delivery of the purchase price) must be made within ninety (90) days after the Investor learns of the Prohibited Transfer, as opposed to the timeframe proscribed in Section 2.2. Such Major Holder shall also reimburse each Investor for any and all reasonable and documented out-of-pocket fees and expenses, including reasonable legal fees and expenses, incurred pursuant to the exercise or the attempted exercise of the Investor's rights under Section 2.2.

2.4 Prohibited Transferees. Notwithstanding the foregoing, no Stockholder shall transfer any of his, her or its Capital Stock to any Competitor (as defined below) of the Company without the prior approval of the Company's Board of Directors. For purposes of this

Section 2.4. "Competitor" means any person or entity that has (i) a development program or commercial products based on small molecule inhibitors of HEF Prolyl Hydroxylase enzymes as human or animal therapeutics or (ii) a program or commercial products based on inhibitors of the HPTPbeta enzyme as human or animal therapeutics. Notwithstanding the foregoing, there shall be no prohibitions on transfers of Capital Stock by any Investor to any Affiliate of such Investor.

3. Exempt Transfers.

3.1 Exempted Transfers. Notwithstanding the foregoing or anything to the contrary herein, the provisions of Sections 2.1 and 2.2 shall not apply: (a) in the case of a Major Holder that is an entity, upon a transfer by such Major Holder to its stockholders, members, partners or other equity holders, (b) to a repurchase of Transfer Stock from a Major Holder by the Company pursuant to an agreement containing vesting and/or repurchase provisions approved by a majority of the Board of Directors, (c) to a pledge of Transfer Stock that creates a mere security interest in the pledged Transfer Stock, provided that the pledgee thereof agrees in writing in advance to be bound by and comply with all applicable provisions of this Agreement to the same extent as if it were the Major Holder making such pledge, or (d) in the case of a Major Holder that is a natural person, upon a transfer of Transfer Stock by such Major Holder made for bona fide estate planning purposes, either during his or her lifetime or on death by will or intestacy to his or her spouse, child (natural or adopted), or any other direct lineal descendant of such Major Holder (or his or her spouse) (all of the foregoing collectively referred to as "family members"), or any other relative/person approved by the Board of Directors of the Company, or any custodian or trustee of any trust, partnership or limited liability company for the benefit of, or the ownership interests of which are owned wholly by, such Major Holder or any such family members; provided that in the case of clauses (a), (c), or (d), the Major Holder shall deliver prior written notice to the Investors of such pledge, gift or transfer and such shares of Transfer Stock shall at all times remain subject to the terms and restrictions set forth in this Agreement and such transferee shall, as a condition to such issuance, deliver a counterpart signature page to this Agreement as confirmation that such transferee shall be bound by all the terms and conditions of this Agreement as a Major Holder (but only with respect to the securities so transferred to the transferee), including the obligations of a Major Holder with respect to Proposed Major Holder Transfers of such Transfer Stock pursuant to Section 2; and provided, further, in the case of any transfer pursuant to clause (a) or (d) above, that such transfer is made pursuant to a transaction in which there is no consideration actually paid for such transfer.

3.2 Exempted Offerings. Notwithstanding the foregoing or anything to the contrary herein, the provisions of (a) Section 2 shall not apply to the sale of any Transfer Stock to the public in an offering pursuant to an effective registration statement under the Securities Act of 1933, as amended (a "Public Offering") and (b) Section 2 shall not apply to the sale of any Transfer Stock pursuant to a Sale of the Company (as defined in that certain Voting Agreement dated as of the date hereof, as it may be amended, restated, supplemented or otherwise modified from time to time), among the Company, the Investors named therein and the Major Holders named therein.

4. Legend. Each certificate representing shares of Capital Stock held by the Stockholders or issued to any permitted transferee in connection with a transfer permitted by Section 3.1 hereof shall be endorsed with the following legend:

THE SALE, PLEDGE, HYPOTHECATION OR TRANSFER OF THE SHARES REPRESENTED BY THIS CERTIFICATE IS SUBJECT TO, AND IN CERTAIN CASES PROHIBITED BY, THE TERMS AND CONDITIONS OF A CERTAIN RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT BY AND AMONG THE STOCKHOLDER, THE COMPANY AND CERTAIN OTHER HOLDERS OF STOCK OF THE COMPANY. COPIES OF SUCH AGREEMENT MAY BE OBTAINED UPON WRITTEN REQUEST TO THE SECRETARY OF THE COMPANY.

Each Stockholder agrees that the Company may instruct its transfer agent to impose transfer restrictions on the shares represented by certificates bearing the legend referred to in this Section 4 to enforce the provisions of this Agreement, and the Company agrees to promptly do so. The legend shall be removed upon termination of this Agreement at the request of the holder.

5. Lock-Up.

5.1 "Market Stand-off" Agreement. Each Stockholder hereby agrees that it will not, without the prior written consent of the managing underwriter, during the period commencing on the date of the final prospectus relating to the registration by the Company of shares of its Common Stock or any other equity securities under the Securities Act on a registration statement on Form S-1 or Form S-3, and ending on the date specified by the Company and the managing underwriter (such period not to exceed (x) one hundred eighty (180) days in the case of the Company's initial public offering ("IPO"), which period may be extended upon the request of the managing underwriter, to the extent required by any FINRA rules, for an additional period of up to fifteen (15) days if the Company issues or proposes to issue an earnings or other public release within fifteen (15) days of the expiration of the 180-day lockup period, or (y) if requested by the managing underwriter and approved by holders of more than the Appropriate Percentage of the shares of Common Stock issued or issuable upon conversion of the Preferred Stock held by the Investors, ninety (90) days in the case of any registration other than the IPO, which period may be extended upon the request of the managing underwriter, to the extent required by any FINRA rules, for an additional period of up to fifteen (15) days if the Company issues or proposes to issue an earnings or other public release within fifteen (15) days of the expiration of the 90-day lockup period), (i) lend; offer; pledge; sell; contract to sell; sell any option or contract to purchase; purchase any option or contract to sell; grant any option, right, or warrant to purchase; or otherwise transfer or dispose of, directly or indirectly, any shares of Common Stock or any securities convertible into or exercisable or exchangeable (directly or indirectly) for Common Stock held immediately before the effective date of the registration statement for such offering or (ii) enter into any swap or other arrangement that transfers to another, in whole or in part, any of the economic consequences of ownership of such securities, whether any such transaction described in clause (i) or (ii) above is to be settled by delivery of Common Stock or other securities, in cash, or otherwise. The foregoing provisions of this Section 5.1 shall not apply to the sale of any shares to an underwriter pursuant to an underwriting agreement, and shall be applicable to the Stockholders only if all officers and directors are subject to the same restrictions and the Company uses

commercially reasonable efforts to obtain a similar agreement from all stockholders individually owning more than one percent (1%) of the Company's outstanding Common Stock (after giving effect to conversion into Common Stock of all outstanding Preferred Stock). The underwriters in connection with such registration are intended third-party beneficiaries of this [Section 5.1](#) and shall have the right, power, and authority to enforce the provisions hereof as though they were a party hereto. Each Stockholder further agrees to execute such agreements as may be reasonably requested by the underwriters in connection with such registration that are consistent with this [Section 5.1](#) or that are necessary to give further effect thereto.

5.2 [Stop Transfer Instructions](#). In order to enforce the foregoing covenant, the Company may impose stop-transfer instructions with respect to the shares of Capital Stock of each Stockholder (and transferees and assignees thereof) until the end of such restricted period.

6. [Miscellaneous](#).

6.1 [Term](#). This Agreement shall automatically expire upon the earlier of immediately prior to the consummation of the Company's IPO and (b) the consummation of a Deemed Liquidation Event (as defined in the Company's Certificate of Incorporation); [provided](#) that the provisions of [Section 5](#) shall survive the expiration of this Agreement until terminated pursuant to the provisions of [Section 6.8](#).

6.2 [Stock Split](#). All references to numbers of shares in this Agreement shall be appropriately adjusted to reflect any stock dividend, split, combination or other recapitalization affecting the Capital Stock occurring after the date of this Agreement.

6.3 [Ownership](#). Each Major Holder represents and warrants that such Major Holder is the sole legal and beneficial owner of the shares of Transfer Stock subject to this Agreement and that no other person or entity has any interest in such shares (other than a community property interest as to which the holder thereof has acknowledged and agreed in writing to the restrictions and obligations hereunder).

6.4 [Reserved.]

6.5 [Notices](#). All notices and other communications given or made pursuant to this Agreement shall be in writing and shall be deemed effectively given upon the earlier of actual receipt or: (a) personal delivery to the party to be notified, (b) when sent, if sent by electronic mail or facsimile during normal business hours of the recipient, and if not sent during normal business hours, then on the recipient's next business day, (c) five (5) days after having been sent by registered or certified mail, return receipt requested, postage prepaid, or (d) one (1) business day after deposit with a nationally recognized overnight courier, freight, prepaid, specifying next business day delivery, with written verification of receipt. All communications shall be sent to the respective parties at their address as set forth on [Schedule A](#) or [Schedule B](#) hereof, as the case may be, or to such email address, facsimile number or address as subsequently modified by written notice given in accordance with this [Section 6.5](#).

If notice is given to the Company, a copy shall also be sent to:

Thompson Hine LLP
312 Walnut Street, 14th Floor
Cincinnati, OH 45202-4089
Attn: David J. Willbrand
david.willbrand@thompsonhine.com

If notice is given to the Investors, a copy shall also be sent to:

Edwards Angell Palmer & Dodge LLP
Attn: Al Sokol
111 Huntington Avenue
Boston, MA 02199 USA
asokol@eapdlaw.com

6.6 Entire Agreement. This Agreement (including the Exhibits and Schedules hereto) amends and restates in its entirety the prior Agreement and constitutes the full and entire understanding and agreement between the parties with respect to the subject matter hereof, and any other written or oral agreement relating to the subject matter hereof existing between the parties are expressly canceled.

6.7 Delays or Omissions. No delay or omission to exercise any right, power or remedy accruing to any party under this Agreement, upon any breach or default of any other party under this Agreement, shall impair any such right, power or remedy of such non-breaching or non-defaulting party nor shall it be construed to be a waiver of any such breach or default, or an acquiescence therein, or of or in any similar breach or default thereafter occurring; nor shall any waiver of any single breach or default be deemed a waiver of any other breach or default theretofore or thereafter occurring. Any waiver, permit, consent or approval of any kind or character on the part of any party of any breach or default under this Agreement, or any waiver on the part of any party of any provisions or conditions of this Agreement, must be in writing and shall be effective only to the extent specifically set forth in such writing. All remedies, either under, this Agreement or by law or otherwise afforded to any party, shall be cumulative and not alternative.

6.8 Amendment: Waiver and Termination. This Agreement may be amended, modified or terminated (other than pursuant to Section 6.1 above) and the observance of any term hereof may be waived (either generally or in a particular instance and either retroactively or prospectively) only by a written instrument executed by (a) the Company, and (b) the holders of more than the Appropriate Percentage of the shares of Common Stock issued or issuable upon conversion of the then outstanding shares of Preferred Stock held by the Stockholders (voting as a single class and on an as-converted basis), provided that any amendment of the provisions of Section 2 shall also require the written consent of Major Holders holding at least a majority of the shares of Transfer Stock then held by all of the Major Holders. Any amendment, modification, termination or waiver so effected shall be binding upon the Company, the Investors, the Major Holders and all of their respective successors and permitted assigns whether or not such party, assignee or other shareholder entered into or approved such amendment, modification, termination or waiver. Notwithstanding the foregoing, (i) this Agreement may not be amended, modified or terminated and the observance of any term hereunder may not be waived with respect to any Stockholder without the written consent of

such Stockholder unless such amendment, modification, termination of waiver applies to all Stockholders in the same fashion and (ii) the consent of the Major Holders shall not be required for any amendment, modification, termination or waiver if such amendment, modification, termination or waiver does not apply to the Major Holders in their capacity as Major Holders. The Company shall give prompt written notice of any amendment, modification or termination hereof or waiver hereunder to any party hereto that did not consent in writing to such amendment, modification, termination or waiver. No waivers of or exceptions to any term, condition or provision of this Agreement, in any one or more instances, shall be deemed to be, or construed as, a further or continuing waiver of any such term, condition or provision.

6.9 Assignment of Rights.

(a) The terms and conditions of this Agreement shall inure to the benefit of, and be binding upon, the respective successors and permitted assigns of the parties. Nothing in this Agreement, express or implied, is intended to confer upon any party other than the parties hereto or their respective successors and permitted assigns any rights, remedies, obligations, or liabilities under or by reason of this Agreement, except as expressly provided in this Agreement.

(b) Any successor or permitted assignee of any Major Holder, including any Prospective Transferee who purchases shares of Transfer Stock in accordance with the terms hereof, shall deliver to the Company and the Investors, as a condition to any transfer or assignment, a counterpart signature page hereto pursuant to which such successor or permitted assignee shall confirm their agreement to be subject to and bound by all of the provisions set forth in this Agreement that were applicable to the predecessor or assignor of such successor or permitted assignee.

(c) The rights of the Investors hereunder are not assignable without the Company's written consent (which shall not be unreasonably withheld, delayed or conditioned), except (i) by an Investor to any Affiliate or (ii) to an assignee or transferee who acquires at least 100,000 shares of Capital Stock (as adjusted for any stock combination, stock split, stock dividend, recapitalization or other similar transaction), it being acknowledged and agreed that any such assignment, including an assignment contemplated by the preceding clauses (i) or (ii) shall be subject to and conditioned upon any such assignee's delivery to the Company and the other Investors of a counterpart signature page hereto pursuant to which such assignee shall confirm their agreement to be subject to and bound by all of the provisions set forth in this Agreement that were applicable to the assignor of such assignee.

(d) Except in connection with an assignment by the Company by operation of law to the acquirer of the Company, the rights and obligations of the Company hereunder may not be assigned under any circumstances.

6.10 Severability. The invalidity or unenforceability of any provision hereof shall in no way affect the validity or enforceability of any other provision.

6.11 Additional Investors. Notwithstanding anything to the contrary contained herein, if the Company issues additional shares of the Company's Preferred Stock after the date hereof, any purchaser of such shares of Preferred Stock may become a party to this Agreement by executing and delivering an additional counterpart signature page to this Agreement and thereafter shall be deemed an "Investor" for all purposes hereunder.

6.12 Governing Law. This Agreement shall be governed by, and construed in accordance with, the laws of the State of Delaware, regardless of the laws that might otherwise govern under applicable principles of conflicts of law.

6.13 Titles and Subtitles. The titles and subtitles used in this Agreement are used for convenience only and are not to be considered in construing or interpreting this Agreement.

6.14 Counterparts: Facsimile or PDF. This Agreement may be executed in two or more counterparts, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument. This Agreement may also be executed and delivered by facsimile or PDF signature and in two or more counterparts, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument.

6.15 Aggregation of Stock. All shares of Capital Stock held or acquired by Affiliated entities or persons shall be aggregated together for the purpose of determining the availability of any rights under this Agreement and such Affiliated persons may apportion such rights as among themselves in any manner they deem appropriate.

6.16 Specific Performance. In addition to any and all other remedies that may be available at law in the event of any breach of this Agreement, each Investor shall be entitled to specific performance of the agreements and obligations of the Company and the Investor hereunder and to such other injunction or other equitable relief as may be granted by a court of competent jurisdiction.

6.17 Additional Major Holders. In the event that after the date of this Agreement, the Company issues shares of Common Stock, or options to purchase Common Stock, to any employee or consultant, which shares or options would collectively constitute with respect to such employee or consultant (taking into account all shares of Common Stock, options and other purchase rights held by such employee or consultant) one percent (1%) or more of the Company's then outstanding Common Stock (treating for this purpose all shares of Common Stock issuable upon exercise of or conversion of outstanding options, warrants or convertible securities, as if exercised or converted), the Company shall, as a condition to such issuance, cause such employee or consultant to execute a counterpart signature page hereto as a Major Holder, and such person shall thereby be bound by, and subject to, all the terms and provisions of this Agreement applicable to a Major Holder.

[Remainder of Page Intentionally Left Blank]

IN WITNESS WHEREOF, the parties have executed this Second Amended and Restated Right of First Refusal and Co-Sale Agreement as of the date first written above.

AKEBIA THERAPEUTICS, INC.

By: /s/ Joseph H. Gardner
Joseph H. Gardner, Ph.D.
President and Chief Executive Officer

Address:

Suite 420,
9987 Carver Road,
Cincinnati, OH 45242

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
FIRST REFUSAL AND CO-SALE AGREEMENT

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NOVARTIS BIOVENTURES LTD.

By: /s/ H. S. Zivi

Name: H. S. Zivi

Title: Deputy Chairman

By: /s/ Hanna Szepietowska

Name: Hanna Szepietowska

Title: Authorized Signatory

NOVARTIS BIOVENTURES LTD.

Attn: Henri Simon Zivi

131 Front Street

Hamilton HM 12

Bermuda

But for mail, to:

Novartis BioVentures Ltd.

Attn: Henri Simon Zivi

PO Box HM 2899

Hamilton HM LX Bermuda

And, also send a copy to:

Novartis Venture Fund

Attn: Campbell Murray

Five Cambridge Center, Suite 603

Cambridge, MA 02142

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
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VENTURE INVESTORS EARLY STAGE FUND IV LIMITED
PARTNERSHIP

By: VIESF IV GP, LLC, its General Partner

By: /s/ Paul M. Weiss

Name: Paul M. Weiss, PhD

Title: Managing Director

Address:

505 South Rosa Road
Madison, WI 53719-1262
Attn: Paul Weiss, Managing Director
Phone: (608) 441-2700
Fax: (608) 441-2727
Email: paul@ventureinvestors.com

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
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TRIATHLON MEDICAL VENTURES FUND, L.P.

By: Triathlon Medical Ventures, LLC

Its: General Partner

By: /s/ John M. Rice

John M. Rice

Managing Partner

Address:

1100 Chiquita Center

250 East Fifth Street

Cincinnati, OH 45202

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
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KEARNY VENTURE PARTNERS, L.P.

By: /s/ Anupam Dalal
Name: Kearny Venture Associates, LLC
Title: its General Partner

Address:

Attn: Anupam Dalal
Kearny Venture Associates LLC
88 Kearny Street, Suite 200
San Francisco, CA 94108-5530

KEARNY VENTURE PARTNERS, L.P. ENTREPRENEURS FUND,
L.P.

By: /s/ Anupam Dalal
Name: Kearny Venture Associates, LLC
Title: its General Partner

Address:

Attn: Anupam Dalal
Kearny Venture Associates LLC
88 Kearny Street, Suite 200
San Francisco, CA 94108-5530

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THOMAS WEISEL HEALTHCARE VENTURE PARTNERS, L.P.

By: Thomas Weisel Capital Management LLC
Title: its Managing Member

By: /s/ Anupam Dalal

Name: DALAL

Title: Partner

Address:

88 Kearny Street, 4th Floor
San Francisco, CA 94108

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
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ATHENIAN VENTURE PARTNERS III L.P.

By: /s/ Karl O. Elderkin

Name: Athenian III, Ltd.

Title: General Partner

Address:

20 East Circle Drive, #37146

Suite 229

Athens, OH 45701

AVP OHIO TECHNOLOGY I L.P.

By: AVP Ohio 1, Ltd.

Its: General Partner

By: /s/ Karl O. Elderkin

Karl O. Elderkin

President

Address:

20 East Circle Drive, #37146

Suite 229

Athens, OH 45701

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
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SIGVION FUND I, LP

By: /s/ J. P. Fairbank
J. P. Fairbank
Founding Partner

Address:

806 West Washington Street, Suite 204
Chicago, IL 60607

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Second Amended and Restated Right of First Refusal and Co-Sale Agreement as of the date first written above.

JOSEPH H. GARDNER

By: /s/ Joseph H. Gardner

Address:

4060 Boomer Road
Cincinnati, OH 45247

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
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ROBERT SHALWITZ

By: /s/ Robert Shalwitz

Address:

2549 Bryden Road
Bexley, OH 43209

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
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FRED SHALWITZ TRUST;
ROBERT SHALWITZ, TRUSTEE

By: /s/ Robert Shalwitz
Name: Robert Shalwitz
Title: Trustee

Address:
2549 Bryden Road
Bexley, OH 43209

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
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BLUE CHIP VALIDATION FUND, LTD.

By: Blue Chip Venture Company, Ltd.

Its: Manager

By: /s/ John McIlwraith

John McIlwraith

Managing Director

Address:

1100 Chiquita Center

250 East Fifth Street

Cincinnati, OH 45202

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
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QCA FIRST FUND II

By: /s/ John Habbert

Name: John Habbert

Title: Manager

Address:

6393 Grand Vista Avenue
Cincinnati, OH 45213

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
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TRI-STATE GROWTH CAPITAL FUND II, L.P.

By: Tri-State Ventures II, LLC
Its: General Partner

By: Fort Washington Investment Advisors, Inc.
Its: Managing Member

By: /s/ Christopher L. Baucom

Christopher L. Baucom
Managing Director

By: /s/ Maribeth S. Rahe

Maribeth S. Rahe
President and Chief Executive Officer

Address:

303 Broadway, Suite 1200
Cincinnati, OH 45202

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
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GITANA FAMILY TRUST, ELIZABETH C. ARMITAGE TRUSTEE

By: /s/ Elizabeth C. Armitage

Elizabeth C. Armitage
Trustee

Address:

2207 Upland Place
Cincinnati, OH 45206

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CITYTECH FUND I, LLC

By: /s/ Robert W. Coy, Jr.

Robert W. Coy, Jr.
President

Address:

30 West 3rd Street, 6th Floor
Cincinnati, OH 45202-3559

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
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IN WITNESS WHEREOF, the parties have executed this Second Amended and Restated Right of First Refusal and Co-Sale Agreement as of the date first written above.

GARDNER FAMILY TRUST,

John D. Gardner TRUSTEE

By: /s/ John D. Gardner
John D. Gardner
Trustee

Address:

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Second Amended and Restated Right of First Refusal and Co-Sale Agreement as of the date first written above.

IAN A. W. HOWES, IRA, STERLING TRUST CUSTODIAN

By: /s/ Ian A. W. Howes

Name: Ian A. W. Howes

Address:

219 Stratford Drive
Chapel Hill, NC 27516

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Second Amended and Restated Right of First Refusal and Co-Sale Agreement as of the date first written above.

Kevin Peters

By: /s/ Kevin Peters

Address:

6100 Miami Rd.
Cincinnati OH, 45243

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
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IN WITNESS WHEREOF, the parties have executed this Second Amended and Restated Right of First Refusal and Co-Sale Agreement as of the date first written above.

Ian A. W. Howes

By: /s/ Ian A. W. Howes

Address:

219 Stratford Drive
Chapel Hill, NC 27816

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Second Amended and Restated Right of First Refusal and Co-Sale Agreement as of the date first written above.

MUNEER A. SATTER REVOCABLE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

THE SATTER FOUNDATION

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

SATTER FAMILY TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

SATTER CHILDREN'S TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

Address:

71 S. Wacker Drive, Suite 500

Chicago, IL 60606

Attn: Muneer A. Satter

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties have executed this Second Amended and Restated Right of First Refusal and Co-Sale Agreement as of the date first written above.

AGECHEM VENTURE FUND L.P.

By: AgeChem Financial Inc., its General Partner

By: /s/ Louise Lacasse

Name: Louise Lacasse

Title: President

Address:

c/o GeneChem Management Inc.
Attn : President
1001 de Maisonneuve Blvd, West.
Suite 920
Montreal, Quebec
H3A 3C8

Acknowledged and agreed to by:

AKEBIA THERAPEUTICS, INC.

By: /s/ Joseph H. Gardner

Joseph H. Gardner, Ph.D.

President and Chief Executive Officer

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
FIRST REFUSAL AND CO-SALE AGREEMENT
CLOSING 1 (EXTENDED)

IN WITNESS WHEREOF, the parties have executed this Second Amended and Restated Right of First Refusal and Co-Sale Agreement as of the date first written above.

DIANE H. JANUSZ TRUST

By: /s/ John M. Janusz

Name: John M. Janusz

Title: Trustee

Address:

7385 Desert Spring Ct.,
West Chester, OH 45069

Acknowledged and agreed to by:

AKEBIA THERAPEUTICS, INC.

By: /s/ Joseph H. Gardner

Joseph H. Gardner, Ph.D.

President and Chief Executive Officer

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
FIRST REFUSAL AND CO-SALE AGREEMENT
CLOSING 1 (EXTENDED)

IN WITNESS WHEREOF, the parties have executed this Second Amended and Restated Right of First Refusal and Co-Sale Agreement as of the date first written above.

THE PROCTER & GAMBLE COMPANY

By: /s/ Jim Prevost
Name: Jim Prevost
Title: VP Acquisitions and Divestitures

Address:

Attn:
One Procter & Gamble Plaza
Cincinnati, OH 45202

Acknowledged and agreed to by:

AKEBIA THERAPEUTICS, INC.

By: /s/ Joseph H. Gardner
Joseph H. Gardner, Ph.D.
President and Chief Executive Officer

SIGNATURE PAGE TO SECOND AMENDED AND RESTATED RIGHT OF
FIRST REFUSAL AND CO-SALE AGREEMENT
CLOSING 1 (EXTENDED)

SCHEDULE A
INVESTORS
(Updated to reflect Closing 1 (Extended))

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock Purchased 1st Closing</u>	<u>Number of Shares of Series B Preferred Stock Purchased 1st Closing (Extended)</u>	<u>Number of Shares of Series B Preferred Stock Purchased 2nd Closing</u>	<u>Number of Shares of Series B Preferred Stock Purchased 3rd Closing</u>
Novartis Bioventures Ltd. Attn: Henri Simon Zivi Novartis BioVentures Ltd. 131 Front Street Hamilton HM 12 Bermuda But for mail, to: Novartis BioVentures Ltd. Attn: Henri Simon Zivi PO Box HM 2899 Hamilton HM LX Bermuda And, also send a copy to: Novartis Venture Fund Attn: Campbell Murray Five Cambridge Center, Suite 603 Cambridge, MA 02142 And Edwards Angell Palmer & Dodge LLP Attn: Al Sokol 111 Huntington Avenue Boston, MA 02199 asokol@eapdlaw.com	0	25,703,116	27,013,221	0		

Schedule A-1

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock Purchased 1st Closing</u>	<u>Number of Shares of Series B Preferred Stock Purchased 1st Closing (Extended)</u>	<u>Number of Shares of Series B Preferred Stock Purchased 2nd Closing</u>	<u>Number of Shares of Series B Preferred Stock Purchased 3rd Closing</u>
Venture Investors Early Stage Fund IV Limited Partnership Attn: Paul Weiss Managing Director 505 South Rosa Road Madison, WI 53719-1262	0	15,743,452	13,506,610	0		
Triathlon Medical Ventures Fund, L.P. Attn: John M. Rice Managing Partner 1100 Chiquita Center 250 East Fifth Street Cincinnati, OH 45202	2,000,000	12,809,546	9,669,049	0		
Kearny Venture Partners, L.P. Attn: Anupam Dalal Kearny Venture Associates LLC 88 Kearny Street, Suite 200 San Francisco, CA 94108-5530	0	4,460,615	3,367,012	0		
Kearny Venture Partners Entrepreneurs Fund, L.P. Attn: Anupam Dalal Kearny Venture Associates LLC 88 Kearny Street, Suite 200 San Francisco, CA 94108-5530	0	90,978	68,673	0		
Thomas Weisel Healthcare Venture Partners, L.P. Attn: Richard Spalding 88 Kearny Street, 4th Floor San Francisco, CA 94108	0	4,551,599	3,435,690	0		

Schedule A-2

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock Purchased 1st Closing</u>	<u>Number of Shares of Series B Preferred Stock Purchased 1st Closing (Extended)</u>	<u>Number of Shares of Series B Preferred Stock Purchased 2nd Closing</u>	<u>Number of Shares of Series B Preferred Stock Purchased 3rd Closing</u>
Athenian Venture Partners III L.P. Attn: Karl O. Elderkin President 20 East Circle Drive #37146 Suite 229 Athens, OH 45701	0	2,296,477	2,251,101	0		
AVP Ohio Technology I L.P. Attn: Karl O Elderkin President 20 East Circle Drive #37146 Suite 229 Athens, OH 45701	0	765,492	900,441	0		
Sigvion Fund 1 LP Attn: J.P. Fairbank Founding Partner 806 W. Washington Street, Suite 204 Chicago, IL 60607	800,000	1,366,069	1,031,151	0		
Joseph H. Gardner 4060 Boomer Road Cincinnati, OH 45247	1,920,300	903,887	900,441	0		
Robert Shalwitz 2549 Bryden Road Bexley, OH 43209	1,371,436	84,953	160,792	0		
Blue Chip Validation Fund, Ltd. Attn: John McIlwraith Managing Director 1100 Chiquita Center 250 East Fifth Street Cincinnati, OH 45202	0	340,209	0	0		

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock Purchased 1st Closing</u>	<u>Number of Shares of Series B Preferred Stock Purchased 1st Closing (Extended)</u>	<u>Number of Shares of Series B Preferred Stock Purchased 2nd Closing</u>	<u>Number of Shares of Series B Preferred Stock Purchased 3rd Closing</u>
QCA First Fund II Attn: John Habbert 1776 Mentor Avenue, MB # 02 Cincinnati, OH 45212	0	337,500	0	0		
Tri-State Growth Capital Fund II, L.P. Attn: Christopher L. Baucom Managing Director 303 Broadway, Suite 1200 Cincinnati, OH 45202	0	1,280,465	966,535	0		
Gitana Family Trust, Elizabeth C. Armitage Trustee Attn: Elizabeth C. Armitage Trustee 2207 Upland Place Cincinnati, OH 45206	0	276,557	64,317	0		
CincyTech Fund I, LLC Attn: Robert W. Coy, Jr. 30 West 3rd Street, 6th Floor Cincinnati, OH 45202-3559	0	1,312,275	990,546	0		
Gardner Family Trust Attn: John Gardner 111 Pine Court Bastrop, TX 78602-7416	0	690,199	566,006	0		
Ian A.W. Howes, IRA, Sterling Trust Custodian 219 Stratford Drive Chapel Hill, NC 27516	0	500,000	450,022	0		
Kevin Peters 6100 Miami Road - Cincinnati, OH 45243	1,060,326	0	112,555	0		

Schedule A-4

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock Purchased 1st Closing</u>	<u>Number of Shares of Series B Preferred Stock Purchased 1st Closing (Extended)</u>	<u>Number of Shares of Series B Preferred Stock Purchased 2nd Closing</u>	<u>Number of Shares of Series B Preferred Stock Purchased 3rd Closing</u>
Muneer A. Satter Revocable Trust 71 S. Wacker Drive, Suite 500 Chicago, IL 60606 Attn: Muneer A. Satter	0	0	9,004,407	0		
The Satter Foundation 71 S. Wacker Drive, Suite 500 Chicago, IL 60606 Attn: Muneer A. Satter	0	0	4,502,203	0		
Satter Family Trust 71 S. Wacker Drive, Suite 500 Chicago, IL 60606 Attn: Muneer A. Satter	0	0	2,251,101	0		
Satter Children's Trust 71 S. Wacker Drive, Suite 500 Chicago, IL 60606 Attn: Muneer A. Satter	0	0	4,502,203	0		
AgeChem Venture Fund L.P. c/o GeneChem Management Inc. Attn : President 1001 de Maisonneuve Blvd, West Suite 920 Montreal, Quebec H3A 3C8	0	0	0	13,506,608		
Diane H. Janusz Trust 7385 Desert Spring Ct., West Chester, OH 45069	0	0	0	112,554		
The Procter & Gamble Company Attn: David Le Neveu and Jim Prevost One Procter & Gamble Plaza Cincinnati, OH 45202	7,204,744	0	0	276,142		

Schedule A-5

**SCHEDULE B
MAJOR HOLDERS**

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>
Joseph H. Gardner 4060 Boomer Road Cincinnati, OH 45247	1,920,300 (plus 2,264,723 options)
Robert Shalwitz 2549 Bryden Road Bexley, OH 43209	1,371,436 (plus 1,937,652 options)
Ian A.W. Howes Address	1,055,639 (plus 160,937 options)
Kevin Peters Address	1,060,326 (plus 156,250 options)

Schedule B-1

**AMENDMENT NUMBER ONE TO
SECOND AMENDED AND RESTATED
RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT**

THIS AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT (the "Amendment") is made and effective as of July 9, 2012, by and among AKEBIA THERAPEUTICS, INC., a Delaware corporation (the "Company"), and those Investors executing and delivering a counterpart signature page hereto. Capitalized terms not defined herein have the meanings given them in that certain Second Amended and Restated Right of First Refusal and Co-Sale Agreement, dated as of April 6, 2011, by and among the Company, the Investors and the Major Holders (as amended, the "Agreement").

WHEREAS, the Company and the Investors desire to amend the Agreement to modify the definition of Preferred Stock and add a definition for the Series X Preferred Stock of the Company; and

WHEREAS, the Investors hold more than the Appropriate Percentage of the shares of Common Stock required to amend the Agreement pursuant to the provisions of Section 6.8 of the Agreement;

NOW, THEREFORE, in consideration of the foregoing recitals and the mutual promises hereinafter set forth and set forth in the Agreement, the parties hereby agree as follows:

1. Amendment.

(a) The definition of "Preferred Stock" shall be deleted in its entirety and replaced with the following: "'Preferred Stock' means all shares of Series A Preferred Stock, Series B Preferred Stock, and Series X Preferred Stock."

(b) A new definition of "Series X Preferred Stock" shall be added to the Agreement and shall read as follows: "'Series X Preferred Stock' shall mean shares of the Series X Preferred Stock of the Company, par value \$0.00001 per share."

2. Miscellaneous Amendments. The Agreement is amended hereby so that any reference therein to the Agreement shall be deemed to be a reference to the Agreement as amended by this Amendment.

3. Continuance of Agreement. Except as specifically amended by this Amendment, the Agreement shall remain in full force and effect.

4. Governing Law. The laws of the State of Delaware govern all matters arising out of or relating to this Amendment, including, without limitation, its interpretation, construction, performance, and enforcement, without giving effect to such state's conflicts of law principles or rules of construction concerning the drafter hereof.

6. Counterparts. This Amendment may be executed in two or more counterparts, including by facsimile or PDF signature, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument.

[Signature Page Follows]

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

AKEBIA THERAPEUTICS, INC.

By: /s/ Joseph H. Gardner

Joseph H. Gardner, Ph.D.

President and Chief Executive Officer

Address:

Suite 420,

9987 Carver Road,

Cincinnati, OH 45242

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

JOSEPH H. GARDNER

By: /s/ JOSEPH H. GARDNER

Address:

4060 Boomer Road
Cincinnati, OH 45247

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

NOVARTIS BIOVENTURES LTD.

By: /s/ H. S. Zivi
Name: H. S. Zivi
Title: Deputy Chairman

By: /s/ Rebecca White
Name: Rebecca White
Title: Authorized Signatory

Address:

NOVARTIS BIOVENTURES LTD.
Attn: Henri Simon Zivi
131 Front Street
Hamilton HM 12
Bermuda

But for mail, to:
Novartis BioVentures Ltd.
Attn: Henri Simon Zivi
PO Box HM 2899
Hamilton HM LX Bermuda

And, also send a copy to:
Novartis Venture Fund
Attn: Campbell Murray
Five Cambridge Center, Suite 603
Cambridge, MA 02142

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

VENTURE INVESTORS EARLY STAGE FUND IV LIMITED
PARTNERSHIP

By: VIESF IV GP, LLC, its General Partner

By: /s/ Paul M. Weiss

Name: Paul M. Weiss, PhD

Title: Managing Director

Address:

505 South Rosa Road

Madison, WI 53719-1262

Attn: Paul Weiss, Managing Director

Phone: (608) 441-2700

Fax: (608) 441-2727

Email: paul@ventureinvestors.com

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

TRIATHLON MEDICAL VENTURES FUND, L.P.

By: Triathlon Medical Ventures, LLC
Its: General Partner

By: /s/ John M. Rice
John M. Rice
Managing Partner

Address:

300 E-Business Way
Suite 200
Cincinnati, OH 45241

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

KEARNY VENTURE PARTNERS, L.P.

By: /s/ Anupam Dalal

Name: Kearny Venture Associates, LLC

Title: its General Partner

Address:

Attn: Anupam Dalal
Kearny Venture Associates LLC
88 Kearny Street, Suite 1800
San Francisco, CA 94108-5530

KEARNY VENTURE PARTNERS ENTREPRENEURS FUND, L.P.

By: /s/ Anupam Dalal

Name: Kearny Venture Associates, LLC

Title: its General Partner

Address:

Attn: Anupam Dalal
Kearny Venture Associates LLC
88 Kearny Street, Suite 1800
San Francisco, CA 94108-5530

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

THOMAS WEISEL HEALTHCARE VENTURE PARTNERS, L.P.

By: Thomas Weisel Capital Management LLC
Title: its Managing Member

By: /s/ Anupam Dalal
Name: Anupam Dalal
Title: Managing Director

Address:
88 Kearny Street, Suite 1800
San Francisco, CA 94108

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

ATHENIAN VENTURE PARTNERS III L.P.

By: /s/ Karl Elderkin
Karl Elderkin, President
Name: Athenian III, Ltd
Title: General Partner

Address:
20 East Circle Drive, #37146
Suite 229
Athens, OH 45701

AVP OHIO TECHNOLOGY I L.P.

By: AVP Ohio I, Ltd.
Its: General Partner

By: /s/ Karl O. Elderkin
Karl O. Elderkin
President

Address:
20 East Circle Drive, #37146
Suite 229
Athens, OH 45701

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

CINCYTECH FUND I, LLC

By: /s/ Robert W. Coy, Jr
Robert W. Coy, Jr
President

Address:

30 West 3rd Street, 6th Floor
Cincinnati, OH 45202-3559

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

GITANA FAMILY TRUST, ELIZABETH C. ARMITAGE TRUSTEE

By: /s/ Elizabeth C. Armitage

Elizabeth C. Armitage
Trustee

Address:

2207 Upland Place
Cincinnati, OH 45206

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

TRI-STATE GROWTH CAPITAL FUND II, L.P.

By: Tri-State Ventures II, LLC
Its: General Partner

By: Fort Washington Investment Advisors, Inc.
Its: Managing Member

By: /s/ Christopher L. Baucom

Christopher L. Baucom
Managing Director

By: /s/ Maribeth S. Rahe

Maribeth S. Rahe
President and Chief Executive Officer

Address:

303 Broadway, Suite 1200
Cincinnati, OH 45202

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

SIGVION FUND I, LP

By: /s/ J.P. Fairbank
J.P. Fairbank
Founding Partner

Address:

1970 N HALSTED ST, 3RD FLOOR
CHICAGO, IL 60614

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

GARDNER FAMILY TRUST, JOHN D. GARDNER TRUSTEE

By: /s/ John D. Gardner

John D. Gardner
Trustee

Address:

111 Pine Court
Bastop, TX 78602

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

MUNEER A. SATTER REVOCABLE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

THE SATTER FOUNDATION

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

SATTER FAMILY TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

SATTER CHILDREN'S TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Investment Advisor

Address:

419 Sheridan Road

Winnetka, IL 60093

Attn: Muneer A. Satter

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

IAN A.W. HOWES, IRA, STERLING TRUST CUSTODIAN

By: /s/ IAN A. W. HOWES

Name: IAN A. W. HOWES

Title: _____

Address:

219 Stratford Dr.

Chapel Hill, NC 27516

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

DIANE H. JANUSZ TRUST

By: /s/ John Janusz

Name: John Janusz

Title: Trustee

Address:

7385 Desert Spring Ct.
West Chester, OH 45069

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

AGECHEM VENTURE FUND L.P.

By: AgeChem Financial Inc., its General Partner

By: /s/ Louis Lacasse

Name: Louis Lacasse

Title: President

Address:

1 Westmount Square, Suite 800
Montreal, Quebec, Canada
H3Z 2P9

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

/s/ Kevin Peters

Kevin Peters

Address:

6100 Miami Rd.

Cincinnati, OH 48243

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

BLUE CHIP VALIDATION FUND, LTD.

By: /s/ John McIlwraith

Name: John McIlwraith

Title: Managing Director

Address:

1100 Chiquita Center

250 East Fifth Street

Cincinnati, OH 45202

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

QCA FIRST FUND II

By: /s/ John Habbert

Name: John Habbert

Title: Manager

Address:

6393 Grand Vista Avenue

Cincinnati, OH 45213

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

/s/ Robert Shalwitz

Robert Shalwitz

Address:

2549 Bryden Road

Bexley, OH 43209

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

FRED SHALWITZ TRUST,
ROBERT SHALWITZ, TRUSTEE

By: /s/ Robert Shalwitz

Name: Robert Shalwitz

Title: Trustee

Address:

2549 Bryden Road
Bexley, OH 43209

SIGNATURE PAGE TO AMENDMENT NUMBER ONE TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

**AMENDMENT NUMBER TWO TO
THE SECOND AMENDED AND RESTATED
RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT**

THIS AMENDMENT NUMBER TWO TO THE SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT (the "Amendment") is made and effective as of May 10, 2013, by and among AKEBIA THERAPEUTICS, INC., a Delaware corporation (the "Company"), and those Investors executing and delivering a counterpart signature page hereto. Capitalized terms not defined herein have the meanings given them in that certain Second Amended and Restated Right of First Refusal and Co-Sale Agreement, dated as of April 6, 2011, as amended by Amendment No. 1 thereto, by and among the Company, the Investors and the Major Holders (as amended, the "Agreement").

WHEREAS, the Company and the Investors desire to amend the Agreement to modify the definitions of Appropriate Percentage and Preferred Stock and add a definition for the Series C Preferred Stock of the Company; and

WHEREAS, the Investors hold more than the Appropriate Percentage of the shares of Common Stock required to amend the Agreement pursuant to the provisions of Section 6.8 of the Agreement;

NOW, THEREFORE, in consideration of the foregoing recitals and the mutual promises hereinafter set forth and set forth in the Agreement, the parties hereby agree as follows:

1. Amendment.

(a) The definition of "Appropriate Percentage" shall be deleted in its entirety and replaced with the following: "Appropriate Percentage" means fifty percent (50%).

(b) The definition of "Preferred Stock" shall be deleted in its entirety and replaced with the following: "'Preferred Stock' means all shares of Series A Preferred Stock, Series B Preferred Stock, Series C Preferred Stock and Series X Preferred Stock."

(c) A new definition of "Series C Preferred Stock" shall be added to the Agreement and shall read as follows: "'Series C Preferred Stock' shall mean shares of the Series C Preferred Stock of the Company, par value \$0.00001 per share."

(d) Schedule A shall be deleted in its entirety and replaced with the Schedule A attached hereto.

2. Miscellaneous Amendments. The Agreement is amended hereby so that any reference therein to the Agreement shall be deemed to be a reference to the Agreement as amended by this Amendment.

3. Continuance of Agreement. Except as specifically amended by this Amendment, the Agreement shall remain in full force and effect.

4. Governing Law. The laws of the State of Delaware govern all matters arising out of or relating to this Amendment, including, without limitation, its interpretation, construction, performance, and enforcement, without giving effect to such state's conflicts of law principles or rules of construction concerning the drafter hereof.

5. Counterparts. This Amendment may be executed in two or more counterparts, including by facsimile or PDF signature, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument.

[Signature Page Follows]

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

AKEBIA THERAPEUTICS, INC.

By: /s/ Joseph H. Gardner
Joseph H. Gardner, Ph.D.
President and Chief Executive Officer

Address:

Suite 420,
9987 Carver Road,
Cincinnati, OH 45242

SIGNATURE PAGE TO AMENDMENT NUMBER TWO TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

AGECHEM VENTURE FUND L.P.

By: /s/ Louis Lacasse

Name: Louis Lacasse

Title: President

Address:

Attn: Louis Lacasse, President
1 Westmount Square, Suite 800
Montreal, Quebec Canada
H3Z 2P9

SIGNATURE PAGE TO AMENDMENT NUMBER TWO TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

ATHENIAN VENTURE PARTNERS III L.P.

By: Athenian III, Ltd.
Its: General Partner

By: /s/ Karl O. Elderkin
Name: Karl O. Elderkin
Title: President

AVP OHIO TECHNOLOGY I L.P.

By: AVP Ohio I, Ltd.
Its: General Partner

By: /s/ Karl O. Elderkin
Name: Karl O. Elderkin
Title: President

Address:

340 West State Street
Unit 29/Suite 137D
Athens, OH 45701

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

BLUE CHIP VALIDATION FUND, LTD
By: Blue Chip Venture Company, LTD
Its: Manager

/s/ John McIlwraith

John McIlwraith
Managing Director

Address:
312 Walnut Street
Suite 1120
Cincinnati, OH 45202

SIGNATURE PAGE TO AMENDMENT NUMBER TWO TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

CINCINNATI CORNERSTONE INVESTORS AKB, LLC

By: /s/ Robert W. Coy
Robert W. Coy, Jr.
President

Address:

30 West 3rd Street, 6th Floor
Cincinnati, OH 45202-3559

SIGNATURE PAGE TO AMENDMENT NUMBER TWO TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

WILLIAM DALY

/s/ William Daly

William Daly

Address:

13 Via Abrazar

San Clemente, CA 92673

SIGNATURE PAGE TO AMENDMENT NUMBER TWO TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

DIANE H. JANUSZ TRUST

By: /s/ John Janusz

Name: John Janusz

Title: Trustee

JOHN JANUSZ

/s/ John Janusz

John Janusz

Address:

7385 Desert Spring Court
West Chester, OH 45069

SIGNATURE PAGE TO AMENDMENT NUMBER TWO TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

FAMILY AKEBIA INVESTMENTS LLC

By: /s/ Milton Berlinski

Name: Milton Berlinski

Title: Managing Member

Address:

1185 Park Avenue #11G

New York, NY 10128

SIGNATURE PAGE TO AMENDMENT NUMBER TWO TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

ALAN FISHMAN

/s/ Alan Fishman

Alan Fishman

Address:

6900 Stonehenge Dr.

Cincinnati, OH 45242-6204

SIGNATURE PAGE TO AMENDMENT NUMBER TWO TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

FRED SHALWITZ TRUST, ROBERT SHALWITZ, TRUSTEE

By: /s/ Robert Shalwitz

Name: Robert Shalwitz

Title: Trustee

Address:

2549 Bryden Road
Bexley, OH 43209

SIGNATURE PAGE TO AMENDMENT NUMBER TWO TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

JOSEPH H. GARDNER

By: /s/ Joseph H. Gardner

Address:

4060 Boomer Road
Cincinnati, OH 45247

SIGNATURE PAGE TO AMENDMENT NUMBER TWO TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

GARDNER FAMILY TRUST, JOHN D. GARDNER TRUSTEE

By: /s/ John D. Gardner

John D. Gardner
Trustee

Address:

111 Pine Court
Bastop, TX 78602

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IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

GITANA FAMILY TRUST, ELIZABETH C. ARMITAGE TRUSTEE

By: /s/ Elizabeth C. Armitage

Elizabeth C. Armitage

Trustee

Address:

2207 Upland Place

Cincinnati, OH 45206

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IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

IAN A.W. HOWES, IRA, STERLING TRUST CUSTODIAN

By: /s/ Ian A. W. Howes

Name: Ian A. W. Howes

Title: Trustee

IAN A. W. HOWES

/s/ Ian A. W. Howes

Ian A. W. Howes

Address:

219 Stratford Drive

Chapel Hill, NC 27516

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

KEARNY VENTURE PARTNERS, L.P.

By: /s/ Anupam Dalal
Name: Kearny Venture Associates, LLC
Title: its General Partner

Address:
Attn: Anupam Dalal
Kearny Venture Associates LLC
88 Kearny Street, Suite 200
San Francisco, CA 94108-5530

KEARNY VENTURE PARTNERS, ENTREPRENEURS FUND, L.P.

By: /s/ Anupam Dalal
Name: Kearny Venture Associates, LLC
Title: its General Partner

Address:
Attn: Anupam Dalal
Kearny Venture Associates LLC
88 Kearny Street, Suite 200
San Francisco, CA 94108-5530

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MCILWRAITH INVESTMENTS, LLC

By: /s/ John McIlwraith

Name: John McIlwraith

Title: Manager

Address:

Attn: John McIlwraith

7680 Foxgate Lane

Cincinnati, OH 45243

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IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

MRK INTERNATIONAL, LLC

By: /s/ Richard L. Kiley

Name: Richard L. Kiley

Title: Principal Member

Address:

7800 Tecumseh Trail

Cincinnati, OH 45243

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IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

NOVARTIS BIOVENTURES LTD.

By: /s/ H. S. Zivi

Name: H. S. Zivi

Title: Deputy Chairman

By: /s/ Rebecca White

Name: Rebecca White

Title: Authorized Signatory

Address:

NOVARTIS BIOVENTURES LTD.

Attn: Henri Simon Zivi

131 Front Street

Hamilton HM 12

Bermuda

But for mail, to:

Novartis BioVentures Ltd.

Attn: Henri Simon Zivi

PO Box HM 2899

Hamilton HM LX Bermuda

And, also send a copy to:

Novartis Venture Fund

Attn: Campbell Murray

Five Cambridge Center, Suite 603

Cambridge, MA 02142

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

KEVIN PETERS

/s/ Kevin Peters

Kevin Peters

Address:

9160 Given Road
Cincinnati, OH 45243

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IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

QCA FIRST FUND II

By: /s/ John Habbert

Name: John Habbert

Title: Manager

Address:

109 Bentwood Ct.

Cincinnati, OH 45241

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MUNEER A. SATTER REVOCABLE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Trustee

THE SATTER FOUNDATION

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Trustee

SATTER FAMILY TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

SATTER CHILDREN'S TRUST I

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

Address:

c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

KRISTEN HAYLER HERTEL REVOCABLE TRUST

By: /s/ Kristen Hayler Hertel

Name: Kristen Hayler Hertel

Title: Trustee

Address:

c/o Satter Investment Management, LLC

676 N. Michigan Avenue, Suite 4000

Chicago, IL 60611

Attn: Muneer A. Satter

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ANNE-CAROLE WITORT INSURANCE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Trustee

ROSE SHEREEN FUQUA INSURANCE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

RABI H. SATTER INSURANCE TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

JOHN WOOD TRUST

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Investment Advisor

Address:

c/o Satter Investment Management, LLC
676 N. Michigan Avenue, Suite 4000
Chicago, IL 60611
Attn: Muneer A. Satter

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

ADBUS SATTER INSURANCE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

GORDON AND BARBARA ANNE HERTEL INSURANCE TRUST

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Title: Trustee

Address:

c/o Satter Investment Management, LLC

676 N. Michigan Avenue, Suite 4000

Chicago, IL 60611

Attn: Muneer A. Satter

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ROBERT SHALWITZ

By: /s/ Robert Shalwitz

Address:

2549 Bryden Road
Bexley, OH 43209

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SIGVION FUND I, LP

By: /s/ J. P. Fairbank
J. P. Fairbank
Founding Partner

Address:

806 West Washington Street, Suite 204
Chicago, IL 60607

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THOMAS WEISEL HEALTHCARE VENTURE PARTNERS, L.P.

By: Thomas Weisel Capital Management LLC
Title: its Managing Member

By: /s/ Anupam Dalal
Name: Anupam Dalal
Title: Managing Director

Address:

88 Kearny Street, 4th Floor
San Francisco, CA 94108

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TRIATHLON MEDICAL VENTURES FUND, L.P.

By: Triathlon Medical Ventures, LLC

Its: General Partner

By: /s/ John M. Rice

John M. Rice

Managing Partner

Address:

300 E-Business Way

Suite 200

Cincinnati, OH 45241

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IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

TRI-STATE GROWTH CAPITAL FUND II, L.P.

By: Tri-State Ventures II, LLC
Its: General Partner

By: Fort Washington Investment Advisors, Inc.
Its: Managing Member

By: /s/ Steve Baker
Name: Steve Baker
Title: Managing Director

By: /s/ Maribeth S. Rahe
Name: Maribeth S. Rahe
Title: President and Chief Executive Officer

Address:

303 Broadway, Suite 1200
Cincinnati, OH 45202

SIGNATURE PAGE TO AMENDMENT NUMBER TWO TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

VENTURE INVESTORS EARLY STAGE FUND IV LIMITED
PARTNERSHIP

By: VIESF IV GP, LLC, its General Partner

By: /s/ Paul M. Weiss

Name: Paul M. Weiss, PhD

Title: Managing Director

Address:

505 South Rosa Road
Madison, WI 53719-1262
Attn: Paul Weiss, Managing Director
Phone: (608) 441-2700
Fax: (608) 441-2727
Email: paul@ventureinvestors.com

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IN WITNESS WHEREOF, the parties hereto have caused this Amendment to be executed as of the date first written above.

JOHN H. WYANT

/s/ John H. Wyant

John H. Wyant

Address:

Blue Chip Venture Company

1120 Scripps Center

312 Walnut Street

Cincinnati, OH 45202

SIGNATURE PAGE TO AMENDMENT NUMBER TWO TO SECOND AMENDED AND RESTATED RIGHT OF FIRST REFUSAL AND CO-SALE AGREEMENT

SCHEDULE A**INVESTORS**

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock</u>	<u>Number of Shares of Series C Preferred Stock</u>
Triathlon Medical Ventures Fund, L.P. Attn: John M. Rice Managing Partner 300 E-Business Way, Suite 200 Cincinnati, OH 45241	20,000.00	128,095.46	124,501.72	118,397.71
Novartis BioVentures Ltd. Attn: Henri Simon Zivi 131 Front Street Hamilton HM 12 Bermuda				
<i>But for mail, to:</i> Novartis BioVentures Ltd. Attn: Henri Simon Zivi PO Box HM 2899 Hamilton HM LX Bermuda				
<i>And, also send a copy to:</i> Novartis Venture Fund Attn: Campbell Murray Five Cambridge Center, Suite 603 Cambridge, MA 02142				
and				
Edwards Angell Palmer & Dodge LLP Attn: Al Sokol 111 Huntington Avenue Boston, MA 02199 asokol@eapdlaw.com	0	257,031.16	347,830.73	708,647.43
Venture Investors Early Stage Fund IV Limited Partnership Attn: Paul Weiss Managing Director 505 South Rosa Road Madison, WI 53719-1262	0	153,434.52	173,915.36	201,811.43
Kearny Venture Partners, L.P. Attn: Anupam Dalal Kearny Venture Associates, LLC 88 Kearny Street, Suite 200 San Francisco, CA 94108-5530	0	44,606.15	43,354.71	308,448.29

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock</u>	<u>Number of Shares of Series C Preferred Stock</u>
Kearny Venture Partners Entrepreneurs Fund, L.P. Attn: Anupam Dalal Kearny Venture Associates, LLC 88 Kearny Street, Suite 200 San Francisco, CA 94108-5530	0	909.78	884.26	6,290.57
Thomas Weisel Healthcare Venture Partners, L.P. 88 Kearny Street, 4th Floor San Francisco, CA 94108	0	45,515.99	44,239.03	74,475.86
The Procter & Gamble Company Attn: David Le Neveu Director, Corporate Acquisitions, Divestitures and Equity Ventures 1 Procter & Gamble Plaza Cincinnati, OH 45202	72,047.44	0	8,475.71	0
Athenian Venture Partners III L.P. Attn: Karl O. Elderkin President Athenian III, Ltd. 340 West State Street Unit 29/Suite 137D Athens, OH 45701	0	22,964.77	31,575.85	124,832.79
AVP Ohio Technology I L.P. Attn: Karl O. Elderkin President AVP Ohio I, Ltd. 340 West State Street Unit 29/Suite 137D Athens, OH 45701	0	7,654.92	9,004.41	24,194.21
Sigvion Fund I, LP Attn: J. P. Fairbank Founding Partner 738 W. Belden Avenue Chicago, IL 60614	8,000.00	13,660.69	13,277.43	21,402.57
Cincinnati Cornerstone Investors AKB, LLC Attn: Robert W. Coy, Jr. President 30 West 3rd Street, 6th Floor Cincinnati, OH 45202-3559	0	13,122.75	12,754.59	118,391.79

Schedule A-2

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock</u>	<u>Number of Shares of Series C Preferred Stock</u>
Tri-State Growth Capital Fund II, L.P. Attn: Steve Baker 303 Broadway, Suite 1200 Cincinnati, OH 45202	0	12,804.65	12,445.41	21,488.36
Blue Chip Validation Fund, Ltd. Attn: John McIlwraith Managing Director 1100 Chiquita Center 250 East Fifth Street Cincinnati, OH 45202	0	3,402.09	0	0
QCA First Fund II Attn: John Habbert 1776 Mentor Avenue, MB #302 Cincinnati, OH 45212	0	3,375.00	0	0
Gitana Family Trust, Elizabeth C. Armitage Trustee Attn: Elizabeth C. Armitage Trustee 2207 Upland Place Cincinnati, OH 45206	0	2,765.57	828.17	1,551.57
Robert Shalwitz 2549 Bryden Road Bexley, OH 43209	127,637.41	849.53	2,070.42	2,500.00
Fred Shalwitz Trust, Robert Shalwitz, Trustee Attn: Robert Shalwitz Trustee 2549 Bryden Road Bexley, OH 43209	0	3,403.13	0	0
Joseph H. Gardner 4060 Boomer Road Cincinnati, OH 45247	161,423.79	9,038.87	11,594.35	26,383.21
Gardner Family Trust, John D. Gardner Trustee Attn: John D. Gardner Trustee 111 Pine Court Bastop, TX 78602	0	6,901.99	7,288.07	2,485.00

Schedule A-3

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock</u>	<u>Number of Shares of Series C Preferred Stock</u>
Ian A. W. Howes, IRA, Sterling Trust Custodian Attn: Ian A. W. Howes Trustee 219 Stratford Drive Chapel Hill, NC 27516	0	5,000.00	5,797.18	7,142.00
Ian A. W. Howes 219 Stratford Drive Chapel Hill, NC 27516	46,925.51	0	0	2,902.21
Kevin Peters 9160 Given Road Cincinnati, OH 45243	46,925.51	0	1,449.29	3,473.00
William Daly 13 Via Abrazar San Clemente, CA 92673	60,810.00	0	0	13,488.86
Muneer A. Satter Revocable Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	115,943.58	182,551.50
John Wood Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	3,571.00
The Satter Foundation c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	57,971.78	191,275.86
Muneer A Satter IRA, Millennium Trust Company, Custodian	0	0	0	14,285.00
Satter Children's Trust I c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	57,971.78	66,275.86

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock</u>	<u>Number of Shares of Series C Preferred Stock</u>
Kristen Hayler Hertel Revocable Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	7,142.00
Anne-Carole Witort Insurance Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	7,142.00
Rose Shereen Fuqua Insurance Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	7,142.00
Rabi H. Satter Insurance Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	7,142.00
Abdus Satter Insurance Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	3,571.00
Gordon and Barbara Anne Hertel Insurance Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	0	3,571.00
Satter Family Trust c/o Satter Investment Management, LLC 676 N. Michigan Avenue, Suite 4000 Chicago, IL 60611 Attn: Muneer A. Satter	0	0	28,985.89	31,351.93
AgeChem Venture Fund L.P. Attn: Louis Lacasse President 1 Westmount Square, Suite 800 Montreal, Quebec, Canada H3Z 2P9	0	0	173,915.35	123,143.86

<u>Name and Address</u>	<u>Number of Shares of Common Stock</u>	<u>Number of Shares of Series A Preferred Stock</u>	<u>Number of Shares of Series B Preferred Stock</u>	<u>Number of Shares of Series C Preferred Stock</u>
Diane H. Janusz Trust Attn: John Janusz Trustee 7385 Desert Spring Court West Chester, OH 45069	0	0	1,449.29	139.00
John Janusz 7385 Desert Spring Court West Chester, OH 45069	0	0	0	1,670.79
MRK International, LLC Attn: Richard L. Kiley Principal Member 7800 Tecumseh Trail Cincinnati, OH 45243	0	0	0	396.00
McIlwraith Investments, LLC Attn: John McIlwraith Manager 7680 Foxgate Lane Cincinnati, OH 45243	0	0	0	4,597.00
John H. Wyant 2337 Grandin Road Cincinnati, OH 45208	0	0	0	1,039.36
Alan Fishman 6900 Stonehenge Drive Cincinnati, OH 45242-6204	0	0	0	1,428.00
Family Akebia Investments LLC Attn: Milton Berlinski 1185 Park Avenue #11G New York, NY 10128	0	0	0	142,858.00
Novo A/S Novo Ventures Tuborg Havnevej 19 DK – 2900 – Hellerup	0	0	0	714,285.00

Schedule A-6

**CAMBRIDGE SCIENCE CENTER AND 245 FIRST STREET
CAMBRIDGE, MASSACHUSETTS**

OFFICE LEASE AGREEMENT

BETWEEN

**MA-RIVERVIEW/245 FIRST STREET, L.L.C. a Delaware limited liability company
("LANDLORD")**

AND

**AKEBIA THERAPEUTICS, INC., a Delaware corporation
("TENANT")**

OFFICE LEASE AGREEMENT

THIS OFFICE LEASE AGREEMENT (this “Lease”) is made and entered into as of December 3, 2013, by and between MA-RIVERVIEW/245 FIRST STREET, L.L.C. a Delaware limited liability company (“Landlord”), and AKEBIA THERAPEUTICS, INC., a Delaware corporation (“Tenant”). The following exhibits and attachments are incorporated into and made a part of this Lease: **Exhibit A** (Outline and Location of Premises), **Exhibit B** (Expenses and Taxes), **Exhibit C** (Work Letter), **Exhibit C-1** (Space Plans), **Exhibit D** (Commencement Letter), **Exhibit E** (Building Rules and Regulations), **Exhibit F** (Additional Provisions), **Exhibit G** (Notice of Lease) and **Exhibit H** (Form of Letter of Credit).

1. **Basic Lease Information.**

- 1.01 “**Buildings**” shall mean those buildings located at 245 First Street, Cambridge, Massachusetts 02142, and commonly known as Cambridge Science Center and 245 First Street comprised of two buildings, the first being the science building (the “**Science Building**”) and the second being the office building (the “**Office Building**”). “**Rentable Square Footage of the Buildings**” is deemed to be 297,631 square feet. “**Rentable Square Footage of the Science Building**” is deemed to be 132,928 square feet, and “**Rentable Square Footage of the Office Building**” is deemed to be 164,703 square feet.
- 1.02 “**Premises**” shall mean the area shown on **Exhibit A** to this Lease. The Premises is located on the 11th floor of the Office Building and known as suite 1100. If the Premises include one or more floors in their entirety, all corridors and restroom facilities located on such full floor(s) shall be considered part of the Premises. The “**Rentable Square Footage of the Premises**” is deemed to be 6,837 square feet of office space on a portion of the 11th floor of the Office Building. Landlord and Tenant stipulate and agree that the Rentable Square Footage of the Buildings, the Rentable Square Footage of the Science Building, the Rentable Square Footage of the Office Building and the Rentable Square Footage of the Premises are correct.
- 1.03 “**Base Rent**”:

<u>Months of Term</u>	<u>Annual Rate Per Square Foot</u>	<u>Monthly Base Rent</u>
Commencement Date - Last day of 12 th full calendar month of the Term	\$ 55.00	\$31,336.25
First day of 13 th full calendar month of the Term - Last day of 24 th full calendar month of the Term	\$ 56.00	\$31,906.00
First day of 25 th full calendar month of the Term - Termination Date	\$ 57.00	\$32,475.75

Notwithstanding anything in this Section of the Lease to the contrary, so long as Tenant is not in Default (as defined in Section 18) under this Lease, Tenant shall be entitled to an abatement of Base Rent in the amount of \$31,336.25 (the "**Abated Base Rent**") for the 1st full calendar month of the Term (as defined in Section 1.06) (the "**Base Rent Abatement Period**"). If Tenant Defaults at any time during the initial Term and fails to cure such Default within any applicable cure period under the Lease, then all unamortized Abated Base Rent (i.e. based upon the amortization of the Abated Base Rent in equal monthly amounts, without interest, during the initial Term) shall immediately become due and payable. The payment by Tenant of the Abated Base Rent in the event of a Default shall not limit or affect any of Landlord's other rights, pursuant to this Lease or at law or in equity. During the Base Rent Abatement Period, only Base Rent shall be abated, and all Additional Rent and other costs and charges specified in this Lease shall remain as due and payable pursuant to the provisions of this Lease.

1.04 Tenant's Pro Rata Share:

(a) "**Tenant's Building Pro Rata Share**" shall mean 4.1511% with respect to the Premises.

(b) "**Tenant's Common Area Pro Rata Share**" shall mean 2.2971% with respect to the Premises.

1.05 "**Base Year**" for Taxes (defined in **Exhibit B**): calendar year 2014; "**Base Year**" for Expenses (defined in **Exhibit B**): calendar year 2014; "**Base Year**" for Common Area Expenses (defined in **Exhibit B**): calendar year 2014.

1.06 "**Term**": The period commencing on the Commencement Date (defined below) and, unless terminated earlier in accordance with this Lease, ending on the last day of the 36th full calendar month following the Commencement Date (the "**Termination Date**"). The "**Commencement Date**" shall mean the date on which the Premises are delivered to Tenant with the Landlord Work (defined in Section 1.14) Substantially Complete (defined in Section 3), free of all tenants and other occupants, and free of liens or encumbrances. The parties anticipate that the Premises shall be delivered to Tenant with the Landlord Work Substantially Complete on or about December 26, 2013 (the "**Target Commencement Date**"). In addition, if Tenant is entitled to register or record a notice or memorandum of this Lease pursuant to the terms in Section 1.16, Landlord and Tenant shall also execute and Tenant may register or record, as appropriate, at Tenant's cost and expense, a Notice of Lease in the form attached as **Exhibit G**.

1.07 Allowance: None.

1.08 "**Security Deposit**": \$125,345.00, as more fully described in Section 6.

1.09 "**Guarantor(s)**": As of the date of this Lease there are no Guarantors.

1.10 “**Broker(s)**”: Richards Barry Joyce & Partners (“**Tenant’s Broker**”), which represented Tenant in connection with this transaction, and Richards Barry Joyce & Partners (“**Landlord’s Broker**”), which represented Landlord in connection with this transaction. Landlord and Tenant hereby specifically acknowledge and agree that the same broker is representing both Tenant and Landlord.

1.11 “**Permitted Use**”: General office use.

1.12 “**Notice Address(es)**”:

Landlord:

MA-Riverview/245 First Street, L.L.C.
c/o Equity Office
125 Summer Street, 17th Floor
Boston, Massachusetts 02110
Attention: Property Manager

Tenant:

Prior to the Commencement Date:

Akebia Therapeutics, Inc.
9987 Carver Road
Suite 420
Cincinnati, Ohio 45242

With copies of any notices to Landlord shall be sent to:

Equity Office
Two North Riverside Plaza
Suite 2100
Chicago, Illinois 60606
Attn: Managing Counsel - Boston Region

From and after the Commencement Date:

Akebia Therapeutics, Inc.
245 First Street
Suite 1100
Cambridge, Massachusetts 02142

and

Equity Office
Two North Riverside Plaza
Suite 2100
Chicago, Illinois 60606
Attn: Lease Administration

1.13 “**Business Day(s)**” are Monday through Friday of each week, exclusive of New Year’s Day, Presidents Day, Memorial Day, Independence Day, Labor Day, Thanksgiving Day and Christmas Day (“**Holidays**”). Landlord may designate additional Holidays that are commonly recognized by other office buildings in the area where the Building is located. “**Building Service Hours**” are 8:00 a.m. to 6:00 p.m. on Business Days and 8:00 a.m. to 1:00 p.m. on Saturdays.

1.14 “**Landlord Work**” means the work that Landlord is obligated to perform in the Premises pursuant to a separate agreement (the “**Work Letter**”) attached to this Lease as **Exhibit C**.

- 1.15 **“Property”** means the Buildings and the parcel(s) of land on which they are located and, at Landlord’s discretion, the parking facilities and other improvements, if any, serving the Office Building or Buildings and the parcel(s) of land on which they are located.
- 1.16 Tenant shall not record this Lease or any memorandum or notice without Landlord’s prior written consent; provided, however, Landlord agrees to consent to the recordation or registration of a memorandum or notice of this Lease, at Tenant’s cost and expense (and in a form reasonably satisfactory to Landlord), if the initial term of this Lease or the initial term plus renewal terms granted exceed, in the aggregate, 7 years. If this Lease is terminated before the Term expires, upon Landlord’s request the parties shall execute, deliver and record an instrument acknowledging the above and the date of the termination of this Lease, and Tenant appoints Landlord its attorney-in-fact in its name and behalf to execute the instrument if Tenant shall fail to execute and deliver the instrument after Landlord’s request therefor within 10 days.
- 1.17 **“Letter of Credit”** is as described in Section 4 of **Exhibit F** attached hereto.

2. Lease Grant.

Landlord hereby leases the Premises to Tenant and Tenant hereby leases the Premises from Landlord. Tenant has the non-exclusive right to use any portions of the Property that are designated by Landlord for the common use of tenants and others (the **“Common Areas”**).

3. Adjustment of Commencement Date; Possession.

3.01. The Landlord Work shall be deemed to be **“Substantially Complete”** on the later of (i) the date that all Landlord Work has been performed in a good and workmanlike manner, other than any minor details of construction, mechanical adjustment or any other similar matter, the non-completion of which does not materially interfere with Tenant’s use of the Premises; and (ii) the date that Landlord receives from the appropriate governmental authorities, with respect to the Landlord Work performed by Landlord or its contractors in the Premises, all approvals necessary for the occupancy of the Premises for the Permitted Use (or would have received such approvals absent any Tenant Delays). If Landlord is delayed in the performance of the Landlord Work as a result of the acts or omissions of Tenant, the Tenant Related Parties (defined in Section 1.3) or their respective contractors or vendors, including, without limitation, changes requested by Tenant to approved plans, Tenant’s failure to comply with any of its obligations under this Lease, or Tenant’s specification of any materials or equipment with long lead times (each a **“Tenant Delay”**), the Landlord Work shall be deemed to be Substantially Complete on the date that Landlord could reasonably have been expected to Substantially Complete the Landlord Work absent any Tenant Delay. Landlord shall use reasonable efforts to notify Tenant, orally or in writing, if any choice specified by Tenant of materials or equipment is a long lead item which may cause a Tenant Delay. If Landlord determines that Landlord will be unable to deliver the Premises in the condition required under Section 1.06 hereof by the Target Commencement Date, then Landlord shall use its reasonable efforts to provide Tenant with notice (orally or in writing) at least 2 weeks prior to the Target Commencement Date of the same

and include in such notice Landlord's projected delivery date. Notwithstanding anything to the contrary in Section 1.06 above, Landlord's failure to Substantially Complete the Landlord Work by the Target Commencement Date (described in Section 1.06) shall not be a default by Landlord or otherwise render Landlord liable for damages except to the extent provided in Section 3.03 hereof. Promptly after the determination of the Commencement Date, Landlord and Tenant shall execute and deliver a commencement letter in the form attached as **Exhibit D** (the "**Commencement Letter**"). Tenant's failure to execute and return the Commencement Letter, or to provide written objection to the statements contained in the Commencement Letter, within 30 days after the date of Tenant's receipt or deemed receipt of the Commencement Letter shall be deemed an approval by Tenant of the statements contained therein.

3.02. Subject to Landlord's obligation to perform Landlord Work in the manner described in Section 3.01 hereof, the Premises are accepted by Tenant in "as is" condition and configuration without any representations or warranties by Landlord. By taking possession of the Premises in the condition and as provided in Section 3.01, Tenant agrees that the Premises are in good order and satisfactory condition. Tenant's acceptance of the Premises shall be subject to Landlord's obligation to correct any of the Landlord Work not performed as required hereunder, which is as set forth on a construction punch list prepared by Landlord and Tenant in accordance with the following terms. Within 15 days after Landlord has provided Tenant with notice (which may be oral or by email) that the Landlord Work is Substantially Complete, Landlord and Tenant shall together conduct an inspection of the Premises and prepare a "punch list" setting forth any portions of the Landlord Work that are not in conformity with the Landlord Work as required by the terms of this Lease. Notwithstanding the foregoing, at the request of Landlord, such construction punch list shall be mutually prepared by Landlord and Tenant prior to the date on which Tenant first begins to move its furniture, equipment or other personal property into the Premises. Landlord, as part of the Landlord Work, shall use good faith efforts to correct all such items within a reasonable time following the completion of the punch list. Notwithstanding the foregoing, Landlord shall be responsible for latent defects in the Landlord Work of which Tenant notifies Landlord to the extent that the correction of such defects is covered under valid and enforceable warranties given Landlord by contractors or subcontractors performing the Landlord Work. Landlord, at its option, may pursue such claims directly or assign any such warranties to Tenant for enforcement. Except as otherwise provided in this Lease, Tenant shall not be permitted to take possession of or enter the Premises prior to the Commencement Date without Landlord's permission. If Tenant takes possession of or enters the Premises before the Commencement Date, Tenant shall be subject to the terms and conditions of this Lease; provided, however, except for the cost of services requested by Tenant (e.g. after hours HVAC service), Tenant shall not be required to pay Rent for any entry or possession before the Commencement Date during which Tenant, with Landlord's approval, has entered, or is in possession of, the Premises for the sole purpose of performing improvements or installing furniture, equipment or other personal property. However, notwithstanding the foregoing but subject to the terms of this Section 3.02, Landlord shall use its reasonable efforts to permit Tenant to enter the Premises after written notice from Landlord, at Tenant's sole risk, at least 15 days prior to the Commencement Date, solely for the purpose of installing equipment, furnishings and other personalty provided that such installations do not interfere with the Landlord Work. The parties agree to cooperate reasonably to coordinate their respective access to and work within the Premises so as to minimize any delay in the performance of the Landlord Work. Landlord may withdraw such permission to enter the Premises prior to the

Commencement Date at any time that Landlord reasonably determines that such entry by Tenant is causing a dangerous situation for Landlord, Tenant or their respective contractors or employees, or if Landlord reasonably determines that such entry by Tenant is hampering or otherwise preventing Landlord from proceeding with the completion of the Landlord Work at the earliest possible date, provided that Landlord agrees to act reasonably in making any such determination in light of the mutual obligation of Landlord and Tenant to cooperate reasonably to coordinate their respective work as set forth above.

3.03. If the Commencement Date has not occurred on or before the Target Commencement Date, Tenant shall be entitled to a rent abatement following the Base Rent Abatement Period of \$1,030.23 for every day in the period beginning on the Target Commencement Date and ending on the Commencement Date. Landlord and Tenant acknowledge and agree that: (i) the determination of the Commencement Date shall take into consideration the effect of any Tenant Delays; and (ii) the Target Commencement Date shall be postponed by the number of days the Commencement Date is delayed due to events of Force Majeure.

4. Rent.

4.01. Tenant shall pay Landlord, without any setoff or deduction, unless expressly set forth in this Lease, all Base Rent and Additional Rent due for the Term (collectively referred to as "**Rent**"), the parties acknowledging that this Lease expressly provides for the abatement of the Abated Base Rent as provided in Section 1.03 hereof. "**Additional Rent**" means all sums (exclusive of Base Rent) that Tenant is required to pay Landlord under this Lease. Tenant shall pay and be liable for all rental, sales and use taxes (but excluding income taxes), if any, imposed upon or measured by Rent. Base Rent and recurring monthly charges of Additional Rent shall be due and payable in advance on the first day of each calendar month without notice or demand, provided that the installment of Base Rent for the first full calendar month of the Term, and the first monthly installment of Additional Rent for Expenses and Taxes, shall be payable upon the execution of this Lease by Tenant. All other items of Rent shall be due and payable by Tenant on or before 30 days after billing by Landlord. Rent shall be made payable to the entity, and sent to the address, Landlord designates and shall be made by good and sufficient check payable in United States of America currency or by other means acceptable to Landlord. If Tenant does not pay any Rent when due hereunder, Tenant shall pay Landlord an administration fee in the amount of \$250.00, provided that Tenant shall be entitled to a grace period of up to 5 Business Days for the first 2 late payments of Rent in a calendar year. In addition, past due Rent shall accrue interest at 10% per annum, and Tenant shall pay Landlord a reasonable fee for any checks returned by Tenant's bank for any reason. Landlord's acceptance of less than the correct amount of Rent shall be considered a payment on account of the oldest obligation due from Tenant hereunder, then to any current Rent then due hereunder, notwithstanding any statement to the contrary contained on or accompanying any such payment from Tenant. Rent for any partial month during the Term shall be prorated. No endorsement or statement on a check or letter accompanying payment shall be considered an accord and satisfaction. Except as specifically set forth in this Lease, Tenant's covenant to pay Rent is independent of every other covenant in this Lease.

4.02. Tenant shall pay Tenant's Building Pro Rata Share of Expenses and Tenant's Common Area Pro Rata Share of Taxes and Common Area Expenses in accordance with **Exhibit B** of this Lease.

5. Compliance with Laws; Use.

The Premises shall be used for the Permitted Use and for no other use whatsoever. Tenant shall comply with all statutes, codes, ordinances, orders, rules and regulations of any municipal or governmental entity whether in effect now or later, including the Americans with Disabilities Act (“**Law(s)**”), regarding the operation of Tenant’s business in the Premises, the use, condition, configuration and occupancy of the Premises and the Buildings’ systems located in or exclusively serving the Premises. However, nothing herein shall require Tenant to comply with Laws or requirements of public authorities which require the installation of new or additional mechanical, electrical, plumbing or fire/life safety systems on a Building-wide basis without reference to the particular use of Tenant or any Alterations performed by or for Tenant (“**Building-Wide Laws**”). Landlord will, at Landlord’s expense (except to the extent properly included in Expenses), perform all acts required to comply with such Building-Wide Laws as the same affect the Premises and the Building. In addition, Tenant shall, at its sole cost and expense, promptly comply with any Laws that relate to the “Base Building” (defined below), but only to the extent such obligations are triggered by Tenant’s use of the Premises, other than for general office use, or Alterations or improvements in the Premises performed or requested by Tenant. “**Base Building**” shall include the structural portions of the Buildings, the public restrooms and the Buildings’ mechanical, electrical and plumbing systems and equipment located in the internal core of the Buildings on the floor or floors on which the Premises are located. Tenant shall promptly provide Landlord with copies of any notices it receives regarding an alleged violation of Law. Tenant shall not exceed the standard density limit for the Buildings. Tenant shall comply with the rules and regulations of the Buildings attached as **Exhibit E** and such other reasonable rules and regulations adopted by Landlord from time to time, including rules and regulations for the performance of Alterations (defined in Section 9.03).

6. Security Deposit.

The Security Deposit, if any, shall be delivered to Landlord upon the execution of this Lease by Tenant and held by Landlord without liability for interest (unless required by Law) as security for the performance of Tenant’s obligations. The Security Deposit is not an advance payment of Rent or a measure of damages. Landlord may from time to time and without prejudice to any other remedy provided in this Lease or by Law, use all or a portion of the Security Deposit to the extent necessary to satisfy past due Rent or to satisfy any other loss or damage resulting from Tenant’s breach under this Lease. If Landlord uses any portion of the Security Deposit, Tenant, within 5 Business Days after receipt or deemed receipt of written demand, shall restore the Security Deposit to its original amount. Landlord shall return any unapplied portion of the Security Deposit to Tenant within 45 days after the later to occur of: (a) determination of the final Rent due from Tenant; or (b) the later to occur of the Termination Date or the date Tenant surrenders the Premises to Landlord in compliance with Section 25. Landlord may assign the Security Deposit to a successor or transferee and, following the assignment, Landlord shall have no further liability for the return of the Security Deposit. Landlord shall not be required to keep the Security Deposit separate from its other accounts.

Within 45 days after the date of this Lease, Tenant shall provide Landlord with an irrevocable letter of credit (the "**Letter of Credit**") in the amount of the Security Deposit which shall conform with and be subject to the terms of Section 3 of **Exhibit F** to this Lease. Notwithstanding anything to the contrary contained herein, promptly upon Landlord's receipt of the Letter of Credit (within 10 Business Days), Landlord shall return the full amount of the unapplied cash Security Deposit to Tenant.

7. Building Services.

7.01. Landlord shall furnish Tenant with the following services: (a) water for use in the Base Building lavatories; (b) customary heat and air conditioning in season during Building Service Hours, although (i) Tenant shall have the right to receive HVAC service during hours other than Building Service Hours by paying Landlord's then standard charge for additional HVAC service and providing such prior notice as is reasonably specified by Landlord, and (ii) if Tenant is permitted to connect any supplemental HVAC units to the Building's condenser water loop or chilled water line, such permission shall be conditioned upon Landlord having adequate excess capacity from time to time and such connection and use shall be subject to Landlord's reasonable approval and reasonable restrictions imposed by Landlord, and Landlord shall have the right to charge Tenant a connection fee and/or a monthly usage fee, as reasonably determined by Landlord; (c) standard janitorial service on Business Days; (d) elevator service; (e) electricity in accordance with the terms and conditions in Section 7.02; (f) access to the Building for Tenant and its employees 24 hours per day/7 days per week, subject to the terms of this Lease and such protective services or monitoring systems, if any, as Landlord may reasonably impose, including, without limitation, sign-in procedures and/or presentation of identification cards; and (g) such other services as Landlord reasonably determines are necessary or appropriate for the Property. If Landlord, at Tenant's request, provides any services which are not Landlord's express obligation under this Lease, including, without limitation, any repairs which are Tenant's responsibility pursuant to Section 9 below, Tenant shall pay Landlord, or such other party designated by Landlord, the cost of providing such service plus a reasonable administrative charge.

7.02. Electricity used by Tenant in the Premises shall be paid for by Tenant by a separate charge payable by Tenant to Landlord. Without the consent of Landlord, Tenant's use of electrical service shall not exceed the Building standard usage, per square foot, as reasonably determined by Landlord, based upon the Building standard electrical design load. For purposes hereof, the Building "electrical standard" is 4.5 watts per usable square foot of connected load to the Premises, exclusive of base Building HVAC. As of the date hereof, Landlord's annual charge for electricity is estimated to be \$1.50 per rentable square foot, subject to change from time to time, payable on a monthly basis. Landlord shall have the right to measure electrical usage by commonly accepted methods, including the installation of measuring devices such as submeters and check meters. If it is determined that Tenant is using electricity in such quantities or during such periods as to cause the total cost of Tenant's electrical usage, on a monthly, per-rentable-square-foot basis, to materially exceed that which Landlord reasonably deems to be standard for the Building, Tenant shall pay Landlord Additional Rent for the cost of such excess electrical usage and, if applicable, for the cost of purchasing and installing the measuring device(s).

7.03. Landlord's failure to furnish, or any interruption, diminishment or termination of services due to the application of Laws, the failure of any equipment, the performance of maintenance, repairs, improvements or alterations, utility interruptions or the occurrence of an event of Force Majeure (defined in Section 26.03) (collectively a "Service Failure") shall not render Landlord liable to Tenant, constitute a constructive eviction of Tenant, give rise to an abatement of Rent, nor relieve Tenant from the obligation to fulfill any covenant or agreement. However, if the Premises, or a material portion of the Premises, are made untenable for a period in excess of 3 consecutive Business Days as a result of a Service Failure that is reasonably within the control of Landlord to correct, then Tenant, as its sole remedy, shall be entitled to receive an abatement of Rent payable hereunder during the period beginning on the 4th consecutive Business Day of the Service Failure and ending on the day the service has been restored. If the entire Premises have not been rendered untenable by the Service Failure, the amount of abatement shall be equitably prorated.

8. Leasehold Improvements.

All improvements in and to the Premises, including any Alterations (defined in Section 9.03) (collectively, "Leasehold Improvements") shall remain upon the Premises at the end of the Term without compensation to Tenant, provided that Tenant, at its expense, shall remove any Cable (defined in Section 9.01 below). In addition, Landlord, by written notice to Tenant at least 30 days prior to the Termination Date, may require Tenant, at Tenant's expense, to remove any Landlord Work or Alterations that, in Landlord's reasonable judgment, are of a nature that would require removal and repair costs that are materially in excess of the removal and repair costs associated with standard office improvements (the Cable and such other items collectively are referred to as "Required Removables"). Required Removables shall include, without limitation, internal stairways, raised floors, personal baths and showers, vaults, rolling file systems and structural alterations and modifications. The Required Removables shall be removed by Tenant before the Termination Date. Tenant shall repair damage caused by the installation or removal of Required Removables. If Tenant fails to perform its obligations in a timely manner, Landlord may perform such work at Tenant's expense. Tenant, at the time it requests approval for a proposed Alteration, including any Landlord Work, as such term may be defined in the Work Letter attached as **Exhibit C**, may request in writing that Landlord advise Tenant whether the Alteration, including any Landlord Work, or any portion thereof, is a Required Removable. Within 10 days after receipt of Tenant's request, Landlord shall advise Tenant in writing as to which portions of the alteration or other improvements are Required Removables. Notwithstanding the foregoing, the Landlord agrees that, except for any Cable, the improvements identified on the Space Plans (as defined in the Work Letter attached as **Exhibit C**), as specifically shown thereon as of the date hereof, shall not be deemed Required Removables.

9. Repairs and Alterations.

9.01. Tenant shall promptly provide Landlord with notice of any conditions in the Premises that are dangerous or in need of maintenance or repair. Tenant, at its sole cost and expense, shall perform all maintenance and repairs to the Premises that are not Landlord's express responsibility under this Lease, and keep the Premises in good condition and repair, reasonable wear and tear excepted. Tenant's repair and maintenance obligations include, without

limitation, repairs to: (a) floor covering; (b) interior partitions; (c) doors; (d) the interior side of demising walls; (e) Alterations (described in Section 9.03); (f) supplemental air conditioning units, kitchens, including hot water heaters, plumbing, and similar facilities exclusively serving Tenant, whether such items are installed by Tenant or are currently existing in the Premises; and (g) electronic, fiber, phone and data cabling and related equipment that is installed by or for the exclusive benefit of Tenant (collectively, "**Cable**"). All repairs and other work performed by Tenant or its contractors, including that involving Cable, shall be subject to the terms of Section 9.03 below. If Tenant fails to make any repairs to the Premises for more than 15 days after written notice from Landlord (although notice shall not be required in an emergency), Landlord may make the repairs, and, within 30 days after demand, Tenant shall pay the reasonable cost of the repairs, together with an administrative charge in an amount equal to 10% of the cost of the repairs. Notwithstanding the foregoing, if the repair to be performed by Tenant cannot reasonably be completed within 15 days after Landlord's notice to Tenant, Landlord shall not exercise its right to make such repair on Tenant's behalf so long as Tenant commences such repair within 5 days after notice from Landlord and is diligently pursuing the same to completion.

9.02. Landlord shall keep and maintain in good repair and working order and perform maintenance upon the: (a) structural elements of the Buildings; (b) mechanical (including HVAC), electrical, plumbing and fire/life safety systems serving the Buildings in general; (c) Common Areas; (d) roof of the Buildings; (e) exterior windows of the Buildings; and (f) elevators serving the Buildings. Landlord shall promptly make repairs for which Landlord is responsible.

9.03. Tenant shall not make alterations, repairs, additions or improvements or install any Cable (collectively referred to as "**Alterations**") without first obtaining the written consent of Landlord in each instance, which consent shall not be unreasonably withheld or delayed. However, Landlord's consent shall not be required for any Alteration that satisfies all of the following criteria (a "**Cosmetic Alteration**"): (a) is of a cosmetic nature such as painting, wallpapering, hanging pictures and installing carpeting; (b) is not visible from the exterior of the Premises or Buildings; (c) will not affect the Base Building (defined in Section 5); and (d) does not require work to be performed inside the walls or above the ceiling of the Premises. Cosmetic Alterations shall be subject to all the other provisions of this Section 9.03. Prior to starting work, Tenant shall furnish Landlord with plans and specifications (which shall be in CAD format if requested by Landlord); names of contractors reasonably acceptable to Landlord (provided that Landlord may designate specific contractors with respect to Base Building and vertical Cable, as may be described more fully below); required permits and approvals; evidence of contractor's and subcontractor's insurance in amounts reasonably required by Landlord and naming Landlord and the managing agent for the Buildings (or any successor(s)) as additional insureds; and with respect to any Alterations performed by or for the benefit of Tenant the cost of which (when taken in the aggregate) is greater than or equal to \$100,000.00, any security for performance in amounts reasonably required by Landlord. Landlord may designate specific contractors with respect to oversight, installation, repair, connection to, and removal of vertical Cable. All Cable shall be clearly marked with adhesive plastic labels (or plastic tags attached to such Cable with wire) to show Tenant's name, suite number, and the purpose of such Cable (i) every 6 feet outside the Premises (specifically including, but not limited to, the electrical room risers and any Common Areas), and (ii) at the termination point(s) of such Cable. Changes to the plans and

specifications must also be submitted to Landlord for its approval. Alterations shall be constructed in a good and workmanlike manner using materials of a quality reasonably approved by Landlord, and Tenant shall ensure that no Alteration impairs any Building system or Landlord's ability to perform its obligations hereunder. Tenant shall reimburse Landlord for any sums paid by Landlord for third party examination of Tenant's plans for non-Cosmetic Alterations. In addition, Tenant shall pay Landlord a fee for Landlord's oversight and coordination of any non-Cosmetic Alterations equal to 5% of the cost of the non-Cosmetic Alterations. Upon completion, Tenant shall furnish "as-built" plans (in CAD format, if requested by Landlord) for non-Cosmetic Alterations, completion affidavits and full and final waivers of lien. Landlord's approval of an Alteration shall not be deemed a representation by Landlord that the Alteration complies with Law.

10. Entry by Landlord.

Landlord may enter the Premises to inspect, show or clean the Premises or to perform or facilitate the performance of repairs, alterations or additions to the Premises or any portion of the Buildings. Except in emergencies or to provide Building services, Landlord shall provide Tenant with reasonable prior verbal notice of entry and shall use reasonable efforts to minimize any interference with Tenant's use of the Premises. Notwithstanding the foregoing, except in emergencies or to provide Building services, Landlord shall provide Tenant with at least 24 hours' prior notice of entry into the Premises, which may be given orally to the entity occupying the Premises, and Tenant shall be entitled to have an employee of Tenant accompany the person(s) entering the Premises. If, however, Tenant does not make an employee available in the Premises at the time indicated in such notice or at such other time as may be mutually agreed upon by Landlord and Tenant, then (i) if the entry is for the purpose of performing work or providing services which have been requested by Tenant and would not otherwise be performed or provided by Landlord, Landlord shall not enter the Premises (unless Tenant otherwise agrees), but (ii) if the entry is for another purpose permitted by this Section, Landlord may enter the Premises. If reasonably necessary, Landlord may temporarily close all or a portion of the Premises to perform repairs, alterations and additions. However, except in emergencies, Landlord will not close the Premises if the work can reasonably be completed on weekends and after Building Service Hours. Except as specifically provided otherwise in this Section 10, entry by Landlord shall not constitute a constructive eviction or entitle Tenant to an abatement or reduction of Rent. If Landlord temporarily closes the Premises as provided above for a period in excess of 3 consecutive Business Days, Tenant, as its sole remedy, shall be entitled to receive a per diem abatement of Base Rent during the period beginning on the 4th consecutive Business Day of closure and ending on the date on which the Premises are returned to Tenant in a tenantable condition. Tenant, however, shall not be entitled to an abatement if the repairs, alterations and/or additions to be performed are required as a result of the acts or omissions of Tenant, its agents, employees or contractors, including, without limitation, a Default by Tenant in its maintenance and repair obligations under the Lease.

Tenant, at its own expense, may provide its own locks to an area within the Premises ("**Secured Area**") containing no more than 5% of the Rentable Area in the Premises. Tenant need not furnish Landlord with a key, but upon the Termination Date or earlier expiration or termination of the Lease or Tenant's right to possession, Tenant shall surrender all such keys to Landlord. If Landlord must gain access to a Secured Area in a non-emergency situation, Landlord shall

contact Tenant, and Landlord and Tenant shall arrange a mutually agreed upon time for Landlord to have such access. Landlord shall comply with all reasonable security measures pertaining to the Secured Area. If Landlord determines in its sole discretion that an emergency in the Building or the Premises, including, without limitation, a suspected fire or flood, requires Landlord to gain access to the Secured Area, Tenant hereby authorizes Landlord to forcibly enter the Secured Area. In such event, Landlord shall have no liability whatsoever to Tenant with respect to such entrance by Landlord, and Tenant shall pay all reasonable expenses incurred by Landlord in repairing or reconstructing any entrance, corridor, door or other portions of the Premises damaged as a result of a forcible entry by Landlord. Landlord shall have no obligation to provide either janitorial service or cleaning in the Secured Area.

11. Assignment and Subletting.

11.01. Except in connection with a Business Transfer (defined in Section 11.04), Tenant shall not assign, sublease, transfer or encumber any interest in this Lease or allow any third party to use any portion of the Premises (collectively or individually, a “**Transfer**”) without the prior written consent of Landlord, which consent shall not be unreasonably withheld, conditioned or delayed if Landlord does not exercise its recapture rights under Section 11.02. Without limitation, it is agreed that Landlord’s consent shall not be considered unreasonably withheld if the proposed transferee is a governmental entity or an occupant of the Building or an occupant of any other buildings within the Cambridge Science Center or if the proposed transferee, whether or not an occupant of the Building or an occupant of any other buildings within the Cambridge Science Center, is in discussions with Landlord regarding the leasing of space within the Building or within any other buildings within the same project. If the entity(ies) which directly or indirectly owns or controls 51% or more of the voting shares/rights of Tenant (other than through the ownership of voting securities listed on a recognized securities exchange) changes as a result of one transaction or one interrelated series of transactions at any time, and such change of ownership or control results in the hiring of all or substantially all new named senior officers of Tenant, then such change of ownership or control shall constitute a Transfer; provided, however, the infusion of additional equity capital in Tenant or an initial public offering of equity securities of Tenant under the Securities Act of 1933, as amended, which results in Tenant’s stock being traded on a national securities exchange, including, but not limited to, the NYSE, the NASDAQ Stock Market or the NASDAQ Small Cap Market System shall not be deemed, in either case, a Transfer. Any Transfer in violation of this Section shall, at Landlord’s option, be deemed a Default by Tenant as described in Section 18, and shall be voidable by Landlord. In no event shall any Transfer, including a Business Transfer, release or relieve Tenant from any obligation under this Lease, and Tenant shall remain primarily liable for the performance of the tenant’s obligations under this Lease, as amended from time to time.

11.02. Tenant shall provide Landlord with financial statements for the proposed transferee (or, in the case of a change of ownership or control that constitutes a Transfer as provided in Section 11.01 above, for the proposed new controlling entity(ies)), a fully executed copy of the proposed assignment, sublease or other Transfer documentation and such other information as Landlord may reasonably request. Within 15 Business Days after receipt of the required information and documentation, Landlord shall either: (a) consent to the Transfer by execution of a consent agreement in a form reasonably designated by Landlord; (b) reasonably refuse to consent to the Transfer in writing; or (c) in the event of an assignment of this Lease or

subletting of more than 25% of the Rentable Square Footage of the Premises for more than 50% of the remaining Term (excluding unexercised options) other than an assignment or sublease consented to by Landlord in clause (a) above, recapture the portion of the Premises that Tenant is proposing to Transfer. Notwithstanding the above, Landlord shall not have the right to recapture the Premises (or applicable portion thereof) in the event of a Business Transfer (defined in Section 11.04). If Landlord exercises its right to recapture, this Lease shall automatically be amended (or terminated if the entire Premises is being assigned or sublet) to delete the applicable portion of the Premises effective on the proposed effective date of the Transfer, although Landlord may require Tenant to execute a reasonable amendment or other document reflecting such reduction or termination. Tenant shall pay Landlord a review fee (that shall include any attorneys' fees) of \$1,500.00 for Landlord's review of any requested Transfer.

11.03. Tenant shall pay Landlord 50% of all rent and other consideration which Tenant receives as a result of a Transfer that is in excess of the Rent payable to Landlord for the portion of the Premises and Term covered by the Transfer. Tenant shall pay Landlord for Landlord's share of the excess within 30 days after Tenant's receipt of the excess. In determining the excess due Landlord, Tenant may deduct from the excess, on a straight-line basis, all reasonable and customary expenses directly incurred by Tenant attributable to the Transfer. If Tenant is in Default, Landlord may require that all sublease payments be made directly to Landlord, in which case Tenant shall receive a credit against Rent in the amount of Tenant's share of payments received by Landlord.

11.04. Tenant may assign this Lease to a successor to Tenant by merger, consolidation or the purchase of substantially all of Tenant's assets, or assign this Lease or sublet all or a portion of the Premises to an Affiliate (defined below), without the consent of Landlord, provided that all of the following conditions are satisfied (a "**Business Transfer**"): (a) Tenant must not be in Default; (b) Tenant must give Landlord written notice at least 15 Business Days before such Transfer; and (c) except in the case of an assignment or sublease to an Affiliate, the Credit Requirement (defined below) must be satisfied. Tenant's notice to Landlord shall include information and documentation evidencing the Business Transfer and showing that each of the above conditions has been satisfied. If requested by Landlord, Tenant's successor shall sign and deliver to Landlord a commercially reasonable form of assumption agreement. "**Affiliate**" shall mean an entity controlled by, controlling or under common control with Tenant. The "**Credit Requirement**" shall be deemed satisfied if, as of the date immediately preceding the date of the Business Transfer, the financial strength of (i) the entity with which Tenant is to merge or consolidate or (ii) the purchaser of substantially all of the assets of Tenant is not less than that of Tenant, as determined (x) based on credit ratings of such entity and Tenant by both Moody's and Standard & Poor's (or by either such agency alone, if applicable ratings by the other agency do not exist), or (y) if such credit ratings do not exist, then in accordance with Moody's KMV RiskCalc (i.e., the on-line software tool offered by Moody's for analyzing credit risk) based on CFO-certified financial statements for such entity and Tenant covering their last two fiscal years ending before the Transfer.

11.05. Notwithstanding anything to the contrary contained in this Section 11, neither Tenant nor any other person having a right to possess, use, or occupy (for convenience, collectively referred to in this subsection as "**Use**") the Premises shall enter into any lease, sublease, license, concession or other agreement for Use of all or any portion of the Premises

which provides for rental or other payment for such Use based, in whole or in part, on the net income or profits derived by any person that leases, possesses, uses, or occupies all or any portion of the Premises (other than an amount based on a fixed percentage or percentages of receipts or sales), and any such purported lease, sublease, license, concession or other agreement shall be absolutely void and ineffective as a transfer of any right or interest in the Use of all or any part of the Premises.

12. Liens.

Tenant shall not permit mechanics' or other liens to be placed upon the Property, Premises or Tenant's leasehold interest in connection with any work or service done or purportedly done by or for the benefit of Tenant or its subtenants or transferees. Tenant shall give Landlord notice at least 15 days prior to the commencement of any work in the Premises to afford Landlord the opportunity, where applicable, to post and record notices of non-responsibility. Tenant, within 20 days of written notice from Landlord, shall fully discharge any lien by settlement, by bonding or by insuring over the lien in the manner prescribed by the applicable lien Law and, if Tenant fails to do so, Tenant shall be deemed in Default under this Lease and, in addition to any other remedies available to Landlord as a result of such Default by Tenant, Landlord, at its option, may bond, insure over or otherwise discharge the lien. Tenant shall reimburse Landlord for any amount paid by Landlord, including, without limitation, reasonable attorneys' fees. Landlord shall have the right to require Tenant to post a performance or payment bond in connection with any work or service done or purportedly done by or for the benefit of Tenant, the cost of which, when taken in the aggregate, is greater than or equal to \$100,000.00. Tenant acknowledges and agrees that all such work or service is being performed for the sole benefit of Tenant and not for the benefit of Landlord.

13. Indemnity and Waiver of Claims.

Except to the extent caused by the negligence or willful misconduct of Landlord or any Landlord Related Parties (defined below), Tenant shall indemnify, defend and hold Landlord and Landlord Related Parties harmless against and from all liabilities, obligations, damages, penalties, claims, actions, costs, charges and expenses, including, without limitation, reasonable attorneys' fees and other professional fees (if and to the extent permitted by Law) (collectively referred to as "**Losses**"), which may be imposed upon, incurred by or asserted against Landlord or any of the Landlord Related Parties by any third party and arising out of or in connection with any damage or injury occurring in the Premises or any acts or omissions (including violations of Law) of Tenant, its trustees, managers, members, principals, beneficiaries, partners, officers, directors, employees and agents (the "**Tenant Related Parties**") or any of Tenant's transferees, contractors or licensees occurring within the Property. Tenant hereby waives all claims against and releases Landlord and its trustees, managers, members, principals, beneficiaries, partners, officers, directors, employees, Mortgagees (defined in Section 23) and agents (the "**Landlord Related Parties**") from all claims for any injury to or death of persons, damage to property or business loss in any manner related to (a) Force Majeure, (b) acts of third parties, (c) the bursting or leaking of any tank, water closet, drain or other pipe, (d) the inadequacy or failure of any security or protective services, personnel or equipment, or (e) any matter not within the reasonable control of Landlord. Notwithstanding the foregoing, except as provided in Section 15 to the contrary, Tenant shall not be required to waive any claims against Landlord where such

loss or damage is due to the negligence or willful misconduct of Landlord or any Landlord Related Parties, provided that in no event shall Landlord or any Landlord Related Party be liable to Tenant for any lost profit, damage to or loss of business or any form of special, indirect or consequential damage.

14. Insurance. Tenant shall maintain the following coverages in the following amounts:

14.01. Commercial General Liability Insurance covering claims of bodily injury, personal injury and property damage arising out of Tenants operations and contractual liabilities, including coverage formerly known as broad form, on an occurrence basis, with minimum primary limits of \$1,000,000 each occurrence and \$2,000,000 annual aggregate (and not more than \$25,000 self-insured retention) and a minimum excess/umbrella limit of \$2,000,000.

14.02. Property insurance covering (i) all office furniture, business and trade fixtures, office equipment, free-standing cabinet work, movable partitions, merchandise and all other items of Tenant's property in the Premises installed by, for, or at the expense of Tenant ("**Tenant's Property**"), and (ii) any Leasehold Improvements installed by or for the benefit of Tenant, whether pursuant to this Lease or pursuant to any prior lease or other agreement to which Tenant was a party ("**Tenant-Insured Improvements**"). Such insurance shall be written on a special cause of loss form for physical loss or damage, for the full replacement cost value (subject to reasonable deductible amounts) without deduction for depreciation of the covered items and in amounts that meet any co-insurance clauses of the policies of insurance, and shall include coverage for damage or other loss caused by fire or other peril, including vandalism and malicious mischief, theft, water damage of any type, including sprinkler leakage, bursting or stoppage of pipes, and explosion, and providing business interruption coverage for a period of one year.

14.03. Worker's Compensation and Employer's Liability or other similar insurance to the extent required by Law.

14.04. Form of Policies. The minimum limits of insurance required to be carried by Tenant shall not limit Tenant's liability. Such insurance shall (i) be issued by an insurance company that has an A.M. Best rating of not less than A-VIII; (ii) be in form and content reasonably acceptable to Landlord; and (ii) provide that it shall not be canceled or materially changed without 30 days' prior notice to Landlord, except that 10 days' prior notice may be given in the case of nonpayment of premiums. Tenant's Commercial General Liability Insurance shall (a) name Landlord, Landlord's managing agent, and any other party designated by Landlord to Tenant in writing ("**Additional Insured Parties**") as additional insureds; and (b) be primary insurance as to all claims thereunder and provide that any insurance carried by Landlord is excess and non-contributing with Tenant's insurance. Landlord shall be designated as a loss payee with respect to Tenant's Property insurance on any Tenant-Insured Improvements. Tenant shall deliver to Landlord, on or before the Commencement Date and at least 15 days before the expiration dates thereof, certificates from Tenant's insurance company on the forms currently designated "**ACORD 28**" (Evidence of Commercial Property Insurance) and "**ACORD 25-S**" (Certificate of Liability Insurance) or the equivalent. Attached to the ACORD 25-S (or equivalent) there shall be an endorsement naming the Additional Insured Parties as additional insureds which shall be binding on Tenant's insurance company and shall expressly require the

insurance company to notify each Additional Insured Party in writing at least 30 days before any termination or material change to the policies, except that 10 days' prior notice may be given in the case of nonpayment of premiums. Upon Landlord's request, Tenant shall deliver to Landlord, in lieu of such certificates, redacted copies of the policies of insurance required to be carried under Section 14.01 showing that the Additional Insured Parties are named as additional insureds.

14.05. Tenant shall maintain such increased amounts of the insurance required to be carried by Tenant under this Section 14, and such other types and amounts of insurance covering the Premises and Tenant's operations therein, as may be reasonably requested by Landlord, but not in excess of the amounts and types of insurance then being required by landlords of buildings comparable to and in the vicinity of the Buildings.

14.06. Landlord shall maintain the following insurance, together with such other insurance coverage as Landlord, in its reasonable judgment, may elect to maintain (collectively, "**Landlord's Insurance**"), the premiums of which will be included in Expenses: (1) Commercial General Liability insurance applicable to the Property, Building and Common Areas providing, on an occurrence basis, a minimum combined single limit of at least \$2,000,000.00; and (2) All Risk Property Insurance or special cause of loss insurance on the Building at replacement cost value as reasonably estimated by Landlord.

15. Subrogation.

Subject to Section 16, each party waives, and shall cause its insurance carrier to waive, any right of recovery against the other for any loss of or damage to property which loss or damage is (or, if the insurance required hereunder had been carried, would have been) covered by insurance. For purposes of this Section 15, any deductible with respect to a party's insurance shall be deemed covered by, and recoverable by such party under, valid and collectable policies of insurance.

16. Casualty Damage.

16.01. If all or any portion of the Premises becomes untenable or inaccessible by fire or other casualty to the Premises or the Common Areas (collectively a "**Casualty**"), Landlord, with reasonable promptness following the Casualty, shall cause a general contractor selected by Landlord to provide Landlord with a written estimate of the amount of time required, using standard working methods, to substantially complete the repair and restoration of the Premises and any Common Areas necessary to provide access to the Premises ("**Completion Estimate**"). Landlord shall promptly forward a copy of the Completion Estimate to Tenant. If the Completion Estimate indicates that the Premises or any Common Areas necessary to provide access to the Premises cannot be made tenable within 270 days from the date the repair is started, then either party shall have the right to terminate this Lease upon written notice to the other within 10 days after Tenant's receipt of the Completion Estimate. Tenant, however, shall not have the right to terminate this Lease if the Casualty was caused by the gross negligence or intentional misconduct of Tenant or any Tenant Related Parties. In addition, Landlord, by notice to Tenant within 90 days after the date of the Casualty, shall have the right to terminate this Lease if: (1) the Premises have been materially damaged (as determined in the reasonable judgment of Landlord) and there is less than 18 months of the Term remaining on the date of the

Casualty; (2) any Mortgagee requires that the insurance proceeds be applied to the payment of the mortgage debt; or (3) a material uninsured loss to the Buildings or Premises occurs (as determined in the reasonable judgment of Landlord).

16.02. If this Lease is not terminated, Landlord shall promptly and diligently, subject to reasonable delays for insurance adjustment or other matters beyond Landlord's reasonable control, restore the Premises and Common Areas. Such restoration shall be to substantially the same condition that existed prior to the Casualty, except for modifications required by Law or any other modifications to the Common Areas deemed reasonably desirable by Landlord. Notwithstanding Section 15, upon written notice from Landlord, Tenant shall assign or endorse over to Landlord (or to any party designated by Landlord) all property insurance proceeds payable to Tenant under Tenant's insurance with respect to any Leasehold Improvements performed by or for the benefit of Tenant; provided if the cost to repair such Leasehold Improvements as reasonably estimated by Landlord exceeds the amount of insurance proceeds received by Landlord from Tenant's insurance carrier, the excess cost of such repairs shall be paid by Tenant to Landlord prior to Landlord's commencement of repairs and any of such amounts so paid but not used by Landlord shall be promptly returned to Tenant after completion. Within 30 days of written demand, Tenant shall also pay Landlord for any additional excess costs that are reasonably determined by Landlord during the performance of the repairs to such Leasehold Improvements to exceed Landlord's original estimate of excess costs. In no event shall Landlord be required to spend more for the restoration of the Premises and Common Areas than the proceeds received by Landlord, whether insurance proceeds or proceeds from Tenant. Landlord shall not be liable for any inconvenience to Tenant, or injury to Tenant's business resulting in any way from the Casualty or the repair thereof. Provided that Tenant is not in Default, during any period of time that all or a material portion of the Premises is rendered untenable as a result of a Casualty, the Rent shall abate for the portion of the Premises that is untenable and not used by Tenant.

17. Condemnation.

Either party may terminate this Lease if any material part of the Premises is taken or condemned for any public or quasi-public use under Law, by eminent domain or private purchase in lieu thereof (a "Taking"). Landlord shall also have the right to terminate this Lease if there is a Taking of any portion of the Buildings or Property which would have a material adverse effect on Landlord's ability to profitably operate the remainder of the Buildings. The terminating party shall provide written notice of termination to the other party within 45 days after it first receives notice of the Taking. The termination shall be effective as of the effective date of any order granting possession to, or vesting legal title in, the condemning authority. If this Lease is not terminated, Base Rent, Tenant's Building Pro Rata Share and Tenant's Common Area Pro Rata Share shall be appropriately adjusted to account for any reduction in the square footage of the Buildings or Premises. All compensation awarded for a Taking shall be the property of Landlord. The right to receive compensation or proceeds is expressly waived by Tenant, provided, however, Tenant may file a separate claim for Tenant's Property and Tenant's reasonable relocation expenses, provided the filing of the claim does not diminish the amount of Landlord's award. If only a part of the Premises is subject to a Taking and this Lease is not terminated, Landlord, with reasonable diligence, will restore the remaining portion of the Premises as nearly as practicable to the condition immediately prior to the Taking.

18. Events of Default.

In addition to any other default specifically described in this Lease, each of the following occurrences shall be a “**Default**”: (a) Tenant’s failure to pay any portion of Rent when due, if the failure continues for 5 Business Days after written notice to Tenant (“**Monetary Default**”); (b) Tenant’s failure (other than a Monetary Default) to comply with any term, provision, condition or covenant of this Lease, if the failure is not cured within 30 days after written notice to Tenant provided, however, if Tenant’s failure to comply cannot reasonably be cured within 30 days, Tenant shall be allowed additional time (not to exceed 60 days) as is reasonably necessary to cure the failure so long as Tenant begins the cure within 30 days and diligently pursues the cure to completion; (c) Tenant permits a Transfer without Landlord’s required approval or otherwise in violation of Section 11 of this Lease; (d) Tenant or any Guarantor becomes insolvent, makes a transfer in fraud of creditors, makes an assignment for the benefit of creditors, admits in writing its inability to pay its debts when due or forfeits or loses its right to conduct business; (e) the leasehold estate is taken by process or operation of Law; (f) in the case of any ground floor or retail Tenant, Tenant does not take possession of or abandons or vacates all or any portion of the Premises; or (g) Tenant is in default beyond any notice and cure period under any other lease or agreement with Landlord at the Buildings or Property that is associated with Tenant’s lease of the Premises under this Lease. If Landlord provides Tenant with notice of Tenant’s failure to comply with any specific provision of this Lease on 3 separate occasions during any 12 month period, Tenant’s subsequent violation of such provision shall, at Landlord’s option, be an incurable Default by Tenant. All notices sent under this Section shall be in satisfaction of, and not in addition to, notice required by Law.

19. Remedies.

19.01. Upon Default, Landlord shall have the right to pursue any one or more of the following remedies:

(a) Terminate this Lease, in which case Tenant shall immediately surrender the Premises to Landlord. If Tenant fails to surrender the Premises, Landlord, in compliance with Law, may enter upon and take possession of the Premises and remove Tenant, Tenant’s Property and any party occupying the Premises. Tenant shall pay Landlord, on demand, all past due Rent and other losses and damages Landlord suffers as a result of Tenant’s Default, including, without limitation, all Costs of Reletting (defined below) and any deficiency that may arise from reletting or the failure to relet the Premises. “**Costs of Reletting**” shall include all reasonable costs and expenses incurred by Landlord in reletting or attempting to relet the Premises, including, without limitation, legal fees, brokerage commissions, the cost of alterations and the value of other concessions or allowances granted to a new tenant.

(b) Terminate Tenant’s right to possession of the Premises and, in compliance with Law, remove Tenant, Tenant’s Property and any parties occupying the Premises. Landlord may (but shall not be obligated to) relet all or any part of the Premises, without notice to Tenant, for such period of time and on such terms and conditions (which may include concessions, free rent and work allowances) as Landlord in its absolute discretion shall determine. Landlord may collect and receive all rents and other income from the reletting. Tenant shall pay Landlord on demand all past due Rent, all Costs of Reletting and any deficiency arising from the reletting or

failure to relet the Premises. The re-entry or taking of possession of the Premises shall not be construed as an election by Landlord to terminate this Lease. Landlord agrees to use reasonable efforts to mitigate damages, provided that those efforts shall not require Landlord to relet the Premises in preference to any other space in the Building or to relet the Premises to any party that Landlord could reasonably reject as a transferee pursuant to Section 11.

19.02. In lieu of calculating damages under Section 19.01, Landlord may elect to receive as damages the sum of (a) all Rent accrued through the date of termination of this Lease or Tenant's right to possession, and (b) an amount equal to the total Rent that Tenant would have been required to pay for the remainder of the Term discounted to present value at the Prime Rate (defined below) then in effect, minus the then present fair rental value of the Premises for the remainder of the Term, similarly discounted, after deducting all anticipated Costs of Reletting. "**Prime Rate**" shall be the per annum interest rate publicly announced as its prime or base rate by a federally insured bank selected by Landlord in the state in which the Buildings are located.

19.03. If Tenant is in Default of any of its non-monetary obligations under this Lease, Landlord shall have the right to perform such obligations. Tenant shall reimburse Landlord for the actual cost of such performance upon demand together with an administrative charge equal to 10% of the cost of the work performed by Landlord. The repossession or re-entering of all or any part of the Premises shall not relieve Tenant of its liabilities and obligations under this Lease. No right or remedy of Landlord shall be exclusive of any other right or remedy. Each right and remedy shall be cumulative and in addition to any other right and remedy now or subsequently available to Landlord at Law or in equity.

20. Limitation of Liability.

NOTWITHSTANDING ANYTHING TO THE CONTRARY CONTAINED IN THIS LEASE, THE LIABILITY OF LANDLORD (AND OF ANY SUCCESSOR LANDLORD) SHALL BE LIMITED TO THE LESSER OF (A) THE INTEREST OF LANDLORD IN THE PROPERTY, OR (B) THE EQUITY INTEREST LANDLORD WOULD HAVE IN THE PROPERTY IF THE PROPERTY WERE ENCUMBERED BY THIRD PARTY DEBT IN AN AMOUNT EQUAL TO 70% OF THE VALUE OF THE PROPERTY. TENANT SHALL LOOK SOLELY TO LANDLORD'S INTEREST IN THE PROPERTY FOR THE RECOVERY OF ANY JUDGMENT OR AWARD AGAINST LANDLORD OR ANY LANDLORD RELATED PARTY. NEITHER LANDLORD NOR ANY LANDLORD RELATED PARTY SHALL BE PERSONALLY LIABLE FOR ANY JUDGMENT OR DEFICIENCY, AND IN NO EVENT SHALL LANDLORD OR ANY LANDLORD RELATED PARTY BE LIABLE TO TENANT FOR ANY LOST PROFIT, DAMAGE TO OR LOSS OF BUSINESS OR ANY FORM OF SPECIAL, INDIRECT OR CONSEQUENTIAL DAMAGE. BEFORE FILING SUIT FOR AN ALLEGED DEFAULT BY LANDLORD, TENANT SHALL GIVE LANDLORD AND THE MORTGAGEE(S) WHOM TENANT HAS BEEN NOTIFIED HOLD MORTGAGES (DEFINED IN SECTION 23 BELOW), NOTICE AND REASONABLE TIME TO CURE THE ALLEGED DEFAULT. WITHOUT LIMITING THE FOREGOING, IN NO EVENT SHALL LANDLORD OR ANY MORTGAGEES OR LANDLORD RELATED PARTIES EVER BE LIABLE FOR ANY CONSEQUENTIAL OR INCIDENTAL DAMAGES OR ANY LOST PROFITS OF TENANT. IN NO EVENT SHALL TENANT BE LIABLE TO LANDLORD FOR ANY LOST PROFIT, DAMAGE TO OR LOSS OF BUSINESS OR ANY FORM OF

SPECIAL, INDIRECT OR CONSEQUENTIAL DAMAGE, EXCEPT AS PROVIDED IN SECTION 22 BELOW. FOR PURPOSES HEREOF, "INTEREST OF LANDLORD IN THE PROPERTY" SHALL INCLUDE RENTS DUE FROM TENANTS, INSURANCE PROCEEDS, AND PROCEEDS FROM CONDEMNATION OR EMINENT DOMAIN PROCEEDINGS (PRIOR TO THE DISTRIBUTION OF SAME TO ANY PARTNER OR SHAREHOLDER OF LANDLORD OR ANY OTHER THIRD PARTY).

21. Relocation.

Landlord, at its expense, at any time before or during the Term, may relocate Tenant from the Premises to space of substantially comparable size and utility ("**Relocation Space**") within the Office Building upon 90 days' prior written notice to Tenant. The Relocation Space shall be built out at Landlord's sole cost and expense and must contain similar finishes (subject to commercial availability) including lighting fixtures, and approximately the same Rentable Square Footage as the Premises (e.g., not less than 90% of the Rentable Square Footage of the Premises) and the same number of work stations, offices, breakrooms and reception areas as are contained in the Premises as of the date Tenant receives Landlord's notice of relocation. From and after the date of the relocation, the Base Rent, Tenant's Building Pro Rata Share and Tenant's Common Area Pro Rata Share shall be adjusted based on the rentable square footage of the Relocation Space. Landlord shall pay Tenant's reasonable costs of relocation, including all costs for moving Tenant's furniture, equipment, supplies and other personal property, as well as the cost of printing and distributing change of address notices to Tenant's customers and one month's supply of stationery showing the new address. Landlord shall also reimburse Tenant for the reasonable cost to install and connect Tenant's Cable in the Relocation Space in the manner such Cable existed in the Premises prior to the relocation.

22. Holding Over.

If Tenant fails to surrender all or any part of the Premises at the termination of this Lease, occupancy of the Premises after termination shall be that of a tenancy at sufferance. Tenant's occupancy shall be subject to all the terms and provisions of this Lease, and Tenant shall pay an amount (on a per month basis without reduction for partial months during the holdover) equal to 150% of the sum of the Base Rent and Additional Rent due for the period immediately preceding the holdover. No holdover by Tenant or payment by Tenant after the termination of this Lease shall be construed to extend the Term or prevent Landlord from immediate recovery of possession of the Premises by summary proceedings or otherwise. If Tenant fails to vacate the Premises within 30 days after Landlord notifies Tenant that Landlord has entered into a lease for the Premises or has received a bona fide offer to lease the Premises and that Landlord will be unable to deliver possession or perform improvements due to Tenant's holdover, and if Landlord is unable to deliver possession of the Premises to a new tenant or to perform improvements for a new tenant as a result of Tenant's holdover, then Tenant shall be liable for all damages, including but not limited to consequential damages, that Landlord suffers from the holdover as a result of Landlord's inability to so deliver possession of the Premises or perform improvements.

23. Subordination to Mortgages; Estoppel Certificate.

23.01. Tenant accepts this Lease subject and subordinate to any mortgage(s), deed(s) of trust, deeds to secure debt, ground lease(s) or other lien(s) now or subsequently arising upon the Premises, the Buildings or the Property, and to renewals, modifications, refinancings and extensions thereof (collectively referred to as a "**Mortgage**"). The party having the benefit of a Mortgage shall be referred to as a "**Mortgagee**". This clause shall be self-operative, but upon request from a Mortgagee, Tenant shall execute a commercially reasonable subordination agreement in favor of the Mortgagee. As an alternative, a Mortgagee shall have the right at any time to subordinate its Mortgage to this Lease. Upon request, Tenant, without charge, shall attorn to any successor to Landlord's interest in this Lease. Landlord and Tenant shall each, within 10 days after receipt of a written request from the other, execute and deliver a commercially reasonable estoppel certificate to those parties as are reasonably requested by the other (including a Mortgagee or prospective purchaser). Without limitation, such estoppel certificate may include a certification as to the status of this Lease, the existence of any defaults and the amount of Rent that is due and payable.

23.02. In the event Mortgagee enforces its rights under the Mortgage, Tenant, at Mortgagee's option, will attorn to Mortgagee or its successor; provided, however, that Mortgagee or its successor shall not be liable for or bound by (i) any payment of any Rent installment which may have been made more than 30 days before the due date of such installment, (ii) any act or omission of or default by Landlord under this Lease (but Mortgagee, or such successor, shall be subject to the continuing obligations of landlord under the Lease to the extent arising from and after such succession to the extent of Mortgagee's, or such successor's, interest in the Property), (iii) any credits, claims, setoffs or defenses which Tenant may have against Landlord or (iv) any obligation under this Lease to maintain a fitness facility at the Buildings, if any. Tenant, upon the reasonable request by Mortgagee or such successor in interest, shall execute and deliver an instrument or instruments confirming such attornment.

24. Notice.

All demands, approvals, consents or notices (collectively referred to as a "**notice**") shall be in writing and delivered by hand or sent by registered, express, or certified mail, with return receipt requested or with delivery confirmation requested from the U.S. postal service, or sent by overnight or same day courier service at the party's respective Notice Address(es) set forth in Section 1; provided, however, notices sent by Landlord regarding general building operational matters may be posted in the building mailroom or the general building newsletter or sent via e-mail to the e-mail address provided by Tenant to Landlord for such purpose. In addition, if the Buildings are closed (whether due to emergency, governmental order or any other reason), then any notice address at the Buildings shall not be deemed a required notice address during such closure, and, unless Tenant has provided an alternative valid notice address to Landlord for use during such closure, any notices sent during such closure may be sent via e-mail or in any other practical manner reasonably designed to ensure receipt by the intended recipient. Each notice shall be deemed to have been received on the earlier to occur of actual delivery or the date on which delivery is refused, or, if Tenant has vacated the Premises or any other Notice Address of Tenant without providing a new Notice Address, 3 days after notice is deposited in the U.S. mail or with a courier service in the manner described above. Either party may, at any time, change its Notice Address (other than to a post office box address) by giving the other party written notice of the new address.

25. Surrender of Premises.

At the termination of this Lease or Tenant's right of possession, Tenant shall remove Tenant's Property from the Premises, and quit and surrender the Premises to Landlord, broom clean, and in good order, condition and repair, ordinary wear and tear, damage which Landlord is obligated to repair hereunder and damage due to Casualty (subject to the terms of Section 16) excepted. If Tenant fails to remove any of Tenant's Property, or to restore the Premises to the required condition, within 2 Business Days after termination of this Lease or Tenant's right to possession, Landlord, at Tenant's sole cost and expense, shall be entitled (but not obligated) to remove and store Tenant's Property and/or perform such restoration of the Premises. Landlord shall not be responsible for the value, preservation or safekeeping of Tenant's Property. Tenant shall pay Landlord, upon demand, the expenses and storage charges incurred. If Tenant fails to remove Tenant's Property from the Premises or storage, within 30 days after notice, Landlord may deem all or any part of Tenant's Property to be abandoned and, at Landlord's option, title to Tenant's Property shall vest in Landlord or Landlord may dispose of Tenant's Property in any manner Landlord deems appropriate.

26. Miscellaneous.

26.01. This Lease shall be interpreted and enforced in accordance with the Laws of the state or commonwealth in which the Buildings are located and Landlord and Tenant hereby irrevocably consent to the jurisdiction and proper venue of such state or commonwealth. If any term or provision of this Lease shall to any extent be void or unenforceable, the remainder of this Lease shall not be affected. If there is more than one Tenant or if Tenant is comprised of more than one party or entity, the obligations imposed upon Tenant shall be joint and several obligations of all the parties and entities, and requests or demands from any one person or entity comprising Tenant shall be deemed to have been made by all such persons or entities. Notices to any one person or entity shall be deemed to have been given to all persons and entities. Tenant represents and warrants to Landlord, and agrees, that each individual executing this Lease on behalf of Tenant is authorized to do so on behalf of Tenant and that the entity(ies) or individual(s) constituting Tenant or Guarantor or which may own or control Tenant or Guarantor or which may be owned or controlled by Tenant or Guarantor are not and at no time will be (i) in violation of any Laws relating to terrorism or money laundering, or (ii) among the individuals or entities identified on any list compiled pursuant to Executive Order 13224 for the purpose of identifying suspected terrorists or on the most current list published by the U.S. Treasury Department Office of Foreign Assets Control at its official website, <http://www.treasury.gov/resource-center/sanctions/SDN-List/Pages/default.aspx> or any replacement website or other replacement official publication of such list.

26.02. If Landlord retains an attorney or institutes legal proceedings due to Tenant's failure to pay Rent when due, then Tenant shall be required to pay Additional Rent in an amount equal to the reasonable attorneys' fees and costs actually incurred by Landlord in connection therewith. Notwithstanding the foregoing, in any action or proceeding between Landlord and Tenant, including any appellate or alternative dispute resolution proceeding, the prevailing party shall be entitled to recover from the non-prevailing party all of its costs and expenses in connection therewith, including, but not limited to, reasonable attorneys' fees actually incurred. Landlord and Tenant hereby waive any right to trial by jury in any proceeding based upon a

breach of this Lease. No failure by either party to declare a default immediately upon its occurrence, nor any delay by either party in taking action for a default, nor Landlord's acceptance of Rent with knowledge of a default by Tenant, shall constitute a waiver of the default, nor shall it constitute an estoppel.

26.03. Whenever a period of time is prescribed for the taking of an action by Landlord or Tenant (other than the payment of the Security Deposit or Rent), the period of time for the performance of such action shall be extended by the number of days that the performance is actually delayed due to strikes, acts of God, shortages of labor or materials, war, terrorist acts, pandemics, civil disturbances and other causes beyond the reasonable control of the performing party ("**Force Majeure**").

26.04. Landlord shall have the right to transfer and assign, in whole or in part, all of its rights and obligations under this Lease and in the Buildings and Property. Upon transfer, Landlord shall be released from any further obligations hereunder and Tenant agrees to look solely to the successor in interest of Landlord for the performance of such obligations, provided that any successor pursuant to a voluntary, third party transfer (but not as part of an involuntary transfer resulting from a foreclosure or deed in lieu thereof) shall have assumed Landlord's obligations under this Lease from and after the date of the transfer.

26.05. Landlord has delivered a copy of this Lease to Tenant for Tenant's review only and the delivery of it does not constitute an offer to Tenant or an option. Tenant represents that it has dealt directly with and only with the Broker (described in Section 1.10) as a broker, agent or finder in connection with this Lease. Tenant shall indemnify and hold Landlord and the Landlord Related Parties harmless from all claims of any other brokers, agents or finders claiming to have represented Tenant in connection with this Lease. Landlord shall indemnify and hold Tenant and the Tenant Related Parties harmless from all claims of any brokers, agents or finders claiming to have represented Landlord in connection with this Lease. Equity Office Properties Management Corp., or such other entity affiliated with Equity Office Properties Management Corp. that is involved in the negotiation of this Lease (each referred to as "**EOPMC**"), represents only the Landlord in this transaction. Any assistance rendered by any agent or employee of EOPMC in connection with this Lease or any subsequent amendment or modification or any other document related hereto has been or will be made as an accommodation to Tenant solely in furtherance of consummating the transaction on behalf of Landlord, and not as agent for Tenant. Landlord agrees to pay a brokerage commission to Tenant's Broker and Landlord's Broker in accordance with the terms of separate commission agreements entered into or to be entered into between Landlord and Tenant's Broker, and Landlord and Landlord's Broker, respectively, provided that in no event shall Landlord be obligated to pay a commission to Tenant's Broker or Landlord's Broker in connection with any extension of the Term or in connection with any additional space that is leased by Tenant pursuant to the terms of this Lease except as may be specifically provided otherwise in such agreement or future agreement between Landlord and Tenant's Broker, and Landlord and Landlord's Broker, respectively.

26.06. Time is of the essence with respect to Tenant's exercise of any expansion, renewal or extension rights granted to Tenant. The expiration of the Term, whether by lapse of time, termination or otherwise, shall not relieve either party of any obligations which accrued prior to or which may continue to accrue after the expiration or termination of this Lease.

26.07. Tenant may peacefully have, hold and enjoy the Premises, subject to the terms of this Lease, provided Tenant pays the Rent and fully performs all of its covenants and agreements. This covenant shall be binding upon Landlord and its successors only during its or their respective periods of ownership of the Buildings.

26.08. This Lease does not grant any rights to light or air over or about the Buildings. Landlord excepts and reserves exclusively to itself any and all rights not specifically granted to Tenant under this Lease. Landlord reserves the right to make changes to the Property, Buildings and Common Areas as Landlord deems appropriate, provided the changes do not materially adversely affect Tenant's ability to use the Premises for the Permitted Use, and Landlord uses reasonable efforts to minimize any material interference with Tenant's use and occupancy of the Premises for the Permitted Use. This Lease constitutes the entire agreement between the parties and supersedes all prior agreements and understandings related to the Premises, including all lease proposals, letters of intent and other documents. Neither party is relying upon any warranty, statement or representation not contained in this Lease. This Lease may be modified only by a written agreement signed by an authorized representative of Landlord and Tenant. Wherever this Lease requires Landlord to provide a customary service or to act in a reasonable manner (whether in incurring an expense, establishing a rule or regulation, providing an approval or consent, or performing any other act), this Lease shall be deemed also to provide that whether such service is customary or such conduct is reasonable shall be determined by reference to the practices of owners of buildings that (i) are comparable to the Buildings in size, age, class, quality and location, and (ii) at Landlord's option, have been, or are being prepared to be, certified under the U.S. Green Building Council's Leadership in Energy and Environmental Design (LEED) rating system or a similar rating system.

26.09. Submission of this Lease by Landlord is not an offer to enter into this Lease but rather is a solicitation for such an offer by Tenant. Landlord shall not be bound by this Lease until Landlord has executed and delivered the same to Tenant. Tenant agrees that its execution of this Lease constitutes a firm offer to enter the same, which may not be withdrawn for a period of 30 days after delivery to Landlord (or such other period as may be expressly provided in any other agreement signed by the parties).

26.10. If Landlord is advised by its counsel at any time that any part of the payments by Tenant to Landlord under this Lease may be characterized as unrelated business income under the United States Internal Revenue Code and its regulations, then Tenant shall enter into any amendment proposed by Landlord to avoid such income, so long as the amendment does not require Tenant to make more payments or accept fewer services from Landlord, than this Lease provides.

26.11. This Lease may be executed in counterparts and shall constitute an agreement binding on all parties notwithstanding that all parties are not signatories to the original or the same counterpart provided that all parties are furnished a copy or copies thereof reflecting the signature of all parties. Transmission of a facsimile or by email of a pdf copy of the signed counterpart of the Lease shall be deemed the equivalent of the delivery of the original, and any party so delivering a facsimile or pdf copy of the signed counterpart of the Lease by email transmission shall in all events deliver to the other party an original signature promptly upon request.

Landlord and Tenant have executed this Lease under seal in two or more counterparts as of the day and year first above written.

LANDLORD:

**MA-RIVERVIEW/245 FIRST STREET, L.L.C., a Delaware
limited liability company**

By: /s/ John Conley

Name: John Conley

Title: Vice President, Asset Managaement

TENANT:

AKEBIA THERAPEUTICS, INC., a Delaware corporation

By: /s/ Jason A. Amello

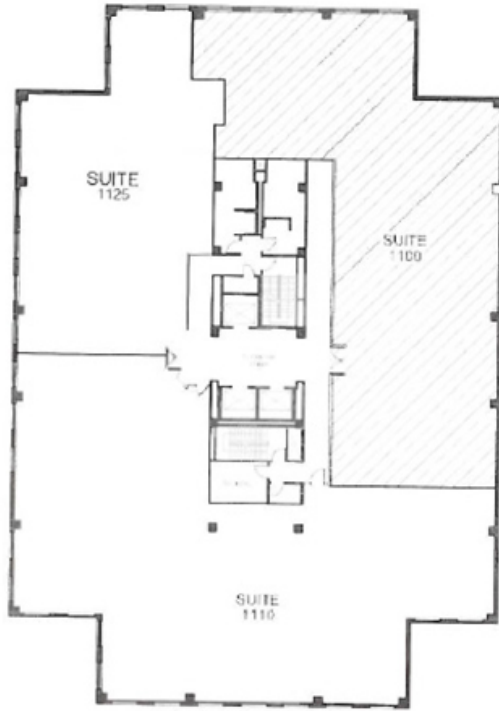
Name: Jason A. Amello

Title: SVP, CFO

EXHIBIT A

OUTLINE AND LOCATION OF PREMISES

This Exhibit is attached to and made a part of the Office Lease Agreement (the "Lease") by and between MA-RIVERVIEW/245 FIRST STREET, L.L.C., a Delaware limited liability company ("Landlord"), and AKEBIA THERAPEUTICS, INC., a Delaware corporation ("Tenant"), for space in the Office Building located at 245 First Street, Cambridge, Massachusetts 02142.



245 First Street
Cambridge, MA
11th Floor



EXHIBIT B

EXPENSES AND TAXES

This Exhibit is attached to and made a part of the Office Lease Agreement (the "Lease") by and between **MA-RIVERVIEW/245 FIRST STREET, L.L.C., a Delaware limited liability company ("Landlord")**, and **AKEBIA THERAPEUTICS, INC., a Delaware corporation ("Tenant")**, for space in the Office Building located at 245 First Street, Cambridge, Massachusetts 02142. Capitalized terms used but not defined herein shall have the meanings given in the Lease.

1. Payments.

1.01. Tenant shall pay (i) Tenant's Building Pro Rata Share of the amount, if any, by which Expenses (defined below) for each calendar year during the Term exceed Expenses for the Base Year (the "**Expense Excess**"), (ii) Tenant's Common Area Pro Rata Share of the amount, if any, by which Taxes (defined below) for each calendar year during the Term exceed Taxes for the Base Year (the "**Tax Excess**") and (iii) Tenant's Common Area Pro Rata Share of the amount, if any, by which Common Area Expenses (defined below) for the each calendar during the Term exceed Common Area Expenses for the Base Year (the "**Common Area Expense Excess**"). If Expenses, Taxes or Common Area Expenses in any calendar year decrease below the amount of Expenses, Taxes or Common Area Expenses for the Base Year, Tenant's Building Pro Rata Share of Expenses and Tenant's Common Area Pro Rata Share of Taxes and Common Area Expenses, as the case may be, for that calendar year shall be \$0. Landlord shall provide Tenant with a good faith estimate of the Expense Excess, Tax Excess and Common Area Expense Excess for each calendar year during the Term. On or before the first day of each month, Tenant shall pay to Landlord a monthly installment equal to one-twelfth of Tenant's Building Pro Rata Share of Landlord's estimate of the Expense Excess and one-twelfth of Tenant's Common Area Pro Rata Share of Landlord's estimate of the Tax Excess and Common Area Expense Excess. If Landlord determines that its good faith estimate of the Expense Excess, the Tax Excess or the Common Area Expense Excess was incorrect by a material amount, Landlord may provide Tenant with a revised estimate, provided that Landlord shall not provide a revised estimate more than twice per calendar year. After its receipt of the revised estimate, Tenant's monthly payments shall be based upon the revised estimate. If Landlord does not provide Tenant with an estimate of the Expense Excess, the Tax Excess or the Common Area Expense Excess by January 1 of a calendar year, Tenant shall continue to pay monthly installments based on the previous year's estimate(s) until Landlord provides Tenant with the new estimate. Upon delivery of the new estimate, an adjustment shall be made for any month for which Tenant paid monthly installments based on the previous year's estimate. Tenant shall pay Landlord the amount of any underpayment within 30 days after receipt of the new estimate. Any overpayment shall be refunded to Tenant within 30 days or credited against the next due future installment(s) of Additional Rent.

1.02. As soon as is practical following the end of each calendar year, Landlord shall furnish Tenant with a statement of the actual Expenses and Expense Excess, the actual Taxes and Tax Excess and the actual Common Area Expenses and Common Area Expense Excess for the prior calendar year. Landlord shall use reasonable efforts to furnish the statement of actual

Expenses on or before June 1 of the calendar year immediately following the calendar year to which the statement applies. If the estimated Expense Excess, the estimated Tax Excess or the estimated Common Area Expense Excess for the prior calendar year is more than the actual Expense Excess, actual Tax Excess and actual Common Area Expense Excess, as the case may be, for the prior calendar year, Landlord shall either provide Tenant with a refund or apply any overpayment by Tenant against Additional Rent due or next becoming due, provided if the Term expires before the determination of the overpayment, Landlord shall refund any overpayment to Tenant after first deducting the amount of Rent due. If the estimated Expense Excess, estimated Tax Excess or estimated Common Area Expense Excess for the prior calendar year is less than the actual Expense Excess, actual Tax Excess or actual Common Area Expense Excess, as the case may be, for such prior year, Tenant shall pay Landlord, within 30 days after its receipt of the statement of Expenses, Taxes or Common Area Expenses, any underpayment for the prior calendar year.

2. Expenses.

2.01. "**Expenses**" means all costs and expenses incurred in each calendar year in connection with operating, maintaining, repairing, and managing the Office Building, including the Common Areas located within the Office Building but excluding the Common Areas located in the Science Building and the shared Common Areas for both the Office Building and the Science Building, and excluding the Common Area Expenses set forth in Section 2.02 below. Landlord agrees to act in a commercially reasonable manner in incurring Expenses, taking into consideration the class and quality of the Office Building. "Expenses" shall include, but not be limited to: (a) all labor and labor related costs, including wages, salaries, bonuses, taxes, insurance, uniforms, training, retirement plans, pension plans and other employee benefits; (b) management fees in an amount equal to 3% of the gross revenues from the Office Building and the Property; (c) the cost of equipping, staffing and operating an on-site and/or off-site management office for the Office Building, provided if the management office services one or more other buildings or properties, the shared costs and expenses of equipping, staffing and operating such management office(s) shall be equitably prorated and apportioned between the Office Building and the other buildings or properties; (d) accounting costs for the Office Building; (e) the cost of services; (f) rental and purchase cost of parts, supplies, tools and equipment; (g) insurance premiums and deductibles; (h) electricity, gas and other utility costs attributable to the Office Building; (i) expenses of periodic routine testing to assure that the Premises and surrounding land are free of hazardous materials, agents or substances, and to assure compliance with codes, regulations and Laws; and (j) the amortized cost of capital improvements (as distinguished from replacement parts or components installed in the ordinary course of business) made subsequent to the Base Year which are: (1) intended to effect economies in the operation or maintenance of the Property, reduce current or future Expenses, enhance the safety or security of the Property or its occupants, or enhance the environmental sustainability of the Property's operations, (2) replacements or modifications of nonstructural items located in the Base Building or Common Areas located within the Office Building that are required to keep the Base Building or Common Areas located within the Office Building in good condition, or (3) required under any Law that is enacted, or first interpreted to apply to the Property, after the date of the Lease. The cost of capital improvements shall be amortized by Landlord over the lesser of the Payback Period (defined below) or the useful life of the capital improvement as reasonably determined by Landlord. The amortized cost of capital

improvements may, at Landlord's option, include actual or imputed interest at the rate that Landlord would reasonably be required to pay to finance the cost of the capital improvement. **"Payback Period"** means the reasonably estimated period of time that it takes for the cost savings resulting from a capital improvement to equal the total cost of the capital improvement. Landlord, by itself or through an affiliate, shall have the right to directly perform, provide and be compensated for any services under the Lease. If Landlord incurs Expenses for the Buildings or Property together with one or more other buildings or properties, whether pursuant to a reciprocal easement agreement, common area agreement or otherwise, the shared costs and expenses shall be equitably prorated and apportioned between the Buildings and Property and the other buildings or properties.

2.02. **"Common Area Expenses"** means all costs and expenses incurred in each calendar year in connection with operating, maintaining, repairing and managing the shared Common Areas of the Buildings and the Property. Landlord agrees to act in commercially reasonable manner in incurring Common Area Expenses, taking into consideration the class and quality of the Common Areas of the Buildings and the Property. Common Area Expenses include, without limitation: (a) security for the shared Common Areas of the Buildings; (b) electricity, gas and other utility costs with respect to the shared Common Areas of the Buildings; (c) repairs to the shared Common Areas of the Buildings; and (d) the amortized cost of capital improvements (as distinguished from replacement parts or components installed in the ordinary course of business) made subsequent to the Commencement Date which are: (1) intended to effect economies in the operation or maintenance of the Property, reduce current or future Common Area Expenses, enhance the safety or security of the Property or its occupants, or enhance the environmental sustainability of the Property's operations, (2) replacements or modifications of nonstructural items located in the Base Building or Common Areas of the Buildings that are required to keep the Base Building or Common Areas of the Buildings in good condition, or (3) required under any Law that is enacted, or first interpreted to apply to the Property, after the date of the Lease. The cost of capital improvements shall be amortized by Landlord over the lesser of the Payback Period (defined in Section 2.01 above) or the useful life of the capital improvement as reasonably determined by Landlord. Common Area Expenses shall not include any costs and expenses incurred with respect to the Common Areas located In the Office Building and the Common Areas located in the Science Building.

2.03. Expenses and Common Area Expenses shall not include: the cost of capital improvements (except as set forth above); depreciation; principal payments of mortgage and other non-operating debts of Landlord; the cost of repairs or other work to the extent Landlord is reimbursed by insurance or condemnation proceeds; costs in connection with leasing space in the Buildings, including brokerage commissions; lease concessions, including rental abatements and construction allowances granted to specific tenants; costs incurred in connection with the sale, financing or refinancing of the Buildings; fines, interest and penalties incurred due to the late payment of Taxes, Expenses or Common Area Expenses; organizational expenses associated with the creation and operation of the entity which constitutes Landlord; ground lease rental; wages, salaries, fees, and fringe benefits (**"Labor Costs"**) paid to executive personnel or officers or partners of Landlord, except that if such individuals provide services directly related to the operation, maintenance or ownership of the Buildings which, if provided directly by a general manager/property manager or its general support staff, would normally be chargeable as an operating expense of a comparable office Building, then an appropriate pro rata share of the

Labor Costs of such individuals that is reflective of the extent to which such individuals are providing such services to the Building may be included in Expenses; costs incurred by Landlord in connection with the correction of defects in design and original construction of the Building or Property; the cost or expense of any services or benefits provided generally to other tenants in the Building and not provided or available to Tenant; sums (other than management fees, it being agreed that the management fees included in Expenses are as described in Section 2.01 above) paid to subsidiaries or other affiliates of Landlord for services on or to the Property, Building and/or Premises, but only to the extent that the costs of such services exceed the competitive cost for such services rendered by persons or entities of similar skill, competence and experience; any general administrative expenses, which costs would not be chargeable to operating expenses of the Building in accordance with generally accepted accounting principles, consistently applied; or any penalties or damages that Landlord pays to Tenant under this Lease or to other tenants in the Buildings under their respective leases.

2.04. If at any time during a calendar year the Office Building is not at least 95% occupied (or a service provided by Landlord to tenants of the Office Building generally is not provided by Landlord to a tenant that provides such service itself, or any tenant of the Office Building is entitled to free rent, rent abatement or the like), Expenses and Common Area Expenses (but not Taxes) shall, at Landlord's option, be determined as if the Office Building had been 95% occupied (and all services provided by Landlord to tenants of the Office Building generally had been provided by Landlord to all tenants, and no tenant of the Office Building had been entitled to free rent, rent abatement or the like) during that calendar year. If Expenses and Common Area Expenses for a calendar year are determined as provided in the prior sentence, Expenses and Common Area Expenses for the Base Year shall also be determined in such manner. Notwithstanding the foregoing, Landlord may calculate the extrapolation of Expenses and Common Area Expenses under this Section based on 100% occupancy and service so long as such percentage is used consistently for each year of the Term. The extrapolation of Expenses and Common Area Expenses under this Section shall be performed in accordance with the methodology specified by the Building Owners and Managers Association.

3. **"Taxes"** shall mean: (a) all real property taxes and other assessments on the Office Building, the Science Building and/or Property, including, but not limited to, gross receipts taxes, assessments for special improvement districts and building improvement districts, governmental charges, fees and assessments for police, fire, traffic mitigation or other governmental service of purported benefit to the Property, taxes and assessments levied in substitution or supplementation in whole or in part of any such taxes and assessments and the Property's share of any real estate taxes and assessments under any reciprocal easement agreement, common area agreement or similar agreement as to the Property; (b) all personal property taxes for property that is owned by Landlord and used in connection with the operation, maintenance and repair of the Property; and (c) all commercially reasonable costs and fees incurred in connection with seeking reductions in any tax liabilities described in (a) and (b), including, without limitation, any such costs incurred by Landlord for compliance, review and appeal of tax liabilities. Without limitation, Taxes shall be determined without regard to any "green building" credit and shall not include any income, capital levy, transfer, capital stock, gift, estate or inheritance tax or any interest or penalties incurred due to the late payment of Taxes. If a change in Taxes is obtained for any year of the Term during which Tenant paid Tenant's Common Area Pro Rata Share of any Tax Excess, then Taxes for that year will be

retroactively adjusted and Landlord shall provide Tenant with a credit, if any, based on the adjustment. Likewise, if a change is obtained for Taxes for the Base Year, Taxes for the Base Year shall be restated and the Tax Excess for all subsequent years shall be recomputed. Tenant shall pay Landlord the amount of Tenant's Common Area Pro Rata Share of any such increase in the Tax Excess within 30 days after Tenant's receipt of a statement from Landlord. For the purpose of determining Taxes for any given calendar year, the amount to be included in Taxes for such year shall be as follows: (1) with respect to any special assessment that is payable in installments, Taxes for such year shall include the amount of the installment (and any interest paid or payable by Landlord) due and payable during such calendar year; and (2) with respect to all other real estate taxes, Taxes for such year shall, at Landlord's election, include either the amount accrued, assessed or otherwise imposed for such calendar year, provided that Landlord's election shall be applied consistently throughout the Term of the Lease.

4. Audit Rights. Within 60 days after receiving Landlord's statement of Expenses or Common Area Expenses as applicable (or, with respect to the Base Year Expenses or Base Year Common Area Expenses, within 60 days after receiving Landlord's initial statement of Expenses or Common Area Expenses for the applicable Base Year, as the case may be) (each such period is referred to as the "**Review Notice Period**"), Tenant may give Landlord written notice ("**Review Notice**") that Tenant intends to review Landlord's records of the Expenses and/or Common Area Expenses for the calendar year (or Base Year, as applicable) to which the statement applies, and within 60 days after sending the Review Notice to Landlord (such period is referred to as the "**Request for Information Period**"), Tenant shall send Landlord a written request identifying, with a reasonable degree of specificity, the information that Tenant desires to review (the "**Request for Information**"). Within a reasonable time after Landlord's receipt of a timely Request for Information and executed Audit Confidentiality Agreement (referenced below), Landlord, as determined by Landlord, shall forward to Tenant, or make available for inspection on site at such location deemed reasonably appropriate by Landlord, such records (or copies thereof) for the applicable calendar year (or Base Year, as applicable) that are reasonably necessary for Tenant to conduct its review of the information appropriately identified in the Request for Information. Within 60 days after any particular records are made available to Tenant (such period is referred to as the "**Objection Period**"), Tenant shall have the right to give Landlord written notice (an "**Objection Notice**") stating in reasonable detail any objection to Landlord's statement of Expenses and/or Common Area Expenses for that year which relates to the records that have been made available to Tenant. If Tenant provides Landlord with a timely Objection Notice, Landlord and Tenant shall work together in good faith to resolve any issues raised in Tenant's Objection Notice. If Landlord and Tenant determine that Expenses and/or Common Area Expenses for the calendar year are less than reported, Landlord shall provide Tenant with a credit against the next installment of Rent in the amount of the overpayment by Tenant. Likewise, if Landlord and Tenant determine that Expenses and/or Common Area Expenses for the calendar year are greater than reported, Tenant shall pay Landlord the amount of any underpayment within 30 days. If Tenant fails to give Landlord an Objection Notice with respect to any records that have been made available to Tenant prior to expiration of the Objection Period applicable to the records which have been provided to Tenant, Tenant shall be deemed to have approved Landlord's statement of Expenses and/or Common Area Expenses with respect to the matters reflected in such records and shall be barred from raising any claims regarding the Expenses and/or Common Area Expenses relating to such records for that year. If Tenant fails to provide Landlord with a Review Notice prior to expiration of the Review Notice

Period or fails to provide Landlord with a Request for Information prior to expiration of the Request for Information Period described above, Tenant shall be deemed to have approved Landlord's statement of Expenses and/or Common Area Expenses and shall be barred from raising any claims regarding the Expenses and/or Common Area Expenses for that year.

If Tenant retains an agent to review Landlord's records, the agent must be with a CPA firm licensed to do business in the state or commonwealth where the Property is located. Tenant shall be solely responsible for all costs, expenses and fees incurred for the audit, and the fees charged cannot be based in whole or in part on a contingency basis. The records and related information obtained by Tenant shall be treated as confidential, and applicable only to the Office Building, by Tenant and its auditors, consultants and other parties reviewing such records on behalf of Tenant (collectively, "**Tenant's Auditors**"), and, prior to making any records available to Tenant or Tenant's Auditors, Landlord may require Tenant and Tenant's Auditors to each execute a reasonable confidentiality agreement ("**Audit Confidentiality Agreement**") in accordance with the foregoing. In no event shall Tenant be permitted to examine Landlord's records or to dispute any statement of Expenses or Common Area Expenses unless Tenant has paid and continues to pay all Rent when due, subject to any applicable notice, grace and cure periods.

EXHIBIT C

WORK LETTER

This Exhibit is attached to and made a part of the Office Lease Agreement (the “**Lease**”) by and between **MA-RIVERVIEW/245 FIRST STREET, L.L.C., a Delaware limited liability company (“Landlord”)**, and **AKEBIA THERAPEUTICS, INC., a Delaware corporation (“Tenant”)**, for space in the Office Building located at 245 First Street, Cambridge, Massachusetts 02142. Capitalized terms used but not defined herein shall have the meanings given in the Lease.

As used in this Work Letter, the “**Premises**” shall be deemed to mean the Premises, as initially defined in the attached Lease.

1. Landlord shall perform the improvements to the Premises as shown on the Fit Plan Option 1 space plans prepared by DBA-W Architects, dated October 17, 2013 (the “**Space Plans**”) attached hereto as **Exhibit C-1**, the parties hereby intending that the Space Plans are depictions of the parties’ agreement on the basic sketch or sketches of the Premises including the Landlord Work (as defined below) and subject to minor modifications as agreed, and not construction drawings. The improvements to be performed by Landlord as shown on the Space Plans are hereinafter referred to as the “**Landlord Work**”. It is agreed that construction of the Landlord Work will be completed at Landlord’s sole cost and expense (subject to the terms of Paragraph 2 below) using Building standard methods, materials, and finishes. Landlord shall enter into a direct contract for the Landlord Work with a general contractor selected by Landlord. In addition, Landlord shall have the right to select and/or approve of any subcontractors used in connection with the Landlord Work. Landlord’s supervision or performance of any work for or on behalf of Tenant shall not be deemed a representation by Landlord that such Space Plans, related space planning, architectural and engineering drawings (collectively, the “**Plans**”), or the revisions thereto comply with applicable insurance requirements, building codes, ordinances, laws or regulations, or that the improvements constructed in accordance with the Plans and any revisions thereto will be adequate for Tenant’s use, it being agreed that Tenant shall be responsible for all elements of the design of Tenant’s plans (including, without limitation, compliance with law, functionality of design, the structural integrity of the design, the configuration of the premises and the placement of Tenant’s furniture, appliances and equipment). Tenant and Landlord agree that Landlord is not responsible for and is not performing any alterations, repairs or improvements in the Premises with respect to the telephone and data cabling, infrastructure (e.g., coring the floors, or making structural alterations to the Premises), or any HVAC supplemental cooling, if any, nor shall Landlord be responsible for purchasing or installing furniture or equipment in the Premises.
2. If Tenant shall request any revisions to the Space Plans or Plans, such revisions shall be subject to approval by Landlord, which approval shall not be unreasonably withheld. If Landlord approves of such revisions to the Space Plans or Plans then, Landlord shall have such revisions prepared at Tenant’s sole cost and expense and Tenant shall reimburse Landlord for the cost of preparing any such revisions to the Space Plans and/or

Plans, plus any applicable state sales or use tax thereon, upon demand. Promptly upon completion of the revisions, Landlord shall notify Tenant in writing of the increased cost in the Landlord Work, if any, resulting from such revisions to the Space Plans and/or Plans. Tenant, within one Business Day, shall notify Landlord in writing whether it desires to proceed with such revisions. In the absence of such written authorization, Landlord shall have the option to continue work on the Premises disregarding the requested revision. Tenant shall be responsible for any Tenant Delay in completion of the Premises resulting from any revision to the Space Plans and/or Plans. If such revisions result in an increase in the cost of Landlord Work, such increased costs, plus any applicable state sales or use tax thereon, shall be payable by Tenant upon demand. Notwithstanding anything herein to the contrary, all revisions to the Space Plans and/or Plans shall be subject to the approval of Landlord.

3. This Exhibit shall not be deemed applicable to any additional space added to the Premises at any time or from time to time, whether by any options under the Lease or otherwise, or to any portion of the original Premises or any additions to the Premises in the event of a renewal or extension of the original Term of the Lease, whether by any options under the Lease or otherwise, unless expressly so provided in the Lease or any amendment or supplement to the Lease.

EXHIBIT D

COMMENCEMENT LETTER

(EXAMPLE)

Date _____

Tenant _____

Address _____

Re: Commencement Letter with respect to that certain Lease dated as of _____, 2013, by and between **MA-RIVERVIEW/245 FIRST STREET, L.L.C., a Delaware limited liability company**, as Landlord, and **AKEBIA THERAPEUTICS, INC., a Delaware corporation**, as Tenant, for **6,837** rentable square feet on the 11th floor of the Office Building located at 245 First Street, Cambridge, Massachusetts 02142.

Lease Id:

Business Unit Number:

Dear _____ :

In accordance with the terms and conditions of the above referenced Lease, Tenant accepts possession of the Premises and acknowledges:

1. The Commencement Date of the Lease is _____ ;
2. The Termination Date of the Lease is _____ .

Please acknowledge the foregoing and your acceptance of possession by signing all 3 counterparts of this Commencement Letter in the space provided and returning 2 fully executed counterparts to my attention. Tenant's failure to execute and return this letter, or to provide written objection to the statements contained in this letter, within 30 days after the date of this letter shall be deemed an approval by Tenant of the statements contained herein.

Sincerely,

Authorized Signatory

Acknowledged and Accepted:

By: _____

Name: _____

Title: _____

Date: _____

EXHIBIT E

BUILDING RULES AND REGULATIONS

This Exhibit is attached to and made a part of the Office Lease Agreement (the "**Lease**") by and between **MA-RIVERVIEW/245 FIRST STREET, L.L.C., a Delaware limited liability company ("Landlord")** and **AKEBIA THERAPEUTICS, INC., a Delaware corporation (Tenant)**", for space in the Office Building located at 245 First Street, Cambridge, Massachusetts 02142. Capitalized terms used but not defined herein shall have the meanings given in the Lease.

The following rules and regulations shall apply, where applicable, to the Premises, the Buildings, the parking facilities (if any), the Property and the appurtenances. In the event of a conflict between the following rules and regulations and the remainder of the terms of the Lease, the remainder of the terms of the Lease shall control.

1. Sidewalks, doorways, vestibules, halls, stairways and other similar areas shall not be obstructed by Tenant or used by Tenant for any purpose other than ingress and egress to and from the Premises. No rubbish, litter, trash, or material shall be placed, emptied, or thrown in those areas. At no time shall Tenant permit Tenant's employees to loiter in Common Areas or elsewhere about the Buildings or Property.
2. Plumbing fixtures and appliances shall be used only for the purposes for which designed and no sweepings, rubbish, rags or other unsuitable material shall be thrown or placed in the fixtures or appliances.
3. No signs, advertisements or notices shall be painted or affixed to windows, doors or other parts of the Buildings, except those of such color, size, style and in such places as are first approved in writing by Landlord. All tenant identification and suite numbers at the entrance to the Premises shall be installed by Landlord, at Tenant's cost and expense, using the standard graphics for the Buildings. Except in connection with the hanging of lightweight pictures and wall decorations, no nails, hooks or screws shall be inserted into any part of the Premises or Buildings except by the Building maintenance personnel without Landlord's prior approval, which approval shall not be unreasonably withheld.
4. Landlord may provide and maintain in the first floor (main lobby) of the Buildings an alphabetical directory board or other directory device listing tenants and no other directory shall be permitted unless previously consented to by Landlord in writing.
5. Tenant shall not place any lock(s) on any door in the Premises or Buildings without Landlord's prior written consent, which consent shall not be unreasonably withheld, and Landlord shall have the right at all times to retain and use keys or other access codes or devices to all locks within and into the Premises. A reasonable number of keys to the locks on the entry doors in the Premises shall be furnished by Landlord to Tenant at Tenant's cost and Tenant shall not make any duplicate keys. All keys shall be returned to Landlord at the expiration or early termination of the Lease.

6. All contractors, contractor's representatives and installation technicians performing work in the Buildings shall be subject to Landlord's prior approval, which approval shall not be unreasonably withheld, and shall be required to comply with Landlord's standard rules, regulations, policies and procedures, which may be revised from time to time. Landlord has no obligation to allow any particular telecommunication service provider to have access to the Buildings or to the Premises. If Landlord permits access, Landlord may condition the access upon the payment to Landlord by the service provider of fees assessed by Landlord in Landlord's sole discretion.

7. Movement in or out of the Buildings of furniture or office equipment, or dispatch or receipt by Tenant of merchandise or materials requiring the use of elevators, stairways, lobby areas or loading dock areas, shall be performed in a manner and restricted to hours reasonably designated by Landlord. Tenant shall obtain Landlord's prior approval by providing a detailed listing of the activity, including the names of any contractors, vendors or delivery companies, which approval shall not be unreasonably withheld. Tenant shall assume all risk for damage, injury or loss in connection with the activity.

8. Landlord shall have the right to approve the weight, size, or location of heavy equipment or articles in and about the Premises, which approval shall not be unreasonably withheld; provided that approval by Landlord shall not relieve Tenant from liability for any damage in connection with such heavy equipment or articles.

9. Corridor doors, when not in use, shall be kept closed.

10. Tenant shall not: (a) make or permit any improper, objectionable or unpleasant noises or odors in the Buildings, or otherwise interfere in any way with other tenants or persons having business with them; (b) solicit business or distribute or cause to be distributed, in any portion of the Buildings, handbills, promotional materials or other advertising; or (c) conduct or permit other activities in the Buildings or Property that might, in Landlord's sole opinion, constitute a nuisance.

11. No animals, except those assisting handicapped persons, shall be brought into the Buildings or kept in or about the Premises.

12. No inflammable, explosive or dangerous fluids or substances shall be used or kept by Tenant in the Premises, Buildings or about the Property, except for those substances as are typically found in similar premises used for general office purposes and are being used by Tenant in a safe manner and in accordance with all applicable Laws. Tenant shall not, without Landlord's prior written consent, use, store, install, spill, remove, release or dispose of, within or about the Premises or any other portion of the Property, any asbestos-containing materials or any solid, liquid or gaseous material now or subsequently considered toxic or hazardous under the provisions of 42 U.S.C. Section 9601 et seq., M.G.L. c. 21C, M.G.L. c. 21E or any other applicable environmental Law which may now or later be in effect. Tenant shall comply with all Laws pertaining to and governing the use of these materials by Tenant and shall remain solely liable for the costs of abatement and removal.

13. Tenant shall not use or occupy the Premises in any manner or for any purpose which might injure the reputation or impair the present or future value of the Premises or the Buildings. Tenant shall not use, or permit any part of the Premises to be used for lodging, sleeping or for any illegal purpose.
14. Tenant shall not take any action which would violate Landlord's labor contracts or which would cause a work stoppage, picketing, labor disruption or dispute or interfere with Landlord's or any other tenant's or occupant's business or with the rights and privileges of any person lawfully in the Buildings (**"Labor Disruption"**). Tenant shall take the actions necessary to resolve the Labor Disruption, and shall have pickets removed and, at the request of Landlord, immediately terminate any work in the Premises that gave rise to the Labor Disruption, until Landlord gives its written consent for the work to resume. Tenant shall have no claim for damages against Landlord or any of the Landlord Related Parties nor shall the Commencement Date be extended as a result of the above actions.
15. Tenant shall not install, operate or maintain in the Premises or in any other area of the Buildings, electrical equipment that would overload the electrical system beyond its capacity for proper, efficient and safe operation as determined solely by Landlord. Tenant shall not furnish cooling or heating to the Premises, including, without limitation, the use of electric or gas heating devices, without Landlord's prior written consent. Tenant shall not use more than its proportionate share of telephone lines and other telecommunication facilities available to service the Buildings.
16. Tenant shall not operate or permit to be operated a coin or token operated vending machine or similar device (including, without limitation, telephones, lockers, toilets, scales, amusement devices and machines for sale of beverages, foods, candy, cigarettes and other goods), except for machines for the exclusive use of Tenant's employees and invitees.
17. Bicycles and other vehicles are not permitted inside the Buildings or on the walkways outside the Buildings, except in areas designated by Landlord.
18. Landlord may from time to time adopt systems and procedures for the security and safety of the Buildings and Property, their occupants, entry, use and contents. Tenant, its agents, employees, contractors, guests and invitees shall comply with Landlord's systems and procedures.
19. Landlord shall have the right to prohibit the use of the name of the Buildings or any other publicity by Tenant that in Landlord's sole opinion may impair the reputation of the Buildings or their desirability. Upon written notice from Landlord, Tenant shall refrain from and discontinue such publicity immediately.
20. Neither Tenant nor its agents, employees, contractors, guests or invitees shall smoke or permit smoking in the Common Areas, unless a portion of the Common Areas have been declared a designated smoking area by Landlord, nor shall the above parties allow smoke from the Premises to emanate into the Common Areas or any other part of the Buildings. Landlord shall have the right to designate the Buildings (including the Premises) as a non-smoking building.

21. Landlord shall have the right to designate and approve standard window coverings for the Premises and to establish rules to assure that the Buildings present a uniform exterior appearance. Tenant shall ensure, to the extent reasonably practicable, that window coverings are closed on windows in the Premises while they are exposed to the direct rays of the sun.

22. Deliveries to and from the Premises shall be made only at the times in the areas and through the entrances and exits reasonably designated by Landlord. Tenant shall not make deliveries to or from the Premises in a manner that might interfere with the use by any other tenant of its premises or of the Common Areas, any pedestrian use, or any use which is inconsistent with good business practice.

23. The work of cleaning personnel shall not be hindered by Tenant after 5:30 p.m., and cleaning work may be done at any time when the offices are vacant. Windows, doors and fixtures may be cleaned at any time. Tenant shall provide adequate waste and rubbish receptacles to prevent unreasonable hardship to the cleaning service.

EXHIBIT F

ADDITIONAL PROVISIONS

This Exhibit is attached to and made a part of the Office Lease Agreement (the "Lease") by and between **MA-RIVERVIEW/245 FIRST STREET, L.L.C., a Delaware limited liability company ("Landlord")**, and **AKEBIA THERAPEUTICS, INC., a Delaware corporation ("Tenant")**, for space in the Office Building located at 245 First Street, Cambridge, Massachusetts 02142. Capitalized terms used but not defined herein shall have the meanings given in the Lease.

1. Parking.

- 1.01 During the Term, Tenant agrees to lease from Landlord and Landlord agrees to lease to Tenant a total of 8 unreserved parking spaces (collectively, the "Spaces"), for the use of Tenant and its employees, in the parking facility owned by Landlord that serves the Buildings (the "Parking Facility"), and if the Parking Facility includes a garage, then such Spaces may be in, or on the roof of, such garage. No deductions or allowances shall be made for days when Tenant or any of its employees does not utilize the Parking Facility or for Tenant utilizing less than all of the Spaces. Tenant shall not have the right to lease or otherwise use more than the number of unreserved Spaces set forth above.
- 1.02 During the initial Term, Tenant shall pay Landlord, as Additional Rent in accordance with Section 4 of the Lease, the sum of \$260.00 per month, plus applicable tax thereon, if any, for each unreserved Space leased by Tenant hereunder, as such rate may be adjusted from time-to-time to reflect the then current rate for parking in the Parking Facility.
- 1.03 Except for particular spaces and areas designated by Landlord for reserved parking, all parking in the Parking Facility shall be on an unreserved, first-come, first-served basis.
- 1.04 Landlord shall not be responsible for money, jewelry, automobiles or other personal property lost in or stolen from the Parking Facility regardless of whether such loss or theft occurs when the Parking Facility is locked or otherwise secured. Except as caused by the negligence or willful misconduct of Landlord and without limiting the terms of the preceding sentence, Landlord shall not be liable for any loss, injury or damage to persons using the Parking Facility or automobiles or other property therein, it being agreed that, to the fullest extent permitted by law, the use of the Spaces shall be at the sole risk of Tenant and its employees.
- 1.05 Landlord shall have the right from time to time to designate the location of the Spaces and to promulgate reasonable rules and regulations regarding the Parking Facility, the Spaces and the use thereof, including, but not limited to, rules and regulations controlling the flow of traffic to and from various parking areas, the angle and direction of parking and the like. Tenant shall comply with and cause its employees to comply with all such rules and regulations as well as all reasonable additions and amendments thereto.

- 1.06 Tenant shall not store or permit its employees to store any automobiles in the Parking Facility without the prior written consent of Landlord. Except for emergency repairs, Tenant and its employees shall not perform any work on any automobiles while located in the Parking Facility or on the Property. If it is necessary for Tenant or its employees to leave an automobile in the Parking Facility overnight, Tenant shall provide Landlord with prior notice thereof designating the license plate number and model of such automobile.
- 1.07 Landlord shall have the right to temporarily close the Parking Facility or certain areas therein in order to perform necessary repairs, maintenance and improvements to the Parking Facility.
- 1.08 Tenant shall not assign or sublease any of the Spaces without the consent of Landlord. Landlord shall have the right to terminate this Parking Agreement with respect to any Spaces that Tenant desires to sublet or assign.
- 1.09 Landlord may elect to provide parking cards or keys to control access to the Parking Facility. In such event, Landlord shall provide Tenant with one card or key for each Space that Tenant is leasing hereunder, provided that Landlord shall have the right to require Tenant or its employees to place a deposit on such access cards or keys and to pay a fee for any lost or damaged cards or keys.
- 1.10 Landlord hereby reserves the right to enter into a management agreement or lease with an entity for the Parking Facility (“**Parking Facility Operator**”). In such event, Tenant, upon request of Landlord, shall enter into a parking agreement with the Parking Facility Operator and pay the Parking Facility Operator the monthly charge established hereunder, and Landlord shall have no liability for claims arising through acts or omissions of the Parking Facility Operator unless caused by Landlord’s negligence or willful misconduct. It is understood and agreed that the identity of the Parking Facility Operator may change from time to time during the Term. In connection therewith, any parking lease or agreement entered into between Tenant and a Parking Facility Operator shall be freely assignable by such Parking Facility Operator or any successors thereto.
- 1.11 Tenant, at its sole cost and expense, shall comply with the applicable terms and conditions of the City of Cambridge Traffic Ordinance, including, without limitation, the PTDM Ordinance. Tenant shall reasonably cooperate with Landlord as reasonably required for Landlord to fulfill its obligations under the PTDM Ordinance.

2. **Initial Suite Signage and Building Directory.** Notwithstanding anything to the contrary contained in Section 3 and Section 4 of **Exhibit E** (Building Rules and Regulations) of the Lease, Landlord, at Landlord’s cost and expense, shall install, for the Tenant as initially named herein, using the standard graphics for the Building, initial Building

standard tenant identification and suite numbers at the entrance to the initial Premises and on the Building directory in the main Building lobby. Thereafter, any additional tenant identification shall be (i) subject to Landlord's prior review and approval thereof, and (ii) installed by Landlord, at Tenant's cost and expense, using the standard graphics for the Building.

3. Letter of Credit.

3.01 **General Provisions.** Within 45 days after the date of this Lease, Tenant shall deliver to Landlord, as collateral for the full performance by Tenant of all of its obligations under this Lease and for all losses and damages Landlord may suffer as a result of Tenant's Failure to Comply (as defined in Section 3.02 below) with one or more provisions of this Lease, a standby, unconditional, irrevocable, transferable letter of credit (the "**Letter of Credit**") in the form of **Exhibit H** hereto and containing the terms required herein or such other form as mutually agreed upon by the parties, in the face amount of \$125,345.00 (the "**Letter of Credit Amount**"), naming Landlord as beneficiary, issued (or confirmed) by Wells Fargo, or a financial institution reasonably acceptable to Landlord permitting multiple and partial draws thereon as provided herein. Tenant shall cause the Letter of Credit to be continuously maintained in effect (whether through replacement, renewal or extension) in the Letter of Credit Amount through the date (the "**Final LC Expiration Date**") that is 60 days after the scheduled expiration date of the Term or any renewal Term. If the Letter of Credit held by Landlord expires earlier than the Final LC Expiration Date (whether by reason of a stated expiration date or a notice of termination or non-renewal given by the issuing bank), Tenant shall deliver a new Letter of Credit or certificate of renewal or extension (a "**Renewal or Replacement LC**") to Landlord not later than 60 days prior to the expiration date of the Letter of Credit then held by Landlord. Any Renewal or Replacement LC shall comply with all of the provisions of this Section 3, shall be irrevocable, transferable and shall remain in effect (or be automatically renewable) through the Final LC Expiration Date upon the same terms as the expiring Letter of Credit or such other terms as may be acceptable to Landlord in its sole discretion. Notwithstanding the foregoing, Landlord will have the right to require Tenant to deliver a Renewal or Replacement LC issued in accordance with the above requirements from a different issuer if any of the following events ("**Issuer Events**") occur: (i) the Tier-1 Capital Ratio (as defined below) of such issuer subsequently falls below five percent (5%), or (ii) if the issuer becomes subject to an FDIC or similar state or federal receivership or conservatorship and the Letter of Credit is repudiated or terminated in connection therewith, or (iii) the issuer is placed on an FDIC "watch list", or (iv) if Landlord analyzes such issuer's capitalization, asset quality, earnings, and/or liquidity and in Landlord's reasonable discretion, disapproves of such issuer's financial wherewithal and ability to remain as the issuer of the Letter of Credit. Tenant shall deliver to Landlord a Renewal or Replacement LC within 10 Business Days following Tenant's receipt (or deemed receipt) from Landlord of notice of the occurrence of such Issuer Event. As used herein, the term "**Tier-1 Capital Ratio**" shall mean, as to an issuer, the ratio of (x) the sum of common

stockholders' equity, non-cumulative perpetual preferred stock, and minority interests in consolidated subsidiaries, less intangible assets, to (y) total assets, all as calculated pursuant to and in accordance with 12 CFR Part 325, such ratio being a measure of such bank's or institution's availability of core capital to satisfy unexpected losses. Upon receipt of a Renewal or Replacement LC, Landlord shall return the original Letter of Credit then held by Landlord to Tenant within 10 Business Days thereafter. In the alternative, Tenant shall have the right to present the Renewal or Replacement LC in person to Landlord at the location where Landlord keeps and maintains its letters of credit in order to effect a simultaneous exchange of the Renewal or Replacement LC with the original Letter of Credit then held by Landlord.

- 3.02 Drawings under Letter of Credit. If any non-curable default on the part of Tenant shall occur under the Lease or if any curable default on the part of Tenant occurs under the Lease and Tenant shall have failed to cure such default within any applicable notice and cure periods provided under the Lease, or as otherwise specifically agreed by Landlord and Tenant pursuant to this Lease or any amendment hereof (any of the foregoing being, for purposes of this Exhibit F, Section 3 only, a "**Failure to Comply**"), Landlord may, without prejudice to any other remedy provided in this Lease or by Law, draw on the Letter of Credit and use all or part of the proceeds to (a) satisfy any amounts due to Landlord from Tenant, and (b) satisfy any other damage, injury, expense or liability caused by Tenant's Failure to Comply. In addition, if Tenant fails to furnish a Renewal or Replacement LC complying with all of the provisions of this Section 3 at least 60 days prior to the stated expiration date of the Letter of Credit then held by Landlord, Landlord may draw upon such Letter of Credit and hold the proceeds thereof (and such proceeds need not be segregated) in accordance with the terms of this Section 3 (the "**LC Proceeds Account**").
- 3.03 Use of Proceeds by Landlord. The proceeds of the Letter of Credit drawn upon because of Tenant's Failure to Comply and in accordance with Section 3.02 above shall constitute Landlord's sole and separate property (and not Tenant's property or the property of Tenant's bankruptcy estate) and Landlord may immediately upon any draw (and without notice to Tenant) apply or offset the proceeds of the Letter of Credit: (a) against any Rent payable by Tenant under this Lease that was not paid when due because of Tenant's Failure to Comply; (b) against all losses and damages that Landlord has suffered or that Landlord reasonably estimates that it may suffer as a result of Tenant's Failure to Comply with one or more provisions of this Lease; (c) against any costs incurred by Landlord in connection with the Lease (including attorneys' fees), provided reimbursement of such costs is provided for under this Lease; and (d) against any other amount that Landlord may spend or become obligated to spend by reason of Tenant's Default, provided the reimbursement of such amount is provided for under this Lease. Provided Tenant has performed all of its obligations under this Lease, Landlord agrees to pay to Tenant within 45 days after the Final LC Expiration Date the amount of any proceeds of the Letter of Credit received by Landlord and not applied as allowed above; provided, that if prior to the Final LC Expiration Date a voluntary

petition is filed by Tenant or any Guarantor, or an involuntary petition is filed against Tenant or any Guarantor by any of Tenant's or Guarantor's creditors, under the Federal Bankruptcy Code, then Landlord shall not be obligated to make such payment in the amount of the unused Letter of Credit proceeds until either all preference issues relating to payments under this Lease have been resolved in such bankruptcy or reorganization case or such bankruptcy or reorganization case has been dismissed, in each case pursuant to a final court order not subject to appeal or any stay pending appeal.

- 3.04 Additional Covenants of Tenant. If, as result of any application or use by Landlord of all or any part of the Letter of Credit as herein provided, the amount of the Letter of Credit shall be less than the Letter of Credit Amount, Tenant shall, within 5 Business Days thereafter, provide Landlord with additional letter(s) of credit in an amount equal to the deficiency (or a replacement letter of credit in the total Letter of Credit Amount), and any such additional (or replacement) letter of credit shall comply with all of the provisions of this Section 3, and if Tenant fails to comply with the foregoing, notwithstanding anything to the contrary contained in this Lease, the same shall constitute a Default by Tenant. Tenant further covenants and warrants that it will neither assign nor encumber the Letter of Credit or any part thereof and that neither Landlord nor its successors or assigns will be bound by any such assignment, encumbrance, attempted assignment or attempted encumbrance.
- 3.05 Nature of Letter of Credit. Landlord and Tenant (a) acknowledge and agree that in no event or circumstance shall the Letter of Credit or any renewal thereof or substitute therefor or any proceeds thereof (including the LC Proceeds Account) be deemed to be or treated as a "security deposit" under any Law applicable to security deposits in the commercial context ("**Security Deposit Laws**"), (b) acknowledge and agree that the Letter of Credit (including any renewal thereof or substitute therefor or any proceeds thereof) is not intended to serve as a security deposit, and the Security Deposit Laws shall have no applicability or relevancy thereto, and (c) waive any and all rights, duties and obligations either party may now or, in the future, will have relating to or arising from the Security Deposit Laws.

EXHIBIT G

NOTICE OF LEASE

Notice is hereby given pursuant to Massachusetts General Laws, Chapter 183, Section 4 of the following lease:

- 1. Landlord: MA-RIVERVIEW/245 FIRST STREET, L.L.C., a Delaware limited liability company
- 2. Tenant: AKEBIA THERAPEUTICS, INC., a Delaware corporation
- 3. Date of Lease: , 2013.
- 4. Premises: 6,837 rentable square feet of space as more particularly described in the Lease on the 11th floor of the office building known as and numbered 245 First Street, Cambridge, Massachusetts 02142. The legal description for the land on which the Building is situated is set forth on **Exhibit A** attached hereto.
- 5. Lease Term: 36 month term.

The foregoing is a summary of certain terms of the Lease for purposes of giving notice thereof, and shall not be deemed to modify or amend the terms of the Lease.

This Notice is executed under seal this day of , , .

LANDLORD: **MA-RIVERVIEW/245 FIRST STREET,
L.L.C., a Delaware limited liability company**

By: _____
Name: _____
Title: _____

TENANT: *[insert name of tenant]*

By: _____
Name: _____
Title: _____

THE COMMONWEALTH OF MASSACHUSETTS

, ss.

On this day of 20 , before me, the undersigned notary public, personally appeared , proved to me through satisfactory evidence of identification, which was photographic identification with signature issued by a federal or state governmental agency, oath or affirmation of a credible witness, personal knowledge of the undersigned, to be the person whose name is signed on the preceding or attached document(s), and acknowledged to me that (he)(she) signed it voluntarily for its stated purpose. (as partner for partnership) (as of corporation), (as of limited liability company), (as attorney in fact for).

Notary Public:

My Commission Expires:

THE COMMONWEALTH OF MASSACHUSETTS

, ss.

On this day of 20 , before me, the undersigned notary public, personally appeared , proved to me through satisfactory evidence of identification, which was photographic identification with signature issued by a federal or state governmental agency, oath or affirmation of a credible witness, personal knowledge of the undersigned, to be the person whose name is signed on the preceding or attached document(s), and acknowledged to me that (he)(she) signed it voluntarily for its stated purpose. (as partner for partnership) (as of corporation), (as of limited liability company), (as attorney in fact for).

Notary Public:

My Commission Expires:

EXHIBIT A

LEGAL DESCRIPTION

G-3

EXHIBIT H

FORM OF LETTER OF CREDIT

This Exhibit is attached to and made a part of the Office Lease Agreement (the "Lease") by and between **MA-RIVERVIEW/245 FIRST STREET, L.L.C., a Delaware limited liability company ("Landlord")** and **AKEBIA THERAPEUTICS, INC., a Delaware corporation ("Tenant")**, for space in the Office Building located at 245 First Street, Cambridge, Massachusetts 02142. Capitalized terms used but not defined herein shall have the meanings given in the Lease.

[Name of Financial Institution]

Irrevocable Standby Letter of Credit
No. _____
Issuance Date: _____
Expiration Date: _____
Applicant: _____

Beneficiary

MA-Riverview/245 First Street, L.L.C.
c/o Equity Office
Summer Street, 17th Floor
Boston, Massachusetts 02110
Attention: Property Manager

Ladies/Gentlemen:

We hereby establish our Irrevocable Standby Letter of Credit in your favor for the account of the above referenced Applicant in the amount of One Hundred Twenty Five Thousand Three Hundred Forty Five and 00/100 U.S. Dollars (\$125,345.00) available for payment at sight by your draft drawn on us when accompanied by the following documents:

1. An original copy of this Irrevocable Standby Letter of Credit.
2. Beneficiary's dated statement purportedly signed by an authorized signatory or agent reading: "This draw in the amount of U.S. Dollars (\$) under your Irrevocable Standby Letter of Credit No. represents funds due and owing to us pursuant to the terms of that certain lease by and between MA-Riverview/245 First Street, L.L.C., a Delaware limited liability company, as landlord, and Akebia Therapeutics, Inc., a Delaware corporation, as tenant, dated November, 2013, and/or any amendment to the lease or any other agreement between such parties related to the lease."

It is a condition of this Irrevocable Standby Letter of Credit that it will be considered automatically renewed for a one year period upon the expiration date set forth above and upon each anniversary of such date, unless at least 60 days prior to such expiration date or applicable anniversary thereof, we notify you in writing, by certified mail return receipt requested or by recognized overnight courier service, that we elect not to so renew this Irrevocable Standby Letter of Credit. A copy of any such notice shall also be sent, in the same manner, to: Equity Office, 2 North Riverside Plaza, Suite 2100, Chicago, Illinois 60606, Attention: Treasury Department. In addition to the foregoing, we understand and agree that you shall be entitled to draw upon this Irrevocable Standby Letter of Credit in accordance with 1 and 2 above in the event that we elect not to renew this Irrevocable Standby Letter of Credit and, in addition, you provide us with a dated statement purportedly signed by an authorized signatory or agent of Beneficiary stating that the Applicant has failed to provide you with an acceptable substitute irrevocable standby letter of credit in accordance with the terms of the above referenced lease. We further acknowledge and agree that: (a) upon receipt of the documentation required herein, we will honor your draws against this Irrevocable Standby Letter of Credit without inquiry into the accuracy of Beneficiary's signed statement and regardless of whether Applicant disputes the content of such statement; (b) this Irrevocable Standby Letter of Credit shall permit partial draws and, in the event you elect to draw upon less than the full stated amount hereof, the stated amount of this Irrevocable Standby Letter of Credit shall be automatically reduced by the amount of such partial draw; and (c) you shall be entitled to transfer your interest in this Irrevocable Standby Letter of Credit from time to time and more than one time without our approval and without charge. In the event of a transfer, we reserve the right to require reasonable evidence of such transfer as a condition to any draw hereunder.

This Irrevocable Standby Letter of Credit is subject to the International Standby Practices (ISP98) ICC Publication No. 590.

We hereby engage with you to honor drafts and documents drawn under and in compliance with the terms of this Irrevocable Standby Letter of Credit.

All communications to us with respect to this Irrevocable Standby Letter of Credit must be addressed to our office located at _____ to the attention of _____.

Very truly yours,

[name]

[title]

AKEBIA THERAPEUTICS, INC.

AMENDED AND RESTATED 2008 EQUITY INCENTIVE PLAN

ARTICLE I

ESTABLISHMENT AND TERM

Section 1.01. Establishment; Definitions. This Plan was originally adopted by the Board effective April 4, 2008, and by the stockholders of the Corporation effective April 4, 2008. This Plan was adopted in this amended and restated form by the Board effective July 28, 2009, and by the stockholders of the Corporation effective July 28, 2009. All capitalized terms used herein are defined herein or in Appendix A attached hereto.

Section 1.02. Term. The Board may suspend or terminate the Plan at any time. Unless sooner terminated, the Plan shall terminate on April 3, 2018, which shall be within ten (10) years from the date the Plan is adopted by the Board or approved by the stockholders of the Corporation, whichever is earlier. No Equity Awards may be granted under the Plan while the Plan is suspended or after it is terminated. Suspension or termination of the Plan shall not impair rights and obligations under any Equity Award granted while the Plan is in effect, except with the consent of the person to whom the Equity Award was granted.

ARTICLE II

STRUCTURE AND PURPOSE

Section 2.01. Section 2.01 Structure of Plan. The Equity Awards issued under the Plan shall be either, in the discretion of the Board, (a) Options granted pursuant to Article VI hereof, including Incentive Stock Options and Non-statutory Stock Options, or (b) Stock bonuses or restricted Stock awards granted pursuant to Article VII hereof. All Options shall be designated as Incentive Stock Options or Non-statutory Stock Options at the time of grant.

Section 2.02. Purpose. The purpose of the Plan is to promote the interests of the Corporation by aligning the interests of selected eligible persons under the Plan with the interests of the stockholders of the Corporation and by providing to such persons an opportunity to obtain the benefits from ownership of the Corporation's Stock through the granting to such persons of Equity Awards. The Corporation, through the use of the Plan, seeks to attract and retain the services of Employees, Directors and Consultants, and to provide additional incentives for such persons apart from the provisions of their employment agreements or other arrangements with the Corporation or its Affiliates.

ARTICLE III

ADMINISTRATION

Section 3.01. Board; Delegation to Committee. The Board shall administer the Plan unless and until the Board delegates administration to a Committee. The Board may delegate administration of the Plan to a Committee composed of two or more members of the Board,

composed solely of Outside Directors or composed, if applicable law permits, of one or more officers of the Corporation. If administration is delegated to a Committee, the Committee shall have, in administering the Plan, all of the powers that were possessed by the Board prior to such delegation, subject, however, to such resolutions, not inconsistent with the provisions of the Plan, as may be adopted from time to time by the Board. If administration is delegated to a Committee, all references in this Plan to the Board shall thereafter be to the Committee. The Board may abolish the Committee at any time and revert in the Board the administration of the Plan.

Section 3.02. Administration. The Board shall have the power, consistent with the express provisions of the Plan:

To determine from time to time which of the eligible persons under the Plan shall be granted Equity Awards;

To determine whether an Equity Award shall be an Incentive Stock Option, a Non-statutory Stock Option, a Stock bonus, a restricted Stock award or a combination of the foregoing;

To approve forms of Equity Award Agreements for use under the Plan;

To determine the number of shares of Stock to be covered by each Equity Award granted hereunder;

To determine how and when each Equity Award shall be granted, the provisions of each Equity Award granted (including, but not limited to, provisions setting forth or relating to exercise price, vesting schedule, vesting acceleration, forfeiture and rights of repurchase), and to provide for any and all other terms and conditions in an Equity Award which are not expressly prohibited by the Plan;

To construe and interpret the Plan and Equity Awards granted under it, and to establish, amend and revoke rules and regulations for the administration of such Plan and Equity Awards;

To correct any defect, omission or inconsistency in the Plan or in any Equity Award Agreement, in a manner and to the extent it shall deem necessary or expedient to make the Plan fully effective;

To amend the Plan or an Equity Award as provided in Article XI; and

Generally, to exercise such powers and to perform such acts as the Board deems necessary or expedient to promote the best interests of the Corporation that are not in conflict with the provisions of the Plan.

Any determination by the Board with respect to the matters referred to above shall be final and conclusive.

ARTICLE IV

ELIGIBILITY

Section 4.01. Persons Eligible for Equity Awards. Incentive Stock Options may be granted only to Employees who meet the definition of “employee” under Section 3401(c) of the Code on the date of grant. Equity Awards other than Incentive Stock Options may be granted only to Employees, Directors or Consultants. The extent to which any such person shall be entitled to be granted Equity Awards pursuant to the Plan shall be determined in the sole and absolute discretion of the Board. Eligibility to participate does not confer upon any Employee any right to be granted Equity Awards and the acceptance of any Equity Award by an Employee is voluntary.

Section 4.02. Other Limitations. If any payment or right accruing to an individual under this Plan (without the application of this Section 4.02), either alone or together with other payments or rights accruing to such individual from the Corporation or an Affiliate of the Corporation (“TOTAL PAYMENTS”), would constitute a “parachute payment” (as defined in Section 280G of the Code), such payment or right shall be reduced to the largest amount or greatest right that will result in no portion of the amount payable or right accruing under this Plan being subject to an excise tax under Section 4999 of the Code or being disallowed as a deduction under Section 280G of the Code, provided that the foregoing shall not apply to the extent provided otherwise in an Equity Award Agreement or in the event the affected individual is party to an agreement with the Corporation or an Affiliate of the Corporation that explicitly provides for an alternate treatment of payments or rights that would constitute “parachute payments.” The determination of whether any reduction in the rights or payments under this Plan is to apply shall be made by the Board in good faith after consultation with the affected individual, and such determination shall be conclusive and binding on such affected individual. The affected individual shall cooperate in good faith with the Board in making such determination and providing the necessary information for this purpose. The foregoing provisions of this Section 4.02 shall apply with respect to any person only if, after reduction for any applicable Federal excise tax imposed by Section 4999 of the Code and Federal income tax imposed by the Code, the Total Payments accruing to such person would be less than the amount of the Total Payments as reduced, if applicable, under the foregoing provisions of this Section 4.02 and after reduction for any applicable Federal income tax imposed by the Code.

ARTICLE V

SHARES SUBJECT TO THE PLAN

Section 5.01. Subject to the provisions of Article VIII relating to adjustments upon changes in Stock, no more than 12,667,667 shares of Stock may be issued pursuant to Equity Awards. The number of shares of Stock reserved for issuance under this Plan may be increased from time to time as permitted by law. If any Equity Award shall for any reason expire, be cancelled, be forfeited or otherwise terminate, in whole or in part, without having been exercised in full, or if shares of Stock are not delivered because an Equity Award is settled in cash or because such shares of Stock are used to satisfy an applicable tax withholding obligation, in whole or in part, or if shares of Stock which originally underlay an Equity Award are

repurchased or otherwise reacquired by the Corporation, the Stock not acquired or delivered or reacquired (as the case may be) under such Equity Award by the holder thereof shall revert to and again become available for issuance under the Plan. The Stock subject to the Plan may be unissued shares or reacquired shares, bought on the market or otherwise.

ARTICLE VI

TERMS OF OPTIONS

Section 6.01. Form of Option. Subject to the provisions of the Plan, each Option shall be in such form and shall contain such terms and conditions as the Board shall determine. The provisions of separate Options need not be identical.

Section 6.02. Term. No Option shall be exercisable after the expiration of ten (10) years from the date it was granted.

Section 6.03. Exercise Price. The exercise price of each Incentive Stock Option shall be not less than one hundred percent (100%) of the Fair Market Value of the Stock subject to the Option on the date the Option is granted. The exercise price of each Non-statutory Stock Option shall be the exercise price determined by the Board. Notwithstanding the foregoing, an Option (whether an Incentive Stock Option or a Non-statutory Stock Option) may be granted with an exercise price lower than that otherwise provided in this Section 6.03 if such Option is granted pursuant to an assumption or substitution for another option in a manner satisfying the provisions of Section 424(a) and Section 409A of the Code.

Section 6.04. Exercise of Options. Subject to the provisions of Section 6.07, an Optionee may at any time prior to the expiration or termination of an Option elect to purchase all or a portion of the Stock subject to such Option which such holder is then entitled to purchase by delivering to the Corporation a completed Stock Purchase Agreement specifying the number of shares of Stock the Participant desires to purchase. An Option may be exercised for whole shares of Stock only. The Stock Purchase Agreement shall be accompanied by payment of the applicable exercise price for Stock being acquired. Subject to the provisions of the Equity Award Agreement, the Corporation shall cause to be delivered to the holder a certificate for the shares of Stock so purchased. If the number of shares so purchased is less than the number of shares of Stock subject to the Option, the Corporation shall deliver to the holder a memorandum of the number of shares in respect of which the Option has been exercised and the number of shares which remain subject to the Option.

Section 6.05. Payment. Except as otherwise provided in the applicable Equity Award Agreement or by the Board, the purchase price of Stock acquired pursuant to an Option shall be paid in cash (by check) at the time the Option is exercised.

Section 6.06. Transferability. An Incentive Stock Option and, unless otherwise provided in an Equity Award Agreement, a Non-statutory Stock Option shall not be transferable except by will or by the laws of descent and distribution, and shall be exercisable during the lifetime of the person to whom the Option is granted only by such person.

Section 6.07. Vesting. Subject to the provisions of the Plan, the Board, in its discretion, shall determine at the time of grant the time when an Option vests, becomes exercisable and shall expire, and such determinations shall be set forth in the applicable Equity Award Agreement. An Option may be subject to such other terms and conditions on the time or times when it may be exercised (which may be based on performance or other criteria) as the Board may deem appropriate, and the Board may provide for early exercise of unvested Options (with the Stock received therefor being itself subject to vesting) if expressly set forth in an Equity Award Agreement. Unless otherwise approved by the Board and set forth in writing by an authorized officer of the Corporation, an Option shall cease vesting upon the Optionee's Termination, regardless of whether or not the Optionee was given requisite notice of Termination of such Optionee's employment by the Corporation or by any Affiliate of the Corporation.

Section 6.08. Termination of Employment or Relationship as a Director or Consultant. An Option will expire immediately upon the Optionee's Termination for Cause. Unless otherwise provided in the Equity Award Agreement relating to an Option, in the event of an Optionee's Termination (for reasons other than Cause, the Optionee's death or the Optionee's Disability), the Optionee may exercise the Option to the extent of the shares in respect of which such Option is exercisable on the date notice of Termination is given to the Optionee by the Corporation or any Affiliate of the Corporation at any time beginning on such date and ending on the earlier of (a) the date one (1) month after such notice of Termination is delivered to the Optionee, or (b) the expiration of the term of the Option as set forth in the Equity Award Agreement. The time period for the exercise of such Options applies regardless of the sufficiency or the length of notice of Termination given by the Corporation or any Affiliate of the Corporation to the Optionee.

Section 6.09. Disability of Optionee. Unless otherwise provided in the Equity Award Agreement relating to an Option, in the event of a Termination as a result of the Optionee's Disability, the Optionee may exercise the Option to the extent of the Shares in respect of which such Option is exercisable on the date notice of Termination is given to the Optionee by the Corporation or any Affiliate of the Corporation at any time beginning on such date and ending on the earlier of (a) the date twelve (12) months after such notice of Termination is delivered to the Optionee, or (b) the expiration of the term of the Option as set forth in the Equity Award Agreement.

Section 6.10. Death of Optionee. Unless otherwise provided in the Equity Award Agreement relating to an Option, in the event of a Termination as a result of the Optionee's death, the Optionee's estate or a person who acquired the right to exercise the Option by bequest or inheritance may exercise the Option to the extent of the Shares in respect of which such Option is exercisable on the date of death at any time beginning on such date and ending on the earlier of (a) the date twelve (12) months after the date of death, or (b) the expiration of the term of the Option as set forth in the Equity Award Agreement.

Section 6.11. Incentive Stock Option Limitations. The following limitations shall apply to a grant of an Incentive Stock Option:

If, at the time of the grant of an Incentive Stock Option, the Optionee owns (or is deemed to own pursuant to Section 424(d) of the Code) equity securities possessing more than ten percent (10%) of the total combined voting power of all classes of equity securities of the Corporation or of any of its Affiliates, the exercise price of such Incentive Stock Option shall be at least one hundred and ten percent (110%) of the Fair Market Value of such Stock on the date of grant and the Incentive Stock Option shall terminate on the date that is within five (5) years after the date of grant.

If the aggregate Fair Market Value (determined as of the time the Incentive Stock Option with respect to such Stock is granted) of Stock with respect to which Incentive Stock Options are exercisable for the first time by the Optionee during any calendar year (under all plans of the Corporation and its Affiliates) exceeds one hundred thousand dollars (\$100,000), the Options or portions thereof that exceed such limit shall be treated as Non-statutory Stock Options.

Section 6.12. Cancellation and Regrant. The Board shall have the authority to effect, at any time and from time to time, (a) the repricing of any outstanding Options under the Plan, or (b) with the consent of the affected holders of Options, the cancellation of any outstanding Options under the Plan and the grant in substitution therefor of new Options under the Plan covering the same or different numbers of shares of Stock and having an exercise price per share as determined by the Board.

Section 6.13. Qualification of Incentive Stock Options. Anything in the Plan to the contrary notwithstanding, no term of the Plan relating to Incentive Stock Options shall be interpreted, amended or altered, nor shall any discretion or authority granted under the Plan be exercised, so as to disqualify the Plan under Section 422 of the Code or, without the consent of the Optionee affected, to disqualify any Incentive Stock Option under Section 422 of the Code.

ARTICLE VII

TERMS OF STOCK BONUSES AND RESTRICTED STOCK AWARDS

Section 7.01. Form of Stock Bonus or Restricted Stock Award. Subject to the provisions of the Plan, each Stock bonus or restricted Stock award shall be in such form and shall contain such terms and conditions as the Board shall determine. The provisions of separate Stock bonuses or restricted Stock awards need not be identical.

Section 7.02. Purchase Price. The purchase price, if any, for any Stock granted as a Stock bonus or restricted Stock award shall be such amount as the Board shall determine and designate in the Equity Award Agreement. Notwithstanding the foregoing, the Board may determine that eligible participants in the Plan may be awarded Stock in consideration for past services rendered to the Corporation or an Affiliate thereof or for the benefit of the Corporation or an Affiliate thereof. Upon the award of any Stock bonus or restricted Stock award and the payment of any purchase price, if applicable, the holder of such Stock bonus or restricted Stock award shall deliver to the Corporation a completed Stock Purchase Agreement.

Section 7.03. Transferability. Unless otherwise provided in the Equity Award Agreement and subject to the provisions of any applicable buy-sell or similar agreements, Stock awarded or purchased pursuant to this Article VII shall not be transferable except by will or by the laws of descent and distribution, or except in connection with a Corporate Transaction, until such time as any vesting restrictions and/or repurchase rights thereon shall lapse.

Section 7.04. Payment. Unless otherwise provided in the applicable Equity Award Agreement, the purchase price, if any, of Stock acquired pursuant to a Stock bonus or restricted Stock award shall be paid in cash (by check) or, in the discretion of the Board, by promissory note (with terms determined by it in its discretion prior to the issuance of any Stock pursuant to such award).

Section 7.05. Vesting. Subject to the provisions of the Plan, the Board, in its discretion, shall determine whether shares of Stock sold or awarded under Article VII of the Plan shall be subject to vesting or to repurchase by the Corporation, and the time or times when such vesting restrictions and/or repurchase rights shall lapse, and such determinations shall be set forth in the applicable Equity Award Agreement. An Equity Award may be subject to such other terms and conditions on the time or times when it may vest (which may be based on performance or other criteria) as the Board may deem appropriate if expressly set forth in an Equity Award Agreement. Unless otherwise approved by the Board and set forth in writing by an authorized officer of the Corporation, a Stock bonus or restricted Stock award shall cease vesting upon the holder's Termination, and (if applicable) the right to acquire any Stock purchasable thereunder which has not been purchased by such time shall terminate, regardless of whether or not the holder was given requisite notice of Termination of such holder's employment by the Corporation or by any Affiliate of the Corporation.

ARTICLE VIII

ADJUSTMENTS UPON CHANGES IN STOCK; CORPORATE TRANSACTIONS

Section 8.01. Change in Stock. If any change is made in the Stock subject to the Plan, through a merger, consolidation, reorganization, recapitalization, reincorporation, stock dividend, dividend in property other than cash, stock split, liquidating dividend, combination of shares, exchange of shares, change in corporate structure or other transaction not involving the receipt of consideration by the Corporation (other than a Corporate Transaction), the Plan will be appropriately adjusted in the class(es) and maximum number of shares subject to the Plan pursuant to Article V, and the outstanding Equity Awards will be appropriately adjusted (to the extent not previously exercised by the holders thereof) in the class(es) and number of shares subject thereto and in the exercise price of such outstanding Equity Awards. If as a result of such event, a holder of an Equity Award would become entitled to a fractional share of Stock or other security, such holder shall have the right to purchase only the next lowest whole number of shares of Stock or other security and no payment or other adjustment will be made with respect to the fractional interest so disregarded. The Board shall make such adjustments at the time of the change in the Stock, whether or not specifically provided for in any outstanding Equity Award. The Board's determination shall be final, binding and conclusive. Notwithstanding the foregoing, any such adjustment shall be made only if and to the extent that such adjustment would not cause any Equity Award intended to qualify as an Incentive Stock Option to fail to so qualify.

Section 8.02. Corporate Transaction. Unless the surviving corporation (or a parent or subsidiary of such corporation) in the Corporate Transaction assumes this Plan or such Equity Award or issues a substitute therefor or unless the Board provides in substitution for any outstanding Equity Award such alternative consideration as it, in good faith, may determine to be equitable in the circumstances, including cash, or unless otherwise provided in the Equity Award Agreement pursuant to which such Equity Award was originally granted, and subject to the provisions of Section 10.01, the following shall apply in the event of a Corporate Transaction:

If such Equity Award is an Option, then it shall terminate upon the effective date of the Corporate Transaction to the extent not exercised prior thereto.

If such Equity Award is a Stock bonus or restricted Stock award, then (i) the vested portion thereof shall survive the Corporate Transaction and shall be subject to the terms and conditions of such Corporate Transaction (including, but not limited to, any terms and conditions applicable to the sale, exchange, conversion or other disposition of such Stock bonus or Restricted Stock award in such Corporation Transaction), and (ii) the unvested portion thereof shall terminate upon the effective date of the Corporate Transaction (provided that in connection with the consummation of such Corporate Transaction, the Corporation shall pay the holder thereof an amount equal to the purchase price (if any) originally paid by such holder for the Stock bonus or Restricted Stock award so terminated).

(c) No Equity Award may be made after the effective date of the Corporate Transaction.

ARTICLE IX

COVENANTS OF THE CORPORATION

Section 9.01. Section 9.01 Reservation of Stock. The Corporation shall reserve from its authorized but unissued Stock the number of shares of Stock issuable pursuant to outstanding Equity Awards.

Section 9.02. Regulatory Authority. The Corporation shall seek to obtain from each regulatory commission or agency having jurisdiction over the Plan such authority as may be required to make an Equity Award and to issue and sell shares of Stock upon the exercise of outstanding Equity Awards, provided that this undertaking shall not require the Corporation to register under the Securities Act or under any applicable state securities laws either the Plan, any Equity Award or any Stock issued or issuable pursuant to any such Equity Award. If, after reasonable efforts, the Corporation is unable to obtain from any such regulatory commission or agency the authority for the lawful grant of any such Equity Award or the lawful issuance and sale of Stock under the Plan, then, as the case may be, the Equity Award so granted shall be nullified or the Corporation shall be relieved from any liability for failure to issue and sell Stock upon exercise of such Equity Awards unless and until such authority is obtained.

ARTICLE X

GENERAL PROVISIONS

Section 10.01. Acceleration of Vesting. Notwithstanding any provision in any Equity Award Agreement, the Board may, in its discretion, accelerate the time at which an Equity Award may first be exercised or the time during which an Equity Award or any part thereof will vest.

Section 10.02. Stockholder Rights. Except as set forth in the Equity Award Agreement, no holder of any Equity Award shall be deemed to be the holder of, or to have any of the rights of a holder with respect to, any Stock subject to such Equity Award unless and until such person has satisfied all requirements for vesting or exercise of the Equity Award pursuant to its terms and the amount due in payment for Stock to be issued pursuant to such Equity Award Agreement, if any, has been paid in full to the Corporation.

Section 10.03. Employment or Other Services. Nothing in the Plan, any Equity Award Agreement or any instrument executed pursuant thereto shall (a) confer upon any Employee or other holder of an Equity Award any right to employment or to continue in the employ of the Corporation or any Affiliate, (b) confer upon any Director or Consultant or other holder of an Equity Award any right to act or to continue acting as a Director or Consultant, (c) affect the right of the Corporation or any Affiliate to terminate the employment of any Employee with or without Cause, (d) affect the right of the Corporation's Board and/or the Corporation's stockholders to remove any Director pursuant to the terms of the Corporation's charter documents and the provisions of applicable law, or (e) affect the right of the Corporation to terminate the relationship of any Consultant pursuant to the terms of such Consultant's agreement with the Corporation or Affiliate.

Section 10.04. Securities Requirements. The Corporation hereby informs each recipient of an Equity Award that the Equity Award and the Stock subject thereto (a) have not been qualified by prospectus and are subject to indefinite holding periods, and (b) are unregistered securities under the Securities Act and under all applicable state securities laws and must be held indefinitely unless they are subsequently registered or qualified thereunder or an exemption from such registration or qualification is available. The grant of any Equity Award and the issuance of any shares of Stock by the Corporation pursuant to an Equity Award is subject to compliance with the laws, rules and regulations of all public agencies and authorities applicable to the issuance and distribution of such Equity Award and/or Stock and to the listing requirements of any stock exchange or exchanges on which the Stock may be listed from time to time. The recipient agrees (a) to comply with all such laws, rules and regulations, (b) to furnish to the Corporation any information, report and/or undertakings required to comply with all such laws, rules and regulations, and (c) to fully cooperate with the Corporation in complying with such laws, rules and regulations. The Corporation may require any person to whom an Equity Award is granted, or any person to whom an Equity Award is transferred, as a condition of exercising or acquiring Stock under any Equity Award, to give written assurances satisfactory to the Corporation (a) as to the matters provided above, (b) as to such person's knowledge and experience in financial and business matters, (c) that he or she is capable of evaluating, alone or together with a purchaser representative, the merits and risks of exercising the Equity Award,

and (d) that such person is acquiring the Stock subject to the Equity Award for such person's own account and not with any view to a distribution of the Stock. The Corporation may, upon advice of counsel to the Corporation, place legends on stock certificates issued under the Plan as such counsel deems necessary or appropriate in order to comply with applicable securities laws, including, but not limited to, legends restricting the transfer of the Stock. Notwithstanding anything to the contrary contained in this Plan or an Equity Award Agreement, no Stock shall be issued to a person pursuant to an Equity Award unless such shares of Stock are then registered under the Securities Act and registered or qualified under all applicable state securities laws, or if such shares are not then so registered or qualified, the Corporation has determined that such issuance would be exempt from the registration requirements of the Securities Act and all applicable state securities laws.

Section 10.05. Tax Withholding. Unless otherwise provided in the applicable Equity Award Agreement or by the Board, the Corporation shall require the holder of an Equity Award to pay in cash (by check) to the Corporation the holder's share of any tax withholding arising under any applicable law by reason of such Equity Award, the vesting thereof or the disposition of Stock subject thereto. Subject to its withholding obligations under applicable law, and notwithstanding any other provision of this Plan, the Corporation does not assume responsibility for the income or other tax consequences for any person who is eligible for or has received an Equity Award under the Plan, and such persons are advised to consult with their own tax advisers with respect to such matters.

Section 10.06. Equity Award Agreement. The grant of any Equity Award is subject to the execution by the recipient of an Equity Award Agreement.

ARTICLE XI

AMENDMENT OF THE PLAN AND EQUITY AWARDS

Section 11.01. Amendment of Plan; Stockholder Approval. The Board may, in its discretion, amend the Plan. Such amendment shall be effective on the date the Board determines, except for amendments that require the approval of the Corporation's stockholders, in which case such amendments shall be effective on the date the Corporation's stockholders approve the amendment. No such amendment shall reduce any outstanding Equity Award without the holder's written consent. The Board may, in its discretion, submit any amendment of the Plan for stockholder approval.

Section 11.02. Changes in Law. The Board may amend the Plan as it deems necessary or advisable to provide eligible Employees, Directors or Consultants with the maximum benefits provided or to be provided under the provisions of the Plan relating to Incentive Stock Options and to bring the Plan or Incentive Stock Options granted under the Plan into compliance therewith. The Board may also, in its discretion, amend the Plan to take into account changes in law and tax and accounting rules, as well as other developments, and to grant Equity Awards that qualify for beneficial treatment under such rules.

APPENDIX A

DEFINITIONS

“**AFFILIATE**” means any parent corporation or subsidiary corporation of the Corporation, whether now or hereafter existing, as those terms are defined in Sections 424(e) and (f) respectively, of the Code.

“**BOARD**” means the Board of Directors of the Corporation.

“**CAUSE**” has the meaning given it in the employment or consulting agreement which governs the relationship between the Corporation and the holder of the Equity Award or, if there is no such definition in any such agreement, means (a) indictment or conviction for either any felony offense or any other crime involving dishonesty, (b) participation in any fraud, theft, embezzlement or other misconduct against the Corporation, (c) intentional damage to any property of the Corporation, (d) breach of the holder’s duties of good faith and fair dealing that are owed to the Corporation, (e) breach or violation of any employment, confidentiality, non-competition, non-solicitation or assignment of inventions agreement, (f) conduct which in the good faith and reasonable determination of the Board demonstrates gross unfitness to serve, (g) failure to comply with the policies of the Corporation that have been approved by the Board, or (h) insubordination or failure to follow the directions of the Board or of the Chief Executive Officer or President of the Corporation.

“**CODE**” means the Internal Revenue Code of 1986, as amended, and the regulations promulgated thereunder.

“**COMMITTEE**” means a Committee appointed by the Board in accordance with Section 3.01 of the Plan.

“**CORPORATION**” means Akebia Therapeutics, Inc., a Delaware corporation, and its successors and assigns.

“**CONSULTANT**” means any person, including an advisor, engaged by the Corporation or an Affiliate to render bona fide consulting services (other than services in connection with the offer or sale of securities in a capital-raising transaction) and who is compensated for such services, provided that the term “Consultant” shall not include Directors who are paid only a director’s fee by the Corporation or who are not compensated by the Corporation for their services as Directors.

“**CORPORATE TRANSACTION**” means a “*Deemed Liquidation Event*” as such term is defined in the Corporation’s charter documentation, as in effect from time to time.

“**DIRECTOR**” means a member of the Board.

“**DISABILITY**” has the meaning given it in the employment or consulting agreement which governs the relationship between the Corporation and the holder of the Equity Award or, if there is no such definition in any such agreement, means any medically determinable physical or mental impairment rendering an individual unable to engage in any substantial gainful activity, which disability can be expected to result in death or which has lasted or can be expected to last for a continuous period of not less than six (6) months.

“**EMPLOYEE**” means any person employed, whether full or part-time, as an employee (including as an officer) by the Corporation or any Affiliate of the Corporation. Neither service as a Director nor payment of a director’s fee by the Corporation shall be sufficient to constitute “*employment*” by the Corporation. However, a Director who is also employed as an employee by the Corporation or an Affiliate shall constitute an Employee hereunder.

“**EQUITY AWARD**” means any right granted under the Plan, including any Option, any Stock bonus or any right to purchase restricted Stock.

“**EQUITY AWARD AGREEMENT**” means a written agreement between the Corporation and a holder of an Equity Award evidencing the terms and conditions of an individual Equity Award grant. Each Equity Award Agreement shall be subject to the terms and conditions of the Plan.

“**FAIR MARKET VALUE**” means, as of any date, the value of the Stock determined as follows:

- If the Stock is listed on any established stock exchange or a national market system, including, but not limited to, the Nasdaq National Market or Nasdaq Small Cap Market, the Fair Market Value of a share of Stock shall be the last sales price for the Stock (or the closing bid, if no sales were reported) as quoted on such system or exchange, as reported in *The Wall Street Journal* or such other source as the Board deems reliable.
- In the absence of an established market for the Stock, the Fair Market Value shall be determined in good faith by the Board, shall take into account appropriate discounts for lack of marketability or due to a minority position, and shall take into account the applicable preferences and privileges of the Corporation’s preferred stock as set forth in the Corporation’s charter documentation, as in effect from time to time.

“**INCENTIVE STOCK OPTION**” means an Option intended to qualify as an incentive stock option within the meaning of Section 422 of the Code and the regulations promulgated thereunder.

“**NON-STATUTORY STOCK OPTION**” means an Option not intended to qualify as an Incentive Stock Option.

“**OPTION**” means a stock option granted pursuant to the Plan.

“**OPTIONEE**” means an Employee, Director or Consultant who holds an outstanding Option.

“**OUTSIDE DIRECTOR**” means a Director who either (a) is not a current Employee of the Corporation or an “*affiliated corporation*” (within the meaning of Treasury regulations promulgated under Section 162(m) of the Code), is not a former Employee of the Corporation or an “*affiliated corporation*” receiving compensation for prior services (other than benefits under a tax qualified pension plan), was not an officer of the Corporation or an “*affiliated corporation*”

at any time, and is not currently receiving direct or indirect remuneration from the Corporation or an “*affiliated corporation*” for services in any capacity other than as a Director, or (b) is otherwise considered an “*outside director*” for purposes of Section 162(m) of the Code.

“**PLAN**” means this Equity Incentive Plan, as amended and restated.

“**SECURITIES ACT**” means the Securities Act of 1933, as amended, and the regulations promulgated thereunder.

“**STOCK**” means the Corporation’s Common Stock, \$0.00001 par value per share, and any security into which such Common Stock may be changed.

“**STOCK PURCHASE AGREEMENT**” means a written agreement between the Corporation and a holder of an Equity Award evidencing the terms and conditions under which such holder shall hold the shares of Stock awarded or purchased under the terms of the Equity Award. Each Stock Purchase Agreement shall be subject to the terms and conditions of the Plan and the Equity Award Agreement that evidenced the bonus, award or Option.

“**TERMINATION**” means the termination of an Employee’s, Director’s or Consultant’s employment or relationship with the Corporation or with any Affiliate of the Corporation.

[End of Amended and Restated 2008 Equity Incentive Plan]

AKEBIA THERAPEUTICS, INC.

AMENDMENT NO. 1 TO

AMENDED AND RESTATED 2008 EQUITY INCENTIVE PLAN

Effective as of May 10, 2013

Section 5.01 is amended such that the number of shares of Stock set forth in the first sentence thereof shall be 957,876.29, which number increases the number of shares of Common Stock reserved thereunder but also reflects the reverse stock split that the Corporation underwent pursuant to the filing of the Corporation's Eight Amended and Restated Certificate of Incorporation, dated May 10, 2013 and in accordance with Section 8.01 of the Corporation's Amended and Restated 2008 Equity Incentive Plan, as amended from time (the "**Plan**").

Capitalized terms not defined herein have the meanings given them in the aforementioned Plan.

[End of Amendment No. 1 to Akebia Therapeutics, Inc. 2008 Equity Incentive Plan]

EXECUTIVE EMPLOYMENT AGREEMENT

THIS EXECUTIVE EMPLOYMENT AGREEMENT (this "Agreement") is made effective as of September 16, 2013, by and between Akebia Therapeutics, Inc., a Delaware corporation (the "Company"), and John P. Butler, an individual resident in the Commonwealth of Massachusetts ("Executive").

1. Employment; Duties; Full Time Employment. The Company hereby agrees to employ Executive, and Executive hereby accepts employment, as President and Chief Executive Officer of the Company, with such employment to commence on September 16, 2013 (the "Commencement Date"). In such capacity, Executive shall perform such Executive duties and exercise such powers for the Company and its subsidiaries as the Board of Directors of the Company (the "Board") may lawfully assign to or vest in Executive from time to time. Executive covenants and agrees that, at all times during the Term (as defined below), Executive shall devote Executive's full business time and efforts to Executive's duties as an employee of the Company and that Executive will not, directly or indirectly, engage or participate in any other business or professional activities during the Term, other than (a) non-conflicting personal investments managed on Executive's personal time, (b) activities for non-profit institutions (including, but not limited to, participating on boards of directors), and (c) activities or commitments set forth and described on Appendix A (Section 1) hereto, provided that such activities do not interfere or conflict with Executive's obligations hereunder.

2. Place of Performance. Executive's principal place of business for the performance of his duties under this Agreement shall be in the greater Boston, Massachusetts metropolitan area. Executive shall also travel as reasonably necessary or appropriate to perform his duties hereunder, including, but not limited to, trailing to the Company's offices in Cincinnati, Ohio. At no time during the Term shall Executive be required to relocate to the Cincinnati, Ohio area or otherwise be required to relocate his residence from the greater Boston, Massachusetts metropolitan area.

3. Term. The Company agrees to employ Executive, and Executive agrees to serve the Company, on an "*at will*" basis, which means that either the Company or Executive may terminate Executive's employment with the Company at any time, with or without Cause, as provided in Section 6 below. The period commencing with the Commencement Date and ending on the effective date of any termination of employment in accordance with the provisions hereof shall constitute the term of this Agreement (the "Term").

4. Compensation and Benefits. During the Term, the Company shall provide Executive with the following compensation and benefits:

(a) Base Salary. The Company shall pay Executive a base salary ("Base Salary") at the rate of \$425,000 per annum (less applicable deductions and withholdings), payable in periodic payments in accordance with the Company's normal payroll practices. During the Term, Executive's compensation shall be reviewed by the Board from time to time and at least once every 12 months. Any increase or decrease in Base Salary (together with the then existing Base Salary) shall serve as the "Base Salary" under this Agreement. Any such decrease in Base Salary, however, shall constitute Good Reason under Section 6(e) of this Agreement.

(b) Discretionary Bonuses. Executive will be eligible to receive an annual bonus in an amount up to 30% of Executive's Base Salary. The exact amount of the actual bonus awarded to Executive for any given year, if any, shall be determined by the Board in its sole discretion based upon its consideration of the Company's performance and Executive's performance against objectives established by the Board for the year, in consultation with Executive. Except as otherwise may be required by applicable law, Executive must remain an employee through the end of the calendar year in order to earn a bonus for that year. Executive will not earn any bonus (including a prorated bonus) for any year if Executive's employment terminates for any reason before the end of such year. Any annual bonus shall be paid by March 15 of the following calendar year.

(c) Stock Ownership. On or by the Effective Date, Executive shall be awarded 350,000 options to acquire shares of the Common Stock of the Company ("Stock Options"), which Stock Options represent approximately 4.3% of the Company's Common Stock outstanding as of the Effective Date on a fully-diluted and as-converted basis. Except for the foregoing Stock Options, and except as otherwise may be determined by the Board from time to time after the date of this Agreement in its discretion Executive shall not have any right to be issued shares of the Company's capital stock or options, warrants or other rights to acquire any capital stock of the Company.

(d) Other Compensation and Benefits. In addition to the compensation specified above in this Section 4, Executive shall be entitled to the following benefits during the Term, all on the terms offered or maintained by the Company to, for or on behalf of its senior executives: vacation, holidays and sick leave, and subject to eligibility therefor, the right to participate in any profit sharing plan, retirement plan, 401(k) plan, group medical plan, group dental plan, group life insurance plan and/or other health or insurance plan maintained by the Company for its senior executives generally and, if applicable, their family members. Executive will be eligible for the paid holidays as are generally made available to employees of the Company.

(e) Vacation. The Executive shall be entitled to four weeks paid vacation per calendar year to be taken at such times as may be approved by the Board (which is more than the normal amount defined in the Akebia Employee Handbook). An aggregate of up to 1 week of unused vacation time may be carried over at the end of a calendar year. Upon termination of the Executive's employment, the Company will pay the Executive for unused vacation at the Executive's Base Salary rate (subject to normal deductions and withholding amounts) on the next regularly scheduled pay date immediately following the termination date, or earlier if required by applicable law.

5. Business Expenses. The Company shall reimburse Executive for all reasonable and necessary business and travel expenses incurred by Executive in the performance of Executive's duties under this Agreement. Such expenses shall be reimbursed in accordance with the Company's guidelines, limits and procedures relating thereto and upon presentation of proper expense vouchers or receipts therefor.

6. Termination.

(a) Termination on Death or Disability. The Term will terminate automatically and immediately upon Executive's death or, upon 30 days prior written notice from the Company, in the event of Executive's Disability. For purposes of this Section 6, "Disability," means that Executive, at the time notice is given, has been unable to substantially perform Executive's duties under this Agreement for not less than sixty (60) work days within a six (6) consecutive month period as a result of Executive's incapacity due to physical or mental illness. Upon any termination for death or Disability, Executive will not be entitled to any further compensation from the Company, including severance pay, pay in lieu of notice or any other such compensation (other than accrued salary and bonus, and accrued and unused vacation, through Executive's last day of employment).

(b) Termination by the Company Without Cause; Termination by the Executive for Good Reason. During the Term, the Company shall be entitled to terminate Executive's employment without Cause (as defined below), and the Executive is also entitled to terminate his employment for Good Reason (as defined below), in which case Executive shall be entitled to receive the following severance benefits (the "Severance Payments"), in addition to accrued salary and bonus, and accrued and unused vacation, through Executive's last day of employment: (i) Executive shall be entitled to severance pay in the form of continuation of Executive's Base Salary in effect on the effective date of termination for a period of twelve (12) months after the date of such termination, to be paid periodically in accordance with the Company's normal payroll practices and subject to standard payroll deductions and withholdings; (ii) if Executive timely elects continued coverage under COBRA, then (A) the Company shall make such COBRA coverage (or equivalent medical benefits after the termination of COBRA) available for at least 18 months following termination and (B) the Company shall pay the COBRA premiums necessary to continue Executive's medical insurance coverage in effect on the termination date for a period of twelve (12) months following Executive's termination (provided that such COBRA continuation and reimbursement shall terminate upon commencement of new employment by an employer that offers health care coverage to its employees); and (iii) a pro-rata portion of the Executive's annual target bonus for the calendar year in which the termination occurs.

Notwithstanding the foregoing, all Severance Payments under this Agreement are conditional on Executive (i) complying with the provisions of Section 7 below, and (ii) delivering prior to receipt of such severance payments, an effective, general release of claims in favor of the Company or its successor, its subsidiaries and their respective directors, officers and stockholders in a form acceptable to the Company or its successor.

In the event that the Company determines that any severance benefit provided hereunder fails to satisfy the distribution requirement of Section 409A(a)(2)(A) of the Internal Revenue Code ("Code") as a result of Section 409A(a)(2)(B)(i) of the Code, then if an accelerated payment of such benefits would cause such benefit not to be subject to the provisions of Section 409A(a)(1) of the Code, the payment of such benefits shall be accelerated to the minimum extent necessary so that the benefit is not subject to the provisions of Section 409A(a)(1) of the Code.

(The payment schedule as revised after the application of the preceding sentence shall be referred to as the “Revised Payment Schedule.”) However, in the event the accelerated payment of such benefits would not avoid the application of Section 409A (a)(1) of the Code, the payment of such benefits shall not be made pursuant to the original payment schedule or the Revised Payment Schedule and instead the payment of such benefits shall be delayed to the minimum extent necessary so that such benefits are not subject to the provisions of Section 409A(a)(1) of the Code. The Board may attach conditions to or adjust the amount paid pursuant to this Section 6(b)(iv) to preserve, as closely as possible, the economic consequences that would have applied in the absence of this Section 6(b)(iv); provided, however, that no such condition or adjustment shall result in the payments being subject to Section 409A(a)(1) of the Code.

Notwithstanding any other provisions in this Agreement, it shall be a prerequisite of any termination by Executive for Good Reason that Executive shall have given the Company written notice within sixty (60) days following the date Executive becomes aware of the event or events giving rise to Good Reason, specifying in reasonable detail the nature and circumstances of such Good Reason, and giving the Company thirty (30) days to cure any such Good Reason prior to any such termination, and if uncured, the termination for Good Reason must occur within ninety (90) days of the end of such cure period.

(c) Termination for Cause; Resignation. The Company may terminate Executive’s employment at any time for Cause, and Executive may resign at any time. Termination for Cause shall be effective on the date the Company gives notice to Executive of such termination in accordance with this Agreement. Resignation by Executive shall be effective on the date Executive gives notice to the Company of such resignation in accordance with this Agreement. In the event of the Company’s termination of the Term for Cause or Executive’s resignation from Executive’s employment, Executive will not be entitled to any further compensation from the Company, including severance pay, pay in lieu of notice or any other such compensation (other than accrued salary and bonus, and accrued and unused vacation, through Executive’s last day of employment).

(d) Cause. For purposes of this Agreement, “Cause” shall mean (i) Executive’s failure to substantially perform Executive’s duties under this Agreement for reasons other than death or Disability, which failure, if curable, is not cured to the reasonable satisfaction of the Board during the thirty (30) day period following written notice of such failure from the Company; (ii) Executive’s material failure or refusal to comply with reasonable written policies, standards and regulations established by the Company from time to time which failure, if curable, is not cured to the reasonable satisfaction of the Board during the thirty (30) day period following written notice of such failure from the Company; (iii) the proven commission by Executive of (x) an act of dishonesty or constituting common law fraud, embezzlement or a felony or (y) any tortious act, unlawful act or malfeasance that causes material harm to the Company’s standing, condition or reputation; or (iv) any material breach by Executive of the provisions of this Agreement, which breach, if curable, is not cured to the reasonable satisfaction of the Board during the thirty (30) day period following written notice of such breach from the Company. The Board (excluding Executive if Executive is at such time a member of the Board) shall in good faith make all determinations relating to termination, including without limitation any determination regarding Cause, pursuant to this Section 6(d).

(e) Good Reason. For purposes of this Agreement, “Good Reason” shall mean any of the following without the consent of the Executive: (i) a material diminution in the Executive’s position, duties or responsibilities from those held by or assigned to the Executive as of the Effective Date, (ii) a reduction of the Executive’s Base Salary, (iii) a material reduction of the Executive’s benefits or bonus/incentive compensation opportunities provided to the Executive as then in effect, so long as he is the only executive to suffer such a reduction, or (iv) requiring the Executive to relocate beyond a fifty (50) mile radius of the Executive’s then current residence.

(f) Removal from any Boards and Positions. Notwithstanding provision in this Agreement, if Executive’s employment is terminated under this Agreement for any reason, Executive shall be deemed to resign (i) from the Board or board of directors of any affiliate of the Company or any other board to which he has been appointed or nominated by or on behalf of the Company, and (ii) from any position with the Company or any affiliate of the Company, including, but not limited to, as an officer of the Company or any of its affiliates.

7. Company Matters: Restrictive Covenants.

(a) Confidential Information. Executive will have access to and will participate in the development of and will be acquainted with confidential or proprietary information and trade secrets related to the business of the Company and its affiliates, including but not limited to (i) customer lists; related records and compilations of information; the identity, lists or descriptions of any new customers, referral sources or organizations; financial statements; cost reports or other financial information; contract proposals or bidding information; business plans; training and operations methods and manuals; personnel records; software programs; reports and correspondence; and management systems, policies or procedures, including related forms and manuals, (ii) information pertaining to future developments such as future marketing or acquisition plans or ideas and potential new business locations; and (iii) all other tangible and intangible property and intellectual property which is used in the business and operations of the Company and its affiliates but not made public. The foregoing is collectively referred to as the “Confidential Information.” The term Confidential Information shall not include any information (x) that is or becomes generally publicly available (other than as a result of violation of this Agreement by Executive), or (y) that Executive receives on a non-confidential basis from a source (other than the Company, its affiliates or their representatives) that is not known by Executive to be bound by an obligation of secrecy or confidentiality to any of the Company or its affiliates. Executive shall not disclose, use or make known for Executive’s or another’s benefit other than for the benefit of the Company and its affiliates any Confidential Information or use such Confidential Information in any way. Upon the termination of Executive’s employment with the Company for any reason, Executive shall immediately return to the Company all Confidential Information in whatever form maintained (including, without limitation, computer discs and other electronic media).

(b) Non-Competition. During Executive’s employment with the Company and for the one-year period immediately following the termination of Executive’s employment with the Company, Executive will not directly or indirectly (whether as an officer, director, stockholder, partner, proprietor, associate, representative, consultant or in any capacity whatsoever) engage in, become financially interested in, be employed by or have any business

connection with any person, corporation, firm, partnership or any other entity whatsoever which competes with the Company in any area where the Company operates, or has operated at any time during Executive's employment with the Company. Notwithstanding the foregoing, the ownership by Executive of not more than three percent (3%) of the shares of stock of any corporation having a class of equity securities actively traded on a national securities exchange or on the Nasdaq Stock Market shall not be deemed to violate the prohibitions this Section 7(b).

(c) Nonsolicitation of Customers. During Executive's employment with the Company and for the one-year period immediately following the termination of Executive's employment with the Company, Executive shall not solicit, directly or indirectly, any customers or prospective customers of the Company with whom Executive had contact on behalf of the Company during Executive's employment with the Company.

(d) Nonsolicitation of Employees. During Executive's employment with the Company and for the one-year period immediately following the termination of Executive's employment with the Company, Executive shall not solicit or hire, directly or indirectly, on Executive's behalf or on behalf of any other person or entity, any person employed by the Company except with the specific written consent of the Company.

(e) Certain Representations. Executive represents that Executive's experience, capabilities and circumstances are such that the provision of this Agreement will not prevent Executive from earning a livelihood. Executive further agrees that the limitations set forth in this Agreement (including, without limitation, the time and territorial limitations) are reasonable and properly required for the adequate protection of the current and future businesses of the Company. Executive further acknowledges that a remedy at law for any breach or threatened breach of the provisions of this Agreement would be inadequate and will cause immediate and irreparable harm to the Company in a manner that cannot be measured nor adequately compensated in damages. Executive further acknowledges that in the event of any such breach and in addition to any and all other remedies that it may have at law or in equity, the Company shall be entitled to seek temporary, preliminary and permanent injunctive relief to restrain such breach by Executive, and the prevailing party in any such proceeding shall be entitled to recover all associated costs and expenses, including reasonable attorneys' fees, from the non-prevailing party. Nothing contained herein shall restrict or limit in any manner the Company's right to seek and obtain any form of relief, legal or equitable, against Executive in an action brought to enforce its rights hereunder.

(f) Intellectual Property. Except as otherwise set forth and described on Appendix A (Section 7(f)) hereto, all ideas, concepts, inventions, improvements, programs, information technology, derivative works, processes, configurations, data, procedures, designs, techniques and other works of authorship and development made, conceived or reduced to practice by Executive, either solely or in collaboration with others, during Executive's employment with the Company, including but not limited to all copyright, trademark, patent, trade secret and intellectual property rights associated therewith, shall become and remain the exclusive property of the Company. Executive hereby assigns to the Company any and all of Executive's right, title and interest in and to any of the foregoing, and Executive waives any claim that Executive may have thereto. Executive will promptly disclose in writing to the Company all such ideas, concepts, inventions, improvements, programs, information technology,

derivative works, processes, configurations, data, procedures, designs, techniques and other works of authorship and development, and will cooperate fully with the Company in confirming and protecting the Company's ownership rights therein. The work product resulting from the Executive's employment with the Company is work made for hire.

(g) Ventures. If, during the Term, Executive is engaged in or associated with the planning or implementing of projects, programs or ventures involving the Company and third parties, all rights in such projects, programs and ventures shall belong to the Company (or the third party, to the extent provided in any agreement between the Company and the third party). Except as formally approved by the Company, Executive shall not be entitled to any interest in such project, program or venture or to any commission, finder's fee or other compensation in connection therewith other than the salary or other compensation to be paid to Executive as provided in this Agreement.

8. Miscellaneous.

(a) Withholding Taxes. The Company may withhold from all salary, bonus or other benefits payable under this Agreement all federal, state, city or other taxes as shall be required pursuant to any law or governmental regulation or ruling.

(b) Entire Agreement; Binding Effect. This Agreement sets forth the entire understanding between the parties as to the Subject matter of this Agreement and supersedes all prior agreements, commitments, representations, writings and discussions between them (whether written or oral) on the subject matters herein; and neither of the parties shall be bound by any obligations, conditions, warranties or representations with respect to the subject matter of the foregoing except as expressly provided herein or therein or as duly set forth on or subsequent to the date hereof in a written instrument signed by the proper and fully authorized representative of the party to be bound hereby. This Agreement is binding on Executive and on the Company and Executive and their respective successors and assigns (whether by assignment, by operation of law or otherwise); provided that neither this Agreement nor any rights or obligations hereunder may be assigned by Executive or the Company without the prior written consent of the other party (except that the Company shall be entitled to assign this Agreement in connection with the sale of all or substantially all of the Company's assets, or a merger or consolidation in which the Company is not the surviving entity).

(c) Absence of Conflict. Executive represents and warrants that Executive's employment by the Company as described herein will not conflict with and will not be constrained by any prior employment or consulting agreement or relationship.

(d) Voluntary Nature of Agreement; Legal Rights. Executive is executing this Agreement voluntarily and without any duress or undue influence by the Company or anyone else. Executive acknowledges that Executive has had the opportunity to consult with an attorney regarding the provisions of this Agreement and has either obtained such advice of counsel or knowingly waived the opportunity to seek such advice. Executive has carefully read this Agreement and has asked any questions needed for Executive to understand the terms, consequences and binding effect of this Agreement and fully understand it, including that Executive is waiving Executive's right to a jury trial.

(e) Waivers. No party shall be deemed to have waived any right, power or privilege under this Agreement or any provisions hereof unless such waiver shall have been duly executed in writing and acknowledged by the party to be charged with such waiver. The failure of any party at any time to insist on performance of any of the provisions of this Agreement shall in no way be construed to be a waiver of such provisions, nor in any way to affect the validity of this Agreement or any part hereof. No waiver of any breach of this Agreement shall be held to be a waiver of any other subsequent breach.

(f) Notices. All notices, approvals, consents, requests or demands required or permitted to be given under this Agreement shall be in writing and shall be deemed sufficiently given (i) upon delivery, if delivered by hand (ii) one business day after transmission, if sent by facsimile (confirmation received) or (iii) one business day after the business day of deposit with a reputable overnight courier for next business day delivery, freight prepaid (signature of receipt obtained). Notice in each case shall be addressed to the party entitled to receive such notice at the following address (or other such addresses as the parties may subsequently designate):

If to the Company:

Akebia Therapeutics, Inc.
Attn: President
9987 Carver Road, Suite 420
Cincinnati, Ohio 45242
Fax: (513) 985-1920

If to the Executive:

John P. Butler
85 Mosher Lane
Marlborough, MA

With copies to:

Steven D. Weatherhead
Bello Black & Welsh LLP
125 Summer Street, Suite 1200
Boston, Massachusetts 02110
Fax:(617) 247-4125

With copies of all notices also to go to Company counsel and the Chair of the Company's Compensation Committee, as follows:

Thompson Hine LLP
312 Walnut Street, Suite 1400
Cincinnati, Ohio 45202
Attn: David J. Willbrand, Esq.
Fax: (513) 241-4771

Akebia Therapeutics, Inc.
Attn: Chair of Compensation Committee
9987 Carver Road, Suite 420
Cincinnati, Ohio 45242
Fax: (513) 985-1920

(g) Governing Law. This Agreement shall be governed by, and construed and enforced in accordance with, the laws of the State of Ohio without regard to conflict of law principles that would result in the application of any law other than the law of the State of Ohio.

(h) Severability. Every provision of this Agreement is intended to be severable from every other provision of this Agreement. If any provision of this Agreement is held to be invalid, illegal or unenforceable, in whole or in part, such invalidity, illegality or unenforceability shall not affect the other provisions of this Agreement; and this Agreement shall be construed as if such invalid, illegal or unenforceable provision had never been contained herein except to the extent that such provision may be construed and modified so as to render it valid, lawful, and enforceable in a manner consistent with the intent of the parties to the extent compatible with the applicable law as it shall then appear.

(i) Effect of Headings. The Section and subsection headings contained herein are for convenience only and shall not affect the construction hereof.

(j) Counterparts. This Agreement may be executed in multiple counterparts, each of which shall be deemed to be an original, and all such counterparts shall constitute but one instrument.

Signature Page Follows

IN WITNESS WHEREOF, the parties hereto have caused this Agreement to be duly executed as of the year and date first written above.

AKEBIA THERAPEUTICS, INC.

By: /s/ Muneer A. Satter

Name: Muneer A. Satter

Its: Chairman of the Board

Executive:

By: /s/ John P. Butler

John P. Butler

Section 1.

- Member of Board of Trustees of the American Kidney Fund
- Potentially a member of the Board of Directors of Relypsa Inc. or Neuralstem Inc. (but not both companies simultaneously)

Section 7(f).

EXECUTIVE EMPLOYMENT AGREEMENT

THIS EXECUTIVE EMPLOYMENT AGREEMENT (this "Agreement") is made effective as of September 23, 2013, by and between Akebia Therapeutics, Inc., a Delaware corporation (the "Company"), and Jason Amello, an individual resident in the Commonwealth of Massachusetts ("Executive").

1. Employment; Duties; Full Time Employment. The Company hereby agrees to employ Executive, and Executive hereby accepts employment, as Chief Financial Officer of the Company, with such employment to commence on September 23, 2013 (the "Commencement Date"). In such capacity, Executive shall perform such executive duties and exercise such powers for the Company and its subsidiaries as the Board of Directors of the Company (the "Board") may lawfully assign to or vest in Executive from time to time. Executive covenants and agrees that, at all times during the Term (as defined below), Executive shall devote Executive's full business time and efforts to Executive's duties as an employee of the Company and that Executive will not, directly or indirectly, engage or participate in any other business or professional activities during the Term, other than (a) non-conflicting personal investments managed on Executive's personal time, (b) activities for non-profit institutions (including, but not limited to, participating on boards of directors), and (c) activities or commitments set forth and described on Appendix A (Section 1) hereto, provided that such activities do not interfere or conflict with Executive's obligations hereunder.

2. Place of Performance. Executive's principal place of business for the performance of his duties under this Agreement shall be in the greater Boston, Massachusetts metropolitan area. Executive shall also travel as reasonably necessary or appropriate to perform his duties hereunder, including, but not limited to, traveling to the Company's offices in Cincinnati, Ohio. At no time during the Term shall Executive be required to relocate to the Cincinnati, Ohio area or otherwise be required to relocate his residence from the greater Boston, Massachusetts metropolitan area.

3. Term. The Company agrees to employ Executive, and Executive agrees to serve the Company, on an "*at will*" basis, which means that either the Company or Executive may terminate Executive's employment with the Company at any time, with or without Cause, as provided in Section 6 below. The period commencing with the Commencement Date and ending on the effective date of any termination of employment in accordance with the provisions hereof shall constitute the term of this Agreement (the "Term").

4. Compensation and Benefits. During the Term, the Company shall provide Executive with the following compensation and benefits:

(a) Base Salary. The Company shall pay Executive a base salary ("Base Salary") at the rate of \$320,000 per annum (less applicable deductions and withholdings), payable in periodic payments in accordance with the Company's normal payroll practices. During the Term, Executive's compensation shall be reviewed by the Board from time to time and at least once every 12 months. Any increase or decrease in Base Salary (together with the then existing Base Salary) shall serve as the "Base Salary" under this Agreement. Any and every decrease in Base Salary, however, shall constitute Good Reason under Section 6(e) of this Agreement.

(b) Discretionary Bonuses. Executive will be eligible to receive an annual bonus in an amount up to 20% of Executive's Base Salary (the "Annual Target Bonus"). The exact amount of the actual bonus awarded to Executive for any given year, if any, shall be determined by the Board in its sole discretion based upon its consideration of the Company's performance and Executive's performance against objectives established by the Board for the year, in consultation with Executive. Except as otherwise may be required by applicable law and except as provided in Section 6(b), Executive must remain an employee through the end of the calendar year in order to earn a bonus for that year. Except as provided in Section 6(b), Executive will not earn any bonus (including a prorated bonus) for any year if Executive's employment terminates for any reason before the end of such year. Any annual bonus shall be paid by March 15 of the following calendar year.

(c) Stock Ownership. On or by the Effective Date, Executive shall be awarded 81,394 options to acquire shares of the Common Stock of the Company ("Stock Options"), which Stock Options represent approximately 1.0% of the Company's Common Stock outstanding as of the Effective Date on a fully-diluted and as-converted basis. Except for the foregoing Stock Options, and except as otherwise may be determined by the Board from time to time after the date of this Agreement in its discretion, Executive shall not have any right to be issued shares of the Company's capital stock or options, warrants or other rights to acquire any capital stock of the Company. Provided, however, Executive will be eligible to participate and participate, commensurate with the Company's senior executives, in any incentive option or stock award plans, deferred compensation plans or other incentive award plans that may be adopted or implemented by the Company.

(d) Other Compensation and Benefits. In addition to the compensation specified above in this Section 4, Executive shall be entitled to the following benefits during the Term, all on the terms offered or maintained by the Company to, for or on behalf of its senior executives: vacation, holidays and sick leave, and subject to eligibility therefor, the right to participate in any profit sharing plan, retirement plan, 401(k) plan, group medical plan, group dental plan, group life insurance plan and/or other health or insurance plan maintained by the Company for its senior executives generally and, if applicable, their family members. Executive will be eligible for the paid holidays as are generally made available to employees of the Company.

(e) Vacation. The Executive shall be entitled to four weeks paid vacation per calendar year to be taken at such times as may be approved by the Board or Chief Executive Officer of the Company (which is more than the normal amount defined in the Akebia Employee Handbook). An aggregate of up to 1 week of unused vacation time may be carried over at the end of a calendar year. Upon termination of the Executive's employment, the Company will pay the Executive for unused vacation at the Executive's Base Salary rate (subject to normal deductions and withholding amounts) on the next regularly scheduled pay date immediately following the termination date, or earlier if required by applicable law.

5. Business Expenses. The Company shall reimburse Executive for all reasonable and necessary business and travel expenses incurred by Executive in the performance of Executive's duties under this Agreement. Such expenses shall be reimbursed in accordance with the Company's guidelines, limits and procedures relating thereto and upon presentation of proper expense vouchers or receipts therefor.

6. Termination.

(a) Termination on Death or Disability. The Term will terminate automatically and immediately upon Executive's death or, upon 30 days prior written notice from the Company, in the event of Executive's Disability. For purposes of this Section 6, "Disability" means that Executive, at the time notice is given, has been unable to substantially perform Executive's duties under this Agreement for not less than sixty (60) work days within a six (6) consecutive month period as a result of Executive's incapacity due to physical or mental illness. Upon any termination for death or Disability, Executive will not be entitled to any further compensation from the Company, including severance pay, pay in lieu of notice or any other such compensation (other than accrued salary and bonus, and accrued and unused vacation, through Executive's last day of employment).

(b) Termination by the Company Without Cause; Termination by the Executive for Good Reason. During the Term, the Company shall be entitled to terminate Executive's employment without Cause (as defined below), and the Executive is also entitled to terminate his employment for Good Reason (as defined below), in which case Executive shall be entitled to receive the following severance benefits (the "Severance Payments"), in addition to accrued salary and bonus, and accrued and unused vacation, through Executive's last day of employment: (i) Executive shall be entitled to severance pay in the form of continuation of Executive's Base Salary in effect on the effective date of termination for a period of twelve (12) months after the date of such termination, to be paid periodically in accordance with the Company's normal payroll practices and subject to standard payroll deductions and withholdings; (ii) if Executive timely elects continued coverage under COBRA, then (A) the Company shall make such COBRA coverage (or equivalent medical benefits after the termination of COBRA) available for at least 18 months following termination and (B) the Company shall pay the COBRA premiums necessary to continue Executive's medical insurance coverage in effect on the termination date for a period of twelve (12) months following Executive's termination (provided that such COBRA continuation and reimbursement shall terminate upon commencement of new employment by an employer that offers health care coverage to its employees); and (iii) a pro-rata portion of the Executive's annual target bonus for the calendar year in which the termination occurs.

Notwithstanding the foregoing, all Severance Payments under this Agreement are conditional on Executive (i) complying with the provisions of Section 7 below, and (ii) delivering prior to receipt of such severance payments, an effective, general release of claims in favor of the Company or its successor, its subsidiaries and their respective directors, officers and stockholders in the form attached hereto as Exhibit 1.

In the event that the Company determines that any severance benefit provided hereunder fails to satisfy the distribution requirement of Section 409A(a)(2)(A) of the Internal Revenue

Code (“Code”) as a result of Section 409A(a)(2)(B)(i) of the Code, then if an accelerated payment of such benefits would cause such benefit not to be subject to the provisions of Section 409A(a)(1) of the Code, the payment of such benefits shall be accelerated to the minimum extent necessary so that the benefit is not subject to the provisions of Section 409A(a)(1) of the Code. (The payment schedule as revised after the application of the preceding sentence shall be referred to as the “Revised Payment Schedule.”) However, in the event the accelerated payment of such benefits would not avoid the application of Section 409A(a)(1) of the Code, the payment of such benefits shall not be made pursuant to the original payment schedule or the Revised Payment Schedule and instead the payment of such benefits shall be delayed to the minimum extent necessary so that such benefits are not subject to the provisions of Section 409A(a)(1) of the Code. The Board may attach conditions to or adjust the amounts paid pursuant to this Section 6 (b) (iv) to preserve, as closely as possible, the economic consequences that would have applied in the absence of this Section 6 (b) (iv) ; provided, however, that no such condition or adjustment shall result in the payments being subject to Section 409A(a)(1) of the Code.

Notwithstanding any other provisions in this Agreement, it shall be a prerequisite of any termination by Executive for Good Reason that Executive shall have given the Company written notice within sixty (60) days following the date Executive becomes aware of the event or events giving rise to Good Reason, specifying in reasonable detail the nature and circumstances of such Good Reason, and giving the Company thirty (30) days to cure any such Good Reason prior to any such termination, and if uncured, the termination for Good Reason must occur within ninety (90) days of the end of such cure period.

(c) Termination for Cause of Resignation. The Company may terminate Executive’s employment at any time for Cause, and Executive may voluntarily resign at any time. Termination for Cause shall be effective on the date the Company gives notice to Executive of such termination in accordance with this Agreement. Voluntary resignation by Executive shall be effective on the date Executive gives notice to the Company of such resignation in accordance with this Agreement. In the event of the Company’s termination of Executive’s employment for Cause or Executive’s voluntary resignation from Executive’s employment, Executive will not be entitled to any further compensation from the Company, including severance pay, pay in lieu of notice or any other such compensation (other than accrued salary and bonus, and accrued and unused vacation, through Executive’s last day of employment).

(d) Cause. For purposes of this Agreement, “Cause” shall mean (i) Executive’s failure to substantially perform Executive’s duties under this Agreement for reasons other than death or Disability, which failure, if curable, is not cured to the reasonable satisfaction of the Board during the thirty (30) day period following written notice of such failure from the Company; (ii) Executive’s material failure or refusal to comply with reasonable written policies, standards and regulations established by the Company from time to time which failure, if curable, is not cured to the reasonable satisfaction of the Board during the thirty (30) day period following written notice of such failure from the Company; (iii) the proven commission by Executive of (x) an act of dishonesty or constituting common law fraud, embezzlement or a felony or (y) any tortious act, unlawful act or malfeasance that causes material harm to the Company’s standing, condition or reputation; or (iv) any material breach by Executive of the provisions of this Agreement, which breach, if curable, is not cured to the reasonable satisfaction

of the Board during the thirty (30) day period following written notice of such breach from the Company. The Board (excluding Executive if Executive is at such time a member of the Board) shall in good faith make all determinations relating to termination, including without limitation any determination regarding Cause, pursuant to this Section 6(d).

(e) Good Reason. For purposes of this Agreement, termination for “Good Reason” shall mean the termination by Executive of his employment following the existence of one or more of the following events arising without the consent of the Executive: (i) a material diminution in the Executive’s position, duties or responsibilities from those held by or assigned to the Executive as of the Effective Date, (ii) a reduction of the Executive’s Base Salary, (iii) a material reduction of the Executive’s benefits or bonus/incentive compensation opportunities provided to the Executive as then in effect, so long as he is the only executive to suffer such a reduction, or (iv) a change in the geographic location where Executive is required to perform services for the Company that is beyond a fifty (50) mile radius of the Executive’s then current residence, except for required travel on the Company’s business to an extent substantially consistent with his business travel obligations under Section 2 above.

(f) Removal from any Boards and Positions. Notwithstanding any other provision in this Agreement, if Executive’s employment is terminated under this Agreement for any reason, Executive shall be deemed to resign (i) from the Board or board of directors of any affiliate of the Company or any other board to which he has been appointed by or on behalf of the Company, and (ii) from any position with the Company or any affiliate of the Company, including, but not limited to, as an officer of the Company or any of its affiliates.

7. Company Matters; Restrictive Covenants.

(a) Confidential Information. Executive will have access to and will participate in the development of and will be acquainted with confidential or proprietary information and trade secrets related to the business of the Company and its affiliates, including but not limited to (i) customer lists; related records and compilations of information; the identity, lists or descriptions of any new customers, referral sources or organizations; financial statements; cost reports or other financial information; contract proposals or bidding information; business plans; training and operations methods and manuals; personnel records; software programs; reports and correspondence; and management systems, policies or procedures, including related forms and manuals, (ii) information pertaining to future developments such as future marketing or acquisition plans or ideas and potential new business locations; and (iii) all other tangible and intangible property and intellectual property which is used in the business and operations of the Company and its affiliates but not made public. The foregoing is collectively referred to as the “Confidential Information.” The term Confidential Information shall not include any information (x) that is or becomes generally publicly available (other than as a result of violation of this Agreement by Executive), or (y) that Executive receives on a non-confidential basis from a source (other than the Company, its affiliates or their representatives) that is not known by

Executive to be bound by an obligation of secrecy or confidentiality to any of the Company or its affiliates. Executive shall not disclose, use or make known for Executive’s or another’s benefit other than for the benefit of the Company and its affiliates any Confidential Information or use such Confidential Information in any way. Upon the termination of

Executive's employment with the Company for any reason, Executive shall immediately return to the Company all Confidential Information in whatever form maintained (including, without limitation, computer discs and other electronic media).

(b) Non-Competition. During Executive's employment with the Company and for the one-year period immediately following the termination of Executive's employment with the Company, Executive will not directly or indirectly (whether as an officer, director, stockholder, partner, proprietor, associate, representative, consultant or in any capacity whatsoever) engage in, become financially interested in, be employed by or have any business connection with any person, corporation, firm, partnership or any other entity whatsoever which is engaged in the development or sale of therapies for anemia or HIF-PH (hypoxia inducible factor prolyl hydroxylase) inhibitors or human protein tyrosine phosphatase beta (HPTPβ) inhibitors or Tie2 stabilizers. Notwithstanding the foregoing, the ownership by Executive of not more than three percent (3%) of the shares of stock of any corporation having a class of equity securities actively traded on a national securities exchange or on the Nasdaq Stock Market shall not be deemed to violate the prohibitions of this Section 7(b).

(c) Nonsolicitation of Customers. During Executive's employment with the Company and for the one-year period immediately following the termination of Executive's employment with the Company, Executive shall not solicit, directly or indirectly, any customers or prospective customers of the Company with whom Executive had contact on behalf of the Company during Executive's employment with the Company.

(d) Nonsolicitation of Employees. During Executive's employment with the Company and for the one-year period immediately following the termination of Executive's employment with the Company, Executive shall not solicit or hire, directly or indirectly, on Executive's behalf or on behalf of any other person or entity, any person employed by the Company except with the specific written consent of the Company.

(e) Certain Representations. Executive represents that Executive's experience, capabilities and circumstances are such that the provisions of this Agreement will not prevent Executive from earning a livelihood. Executive further agrees that the limitations set forth in this Agreement (including, without limitation, the time and territorial limitations) are reasonable and properly required for the adequate protection of the current and future businesses of the Company. Executive further acknowledges that a remedy at law for any breach or threatened breach of the provisions of this Agreement would be inadequate and will cause immediate and irreparable harm to the Company in a manner that cannot be measured nor adequately compensated in damages. Executive further acknowledges that in the event of any such breach and in addition to any and all other remedies that it may have at law or in equity, the Company shall be entitled to seek temporary, preliminary and permanent injunctive relief to restrain such breach by Executive, and the prevailing party in any such proceeding shall be entitled to recover all associated costs and expenses, including reasonable attorneys' fees, from the non-prevailing party. Nothing contained herein shall restrict or limit in any manner the Company's right to seek and obtain any form of relief, legal or equitable, against Executive in an action brought to enforce its rights hereunder.

(f) Intellectual Property. Except as otherwise set forth and described on Appendix A Section 7(f), hereto, all ideas, concepts, inventions, improvements, programs, information technology, derivative works, processes, configurations, data, procedures, designs, techniques and other works of authorship and development made, conceived or reduced to practice by Executive, either solely or in collaboration with others, during Executive's employment with the Company, including but not limited to all copyright, trademark, patent, trade secret and intellectual property rights associated therewith, shall become and remain the exclusive property of the Company. Executive hereby assigns to the Company any and all of Executive's right, title and interest in and to any of the foregoing, and Executive waives any claim that Executive may have thereto. Executive will promptly disclose in writing to the Company all such ideas, concepts, inventions, improvements, programs, information technology, derivative works, processes, configurations, data, procedures, designs, techniques and other works of authorship and development, and will cooperate fully with the Company in confirming and protecting the Company's ownership rights therein. The work product resulting from the Executive's employment with the Company is work made for hire.

(g) Ventures. If, during the Term, Executive is engaged in or associated with the planning or implementing of projects, programs or ventures involving the Company and third parties, all rights in such projects, programs and ventures shall belong to the Company (or the third party, to the extent provided in any agreement between the Company and the third party). Except as formally approved by the Company, Executive shall not be entitled to any interest in such project, program or venture or to any commission, finder's fee or other compensation in connection therewith other than the salary or other compensation to be paid to Executive as provided in this Agreement.

8. Miscellaneous.

(a) Withholding Taxes. The Company may withhold from all salary, bonus or other benefits payable under this Agreement all federal, state, city or other taxes as shall be required pursuant to any law or governmental regulation or ruling.

(b) Entire Agreement; Binding Effect. This Agreement sets forth the entire understanding between the parties as to the subject matter of this Agreement and supersedes all prior agreements, commitments, representations, writings and discussions between them (whether written or oral) on the subject matters herein; and neither of the parties shall be bound by any obligations, conditions, warranties or representations with respect to the subject matter of the foregoing except as expressly provided herein or therein or as duly set forth on or subsequent to the date hereof in a written instrument signed by the proper and fully authorized representative of the party to be bound hereby. This Agreement is binding on Executive and on the Company and Executive and their respective successors and assigns (whether by assignment, by operation of law or otherwise); provided that neither this Agreement nor any rights or obligations hereunder may be assigned by Executive or the Company without the prior written consent of the other party (except that the Company shall be entitled to assign this Agreement in connection with the sale of all or substantially all of the Company's assets, or a merger or consolidation in which the Company is not the surviving entity).

(c) Absence of Conflict. Executive represents and warrants that Executive's employment by the Company as described herein will not conflict with and will not be constrained by any prior employment or consulting agreement or relationship.

(d) Voluntary Nature of Agreement; Legal Rights. Executive is executing this Agreement voluntarily and without any duress or undue influence by the Company or anyone else. Executive acknowledges that Executive has had the opportunity to consult with an attorney regarding the provisions of this Agreement and has either obtained such advice of counsel or knowingly waived the opportunity to seek such advice. Executive has carefully read this Agreement and has asked any questions needed for Executive to understand the terms, consequences and binding effect of this Agreement and fully understands it.

(e) Waivers. No party shall be deemed to have waived any right, power or privilege under this Agreement or any provisions hereof unless such waiver shall have been duly executed in writing and acknowledged by the party to be charged with such waiver. The failure of any party at any time to insist on performance of any of the provisions of this Agreement shall in no way be construed to be a waiver of such provisions, nor in any way to affect the validity of this Agreement or any part hereof. No waiver of any breach of this Agreement shall be held to be a waiver of any other subsequent breach.

(f) Notices. All notices, approvals, consents, requests or demands required or permitted to be given under this Agreement shall be in writing and shall be deemed sufficiently given (i) upon delivery, if delivered by hand (ii) one business day after transmission, if sent by facsimile (confirmation received) or (iii) one business day after the business day of deposit with a reputable overnight courier for next business day delivery, freight prepaid (signature of receipt obtained). Notice in each case shall be addressed to the party entitled to receive such notice at the following address (or other such addresses as the parties may subsequently designate):

If to the Company:

Akebia Therapeutics, Inc.
Attn: President
9987 Carver Road, Suite 420
Cincinnati, Ohio 45242
Fax: (513) 985-1920

If to the Executive:

Jason Amello
1700 Beacon Street
Newton, MA 02468

With copies of all notices also to go to Executive's counsel, as follows:

Lawrence J. Casey
Shilepsky Hartley Robb Casey Michon
155 Seaport Boulevard, 11th Floor
Boston, MA 02210-2698
Email: lcasev@shilepsky.com
Fax: (617) 447-2800

With copies of all notices also to go to Company counsel and the Chair of the Company's Compensation Committee, as follows:

Thompson Hine LLP
312 Walnut Street, Suite 1400
Cincinnati, Ohio 45202
Attn: David J. Willbrand, Esq.
Fax: (513)241-4771

Akebia Therapeutics, Inc.
Attn: Chair of Compensation Committee
9987 Carver Road, Suite 420
Cincinnati, Ohio 45242
Fax:(513)985-1920

(g) Governing Law. This Agreement shall be governed by, and construed and enforced in accordance with, the laws of the Commonwealth of Massachusetts without regard to conflict of law principles that would result in the application of any law other than the law of the Commonwealth of Massachusetts.

(h) Severability. Every provision of this Agreement is intended to be severable from every other provision of this Agreement. If any provision of this Agreement is held to be invalid, illegal or unenforceable, in whole or in part, such invalidity, illegality or unenforceability shall not affect the other provisions of this Agreement; and this Agreement shall be construed as if such invalid, illegal or unenforceable provision had never been contained herein except to the extent that such provision may be construed and modified so as to render it valid, lawful, and enforceable in a manner consistent with the intent of the parties to the extent compatible with the applicable law as it shall then appear.

(i) Effect of Headings. The Section and subsection headings contained herein are for convenience only and shall not affect the construction hereof.

(j) Counterparts. This Agreement may be executed in multiple counterparts, each of which shall be deemed to be an original, and all such counterparts shall constitute but one instrument

Signature Page Follows

IN WITNESS WHEREOF, the parties hereto have caused this Agreement to be duly executed as of the year and date first written above.

AKEBIA THERAPEUTICS, INC.

By: /s/ John P. Butler

Name: John P. Butler

Its: CEO

Executive:

By: /s/ Jason Amello

Jason Amello

Appendix A

(Section 1)

- (1) Participating on boards of directors of public or privately held companies subject to prior written approval of the Board.
- (2) Serving on any committees, boards, or organizations associated with Public Accountancy.

EXHIBIT 1

“Release”

Executive, on behalf of himself and anyone claiming by, through, or under him (including, but not limited to, his heirs, executors, administrators, attorneys, successors, assigns and agents), fully settles, releases and forever discharges the Company, and its present and former affiliates, related persons, parents, subsidiaries, predecessors, partners, principals, officers, directors, stockholders, managers, members, agents, representatives, attorneys, insurers, successors and assigns of and from any and all past or, present liability, claims, rights, demands, obligations, controversies, damages, costs, expenses (including reasonable attorneys' fees), actions, causes of actions or compensation of any nature whatsoever whether known or unknown as of the date of execution of this Release, arising directly or indirectly, up to and including the day hereof, out of or related to the engagement of Executive by the Company, the performance of services by Executive for or on behalf of the Company, or the termination of Executive's engagement by the Company, including, but not limited to, any claims which have been or could have been brought for discrimination under federal, state or local law, as well as any claims or causes of action under any law dealing with employment torts, intentional torts, ERISA, wrongful discharge, retaliation, breach of contract, implied contract, promissory estoppel, wage and hour violations, violation of public policy or personal injury, as well as claims for wages, overtime pay, vacation pay, commissions, bonuses, profit sharing, expenses, benefits, termination pay, severance pay, reasonable notice or pay in lieu of such notice.

This Release does not include a release of any rights and claims to any benefits to which Executive might be entitled under the terms of any employee benefit plan maintained by the Company in which Executive is a participant. This Release also does not include a release or waiver of any rights or claims Executive has, or might subsequently have, in Executive's capacity as a stockholder of the Company. In addition, this Release shall not release the Company from its continuing obligation to honor the terms of the Employment Agreement. However, this Release shall remain in full force and effect regardless of any claim by Executive that the Company failed to honor the terms of the Employment Agreement. In the event of any such dispute, Executive's sole remedy against the Company shall be to enforce the terms of the Employment Agreement. Executive is also not waiving, and nothing in this Release is intended to waive, any right to defense and indemnification from the Company or its insurers or coverage under any directors and officers insurance coverage, if any, provided by the Company, to which Executive might be entitled. Executive is also not waiving, and nothing in this Release is intended to waive, any claims Executive may have for unemployment insurance or workers' compensation benefits, state disability compensation, claims for any vested benefits under any Company-sponsored benefit plan, or any claims that, as a matter of law, may not be released by private agreement.

Notwithstanding this release of claims, the Company shall provide for the defense and, if applicable, satisfaction of any judgments rendered by any liability, claims, rights, demands, obligations, controversies, damages, costs, expenses (including reasonable attorneys' fees), actions, causes of actions or compensation of any nature whatsoever asserted by a third party against Executive related to or arising out of the Employment Agreement, whether known or unknown as of the date of execution of this Release.

The Company, on behalf of itself or anyone claiming by, through, or under it (including, but not limited to, its present and former affiliates, related persons, parents, subsidiaries, predecessors, partners, principals, officers, directors, stockholders, managers, members, agents, representatives, attorneys, insurers, successors or assigns) fully settles, releases and forever discharges Executive, and his heirs, executors, administrators, attorneys, successors, assigns and agents, of and from any and all past, present or future liability, claims, rights, demands, obligations, controversies, damages, costs, expenses (including reasonable attorneys' fees), actions, causes of actions or compensation of any nature whatsoever, arising directly or indirectly, up to and including the day hereof, out of or related to the engagement of Executive by the Company, the performance of services by Executive for or on behalf of the Company, or the termination of Executive's engagement by the Company.

Neither Executive, on the one hand, nor the any of the Company's officers or directors, on the other hand, shall make any statements or remarks which are disparaging toward the other.

This release does not constitute an admission of liability by any party hereto, and such liability is expressly denied by those released.

November 13, 2013

Nicole R. Hadas
20 Pilgrim Drive
Winchester, Massachusetts 01890

Dear Nicole:

It is my pleasure to extend the following offer of employment to you on behalf of Akebia Therapeutics, Inc. ("Akebia" or "the Company"). Akebia is proud of its achievements to date and we are looking to individuals such as yourself to play a supporting role in advancing our exciting drug products. We are convinced that you can make an immediate impact, and will be a productive member of our team.

This offer letter confirms to you Akebia's offer of employment (the "Offer Letter"). The terms of our offer are as follows:

1. **Position:** The Vice President, General Counsel and Corporate Secretary position is a full-time, salaried position reporting to Mr. John P. Butler, President and Chief Executive Officer. The primary responsibilities will be to lead all legal activities for the company.
2. **Start Date:** Subject to satisfactory background and reference checks, your employment will begin on a date that is mutually agreeable between you and the Company.
3. **Compensation:** Your compensation will include the following:
 - (i) A base salary of \$290,000.00 per year, paid in accordance with the Company's normal payroll practices.
 - (ii) Additional incentive compensation, which includes:
 - (a) An annual bonus in an amount up to 20% of your base salary. The exact amount of the actual bonus awarded for any given year, if any, shall be determined by your manager and the Board of Directors based upon their consideration of the Company's performance and your performance against objectives, and paid in accordance with the Company's bonus plan; and
 - (b) Subject to approval by the Company's Board of Directors and in accordance with the Company's Amended and Restated 2008 Equity Incentive Plan, as amended (the "Option Plan"), options to acquire shares of the Company's common stock representing approximately 0.50% of the Company's common stock outstanding on a fully-diluted and as-converted basis.
4. **Performance/Salary Reviews:** The current policy, subject to change without notice, is that reviews are conducted on an annual basis. See the Akebia Employee Handbook for more details regarding policies and expectations for employees of Akebia.

5. **Benefits:** As a full time employee you will be entitled to participate in all applicable benefit programs as currently, or prospectively, offered by Akebia. The Company is committed to providing comprehensive and competitive benefits to its employees. The Company has a plan to provide for health care and dental insurance and as an employee you and your family will be eligible to join those plans. The Company also provides a 401K plan allowing employees to place pre-tax dollars in a retirement account up to the maximum permitted by law. We will provide more detailed information regarding additions to our benefit plans when these modifications are completed. The Company will provide twenty (20) days of paid time off (PTO) per calendar year, plus holidays, as defined and allocated in the Akebia Employee Handbook. We will provide more detailed information regarding these plans and any future additions/modifications to our benefit plans when available. In addition to our "hard" benefits we offer a host of "soft" benefits such as a fun, flexible and stimulating work environment and the rewards of developing important new medicines.
6. **Taxes:** All forms of compensation referred to in this Offer Letter are subject to reduction to reflect applicable withholding and payroll taxes and other deductions required by law. You hereby acknowledge that the Company does not have a duty to design its compensation policies in a manner that minimizes your tax liabilities, and you will not make any claim against the Company or its Board of Directors related to tax liabilities arising from your compensation.
7. **Contingencies:** Additionally, this offer is contingent upon your submission of appropriate documentation for verification purposes in order that the Company may be in compliance with the Immigration and Reform Control Act of 1986, as amended.
8. **Employment at Will:** The parties hereto recognize that this offer of employment is not intended to create a contract of employment and both you and Akebia retain the right to terminate the employment relationship at any time without cause. However, if you are terminated by the Company without Cause (as defined below) or if you resign from the Company for Good Reason (as defined below), you shall be entitled to receive the following severance benefits, in addition to accrued salary and bonus and accrued and unused vacation through your last day of employment:
 - (i) Severance pay in the form of continuation of your base salary in effect on the effective date of termination for a period of six (6) months after the date of such termination, to be paid periodically in accordance with the Company's normal payroll practices and subject to standard payroll deductions and withholdings;
 - (ii) If you timely elect continued coverage under COBRA, then (A) the Company shall make such COBRA coverage available for at least 18 months following termination and (B) the Company shall pay the COBRA premiums necessary to continue your medical and/or dental insurance coverage in effect on the termination date for a period of six (6) months following your termination (provided that such COBRA reimbursement shall terminate upon commencement of new employment by an employer that offers health care coverage to its employees); and

- (iii) In the event of a sale of the company and you are terminated by the acquirer or you resign for Good Reason then, in addition to the severance benefits in Sections 8(i) and 8(ii) hereof, your unvested options as of the effective date of your termination or resignation for Good Reason (as the case may be) shall become fully vested; *provided, however*, that in the event of a conflict between the terms of this Section 8(iii), the terms of the Option Plan, and/or the terms of your Equity Award Agreement, the terms most favorable to you shall govern.
 - (iv) For purposes of this Offer Letter, "Good Reason" shall mean the occurrence of any of the following without your consent: (a) a material diminution in your position, duties or responsibilities, (b) a reduction of your base salary, (c) a material reduction of your benefits or bonus/incentive compensation opportunities, so long as you are the only employee to suffer such a reduction, or (d) relocation of the Akebia corporate office to a location more than fifty (50) miles from Cambridge, Massachusetts.
 - (v) For purposes of this Offer Letter, "Cause" shall mean: (a) your failure to substantially perform your duties under this Agreement for reasons other than death or disability, which failure, if curable, is not cured to the reasonable satisfaction of the Chief Executive Officer during the thirty (30) day period following written notice of such failure from the Company; (b) your material failure or refusal to comply with reasonable written policies, standards and regulations established by the Company from time to time which failure, if curable, is not cured to the reasonable satisfaction of the Chief Executive Officer during the thirty (30) day period following written notice of such failure from the Company; (c) the proven commission by you of (x) an act of dishonesty or constituting common law fraud, embezzlement or a felony or (y) any tortious act, unlawful act or malfeasance that causes material harm to the Company's standing, condition or reputation; or (d) any material breach by you of the provisions of this Agreement, which breach, if curable, is not cured to the reasonable satisfaction of the Chief Executive Officer during the thirty (30) day period following written notice of such breach from the Company. The Company, through the Chief Executive Officer, shall in good faith make all determinations relating to termination including, without limitation, any determination regarding Cause, pursuant to this Section 8(v).
9. **Covenants, Company Matters - Confidentiality and Assignment of Rights:** As a condition of your employment, you will execute an Employee Agreement containing standard confidentiality and invention assignment provisions.
10. **Interpretation, Amendment and Enforcement:** This Offer Letter, together with the Employee Agreement, constitutes the complete agreement between you and the Company, contain all of the terms of your employment with the Company and supersede any prior agreements, representations or understandings (whether written, oral or implied) between you and the Company. The terms of this Offer Letter and the resolution of any disputes as to the meaning, effect, performance or validity of this Offer Letter or arising out of, related to, or in any way connected with, this Offer Letter, your

employment with the Company or any other relationship between you and the Company (the "Disputes") will be governed by Massachusetts law excluding laws relating to conflicts or choice of law. You and the Company submit to the exclusive personal jurisdiction of the federal and state courts located in the Commonwealth of Massachusetts in connection with any Dispute or any claim related to any Dispute.

To indicate acceptance, please sign and return a copy of this letter (via fax, pdf or regular mail).

Nikki, we look forward to you joining Akebia. Teamwork, quality people, and a business focus, are all critical to Akebia's future success. We are confident that you will play an important role in our success over the coming year and well into the future.

Very truly yours,

Akebia Therapeutics, Inc.

By: /s/ John P. Butler

Name: John P. Butler

Title: President and CEO

cc: J. Amello

Response requested by November 20, 2013

The undersigned accepts the above employment offer and agrees that it contains the terms of employment with Akebia Therapeutics, Inc. By accepting this offer of employment, the undersigned is acknowledging that no prior employment obligations or other contractual restrictions exist which preclude employment with Akebia Therapeutics, Inc. It is further understood that this offer is confidential and disclosure of any of the terms and conditions contained herein constitute grounds for termination of employment or withdrawal of this offer.

Accepted:

/s/ Nicole R. Hadas

Nicole R. Hadas

Date: November 19, 2013

EXECUTIVE EMPLOYMENT AGREEMENT

THIS EXECUTIVE EMPLOYMENT AGREEMENT (this "Agreement") is made as of April 6, 2011 (the "Effective Date") by and between Akebia Therapeutics, Inc., a Delaware corporation (the "Company"), and Dr. Robert Shalwitz, an individual resident in the State of Ohio ("Executive").

1. Employment; Duties; Full Time Employment.

(a) General. The Company hereby agrees to continue to employ Executive, and Executive hereby accepts continued employment, as Chief Medical Officer and Vice President of the Company. The Company and Executive acknowledge and agree that Executive's employment commenced on May 2, 2007 (the "Commencement Date"). In such capacity, Executive shall perform such executive duties and exercise such powers for the Company and its subsidiaries as the Board of Directors of the Company (the "Board") may lawfully assign to or vest in Executive from time to time. Executive covenants and agrees that, at all times during the Term (as defined below), Executive shall devote Executive's full business time and efforts to Executive's duties as an employee of the Company and that Executive will not, directly or indirectly, engage or participate in any other business or professional activities during the Term, other than non-conflicting personal investments managed on Executive's personal time and activities for non-profit institutions, provided that such activities do not interfere or conflict with Executive's obligations hereunder.

(b) Location. Executive acknowledges and agrees that the Company is currently located in Cincinnati, however the Company will shortly be evaluating whether the Company should relocate to (or co-locate in) Boston or San Francisco. Subject to the balance of this subsection, Executive is expected to spend a minimum of two (2) working days a week (exclusive of travel time) in Cincinnati or, Boston or San Francisco (if the Company does relocate or co-locate to Boston or San Francisco or other relevant city), as appropriate, to perform his services for the Company. However, for any week that Executive is already traveling for Company business to a destination other than where the Company is then located, the requirement in the previous sentence shall not apply. If the Company adopts a plan for relocation or co-location as provided in this Section 1(b), and Executive is selected as a candidate for such relocation or co-location, then (i) Executive shall relocate or co-locate in accordance with the plan, and (ii) subject to Section 4 (*Business Expenses*) and to the foregoing plan, the Company shall pay the out-of-pocket costs of renting an apartment in support of the plan (the selection of which apartment shall be subject to the Company's prior written consent) and shall reimburse reasonable out-of-pocket travel costs directly related to the relocation and/or co-location. The weekly schedule in effect from time-to-time can be adjusted with the verbal agreement of the CEO based on business needs and the need for business related travel to various destinations. Notwithstanding the foregoing, Executive's obligation under this Section 1(b) to relocate (or co-locate) shall expire on the first anniversary of the date hereof unless by then the Company has adopted a relocation or co-location plan.

2. Term. The Company agrees to continue to employ Executive, and Executive agrees to continue to serve the Company, on an "*at will*" basis, which means that, subject to the payment obligations imposed on the Company pursuant to this Agreement, either the Company

or Executive may terminate Executive's employment with the Company at any time, with or without Cause, as provided in Section 5 below. The period commencing with the Commencement Date and ending on the effective date of any termination of employment in accordance with the provisions hereof shall constitute the term of this Agreement (the "Term").

3. Compensation and Benefits. From and after the Effective Date, the Company shall provide Executive with the following compensation and benefits:

(a) Base Salary. The Company shall pay Executive a base salary ("Base Salary") at the rate of \$269,280 per annum (less applicable deductions and withholdings), payable in periodic payments in accordance with the Company's normal payroll practices. During the Term, Executive's compensation shall be reviewed by the Board from time to time and at least once every 12 months. Any increase or decrease in Base Salary (together with the then existing Base Salary) shall serve as the "Base Salary" under this Agreement.

(b) Discretionary Bonuses. The Executive shall be eligible to participate in all bonus or similar incentive plans adopted by the Board including, without limitation, an incentive compensation plan with a yearly performance based cash bonus of up to a maximum of 20% of the Executive's Base Salary (the "Target Bonus"). Under the current incentive compensation plan, the Board reviews the Company's and the Executive's performance for the 12 months ending June 30th and pays any bonus to be awarded during the month of July. Executive will not accrue or earn any bonus (including a prorated bonus) for any bonus evaluation period if Executive's employment terminates for any reason before the end of such period. See also Section 5 for rules relating to payment of bonus following termination.

(c) Stock Ownership. The provisions of this Agreement shall not alter or modify the terms (including, without limitation, any vesting provisions) of any stock options or restricted stock granted to the Executive by the Company or which the Executive has purchased from the Company in connection with the Company's fundraising activities, which shall be governed in all respects by the equity compensation or other related plan pursuant to which such options/restricted stock are granted and the respective stock option/restricted stock agreement(s) evidencing the same or, as applicable, the investment/stock purchase documents under which such stock was acquired (the "Governing Documents"). Except for the above, any rights afforded the Executive as part of a purchase of Preferred Stock, and except as otherwise may be determined by the Board from time to time after the date of this Agreement in its discretion, Executive shall not have any right to be issued shares of the Company's capital stock or options, warrants or other rights to acquire any capital stock of the Company.

(d) Other Compensation and Benefits. In addition to the compensation specified above in this Section 3, Executive shall be entitled to the following benefits during the Term, all on the terms offered or maintained by the Company to, for or on behalf of its senior executives: vacation (on the terms in 3(e) below), holidays and sick leave, and subject to eligibility therefor, the right to participate in any profit sharing plan, retirement plan, 401(k) plan, group medical plan, group dental plan, and/or other health or insurance plan maintained by the Company for its senior executives generally and, if applicable, their family members. Executive will also be eligible for the paid holidays as are generally made available to employees of the Company. The Company agrees to pay 100% of the premium associated with the Executive's and 50% of the premium associated with the Executive's spouse's/eligible dependents' participation in the Company-sponsored group medical and dental insurance plan.

(e) Vacation. The Executive shall be entitled to four weeks paid vacation per calendar year to be taken at such times as may be approved by the Board (which is more than the normal amount defined in the Akebia Employee Handbook). An aggregate of up to 1 week of unused vacation time may be carried over at the end of a calendar year. Upon termination of the Executive's employment, the Company will pay the Executive for unused vacation at the Executive's Base Salary rate (subject to normal deductions and withholding amounts) on the next regularly scheduled pay date immediately following the termination date.

4. Business Expenses. The Company shall reimburse Executive for all reasonable and necessary business and travel expenses incurred by Executive in the performance of Executive's duties under this Agreement. Such expenses shall be reimbursed in accordance with the Company's guidelines, limits and procedures relating thereto and upon presentation of proper expense vouchers or receipts therefor.

5. Termination. This Agreement and the Executive's employment by the Company shall or may be terminated, as the case may be, as set forth in this Section 5. See also Section 2.

Upon termination of Executive's employment with the Company, all salary and bonuses that have accrued through the date of termination plus an amount equal to Executive's unused vacation through the termination date shall be paid (subject to normal deductions and withholding amounts) on the next regularly scheduled payroll date of the Company following the date of termination. See Section 3(b) regarding when bonuses are considered to have been accrued.

Subject to Executive's (i) complying with the provisions of Section 6 below, and (ii) delivering to the Company prior to the 53rd day after the date of termination an effective, general release of claims in favor of the Company or its successor, its subsidiaries and their respective directors, officers and stockholders in a form acceptable to the Company or its successor, the Executive may also be entitled to receive additional compensation as set forth below.

(a) Termination on Death or Disability. The Term will terminate automatically and immediately upon Executive's death or, upon 30 days prior written notice from the Company, in the event of Executive's Disability. For purposes of this Section 5, Executive shall be deemed to be under a "Disability" when a qualified medical doctor mutually acceptable to the Company and the Executive or the Executive's personal representative shall have certified in writing that: (A) the Executive is unable, because of a medically determinable physical or mental disability, to substantially perform all of the Executive's duties, with or without a reasonable accommodation, for more than an aggregate of one hundred and eighty (180) calendar days in any 365 day period; or (B) by reason of mental or physical disability, it is unlikely that the Executive will be able, within an aggregate one hundred and eighty (180) calendar days within any 365 day period, to substantially resume all business duties and responsibilities in which the Executive was previously engaged and otherwise discharge the Executive's duties under this Agreement.

(b) Severance Benefits on Termination. The Executive's employment may be terminated at any time for any reason; provided, however, that if the Executive's employment is terminated (1) by the Company for any reason other than those listed in Sections 5(a) or 5(c) below; (2) by the Executive for "Good Reason" (as defined in Section 5(d) below); or (3) as a result of the events particularly described in Section 5(e) below, then the Executive, if he complies with the requirements of subsection (ii) of the third paragraph of Section 5 above (in the sentence commencing with the words "Subject to Executive's"), and the Executive has not revoked the release provided thereby, shall be entitled to receive the following severance benefits from the Company (or an acquiror/NewCo, if applicable) (collectively, the "Severance Benefits") (subject to applicable deductions and mandatory withholding, including federal, state and local income taxes, as well as FICA and other applicable withholding):

(i) Executive shall be entitled to severance in the form of continuation of Executive's Base Salary in effect on the effective date of termination for a period of six months, to be paid periodically in accordance with the Company's normal payroll practices and subject to standard payroll deductions and withholdings, commencing on the next regularly scheduled payroll date of the Company on or after the 61st day after the date of termination.

(ii) If Executive timely elects continued coverage under COBRA, then (A) the Company shall make such COBRA coverage (or equivalent medical benefits after the termination of COBRA) available for at least 24 months following termination and (B) the Company shall pay the COBRA premiums necessary to continue Executive's medical insurance coverage in effect on the termination date with respect to a period of six months following Executive's termination (provided that such COBRA continuation and reimbursement shall terminate upon commencement of new employment by an employer that offers health care coverage to its employees), with payment of the reimbursement to be made on the 61st day after the date of termination with respect to any such month that ends on or before such day and on the last day of each such month that ends after such day.

(iii) If Executive timely elects continued coverage, without regard to whether the Executive has delivered the release provided for in subsection (ii) of the second paragraph of Section 5 above, the Company will continue for a period of six months his Company-sponsored group insurance benefits (other than health insurance) that Executive participated in immediately prior to the termination date, if any, including without limitation life insurance (if then-applicable), accidental death & dismemberment insurance, long-term disability insurance, short-term disability insurance, supplemental disability insurance, and long-term care insurance (the "Benefits Payment").

(c) Termination by the Company for Cause. Notwithstanding any provision of this Agreement, in no event shall the Executive be entitled to receive any Severance Benefits hereunder in the event the Company elects to terminate the employment of the Executive for "cause," as hereinafter defined. For purposes of this Agreement, "cause" shall mean (i) Executive's failure to substantially perform Executive's duties under this Agreement for reasons other than death or Disability, which failure, if curable, is not cured to the reasonable satisfaction of the Board during the fifteen (15) day period following written notice of such failure from the Company; (ii) Executive's material failure or refusal to comply with reasonable written policies, standards and regulations established by the Company from time to time which

failure, if curable, is not cured to the reasonable satisfaction of the Board during the fifteen (15) day period following written notice of such failure from the Company; (iii) the commission by Executive of (x) an act of dishonesty or constituting common law fraud, embezzlement or a felony or (y) any tortious act, unlawful act or malfeasance that causes or reasonably would cause (for example, if it became publicly known) material harm to the Company's standing, condition or reputation; or (iv) any material breach by Executive of the provisions of this Agreement, which breach, if curable, is not cured to the reasonable satisfaction of the Board during the fifteen (15) day period following written notice of such breach from the Company. The Board (excluding Executive if Executive is at such time a member of the Board) shall make all determinations relating to termination, including without limitation any determination regarding cause, pursuant to this Section 5(c).

(d) Good Reason. For purposes of this Agreement, "Good Reason" shall mean any of the following without the consent of the Executive: (i) a material diminution in the Executive's position, duties or responsibilities from those held by or assigned to the Executive as of the Effective Date, (ii) a reduction of the Executive's Base Salary, or (iii) a material reduction of the Executive's benefits or bonus/incentive compensation opportunities provided to the Executive as then in effect, so long as he is the only executive to suffer such a reduction.

(e) Change of Control Termination. The Executive and the Company agree that the Executive shall be paid the Severance Benefits specified in Section 5(b)(i), (ii) and (iii) above if the Executive's employment terminates in connection with or within 6 months following a change of control (as described below) because either (i) the acquiror/NewCo does not offer the Executive employment on at least materially comparable compensation terms and benefits (including severance obligations) to those that are provided to the Executive pursuant to this Agreement; or (ii) such terms are initially offered and accepted, but within 6 months following the change of control the Executive's employment with the acquiror/NewCo is terminated as a result of any of the events described in Section 5(b). For purposes of this Agreement a change of control shall be deemed to have occurred upon a transfer (or license on an exclusive basis) of all or substantially all of the assets of the Company or the transfer of ownership of more than a majority of the securities of the Company, whether in a single transaction or series of separate transactions, other than in connection with fundraising activities of the Company, including without limitation a transaction in which a portion of the assets of the Company are transferred to an acquiror and the Company does not continue as a going concern during the 6 months thereafter, or its remaining assets are moved following such transfer to an acquiror to a NewCo (regardless of whether the NewCo stockholders are existing Company stockholders) and such Newco does not continue as a going concern during the 6 months after such transfer to such acquiror. If a party obtains an option to close a transaction, the transaction will not be considered as having occurred until such option is exercised and the transaction thereafter closed.

(f) Termination by Executive other than for Good Reason. If the Executive terminates his employment at any time for any reason other than for Good Reason, the Executive shall not be entitled to any of the Severance Benefits and shall provide the Company with 14 days notice of such termination.

6. Company Matters; Restrictive Covenants.

(a) Confidential Information. Executive will have access to and will participate in the development of and will be acquainted with confidential or proprietary information and trade secrets related to the business of the Company and its affiliates, including but not limited to (i) customer lists; related records and compilations of information; the identity, lists or descriptions of any customers, referral sources or organizations; financial statements; cost reports or other financial information; contract proposals or bidding information; business plans; training and operations methods and manuals; personnel records; software programs; reports and correspondence; and management systems, policies or procedures, including related forms and manuals, (ii) information pertaining to future developments such as future marketing or acquisition plans or ideas and potential new business locations; and (iii) all other tangible and intangible property and intellectual property which is used in the business and operations of the Company and its affiliates but not made public. The foregoing is collectively referred to as the "Confidential Information." The term Confidential Information shall not include any information (x) that is or becomes generally publicly available (other than as a result of violation of this Agreement by Executive), or (y) that Executive receives on a non-confidential basis from a source (other than the Company, its affiliates or their representatives) that is not known by Executive to be bound by an obligation of secrecy or confidentiality to any of the Company or its affiliates. Executive shall not disclose, use or make known any Confidential Information for Executive's or another's benefit or use such Confidential Information in any way other than for the benefit of and use on behalf of the Company and its affiliates. Upon the termination of Executive's employment with the Company for any reason, Executive shall immediately return to the Company all Confidential Information in whatever form maintained (including, without limitation, computer discs and other electronic media).

(b) Non-Competition. During Executive's employment with the Company and for the one-year period immediately following the termination of Executive's employment with the Company, Executive will not directly or indirectly (whether as an officer, director, stockholder, partner, proprietor, associate, representative, consultant or in any capacity whatsoever) engage in, become financially interested in, be employed by or have any business connection with any person, corporation, firm, partnership or any other entity whatsoever which competes with the Company in any area where the Company operates, or has operated at any time during Executive's employment with the Company, or any area the Company has planned to expand into at any time during Executive's employment with the Company.

(c) Nonsolicitation of Customers. During Executive's employment with the Company and for the one-year period immediately following the termination of Executive's employment with the Company, Executive shall not solicit, directly or indirectly, any customers of the Company or of its affiliates who or which were customers of the Company at any time during Executive's employment with the Company, nor shall Executive solicit any potential customers of the Company or of its affiliates with whom Executive had contact on behalf of the Company or its affiliates during Executive's employment with the Company.

(d) Nonsolicitation of Employees. During Executive's employment with the Company and for the one-year period immediately following the termination of Executive's

employment with the Company, Executive shall not solicit or hire, directly or indirectly, on Executive's behalf or on behalf of any other person or entity, any person employed by the Company or its affiliates during the prior six month period except with the specific written consent of the Company.

(e) Certain Representations. Executive represents that Executive's experience, capabilities and circumstances are such that the provisions of this Agreement will not prevent Executive from earning a livelihood. Executive further agrees that the limitations set forth in this Agreement (including, without limitation, the time and territorial limitations) are reasonable and properly required for the adequate protection of the current and future businesses of the Company and its affiliates. Executive further acknowledges that a remedy at law for any breach or threatened breach of the provisions of this Agreement would be inadequate and will cause immediate and irreparable harm to the Company or its affiliates in a manner that cannot be measured nor adequately compensated in damages. Executive further acknowledges that in the event of any such breach and in addition to any and all other remedies that it may have at law or in equity, the Company or its affiliates shall be entitled to temporary, preliminary and permanent injunctive relief to restrain such breach by Executive and to recover all costs and expenses, including reasonable attorneys' fees, of any proceedings brought to obtain such injunctive relief. Nothing contained herein shall restrict or limit in any manner the Company's right to seek and obtain any form of relief, legal or equitable, against Executive in an action brought to enforce its rights hereunder.

(f) Intellectual Property. All ideas, concepts, inventions, improvements, programs, information technology, derivative works, processes, configurations, data, procedures, designs, techniques and other works of authorship and development made, conceived or reduced to practice by Executive, either solely or in collaboration with others, during Executive's employment with the Company, including but not limited to all copyright, trademark, patent, trade secret and intellectual property rights associated therewith, shall become and remain the exclusive property of the Company. Executive hereby assigns to the Company any and all of Executive's right, title and interest in and to any of the foregoing, and Executive waives any claim that Executive may have thereto. Executive will promptly disclose in writing to the Company all such ideas, concepts, inventions, improvements, programs, information technology, derivative works, processes, configurations, data, procedures, designs, techniques and other works of authorship and development, and will cooperate fully with the Company in confirming and protecting the Company's ownership rights therein. The work product resulting from the Executive's employment with the Company is work made for hire. For clarity, however, nothing in this Agreement is intended to assign ownership to the Company of any of the foregoing that is developed by the Executive entirely on the Executive's own time without using the Company's equipment, supplies, facility or trade secret information and that does not relate to the Company's business or research or development, as conducted or as it might be conducted or formulated.

(g) Ventures. If, during the Term, Executive is engaged in or associated with the planning or implementing of projects, programs or ventures involving the Company and third parties, all rights in such projects, programs and ventures shall belong to the Company (or the third party, to the extent provided in any agreement between the Company and the third party). Except as formally approved by the Company, Executive shall not be entitled to any interest in

such project, program or venture or to any commission, finder's fee or other compensation in connection therewith other than the salary or other compensation to be paid to Executive as provided in this Agreement.

(h) Resignation on Termination. On termination of Executive's employment, Executive shall immediately (and with contemporaneous effect) resign any directorships, offices or other positions that Executive may hold in the Company or any of its affiliates, unless otherwise requested by the Board.

7. Miscellaneous.

(a) Withholding Taxes. The Company may withhold from all salary, bonus or other benefits payable under this Agreement all federal, state, city or other taxes as shall be required pursuant to any law or governmental regulation or ruling.

(b) Entire Agreement; Binding Effect. This Agreement sets forth the entire understanding between the parties as to the subject matter of this Agreement and supersedes all prior agreements, commitments, representations, writings and discussions between them (whether written or oral) on the subject matters herein, except for the Governing Documents. Neither of the parties shall be bound by any obligations, conditions, warranties or representations with respect to the subject matter of the foregoing except as expressly provided herein or therein or as duly set forth on or subsequent to the date hereof in a written instrument signed by the proper and fully authorized representative of the party to be bound hereby. This Agreement is binding on Executive and on the Company and their respective successors and assigns (whether by assignment, by operation of law or otherwise); provided that neither this Agreement nor any rights or obligations hereunder may be assigned by Executive or the Company without the prior written consent of the other party (except that the Company shall be entitled to assign this Agreement in connection with the sale of all or substantially all of the Company's assets or stock, or a merger or consolidation in which the Company is not the surviving entity). The Governing Documents pertaining to equity awards and stock acquisitions shall not be affected by this Agreement and shall continue in full force and effect.

(c) Absence of Conflict. Executive represents and warrants that Executive's employment by the Company as described herein will not conflict with and will not be constrained by any prior employment or consulting agreement or relationship.

(d) Voluntary Nature of Agreement; Legal Rights. Executive is executing this Agreement voluntarily and without any duress or undue influence by the Company or anyone else. Executive acknowledges that Executive has had the opportunity to consult with an attorney regarding the provisions of this Agreement and has either obtained such advice of counsel or knowingly waived the opportunity to seek such advice. Executive has carefully read this Agreement and has asked any questions needed for Executive to understand the terms, consequences and binding effect of this Agreement and fully understand it, including that Executive is waiving Executive's right to a jury trial.

(e) Waivers. No party shall be deemed to have waived any right, power or privilege under this Agreement or any provisions hereof unless such waiver shall have been duly

executed in writing and acknowledged by the party to be charged with such waiver. The failure of any party at any time to insist on performance of any of the provisions of this Agreement shall in no way be construed to be a waiver of such provisions, nor in any way to affect the validity of this Agreement or any part hereof. No waiver of any breach of this Agreement shall be held to be a waiver of any other subsequent breach.

(f) Notices. All notices, approvals, consents, requests or demands required or permitted to be given under this Agreement shall be in writing and shall be deemed sufficiently given (i) upon delivery, if delivered by hand (ii) one business day after transmission, if sent by facsimile (confirmation received) or (iii) one business day after the business day of deposit with a reputable overnight courier for next business day delivery, freight prepaid (signature of receipt obtained). Notice in each case shall be addressed to the party entitled to receive such notice at the following address (or other such addresses as the parties may subsequently designate):

The Company:

Akebia Therapeutics, Inc.
Attn: President
9987 Carver Road, Suite 420
Cincinnati, Ohio 45242
Fax: 513.985.1920

With copies to Company counsel and the Chair of the Company's Compensation Committee, as follows:

Thompson Hine LLP
312 Walnut Street, Suite 1400
Cincinnati, Ohio 45202
Attn: David J. Willbrand, Esq.
Fax: (513) 241-4771

Akebia Therapeutics, Inc.
Attn: Chair of Compensation Committee
9987 Carver Road, Suite 420
Cincinnati, Ohio 45242
Fax: 513.985.1920

Executive:

Robert Shalwitz, M.D.
2549 Bryden Rd
Bexley, OH 43209

(g) Governing Law. This Agreement shall be governed by, and construed and enforced in accordance with, the laws of Ohio as they apply to contracts entered into and wholly to be performed therein by residents thereof.

(h) Severability. Every provision of this Agreement is intended to be severable from every other provision of this Agreement. If any provision of this Agreement is held to be invalid, illegal or unenforceable, in whole or in part, such invalidity, illegality or unenforceability shall not affect the other provisions of this Agreement; and this Agreement shall be construed as if such invalid, illegal or unenforceable provision had never been contained herein except to the extent that such provision may be construed and modified so as to render it valid, lawful, and enforceable in a manner consistent with the intent of the parties to the extent compatible with the applicable law as it shall then appear.

(i) 409A Matters. This Agreement is intended to comply with the requirements of Section 409A of the Internal Revenue Code of 1986, as amended and the Treasury Regulations and other applicable guidance thereunder ("Section 409A"). To the extent that there is any ambiguity as to whether this Agreement (or any of its provisions) contravenes one or more requirements of Section 409A, such provision shall be interpreted and applied in a manner that does not result in a Section 409A violation. Without limiting the generality of the above:

(i) For clarity, the Severance Benefits are only payable upon a termination of the Executive's employment that constitutes a "separation from service" as defined in Section 409A, or the Executive's death.

(ii) The Severance Benefits shall be deemed to be series of separate payments, with each installment being treated as a separate payment.

(iii) The time and form of payment of any compensation may not be deferred or accelerated to the extent it would result in an impermissible acceleration or deferral under Section 409A.

(iv) To the extent this Agreement contains payments which are subject to Section 409A (as opposed to exempt from Section 409A), the Employee's rights to such payments are not subject to assignment, anticipation, alienation, sale, transfer, pledge, encumbrance, attachment or garnishment and, where applicable, may only be transferred by will or the laws of descent and distribution.

(v) If the Employee is a "specified employee" (as defined in Section 409A) on the termination date and a delayed payment is required by Section 409A to avoid a prohibited distribution under Section 409A, then no Severance Benefits that constitute "nonqualified deferred compensation" under Section 409A shall be paid until the earlier of (i) the first day of the 7th month following the date of Employee's "separation from service" as defined in Section 409A, or (ii) the date of Employee's death. Upon the expiration of the applicable deferral period, all payments deferred under this clause shall be paid in a lump sum and any remaining severance benefits shall be paid per the schedule specified in this Agreement.

(j) Effect of Headings. The Section and subsection headings contained herein are for convenience only and shall not affect the construction hereof.

(k) Counterparts. This Agreement may be executed in multiple counterparts, each of which shall be deemed to be an original, and all such counterparts shall constitute but one instrument.

Signature Page Follows

IN WITNESS WHEREOF, the parties hereto have caused this Agreement to be duly executed as of the year and date first written above.

AKEBIA THERAPEUTICS, INC.

By: /s/ Joseph Gardner

Name: Joseph Gardner

Title: President and Chief Executive Officer

EXECUTIVE

/s/ Robert Shalwitz, M.D.

Robert Shalwitz, M.D.

[Signature page to Shalwitz's Employment Agreement]



Phone: 513-985-1920

Fax: 513-985-0999

www.akebia.com

Akebia Therapeutics, Inc.

9987 Carver Road

Suite 420

Cincinnati, OH 45242

SENT VIA EMAIL

January 7, 2012

William Daly
13 Via Abrazar
San Clemente, CA 92673
949-498-8220

Dear Bill:

It is my pleasure to extend the following offer of employment to you on behalf of Akebia Therapeutics, Inc. Akebia is proud of its achievements to date and we are looking to individuals such as yourself to play a key role in partnering our exciting drug products. We are convinced that you can make an immediate impact, and we are excited about your future growth potential. In short, we want you on our team.

This letter confirms to you Akebia's offer of employment, the terms of our offer are as follows:

1. Position/Expectations: Senior Vice President of Business Development, Akebia Therapeutics, Inc., reporting to Joseph Gardner, Chief Executive Officer. You will be overall business development leader for our anemia program, including our current lead AKB-6548, and our HPTP β program for vascular leak on behalf of Aerpio Therapeutics (a "spin out" company from Akebia). Your responsibilities will include all partnering and sale of asset activities on both AKB-6548 and AKB-9778. You will be working closely and collaboratively with Ian Howes, CFO in this regard. As a commuting employee you are expected to spend at least 3 days a week in Cincinnati for the first four months of employment (the start up period). Commuting costs will be covered by the Company in a manner consistent with other commuting executives. The commuting schedule can be adjusted, both during and subsequent to the start up period, based on business needs with the verbal agreement of Joseph Gardner, CEO. Your start date will be as soon as feasible, preferably on or around January 23, 2012.

2. Compensation:

(i) A base salary of \$275,000 per year, paid monthly.

(ii) In addition to your base salary, you will receive Incentive Compensation (pending Board approval):

(a) Incentive Compensation will include performance based cash bonuses up to a maximum of 20% of salary per year, and participation in the company stock option plan with a target restricted stock allocation of 3,004,705 shares (corresponding to approximately 0.75% of total company shares, with vesting terms as described in the Company Equity Award agreement) subject to Board approval.

3. Performance / Salary Reviews: The current policy, subject to change without notice, is that reviews are conducted on an annual basis. See Employee Handbook for more details regarding policies and expectations for employees of Akebia Therapeutics, Inc.

4. Benefits: You will be entitled to participate in all applicable benefit programs as currently, or prospectively, offered by Akebia Therapeutics, Inc. The Company is committed to providing comprehensive and competitive benefits to its employees. The Company has a plan to provide for health care and dental insurance, and as an employee you and your family will be eligible to join those plans. The Company has a plan in place to provide disability insurance and is developing a benefit plan that will include life insurance. The Company also provides a 401K plan allowing employees to place pre-tax dollars in a retirement account up to the maximum permitted by law. We will provide more detailed information regarding these plans and any future additions/modifications to our benefit plans when available. The Company will provide twenty (20) days of paid time off (PTO) per calendar year, plus holidays, as defined and allocated in the Akebia Employee Handbook. The number of vacation days available to you in 2012 is on a prorated basis depending on start date and percent year remaining. In addition to our "hard" benefits we offer a host of "soft" benefits such as a fun, flexible and stimulating work environment and the rewards of developing important new medicines.

5. Contingencies: Additionally, this offer is contingent upon your (i) submission of appropriate documentation for verification purposes in order that the Company may be in compliance with the immigration and Reform Control Act of 1986, as amended. Subsequent to accepting our offer Akebia expects all employees to complete and have in force an Employee Agreement which will be sent in a subsequent mailing. The Employee contract will specify "the following benefit relative to severance pay- 3 months severance (salary and benefits) for the first 6 months. Then 6 months severance after the employee achieves 6 months of employee history (assuming that termination/separation from Company is not "for cause").

6. Employment at Will: The parties hereto recognize that this offer of employment is not intended to create a contract of employment and both Akebia and the employee retain the right to terminate the employment relationship at any time without cause.

7. Covenants, Company Matters - Confidentiality and Assignment of Rights: Employee shall not disclose, use or make known for Employee's or another's benefit other than for the benefit of the Company and its affiliates any Company Confidential Information (as further defined and elaborated in the Employee Agreement).

All ideas, concepts, inventions, improvements, programs, information technology, derivative works, processes, configurations, data, procedures, designs, techniques and other works of authorship, conceived or reduced to practice by Employee, either solely or in collaboration with others, during Employee's employment with the Company, including but not limited to all

Akebia Therapeutics, Inc.

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copyright, trademark, patent, trade secret and intellectual property rights associated therewith, shall become and remain the exclusive property of the Company. Employee hereby assigns to the Company any and all of Employee's right, title and interest in and to any of the foregoing, and Employee waives any claim that Employee may have thereto (as further defined and elaborated in the Employee Agreement).

To indicate acceptance, please sign and return a copy of this letter (via fax, pdf or regular mail).

Bill we look forward to you joining Akebia Therapeutics, Inc. Teamwork, quality people, and a business focus, are all critical to Akebia's future success. We are confident that you will play an important role in our success over the coming year and well into the future.

Very truly yours,

Akebia Therapeutics, Inc.

By: /s/ Joseph H. Gardner
Name: Joseph H. Gardner
Title: President and Chief Executive Officer

cc: Ian Howes, Chief Financial Officer

The undersigned accepts the above employment offer, agrees that it contains partial terms of employment with Akebia Therapeutics, Inc. By accepting this offer of employment, the undersigned is acknowledging that no prior employment obligations or other contractual restrictions exist which preclude employment with Akebia Therapeutics, Inc. It is further understood that this offer is confidential and disclosure of any of the terms and conditions contained herein constitute grounds for termination of employment or withdrawal of this offer.

Accepted:

/s/ William Daly
William Daly

1/8/2012
Date:

Akebia Therapeutics, Inc.

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EXECUTIVE EMPLOYMENT AGREEMENT

THIS EXECUTIVE EMPLOYMENT AGREEMENT (this "Agreement") is made as of May 2, 2007, by and between Akebia Therapeutics, Inc., a Delaware corporation (the "Company"), and Joseph H. Gardner, an individual resident in the State of **Ohio** ("Executive").

1. Employment; Duties; Full Time Employment. The Company hereby agrees to employ Executive, and Executive hereby accepts employment, as President and CEO of the Company, with such employment to commence on May 1st 2007, (the "Commencement Date"). In such capacity, Executive shall perform such executive duties and exercise such powers for the Company and its subsidiaries as the Board of Directors of the Company (the "Board") may lawfully assign to or vest in Executive from time to time. Executive covenants and agrees that, at all times during the Term (as defined below), Executive shall devote Executive's full business time and efforts to Executive's duties as an employee of the Company and that Executive will not, directly or indirectly, engage or participate in any other business or professional activities during the Term, other than (a) non-conflicting personal investments managed on Executive's personal time, (b) activities for non-profit institutions (including, but not limited to, participating on boards of directors), and (c) activities or commitments set forth and described on Appendix A (Section 1) hereto, provided that such activities do not interfere or conflict with Executive's obligations hereunder.

2. Term. The Company agrees to employ Executive, and Executive agrees to serve the Company, on an "*at will*" basis, which means that either the Company or Executive may terminate Executive's employment with the Company at any time, with or without Cause, as provided in Section 5 below. The period commencing with the Commencement Date and ending on the effective date of any termination of employment in accordance with the provisions hereof shall constitute the term of this Agreement (the "Term").

3. Compensation and Benefits. During the Term, the Company shall provide Executive with the following compensation and benefits:

(a) Base Salary. The Company shall pay Executive a base salary ("Base Salary") at the rate of \$198,000.00 per annum (less applicable deductions and withholdings), payable in periodic payments in accordance with the Company's normal payroll practices. During the Term, Executive's compensation shall be reviewed by the Board from time to time and at least once every 12 months. Any increase or decrease in Base Salary (together with the then existing Base Salary) shall serve as the "Base Salary" under this Agreement.

(b) Discretionary Bonuses. Executive will be eligible for discretionary bonuses on both a quarterly and annual basis in amounts to be determined by the Board in its sole discretion. The exact amount of the actual bonus awarded to Executive for any given quarter or year, if any, shall be determined by the Board in its sole discretion based upon its consideration of the Company's performance and Executive's performance against objectives established by the Board for the quarter or year, as the case may be, in consultation with Executive. Executive must remain an active employee through the end of the calendar quarter or year in order to earn a bonus for that quarter or year. Executive will not earn any bonus (including a prorated bonus) for any quarter or year if Executive's employment terminates for any reason before the end of such respective period.

(c) Stock Ownership. Executive has been issued an aggregate of 800,000 shares of Common Stock of the Company (the "Acquisition Stock"). Except for the foregoing Acquisition Stock, and except as otherwise may be determined by the Board from time to time after the date of this Agreement in its discretion, Executive shall not have any right to be issued shares of the Company's capital stock or options, warrants or other rights to acquire any capital stock of the Company.

(d) Other Compensation and Benefits. In addition to the compensation specified above in this Section 3, Executive shall be entitled to the following benefits during the Term, all on the terms offered or maintained by the Company to, for or on behalf of its senior executives: vacation, holidays and sick leave, and subject to eligibility therefor, the right to participate in any profit sharing plan, retirement plan, 401(k) plan, group medical plan, group dental plan, group life insurance plan and/or other health or insurance plan maintained by the Company for its senior executives generally and, if applicable, their family members. Executive will be eligible for the paid holidays as are generally made available to employees of the Company.

4. Business Expenses. The Company shall reimburse Executive for all reasonable and necessary business and travel expenses incurred by Executive in the performance of Executive's duties under this Agreement. Such expenses shall be reimbursed in accordance with the Company's guidelines, limits and procedures relating thereto and upon presentation of proper expense vouchers or receipts therefor.

5. Termination.

(a) Termination on Death or Disability. The Term will terminate automatically and immediately upon Executive's death or, upon 30 days prior written notice from the Company, in the event of Executive's Disability. For purposes of this Section 5, "Disability" means that Executive, at the time notice is given, has been unable to substantially perform Executive's duties under this Agreement for not less than sixty (60) work days within a six (6) consecutive month period as a result of Executive's incapacity due to physical or mental illness. Upon any termination for death or Disability, Executive will not be entitled to any further compensation from the Company, including severance pay, pay in lieu of notice or any other such compensation (other than accrued salary and bonus, and accrued and unused vacation, through Executive's last day of employment).

(b) Termination Without Cause. During the Term, the Company shall be entitled to terminate Executive's employment without Cause (as defined below), in which case Executive shall be entitled to receive the following severance benefits (in addition to accrued salary and bonus, and accrued and unused vacation, through Executive's last day of employment): (i) Executive shall be entitled to severance pay in the form of continuation of Executive's Base Salary in effect on the effective date of termination for a period of three months after the date of such termination, to be paid periodically in accordance with the Company's normal payroll practices and subject to standard payroll deductions and withholdings; and (ii) if Executive timely elects continued coverage under COBRA, then (A) the Company shall make such

COBRA coverage (or equivalent medical benefits after the termination of COBRA) available for at least 24 months following termination and (B) the Company shall pay the COBRA premiums necessary to continue Executive's medical insurance coverage in effect on the termination date for a period of three months following Executive's termination (provided that such COBRA continuation and reimbursement shall terminate upon commencement of new employment by an employer that offers health care coverage to its employees). Notwithstanding the foregoing, all severance benefits contemplated by hereunder are conditional on Executive (i) complying with the provisions of Section 6 below, and (ii) delivering prior to receipt of such severance benefits, an effective, general release of claims in favor of the Company or its successor, its subsidiaries and their respective directors, officers and stockholders in a form acceptable to the Company or its successor. In the event that the Company determines that any severance benefit provided hereunder fails to satisfy the distribution requirement of Section 409A(a)(2)(A) of the Internal Revenue Code ("Code") as a result of Section 409A(a)(2)(B)(i) of the Code, then if an accelerated payment of such benefits would cause such benefit not to be subject to the provisions of Section 409A(a)(1) of the Code, the payment of such benefits shall be accelerated to the minimum extent necessary so that the benefit is not subject to the provisions of Section 409A(a)(1) of the Code. (The payment schedule as revised after the application of the preceding sentence shall be referred to as the "Revised Payment Schedule.") However, in the event the accelerated payment of such benefits would not avoid the application of Section 409A(a)(1) of the Code, the payment of such benefits shall not be made pursuant to the original payment schedule or the Revised Payment Schedule and instead the payment of such benefits shall be delayed to the minimum extent necessary so that such benefits are not subject to the provisions of Section 409A(a)(1) of the Code. The Board may attach conditions to or adjust the amounts paid pursuant to this Section 5(b)(iv) to preserve, as closely as possible, the economic consequences that would have applied in the absence of this Section 5(b)(iv); provided, however, that no such condition or adjustment shall result in the payments being subject to Section 409A(a)(1) of the Code.

(c) Termination for Cause; Resignation. The Company may terminate Executive's employment at any time for Cause, and Executive may resign at any time. Termination for Cause shall be effective on the date the Company gives notice to Executive of such termination in accordance with this Agreement. Resignation by Executive shall be effective on the date Executive gives notice to the Company of such resignation in accordance with this Agreement. In the event of the Company's termination of the Term for Cause or Executive's resignation from Executive's employment Executive will not be entitled to any further compensation from the Company, including severance pay, pay in lieu of notice or any other such compensation (other than accrued salary and bonus, and accrued and unused vacation, through Executive's last day of employment).

(d) Cause. For purposes of this Agreement, "Cause" shall mean (i) Executive's failure to substantially perform Executive's duties under this Agreement for reasons other than death or Disability, which failure, if curable, is not cured to the reasonable satisfaction of the Board during the fifteen (15) day period following written notice of such failure from the Company; (ii) Executive's material failure or refusal to comply with reasonable written policies, standards and regulations established by the Company from time to time which failure, if curable, is not cured to the reasonable satisfaction of the Board during the fifteen (15) day period following written notice of such failure from the Company; (iii) the commission by Executive of

(x) an act of dishonesty or constituting common law fraud, embezzlement or a felony or (y) any tortious act, unlawful act or malfeasance that causes or reasonably would cause (for example, if it became publicly known) material harm to the Company's standing, condition or reputation; or (iv) any breach by Executive of the provisions of this Agreement, which breach, if curable, is not cured to the reasonable satisfaction of the Board during the fifteen (15) day period following written notice of such breach from the Company. The Board (excluding Executive if Executive is at such time a member of the Board) shall make all determinations relating to termination, including without limitation any determination regarding Cause, pursuant to this Section 5(d).

6. Company Matters; Restrictive Covenants.

(a) Confidential Information. Executive will have access to and will participate in the development of and will be acquainted with confidential or proprietary information and trade secrets related to the business of the Company and its affiliates, including but not limited to (i) customer lists; related records and compilations of information; the identity, lists or descriptions of any new customers, referral sources or organizations; financial statements; cost reports or other financial information; contract proposals or bidding information; business plans; training and operations methods and manuals; personnel records; software programs; reports and correspondence; and management systems, policies or procedures, including related forms and manuals, (ii) information pertaining to future developments such as future marketing or acquisition plans or ideas and potential new business locations; and (iii) all other tangible and intangible property and intellectual property which is used in the business and operations of the Company and its affiliates but not made public. The foregoing is collectively referred to as the "Confidential Information." The term Confidential Information shall not include any information (x) that is or becomes generally publicly available (other than as a result of violation of this Agreement by Executive), or (y) that Executive receives on a non-confidential basis from a source (other than the Company, its affiliates or their representatives) that is not known by Executive to be bound by an obligation of secrecy or confidentiality to any of the Company or its affiliates. Executive shall not disclose, use or make known for Executive's or another's benefit other than for the benefit of the Company and its affiliates any Confidential Information or use such Confidential Information in any way. Upon the termination of Executive's employment with the Company for any reason, Executive shall immediately return to the Company all Confidential Information in whatever form maintained (including, without limitation, computer discs and other electronic media).

(b) Non-Competition. During Executive's employment with the Company and for the one-year period immediately following the termination of Executive's employment with the Company, Executive will not directly or indirectly (whether as an officer, director, stockholder, partner, proprietor, associate, representative, consultant or in any capacity whatsoever) engage in, become financially interested in, be employed by or have any business connection with any person, corporation, firm, partnership or any other entity whatsoever which competes with the Company in any area where the Company operates, or has operated at any time during Executive's employment with the Company, or any area the Company has planned to expand into at any time during Executive's employment with the Company.

(c) Nonsolicitation of Customers. During Executive's employment with the Company and for the one-year period immediately following the termination of Executive's

employment with the Company, Executive shall not solicit, directly or indirectly, any customers of the Company or of its affiliates who or which were customers of the Company at any time during Executive's employment with the Company, nor shall Executive solicit any potential customers of the Company or of its affiliates with whom Executive had contact on behalf of the Company or its affiliates during Executive's employment with the Company.

(d) Nonsolicitation of Employees. During Executive's employment with the Company and for the one-year period immediately following the termination of Executive's employment with the Company, Executive shall not solicit or hire, directly or indirectly, on Executive's behalf or on behalf of any other person or entity, any person employed by the Company or its affiliates except with the specific written consent of the Company.

(e) Certain Representations. Executive represents that Executive's experience, capabilities and circumstances are such that the provisions of this Agreement will not prevent Executive from earning a livelihood. Executive further agrees that the limitations set forth in this Agreement (including, without limitation, the time and territorial limitations) are reasonable and properly required for the adequate protection of the current and future businesses of the Company and its affiliates. Executive further acknowledges that a remedy at law for any breach or threatened breach of the provisions of this Agreement would be inadequate and will cause immediate and irreparable harm to the Company or its affiliates in a manner that cannot be measured nor adequately compensated in damages. Executive further acknowledges that in the event of any such breach and in addition to any and all other remedies that it may have at law or in equity, the Company or its affiliates shall be entitled to temporary, preliminary and permanent injunctive relief to restrain such breach by Executive, and to recover all costs and expenses, including reasonable attorneys' fees, of any proceedings brought to obtain such injunctive relief. Nothing contained herein shall restrict or limit in any manner the Company's right to seek and obtain any form of relief, legal or equitable, against Executive in an action brought to enforce its rights hereunder.

(f) Intellectual Property. Except as otherwise set forth and described on Appendix A (Section 6(f)) hereto, all ideas, concepts, inventions, improvements, programs, information technology, derivative works, processes, configurations, data, procedures, designs, techniques and other works of authorship and development made, conceived or reduced to practice by Executive, either solely or in collaboration with others, during Executive's employment with the Company, including but not limited to all copyright, trademark, patent, trade secret and intellectual property rights associated therewith, shall become and remain the exclusive property of the Company. Executive hereby assigns to the Company any and all of Executive's right, title and interest in and to any of the foregoing, and Executive waives any claim that Executive may have thereto. Executive will promptly disclose in writing to the Company all such ideas, concepts, inventions, improvements, programs, information technology, derivative works, processes, configurations, data, procedures, designs, techniques and other works of authorship and development, and will cooperate fully with the Company in confirming and protecting the Company's ownership rights therein. The work product resulting from the Executive's employment with the Company is work made for hire.

(g) Ventures. If, during the Term, Executive is engaged in or associated with the planning or implementing of projects, programs or ventures involving the Company and third

parties, all rights in such projects, programs and ventures shall belong to the Company (or the third party, to the extent provided in any agreement between the Company and the third party). Except as formally approved by the Company, Executive shall not be entitled to any interest in such project, program or venture or to any commission, finder's fee or other compensation in connection therewith other than the salary or other compensation to be paid to Executive as provided in this Agreement.

(h) Resignation on Termination. On termination of Executive's employment Executive shall immediately (and with contemporaneous effect) resign any directorships, offices or other positions that Executive may hold in the Company or any of its affiliates, unless otherwise requested by the Board.

7. Miscellaneous.

(a) Withholding Taxes. The Company may withhold from all salary, bonus or other benefits payable under this Agreement all federal, state, city or other taxes as shall be required pursuant to any law or governmental regulation or ruling.

(b) Entire Agreement; Binding Effect. This Agreement sets forth the entire understanding between the parties as to the subject matter of this Agreement and supersedes all prior agreements, commitments, representations, writings and discussions between them (whether written or oral) on the subject matters herein; and neither of the parties shall be bound by any obligations, conditions, warranties or representations with respect to the subject matter of the foregoing except as expressly provided herein or therein or as duly set forth on or subsequent to the date hereof in a written instrument signed by the proper and fully authorized representative of the party to be bound hereby. This Agreement is binding on Executive and on the Company and Executive and their respective successors and assigns (whether by assignment, by operation of law or otherwise); provided that neither this Agreement nor any rights or obligations hereunder may be assigned by Executive or the Company without the prior written consent of the other party (except that the Company shall be entitled to assign this Agreement in connection with the sale of all or substantially all of the Company's assets, or a merger or consolidation in which the Company is not the surviving entity).

(c) Absence of Conflict. Executive represents and warrants that Executive's employment by the Company as described herein will not conflict with and will not be constrained by any prior employment or consulting agreement or relationship.

(d) Voluntary Nature of Agreement; Legal Rights. Executive is executing this Agreement voluntarily and without any duress or undue influence by the Company or anyone else. Executive acknowledges that Executive has had the opportunity to consult with an attorney regarding the provisions of this Agreement and has either obtained such advice of counsel or knowingly waived the opportunity to seek such advice. Executive has carefully read this Agreement and has asked any questions needed for Executive to understand the terms, consequences and binding effect of this Agreement and fully understand it, including that Executive is waiving Executive's right to a jury trial.

(e) Waivers. No party shall be deemed to have waived any right, power or privilege under this Agreement or any provisions hereof unless such waiver shall have been duly executed in writing and acknowledged by the party to be charged with such waiver. The failure of any party at any time to insist on performance of any of the provisions of this Agreement shall in no way be construed to be a waiver of such provisions, nor in any way to affect the validity of this Agreement or any part hereof. No waiver of any breach of this Agreement shall be held to be a waiver of any other subsequent breach.

(f) Notices. All notices, approvals, consents, requests or demands required or permitted to be given under this Agreement shall be in writing and shall be deemed sufficiently given (i) upon delivery, if delivered by hand (ii) one business day after transmission, if sent by facsimile (confirmation received) or (iii) one business day after the business day of deposit with a reputable overnight courier for next business day delivery, freight prepaid (signature of receipt obtained). Notice in each case shall be addressed to the party entitled to receive such notice at the following address (or other such addresses as the parties may subsequently designate):

The Company:
Akebia Therapeutics, Inc.
250 East 5th Street
Cincinnati, Ohio 45202
Attn: Chairman of the Board
Fax:

With a copy to:
David Willbrand
Thompson Hine LLP
312 Walnut Street, Suite 1400
Cincinnati, Ohio 45202
Attn: David J. Willbrand, Esq.
Fax: (513) 241-4771

Executive:
Joseph H. Gardner
4060 Boomer Rd
Cincinnati, Ohio 45247

(g) Governing Law. This Agreement shall be governed by, and construed and enforced in accordance with, the laws of Ohio as they apply to contracts entered into and wholly to be performed therein by residents thereof.

(h) Severability. Every provision of this Agreement is intended to be severable from every other provision of this Agreement. If any provision of this Agreement is held to be invalid, illegal or unenforceable, in whole or in part, such invalidity, illegality or unenforceability shall not affect the other provisions of this Agreement; and this Agreement shall be construed as if such invalid, illegal or unenforceable provision had never been contained herein except to the extent that such provision may be construed and modified so as to render it valid, lawful, and enforceable in a manner consistent with the intent of the parties to the extent compatible with the applicable law as it shall then appear.

(i) Effect of Headings. The Section and subsection headings contained herein are for convenience only and shall not affect the construction hereof.

(j) Counterparts. This Agreement may be executed in multiple counterparts, each of which shall be deemed to be an original, and all such counterparts shall constitute but one instrument.

Signature Page Follows

IN WITNESS WHEREOF, the parties hereto have caused this Agreement to be duly executed as of the year and date first written above.

AKEBIA THERAPEUTICS, INC.

By: /s/ John Rice

John Rice, Ph.D.

Chairman of the Board

Executive:

/s/ Joseph H. Gardner

Joseph H. Gardner, Ph.D.

President and CEO

Appendix A

[JG]

Section 1.

Executive will continue to volunteer as a Director on the Board of the Juvenile Diabetes Research Foundation International (“JDRF”). Executive will continue to advise the JDRF Board on research and development programs directed at curing Type 1 diabetes and its complications. If occasions arise that represent a potential conflict of interest with or competition with the Company, Executive will recuse himself from the JDRF responsibilities to insure that the Company’s business interests are appropriately carried forward and protected.

In the event that a for-profit company requests the participation of Executive on its board of directors or board of advisors, Executive may so participate if he first receives the prior written consent of the Chairman of the Board, and in any event, if occasions arise that represent a potential conflict of interest with or competition with the Company, Executive will recuse himself therefrom to insure that the Company’s business interests are appropriately carried forward and protected.

In addition, there may be occasions where Executive is requested to consult on small projects on behalf of various venture capital firms. Executive shall disclose all such activities to the Board, and Executive agrees that (i) these activities must not and shall not interfere with the operation and progress of Company projects, and (ii) in any event, time devoted by Executive to such projects will not exceed 5% of Executive’s time in any calendar year period.

Section 6(f).

As part of his duties for the Company, Executive will evaluate various academic projects which are funded and owned by universities. In discussions with these academic collaborators, Executive may occasionally contribute ideas to these academic projects, with the understanding that these ideas will be incorporated into the university intellectual property. In such instances, Executive will make every reasonable effort to negotiate a “*first right to negotiate a license*” to the related university intellectual property on behalf of the Company, but Executive cannot guarantee that he will be successful in negotiating such a right in every instance.

**Amendment No. 1 to
EXECUTIVE EMPLOYMENT AGREEMENT**

This Amendment No. 1 (this "Amendment") to EXECUTIVE EMPLOYMENT AGREEMENT (the "Agreement") is made as of April 6, 2011, by and between Akebia Therapeutics, Inc., a Delaware corporation (the "Company"), and Joseph Gardner, an individual resident in the State of Ohio ("Executive").

Whereas, the Company and Executive entered into the Agreement on May 2, 2007, and now wish to amend it in certain respects, *now therefore*, it is agreed as follows:

1. Section 1 is hereby amended to renumber its existing text to be a new subsection "(a)" (so it is depicted as reading in its beginning: "(a) General. The company hereby agrees to employ . . ."), and also to add the following subsection (b):

(b) Location. Executive acknowledges and agrees that the Company is currently located in Cincinnati, although the Company will shortly be evaluating whether the Company should relocate to (or co-locate in) Boston or San Francisco. Subject to the balance of this subsection, Executive is expected to spend a minimum of four (4) working days a week (exclusive of travel time) in Cincinnati, or Boston or San Francisco (if the Company does relocate or co-locate to Boston or San Francisco or other relevant city), as appropriate, to perform his services for the Company. However, for any week that Executive is already traveling on Company business to a destination other than where the Company is then located, the requirement in the previous sentence shall not apply. If the Company adopts a plan for such relocation or co-location as provided in this Section 1(b), and Executive is selected as a candidate for relocation or co-location, then (i) Executive shall relocate or co-locate in accordance with the plan, and (ii) subject to Section 4 (*Business Expenses*) and to the foregoing plan, the Company shall pay the out-of-pocket costs of renting an apartment in support of the plan (the selection of which apartment shall be subject to the Company's prior written consent) and shall reimburse reasonable out-of-pocket travel costs directly related to the relocation and/or co-location. The weekly schedule in effect from time-to-time can be adjusted with the verbal agreement of the Chairman of the Compensation Committee based on business needs and the need for business related travel to various destinations. Notwithstanding the foregoing, Executive's obligation under this Section 1(b) to relocate (or co-locate) shall expire on the first anniversary of the date hereof unless by then the Company has adopted a relocation or co-location plan.

2. Section 3(a) is hereby amended and restated in its entirety, to read as follows:

(a) Base Salary. The Company shall pay Executive a base salary ("Base Salary") at the rate of \$275,000.00 per annum (less applicable deductions and withholdings), payable in periodic payments in accordance with the Company's normal payroll practices. During the Term, Executive's compensation shall be reviewed by the Board from time to time and at least once every 12 months. Any increase or decrease in Base Salary (together with the then existing Base Salary) shall serve as the "Base Salary" under this Agreement.

3. Section 3 is further amended by adding the following at its end:

(e) Vacation. The Executive shall be entitled to four weeks paid vacation per calendar year to be taken at such times as may be approved by the Board (which is more than the normal amount defined in the Akebia Employee Handbook). An aggregate of up to 1 week of unused vacation time may be carried over at the end of a calendar year. Upon termination of the Executive's employment, the Company will pay the Executive for unused vacation at the Executive's Base Salary rate (subject to normal deductions and withholding amounts) on the next regularly scheduled pay date immediately following the termination date.

4. Section 5(b) is hereby amended and restated in its entirety to read as follows:

(b) Termination Without Cause or for Good Cause. During the Term, the Company shall be entitled to terminate Executive's employment without Cause (as defined below), and the Executive is also entitled to terminate his employment for Good Reason (as defined below), in which case Executive shall be entitled to receive the following severance benefits (the "Severance Payments"), in addition to accrued salary and bonus, and accrued and unused vacation, through Executive's last day of employment: (i) Executive shall be entitled to severance pay in the form of continuation of Executive's Base Salary in effect on the effective date of termination for a period of six months, to be paid periodically in accordance with the Company's normal payroll practices and subject to standard payroll deductions and withholdings, commencing on the next regularly scheduled payroll date of the Company on or after the 61st day after the date of termination; and (ii) if Executive timely elects continued coverage under COBRA, then (A) the Company shall make such COBRA coverage (or equivalent medical benefits after the termination of COBRA) available for at least 24 months following termination and (B) the Company shall reimburse Executive for the COBRA premiums necessary to continue Executive's medical insurance coverage in effect on the termination date with respect to a period of six months following Executive's termination (provided that such COBRA continuation and reimbursement shall terminate upon commencement of new employment by an employer that offers health care coverage to its employees), with payment of the reimbursement to be made on the 61st day after the date of termination with respect to any such month that ends on or before such day and on the last day of each month that ends after such day.

Notwithstanding the foregoing, all Severance Payments under this Agreement are conditional on Executive (i) complying with the provisions of Section 6 below, and (ii) delivering prior to the 53rd day after the date of termination an effective, general release of claims in favor of the Company or its successor, its subsidiaries and their respective directors, officers and stockholders in a form acceptable to the Company or its successor, and not thereafter revoking such release.

The Board may attach conditions to or adjust the amounts paid pursuant to this Section 5(b) to preserve, as closely as possible, the economic consequences that would have applied in the absence of this Section 5(b); provided, however, that no such condition or adjustment shall result in the payments being subject to Section 409A(a)(1) of the Code.

5. Section 5 is further amended by adding the following new subparagraphs (e) and (f):

(e) Good Reason. For purposes of this Agreement, “Good Reason” shall mean any of the following without the consent of the Executive: (i) a material diminution in the Executive’s position, duties or responsibilities from those held by or assigned to the Executive as of the Effective Date, (ii) a reduction of the Executive’s Base Salary, or (iii) a material reduction of the Executive’s benefits or bonus/incentive compensation opportunities provided to the Executive as then in effect, so long as he is the only executive to suffer such a reduction.

(f) Change of Control Termination. The Executive and the Company agree that the Executive shall be paid the Severance Payments if the Executive’s employment terminates in connection with or within 6 months following a change of control (as described below) because either (i) the acquiror/NewCo does not offer the Executive employment on at least materially comparable compensation terms and benefits (including severance obligations) to those that are provided to the Executive pursuant to this Agreement; or (ii) such terms are initially offered and accepted, but within 6 months following the change of control the Executive’s employment with the acquiror/NewCo is terminated as a result of any of the events described in Section 5(b). For purposes of this Agreement a change of control shall be deemed to have occurred upon a transfer (or license on an exclusive basis) of all or substantially all of the assets of the Company or the transfer of ownership of more than a majority of the securities of the Company, whether in a single transaction or series of separate transactions, other than in connection with fundraising activities of the Company, including without limitation a transaction in which a portion of the assets of the Company are transferred to an acquiror and the Company does not continue as a going concern during the 6 months thereafter, or its remaining assets are moved following such transfer to an acquiror to a NewCo (regardless of whether the NewCo stockholders are existing Company stockholders) and such Newco does not continue as a going concern during the 6 months after such transfer to such acquiror. If a party obtains an option to close a transaction, the transaction will not be considered as having occurred until such option is exercised and the transaction thereafter closed.

6. Section 7(f) is amended and restated to read in its entirety as follows:

(f) Notices. All notices, approvals, consents, requests or demands required or permitted to be given under this Agreement shall be in writing and shall be deemed sufficiently given (i) upon delivery, if delivered by hand (ii) one business day after transmission, if sent by facsimile (confirmation received) or (iii) one business day after the business day of deposit with a reputable overnight courier for next business day delivery, freight prepaid (signature of receipt obtained). Notice in each case shall be addressed to the party entitled to receive such notice at the following address (or other such addresses as the parties may subsequently designate):

If to the Company:

Akebia Therapeutics, Inc.
Attn: President
9987 Carver Road, Suite 420
Cincinnati, Ohio 45242
Fax: 513.985.1920

If to the Executive:

Joseph Gardner
4060 Boomer Road
Cincinnati, Ohio 45247
Fax:

With copies of all notices also to go to Company counsel and the Chair of the Company's Compensation Committee, as follows:

Thompson Hine LLP
312 Walnut Street, Suite 1400
Cincinnati, Ohio 45202
Attn: David J. Willbrand, Esq.
Fax: (513) 241-4771

Akebia Therapeutics, Inc.
Attn: Chair of Compensation Committee
9987 Carver Road, Suite 420
Cincinnati, Ohio 45242
Fax: 513.985.1920

7. Section 7 is further amended by the addition of the following new subparagraph (k):

(k) If the Executive is a "specified employee" (as defined in Section 409A) on the termination date referenced in Section 5(b) and a delayed payment is required by Section 409A to avoid a prohibited distribution under Section 409A, then no Severance Payments that constitute "non-qualified deferred compensation" under Section 409A shall be paid until the earlier of (i) the first day of the 7th month following the date of Executive's "separation from service" as defined in Section 409A, or (ii) the date of Executive's death. Upon the expiration of the applicable deferral period, all payments deferred under this clause shall be paid in a lump sum and any remaining severance benefits shall be paid per the schedule specified in this Agreement.

8. Miscellaneous. This Agreement may be executed in multiple counterparts, each of which shall be deemed to be an original, and all such counterparts shall constitute but one instrument.

[signature page to follow]

IN WITNESS WHEREOF, the parties hereto have caused this Agreement to be duly executed as of the year and date first written above.

AKEBIA THERAPEUTICS, INC.

By: /s/ John M. Rice

Name: John M. Rice

Its: Chairman

Executive:

/s/ Joseph Gardner

Joseph Gardner

[Signature page to Gardner's Amendment No. 1 to Employment Agreement]

CONSULTING AGREEMENT

THIS CONSULTING AGREEMENT (this "Agreement") is made as of September 15, 2013, by and between Akebia Therapeutics, Inc., a Delaware corporation (the "Company"), and Joseph H. Gardner ("Gardner").

WHEREAS, the Company desires to engage Gardner, and Gardner desires to serve the Company, as a consultant in accordance with the terms and subject to the conditions set forth in this Agreement.

NOW, THEREFORE, in consideration of the mutual covenants and agreements set forth in this Agreement, the Company and Gardner hereby agree as follows:

1. Retention; Contract Period. The Company will retain Gardner, and Gardner will make himself available to serve the Company, as a consultant on the terms and subject to the conditions set forth in this Agreement. The term of Gardner's services will commence as of September 15, 2013 (the "Effective Date") and, subject to Section 5, will continue until terminated in accordance with this Agreement (the "Contract Period").

2. Nature of Service. During the Contract Period, Gardner will make himself generally available to consult with the Company on an as-needed basis regarding any matters on which the Company's chief executive officer or any other representative of the Company requests his advice, including, but not limited to, matters pertaining to the Company's intellectual property and management, provided that such requested assistance shall not exceed or require more than 20 hours per month; provided, however, that if occasions arise that represent a potential conflict of interest or competition with Aerpio Therapeutics, Inc. or its successors, as determined by Gardner in his sole and absolute discretion, then Gardner will recuse himself from the particular services and such recusal shall be permissible.

3. Compensation. As compensation for Gardner's services, the Company award Gardner 19,398 shares of the Company's common stock (the "Stock Award") subject to the same vesting schedule afforded to the Directors of the Company and attached hereto as Exhibit A.

4. Reimbursement for Expenses. The Company will reimburse Gardner for all reasonable, ordinary and necessary business expenses incurred by him in the performance of his duties, provided that Gardner provides documentation evidencing such expenses as may be reasonably requested by the Company.

5. Termination.

(a) Death. This Agreement will terminate immediately upon Gardner's death.

(b) For Cause. The Company may terminate this Agreement for "Cause" if Gardner:

(i) materially breaches this Agreement or any other agreement between the Company and him, and does not cure such breach within 30 days of receipt of written notice from the Company specifying the breach and referring to the Company's right to terminate this Agreement for Cause;

(ii) is convicted for or pleads *nolo contendere* to any felony involving moral turpitude;

(iii) commits an act or series of acts of gross misconduct, gross negligence, fraudulent conduct, or misappropriation of funds or property of the Company or any of its affiliates in the course of performing his services under this Agreement or any other agreement between the Company and him; or

(iv) does not respond in a reasonably timely manner when called upon to provide services to the Company.

Subject to the notice period provided in clause (i) above, any termination of this Agreement for Cause will be effective immediately upon the Company giving notice of termination to Gardner.

(c) Without Cause. The Company may terminate this Agreement for any reason or for no reason, with or without "Cause," upon 30 days' prior written notice to Gardner. On and after the second anniversary of this Agreement, Gardner may terminate this Agreement for any reason or for no reason, with or without "Cause," upon 30 days' prior written notice to the Company.

6. Payments Upon Termination. Upon any termination of this Agreement for any reason, the Company will pay to Gardner all reimbursable expenses that were unpaid through the date of termination but will not be obligated to make any further payment or be obligated to provide any further benefits to Gardner. The compensation due or paid by the Company to Gardner shall be non-forfeitable and shall not be subject to set off or reduced in any manner.

7. Confidentiality. Gardner will have access to and will participate in the development of and will be acquainted with confidential or proprietary information and trade secrets related to the business of the Company and its affiliates, including but not limited to (i) customer lists; related records and compilations of information; the identity, lists or descriptions of any new customers, referral sources or organizations; financial statements; cost reports or other financial information; contract proposals or bidding information; business plans; training and operations methods and manuals; personnel records; software programs; reports and correspondence; and management systems, policies or procedures, including related forms and manuals, (ii) information pertaining to future developments such as future marketing or acquisition plans or ideas and potential new business locations; and (iii) all other tangible and intangible property and intellectual property which is used in the business and operations of the Company and its affiliates but not made public. The foregoing is collectively referred to as the "Confidential Information." The term Confidential Information shall not include any information (x) that is or becomes generally publicly available (other than as a result of violation of this Agreement by Gardner), or (y) that Gardner receives on a non-confidential basis from a source (other than the Company, its affiliates or their representatives) that is not known by Gardner to be bound by an obligation of secrecy or confidentiality to any of the Company or its affiliates. Gardner shall not disclose, use or make known for his or another's benefit other than for the

benefit of the Company and its affiliates any Confidential Information or use such Confidential Information in any way. Upon the termination of this Agreement or Gardner's engagement with the Company for any reason, Gardner shall immediately return to the Company all Confidential Information in whatever form maintained (including, without limitation, computer discs and other electronic media). Notwithstanding the foregoing, Gardner is permitted to provide this Agreement to Aerpio Therapeutics, Inc. or its successors.

8. Return of Records. At the end of the Contract Period, Gardner will deliver to the Company any and all information, data, lists, property, records, reports, memoranda, and notes that are in his possession or under his control or that were prepared or acquired in the course of performing his services under this Agreement, together with all equipment and other property that belongs to the Company (collectively, "Company Property"). Gardner agrees not to take with him any such Company Property.

9. Intellectual Property. Gardner agrees to disclose and hereby assigns to the Company, or its respective nominee, all rights to every discovery, invention, improvement, innovation, design, and other definite and useful idea or compilation of information of value (the "Intellectual Property") that Gardner may make or originate, individually or with others, at any time during the Contract Period, resulting from Gardner's performance of services for the Company. Gardner will fully cooperate with the Company, at any time during, or within six months after, the Contract Period and at the Company's cost, in securing, in the name of the Company or its designees, rights with respect to the Intellectual Property.

10. Independent Contractor. Gardner acknowledges and agrees that his status at all times shall be that of independent contractor, and that he may not, at any time, act as an employee, agent, or representative for or on behalf of the Company, for any purpose or transaction, and may not bind or otherwise obligate the Company in any manner whatsoever. In recognition of Gardner's status as an independent contractor, he hereby waives any rights as an employee or deemed employee of the Company. In addition, while performing the services contemplated by this Agreement, Gardner shall be responsible for complying with all applicable federal, state, and local laws, ordinances, and regulations related to such services performed hereunder.

11. Taxes. Gardner shall pay directly all taxes associated with the compensation he receives under this Agreement. Gardner acknowledges the separate responsibility for the payment of all such taxes, and agrees to indemnify the Company and hold the Company harmless from and against any and all liability, claims, costs, and expenses that any of them may suffer or incur arising out of any failure by Gardner to pay promptly any such tax as required by any applicable law.

12. Relationship With Others. The parties agree that the profitability and goodwill of the Company depends on continued amicable relations with its customers and suppliers, and Gardner agrees that he will not cause, request, or advise any customers or suppliers of the Company to curtail or cancel their business with the Company.

13. Remedies. In addition to other remedies provided by law or equity, upon a breach by Gardner of any of the covenants contained in this Agreement, the Company will be entitled to have a court of competent jurisdiction enter a temporary restraining order, a temporary or permanent injunction, and/or other injunctive relief, all without any showing of irreparable harm or damage, prohibiting any further breach of such covenants.

14. Assignment; Binding Effect. This Agreement may not be assigned, except upon the written consent of the other party hereto; provided that the Company may assign this Agreement to any of its affiliates without the consent of Gardner. This Agreement will be binding upon and inure to the benefit of Gardner and the Company and their permitted assigns.

15. Entire Agreement; Amendments; Waivers. This Agreement contains the entire agreement between the parties with respect to the subject matter hereof and will supersede any prior agreement between the Company and Gardner relating to the subject matter hereof that may be in effect on the Effective Date. This Agreement may not be amended orally but only by a written agreement signed by Gardner and the Company. The terms or covenants of this Agreement may be waived only by a written instrument specifically referring to this Agreement, executed by the party waiving compliance. The failure of Gardner or the Company at any time to require performance of any of obligations under this Agreement will in no manner affect the other party's right to enforce any provisions of this Agreement at a subsequent time, and the waiver by either party of any right arising out of any breach will not be construed as a waiver of any right arising out of any subsequent breach.

16. Notices. Any notice, request, or instruction to be given under this Agreement will be deemed to have been given (a) when it is delivered, (b) the day after it is sent by overnight courier, or (c) when it is sent by facsimile or email, with confirmation of receipt, addressed as follows (or to such other addresses as may be designated by written notice to the other party):

If to the Company:

Akebia Therapeutics
Attn: Chief Executive Officer
Suite 420 Carver Road
Cincinnati, OH 45242
Phone No.: 513-985-1921
Fax No.: 513-985-0999

With a copy to:

Thompson Hine
312 Walnut Street, 14th Floor
Cincinnati, Ohio 45202
Fax: 513-241-4771
Attn: David J. Willbrand

If to Gardner:

Joseph H. Gardner
4060 Boomer Road
Cincinnati, OH 45247

With a copy to:

Dinsmore & Shohl LLP
Attn: Lee M. Stautberg, Esq.
255 East Fifth Street
Suite 1900
Cincinnati, Ohio 45202

17. Severability. Any provision of this Agreement that is prohibited or unenforceable will be ineffective to the extent, but only to the extent, of such prohibition or unenforceability without invalidating the remaining portions of this Agreement and such remaining portions will continue to be in full force and effect.

18. Governing Law. The provisions of this Agreement will be governed by and construed in accordance with the laws of the State of Ohio, notwithstanding any conflict of law provision to the contrary.

19. Counterparts. This Agreement may be executed in two counterparts, each of which will be deemed an original, but all of which together will constitute but one and the same instrument.

(signature page follows)

IN WITNESS WHEREOF, the parties have executed this Agreement as of the date first written above to become effective as of the Effective Date.

AKEBIA THERAPEUTICS, INC.

By: /s/ Muneer A. Satter
Name: Muneer A. Satter
Title: Chairman

/s/ Joseph H. Gardner
JOSEPH H. GARDNER

SEPARATION AGREEMENT

This Separation Agreement (“Agreement”) is made as of September 15, 2013 (but in no case before the Termination Date), by and between Akebia Therapeutics, Inc., a Delaware corporation (the “Company”), and Joseph H. Gardner (“Employee”) under the following circumstances:

A. The Company and Employee have agreed to sever their employment relationship as of September 15, 2013 (the “Termination Date”).

B. The Company has proposed, and Employee has agreed to, certain individualized separation benefits in connection with his termination from employment.

NOW, THEREFORE, the parties agree, in consideration of the provisions and payments described below, contract, covenant and agree as follows:

1. On or by the Termination Date, Employee shall be awarded 21,153 shares of the Common Stock of the Company (“New Shares”), such that those shares of Company Common Stock owned by Employee together with all options to purchase Company Common Stock (including, but not limited to, the New Shares and the stock granted pursuant to the Consulting Agreement by and between the Company and Employee of even date herewith) shall collectively represent, in the aggregate, approximately 2.5% of the Company’s Common Stock outstanding as of the Termination Date on a fully-diluted and as-converted basis (all of the foregoing, “Employee’s Common Equity”). All of Employee’s Common Equity shall be fully-accelerated and fully-vested as of the Termination Date, exercisable in accordance with the provisions of the respective award agreements relating to each such award and otherwise subject to the terms and conditions of each such applicable award agreement, including, but not limited to, those provisions relating to expiration. Except for the foregoing Employee’s Common Equity, and except as otherwise may be determined by the Board of Directors of the Company (the “Board”) from time to time after the date of this Agreement in its discretion, Executive shall not have any right to be issued shares of the Company’s capital stock or options, warrants or other rights to acquire any capital stock of the Company.

In connection with the Company’s May 2013 Series C Preferred Stock financing, the Board of Directors of the Company (the “Board”) approved a “Carve-Out” plan for Company employees, as generally described in the document attached hereto as Exhibit A. It is anticipated that such plan shall accommodate and allow for the Board to make (i) discretionary awards thereunder to eligible participants, as well as (ii) awards thereunder that are based upon the relative incentive equity holdings of eligible participants. Employee shall be entitled to participate in the latter awards, and shall participate therein on the same proportional basis and in accordance with the same proportional principles as the other eligible participants.

2. The Company agrees to pay Employee for all Paid Time Off (“PTO”) accrued but unused as of the Termination Date. Payment for accrued but unused PTO will be made in a lump sum on the first payroll date that is at least eight (8) days after the Employee signs and does not revoke this Agreement. Employee will not accrue additional PTO after the Termination Date.

3. The Company (i) shall make such COBRA coverage (or equivalent medical benefits after the termination of COBRA) available for at least 18 months after the Termination Date and (ii) shall pay the actual premiums to continue such medical insurance coverage during the Severance Period (provided that such COBRA continuation shall terminate upon commencement of new employment by an employer that offers health care coverage to its employees).

4. The Company agrees not to contest Employee's application for unemployment benefits.

5. Employee agrees that the separation benefits provided by this Agreement are in excess of any separation benefits for which he might have been eligible, or entitled to, under Company policy or practice, and that he waives the right to receive any additional payment under any such Company policy or practice. Employee further agrees that the separation benefits described in Sections 1-4 of this Agreement are valid and sufficient consideration for the releases and waivers provided by Employee under this Agreement.

6. For and in consideration of the separation benefits provided by this Agreement, Employee on behalf of himself or anyone claiming by, through, or under him (including without limitation his heirs, executors, administrators, attorneys, successors, assigns, and agents), fully settles, releases, and forever discharges the Company, and its present and former Affiliates (which term, for the purposes of this Agreement shall include, without limitation, Aerpio Therapeutics, Inc.), related persons, associations, corporations, entities, parents, subsidiaries, predecessors, partners, principals, officers, directors, shareholders, agents, attorneys, insurers, successors and assigns (collectively "Released Parties"), of and from any and all past, present or future liability, claims, rights, demands, obligations, controversies, damages, costs, expenses (including reasonable attorneys' fees), actions, causes of actions, or compensation of any nature whatsoever, known or unknown, arising, directly or indirectly, up to and including the day Employee signs this Agreement, out of or related to his employment or his termination from employment with the Company, including, but not limited to, any claims which have been or could have been brought for discrimination under federal, state, or local law, as well as any claims or causes of action under any law dealing with employment torts, intentional torts, employee benefits, wrongful discharge, retaliation, breach of contract, implied contract, promissory estoppel, wage and hour violations, violation of public policy, workers' compensation or personal injury, as well as claims for wages, overtime pay, vacation pay, commissions, bonuses, profit sharing, expenses, benefits, termination pay, separation pay, reasonable notice or pay in lieu of such notice, and including all claims and benefits that are or may be available to Employee under his Employment Agreement by and between Employee and the Company dated May 2, 2007 (Employee's "Employment Agreement").

7. Employee further expressly and specifically waives any and all rights or claims under the Age Discrimination in Employment Act of 1967 and the Older Workers Benefit Protection Act (collectively the "Act"), and acknowledges and agrees that this waiver of any right or claim under the Act (the "Waiver") is knowing and voluntary, and specifically

agrees as follows: (a) that this Agreement and this Waiver is written in a manner which Employee understands; (b) that this Waiver specifically relates to rights or claims under the Act; (c) that Employee does not waive any rights or claims under the Act that may arise after the date of execution of this Agreement; (d) that Employee waives rights or claims under the Act in exchange for consideration in addition to anything of value to which Employee is already entitled; and (e) that Employee is advised in writing to consult with an attorney prior to executing this Agreement.

8. Employee acknowledges and understands that to obtain the benefits herein, Employee must accept this Agreement by signing within twenty-one days of receipt. Employee further acknowledges and understands that Employee may revoke acceptance of this Agreement within seven (7) days of such acceptance.

9. Employee covenants not to sue the Released Parties with respect to any claim released pursuant to this Agreement. Moreover, Employee agrees that in the event Employee violates this covenant, Employee will pay all expenses and costs incurred by Released Parties in defending against such lawsuit, administrative charge, or complaint. Nothing in this paragraph or in this Agreement is intended or shall be deemed to prohibit Employee from participating, or cooperating with the Equal Employment Opportunity Commission (the "EEOC"), in any action brought by the EEOC. Employee agrees and acknowledges, however, that he is not entitled to and will not seek or permit anyone to seek on his behalf any personal, equitable or monetary relief in any such action.

10. Employee also agrees to return all Company property on or before the Termination Date.

11. Employee agrees that the provisions of and obligations contained in Section 6 of the Employment Agreement survive this Agreement and are incorporated into this Agreement and shall remain in full force and effect, but all remaining provisions of the Employment Agreement are hereby irrevocably extinguished and cancelled. Notwithstanding the foregoing, Employee's performance of his duties to Aerpio Therapeutics, Inc. or its successors shall be permissible under the Employment Agreement and any other agreement with the Company.

12. The parties agree not to make any statements that disparage the other party, its respective Affiliates, employees, officers, directors, products or services. Notwithstanding the foregoing, statements made in the course of sworn testimony in administrative, judicial or arbitral proceedings (including, without limitation, depositions in connection with such proceedings) shall not be subject to this Section 12. Employee and the Company agree that any violation of this Section 12 will constitute a material breach of this Agreement.

13. In connection with the termination of the employment relationship, Employee hereby resigns from the Board of Directors of the Company, effective as of the Termination Date. Moreover, from and after the Termination Date, Employee will proactively, collaboratively and in good faith cooperate in facilitating the transition to the Company of all relevant corporate, business and operational matters, including but not limited to all banking and other financial accounts as well as all corporate and commercial relationships.

14. Employee agrees that this Agreement and each of its terms and conditions are and shall remain confidential. Employee also agrees that except for discussions with his financial counsel, spouse, and/or legal counsel, he will not disclose the existence of the Agreement or any of its terms and conditions, unless required to do so by law. Employee agrees that any violation of this policy by him will result in the forfeiture of any and all claims or entitlements under the Agreement.

15. This Agreement does not constitute an admission by the Company that it has violated any contract, law, or regulation, or in any way infringed Employee's rights or privileges.

16. The provisions of this Agreement are divisible. If any provisions shall be deemed invalid or unenforceable, it shall not affect the applicability or validity of any other provision of this Agreement, but rather such provision shall be amended to the extent necessary to render it valid and enforceable.

17. The terms of this Agreement represent the entire agreement between the parties and the only consideration for signing this Agreement. No other promises or agreements of any kind have been made to or with the parties to cause them to execute this Agreement. The parties state that they have carefully read this Agreement, that its contents have been fully explained to them; that they have been given adequate time to consider the Agreement; that they have had full opportunity to review its contents with their own legal counsel; and that they know and understand its contents and its legal effect, including, but not limited to, its binding effect, and that they sign this Agreement as their own free act and deed.

18. This Agreement, together with the award agreement relating to the Stock Options, contains the complete understanding between the parties with regard to the subject matter hereof. The terms of this Agreement may not be changed, amended or waived except by another written agreement signed by both parties.

19. This Agreement shall be construed, interpreted and applied in accordance with the law of the State of Ohio.

20. Notices. All notices, approvals, consents, requests or demands required or permitted to be given under this Agreement shall be in writing and shall be deemed sufficiently given (i) upon delivery, if delivered by hand (ii) one business day after transmission, if sent by facsimile (confirmation received) or (iii) one business day after the business day of deposit with a reputable overnight courier for next business day delivery, freight prepaid (signature of receipt obtained). Notice in each case shall be addressed to the party entitled to receive such notice at the following address (or other such addresses as the parties may subsequently designate):

If to the Company:

Akebia Therapeutics, Inc.
Attn: President
9987 Carver Road, Suite 420
Cincinnati, Ohio 45242
Fax:

If to the Executive:

Joseph Gardner
4060 Boomer Road
Cincinnati, Ohio 45247
Fax:

With a copy to:

Dinsmore & Shohl LLP
255 E. Fifth Street
Cincinnati, Ohio 45202
Fax: 513-977-8141
Attn: Lee M. Stautberg, Esq.

With copies of all notices also to go to Company counsel as follows:

Thompson Hine
312 Walnut Street, 14th Floor
Cincinnati, Ohio 45202
Fax: 513-241-4771
Attn: David J. Willbrand

- Signature Page Follows -

IN WITNESS WHEREOF, the parties hereto have caused this Agreement to be duly executed as of the year and date first written above.

Akebia Therapeutics, Inc.

/s/ Muneer A. Satter

By: Muneer A. Satter
Its: Chairman

Employee:

/s/ Joseph H. Gardner

Joseph H. Gardner

ACKNOWLEDGMENT

Employee, in connection with his execution of this Agreement, acknowledges the following:

1. that he is waiving rights or claims arising under the Age Discrimination in Employment Act;
2. that he has been advised by the Company to consult with an attorney prior to executing this Separation Agreement;
3. that he has had a period of up to 21 days in which to consider this Agreement;

4. that for a period of 7 days following execution of this Agreement, he may revoke the Agreement, and that the Agreement shall not become effective or enforceable until the 7-day revocation period has expired.

Date: Sept. 6, 2013

Joseph H. Gardner

/s/ Joseph H. Gardner

Akebia Therapeutics, Inc.

/s/ Muneer A. Satter

By: Muneer A. Satter

Its: Chairman

Date: Sept. 17, 2013

Akebia Therapeutics, Inc.

AMENDED AND RESTATED PARTIAL RECOURSE PROMISSORY NOTE

\$16,804.50

Cincinnati, Ohio
May 9, 2013

FOR VALUE RECEIVED, Joseph Gardner (“**Borrower**”) promises to pay to Akebia Therapeutics, Inc., a Delaware corporation (“**Lender**”), or order, the principal sum of \$16,804.50 with interest as set forth below, both principal and interest payable in lawful money of the United States of America, at such place as Lender may designate in writing.

The principal and interest shall be due and payable as follows:

Interest shall accrue at the rate of six percent (3%) per annum from the date hereof up to and through the Maturity date (as defined herein). The entire aggregate unpaid principal balance and interest shall be due and payable on the first to occur of (a) the consummation of Lender’s first underwritten public offering of its Common Stock (other than a registration statement relating either to the sale of securities to employees of Lender pursuant to its stock option, stock purchase or similar plan or an SEC Rule 145 transaction); (b) the consummation of a “Deemed Liquidation Event” and distribution of proceeds to or escrow for the benefit of the stockholders of Lender in accordance with Lender’s certificate of incorporation as in effect and amended from time to time; or (c) May 9, 2023 (the date of whichever such event occurs first being the “**Maturity Date**” of the Note).

The Note may be prepaid in full or in part at any time without penalty or premium; provided, however, that partial prepayments shall be applied first to the payment of interest accrued to the date of such prepayment and then to the payment of principal.

All parties to this Note, including maker and any sureties, endorsers or guarantors, hereby waive protest, presentment, notice of dishonor and notice of acceleration of maturity and agree to continue to remain bound for the payment of principal, interest and all other sums due under this Note, notwithstanding any change or changes by way of any extension or extensions of time for the payment of principal and interest; and all such parties waive all and every kind of notice of such change or changes and agree that the same may be made without notice or consent of any of them.

As an inducement for Lender to accept from Borrower this Note and as collateral security for the payment of any and all indebtedness and liabilities whatsoever of Borrower to Lender evidenced by this Note, the parties hereto have executed a certain Amended and Restated Stock Pledge Agreement of even date herewith (the “**Pledge Agreement**”), substantially in the form attached hereto as Exhibit A, pursuant to which Borrower has delivered, assigned and pledged to Lender and has granted to Lender a first priority security interest in 11,203 shares of Common Stock of Lender owned by Borrower (the “**Stock**”).

Upon default of Borrower in the payment of any indebtedness under this Note, Lender's sole recourse with respect to fifty percent (50%) of the sum of (a) unpaid principal of this Note, (b) accrued but unpaid interest on this Note, and (c) collection costs including attorneys' fees in connection therewith (the "**Non-Recourse Portion**") shall be to exercise its rights under the Pledge Agreement. Liability of Borrower under the Non-Recourse Portion of this Note is limited to the shares held by Lender pursuant to the Pledge Agreement, and in no event shall Borrower be liable on the Non-Recourse Portion of this Note for any deficiency resulting from any sale of shares pursuant to the Pledge Agreement, nor shall any action or proceeding be brought by Lender against Borrower to recover judgment against Borrower upon the Non-Recourse Portion of this Note or the Pledge Agreement. Upon default of Borrower in the payment of any indebtedness under this Note, Borrower shall be fully liable for all amounts due under this Note other than the Non-Recourse Portion.

At the sole and absolute discretion of Borrower, Borrower may elect to repay some or all of the amounts due and owing hereunder, at any time and from time to time, whether in the event of Default or otherwise, and without the requirement of Lender's consent or approval, by putting to Lender that number of shares of Stock equal to the amount of such repayment, based upon a price per share of the Stock equal to the fair market value thereof as determined in the most recent third-party appraisal thereof.

This Note amends, restates, and supersedes in all respects that certain Partial Recourse Promissory Note by Joseph Gardner to Lender dated as of October 15, 2009, with respect to the time period beginning on May 9, 2013 and ending on the Maturity Date.

This Note is to be governed and construed in accordance with the laws of the State of Delaware.

IN TESTIMONY WHEREOF, the undersigned has executed this instrument the day and year first above written.

/s/ Joseph H. Gardner

Borrower (Print Name): Joseph H. Gardner

ACKNOWLEDGED AND ACCEPTED:

AKEBIA THERAPEUTICS, INC.

By: /s/ Kevin Peters

Name: Kevin Peters

Title: CSO, SVP R&D

Stock Pledge Agreement

Akebia Therapeutics, Inc.

AMENDED AND RESTATED STOCK PLEDGE AGREEMENT

THIS AMENDED AND RESTATED STOCK PLEDGE AGREEMENT (the "**Agreement**") is made as of this 9th day of May, 2013, by and between Joseph Gardner ("**Pledgor**"), and Akebia Therapeutics, Inc., a Delaware corporation ("**Lender**").

WHEREAS, Lender has extended a loan to Pledgor in the principal amount of \$16,804.50 (the "**Loan**"), which Loan is evidenced by a promissory note in favor of Lender attached hereto as **Exhibit A** (the "**Note**"); and

WHEREAS, to secure the payment and performance of all obligations under the Note, Pledgor wishes to pledge to Lender all of Pledgor's right, title and interest in the capital stock of Lender owned by Pledgor and listed on **Exhibit B** hereto (the "**Stock**").

NOW, THEREFORE, the parties hereto agree as follows:

1. **Warranty.** Pledgor hereby represents and warrants to Lender that except for the security interest created hereby, Pledgor owns the Stock free and clear of all liens, charges and encumbrances, that the Stock is duly issued, fully paid and nonassessable, and that Pledgor has the unencumbered right to pledge the Stock.

2. **Security Interest.** Pledgor hereby unconditionally grants and assigns to Lender, its successors and assigns, a continuing security interest in the security title to the Stock. Pledgor has delivered to and deposited with Lender herewith all of Pledgor's right, title and interest in and to the Stock, together with certificates representing the Stock and stock powers endorsed in blank by Pledgor, as security for payment and performance of all obligations of Pledgor to Lender under the Note or any extension, renewal, amendment or modification of the Note, however created, acquired, arising or evidenced, whether direct or indirect, absolute or contingent, now or hereafter existing, or due or to become due. Beneficial ownership of the Stock, including, without limitation, all voting, consensual and dividend rights, shall remain in Pledgor until the occurrence of a Default under the terms hereof (as defined in Section 4 below).

3. **Additional Shares.** In the event that, during the term of this Agreement:

(a) any stock dividend, stock split, reclassification, readjustment or other change is declared or made in the capital structure of Lender, all new, substituted and additional shares, or other securities, issued by reason of any such change and received by Pledgor or to which Pledgor shall be entitled shall be immediately delivered to Lender, together with stock powers endorsed in blank by Pledgor, and shall thereupon constitute Stock to be held by Lender under the terms of this Agreement; and

(b) subscriptions, warrants or any other rights or options are issued in connection with the Stock, all new stock or other securities acquired through such subscriptions, warrants, rights or options by Pledgor shall be immediately delivered to Lender and shall thereupon constitute Stock to be held by Lender under the terms of this Agreement.

4. Default. Failure of Pledgor to pay any amount of principal or interest when due pursuant to the terms of the Note or a default by Pledgor under this Agreement shall constitute a default under the terms of this Agreement (any of such occurrences being hereinafter referred to as a “**Default**”). Upon the occurrence of a Default, Lender may take the actions described in the following sentence and thereafter, or may elect, as its sole recourse hereunder and under the Note and full remedy hereunder and thereunder, in full settlement and repayment of all amounts due and owing under the Note (the “**Obligations**”), and without the requirement of Pledgor’s consent or approval, to redeem that number of shares of Stock equal to the amount of the Obligations (or, if the Obligations exceed the total value of the Stock, then all of the Stock), based upon a price per share of the Stock equal to the fair market value thereof as determined in the most recent third-party appraisal thereof. Alternatively, Lender may sell or make other commercially reasonable disposition of the Stock or any portion thereof after ten (10) business days’ written notice to Pledgor, and Lender may purchase the Stock or any portion thereof at any public sale. The proceeds of the public or private sale or other disposition shall be applied (i) to the costs incurred in connection with the sale; (ii) to any unpaid interest which may have accrued on any obligations secured hereby; (iii) to any unpaid principal; and (iv) to damages incurred by Lender by reason of any breach of the obligations secured against hereby, in such order as Lender may determine but in any event the proceeds shall be applied first to the Non-Recourse Portion of the Note (as defined in the Note) and then to the balance of the sums due under the Note, and any remaining proceeds shall be paid over to Pledgor or others as law provides. Pledgor shall not be liable to Lender for any deficiency in the Non-Recourse Portion of the Note in the event the proceeds of the sale or other disposition of the Stock are insufficient to pay such expenses, interest, principal, obligations and damages.

5. Additional Rights of Secured Parties. In addition to other rights and privileges under this Agreement, Lender shall have the rights, powers and privileges of secured parties under the Uniform Commercial Code.

6. Return of Stock to Pledgor. Upon payment in full of all principal and interest on the Note, Lender shall return to Pledgor all of the then remaining Stock and all rights received by Lender as agent for Pledgor as a result of its possessory interest in the Stock.

7. Voting Rights. Pledgor shall retain all rights to vote the Stock until such time as Lender either cancels or sells the Stock after a Default under the Note.

8. Notices. All notices and other communications required or permitted hereunder shall be in writing and, if mailed by prepaid certified mail, shall be deemed to have been received on the earlier of the date shown on the receipt or three (3) business days after the postmarked date thereof. In addition, notices hereunder may be delivered by hand, by facsimile or by email, in which event such notice shall be deemed effective when delivered. Notice of change of address for notice shall also be governed by this Section. Notices shall be addressed as follows:

If to Pledgor: Joseph Gardner
4060 Boomer Road
Cincinnati, Ohio 45247

If to Lender: Akebia Therapeutics, Inc.
Attention: Kevin Peters
9987 Carver Road, Suite 420
Cincinnati, Ohio 45242

With a copy to:
Thompson Hine LLP
Attention: David J. Willbrand
312 Walnut Street, Suite 1400
Cincinnati, Ohio 45202
Facsimile: (513) 241-4771
Email: David.Willbrand@ThompsonHine.com

9. Binding Agreement. This Agreement amends, restates and supersedes in all respects the Stock Pledge Agreement between Pledgor and Lender dated as of October 15, 2009. The provisions of this Agreement shall be construed and interpreted, and all rights and obligations of the parties hereto determined, in accordance with the laws of the State of Delaware. This Agreement, together with all documents referred to herein, constitutes the entire agreement between Pledgor and Lender with respect to the matters addressed herein and may not be modified except by a writing executed by Lender and Pledgor. This Agreement may be executed in multiple counterparts and by facsimile or PDF, each of which shall be deemed an original but all of which, taken together, shall constitute one and the same instrument.

10. Severability. If any paragraph or part thereof shall for any reason be held or adjudged to be invalid, illegal or unenforceable by any court of competent jurisdiction, such paragraph or part thereof so adjudicated invalid, illegal or unenforceable shall be deemed separate, distinct and independent, and the remainder of this Agreement shall remain in full force and effect and shall not be affected by such holding or adjudication.

11. Assignability. This Agreement, and the rights and obligations of Lender hereunder, may be assigned by Lender to any person or entity to which the Note is transferred by Lender, and such transferee shall be deemed the "Lender" for purposes of this Agreement; provided that the transferee provides written notice of such assignment to Pledgor and agrees to be bound by the terms of this Agreement.

Signature Page Follows

Exhibit B
STOCK CERTIFICATE NUMBERS

<u>Number</u>	<u>Owner</u>	<u>Class of Shares</u>	<u>Number of Shares Represented</u>
—	Joseph Gardner	Common	11,203

Akebia Therapeutics, Inc.

AMENDED AND RESTATED PARTIAL RECOURSE PROMISSORY NOTE

\$96,026.84

Cincinnati, Ohio
June 15, 2013

FOR VALUE RECEIVED, Joseph Gardner (“**Borrower**”) promises to pay to Akebia Therapeutics, Inc., a Delaware corporation (“**Lender**”), or order, the principal sum of \$96,026.84 with interest as set forth below, both principal and interest payable in lawful money of the United States of America, at such place as Lender may designate in writing.

The principal and interest shall be due and payable as follows:

Interest shall accrue at the rate of six percent (3%) per annum from the date hereof up to and through the Maturity date (as defined herein). The entire aggregate unpaid principal balance and interest shall be due and payable on the first to occur of (a) the consummation of Lender’s first underwritten public offering of its Common Stock (other than a registration statement relating either to the sale of securities to employees of Lender pursuant to its stock option, stock purchase or similar plan or an SEC Rule 145 transaction); (b) the consummation of a “**Deemed Liquidation Event**” and distribution of proceeds to or escrow for the benefit of the stockholders of Lender in accordance with Lender’s certificate of incorporation as in effect and amended from time to time; or (c) June 15, 2023 (the date of whichever such event occurs first being the “**Maturity Date**” of the Note).

The Note may be prepaid in full or in part at any time without penalty or premium; provided, however, that partial prepayments shall be applied first to the payment of interest accrued to the date of such prepayment and then to the payment of principal.

All parties to this Note, including maker and any sureties, endorsers or guarantors, hereby waive protest, presentment, notice of dishonor and notice of acceleration of maturity and agree to continue to remain bound for the payment of principal, interest and all other sums due under this Note, notwithstanding any change or changes by way of any extension or extensions of time for the payment of principal and interest; and all such parties waive all and every kind of notice of such change or changes and agree that the same may be made without notice or consent of any of them.

As an inducement for Lender to accept from Borrower this Note and as collateral security for the payment of any and all indebtedness and liabilities whatsoever of Borrower to Lender evidenced by this Note, the parties hereto have executed a certain Stock Pledge Agreement of even date herewith (the “**Pledge Agreement**”), pursuant to which Borrower has delivered, assigned and pledged to Lender and has granted to Lender a first priority security interest in 6,401,788 shares of Common Stock of Lender owned by Borrower (the “**Stock**”).

Upon default of Borrower in the payment of any indebtedness under this Note, Lender’s sole recourse with respect to fifty percent (50%) of the sum of (a) unpaid principal of this Note, (b) accrued but unpaid interest on this Note, and (c) collection costs including attorneys’ fees in

connection therewith (the “**Non-Recourse Portion**”) shall be to exercise its rights under the Pledge Agreement. Liability of Borrower under the Non-Recourse Portion of this Note is limited to the shares held by Lender pursuant to the Pledge Agreement, and in no event shall Borrower be liable on the Non-Recourse Portion of this Note for any deficiency resulting from any sale of shares pursuant to the Pledge Agreement, nor shall any action or proceeding be brought by Lender against Borrower to recover judgment against Borrower upon the Non-Recourse Portion of this Note or the Pledge Agreement. Upon default of Borrower in the payment of any indebtedness under this Note, Borrower shall be fully liable for all amounts due under this Note other than the Non-Recourse Portion.

At the sole and absolute discretion of Borrower, Borrower may elect to repay some or all of the amounts due and owing hereunder, at any time and from time to time, whether in the event of Default or otherwise, and without the requirement of Lender’s consent or approval, by putting to Lender that number of shares of Stock equal to the amount of such repayment, based upon a price per share of the Stock equal to the fair market value thereof as determined in the most recent third-party appraisal thereof.

This Note amends, restates, and supersedes in all respects that certain Partial Recourse Promissory Note by Joseph Gardner to Lender dated as of June 15, 2011 with respect to the time period beginning on June 15, 2013 and ending on the Maturity Date.

This Note is to be governed and construed in accordance with the laws of the State of Delaware.

IN TESTIMONY WHEREOF, the undersigned has executed this instrument the day and year first above written.

/s/ Joseph H. Gardner

Borrower (Print Name): Joseph Gardner

AMENDED AND RESTATED STOCK PLEDGE AGREEMENT

THIS AMENDED AND RESTATED STOCK PLEDGE AGREEMENT (the "**Agreement**") is made as of this 15th day of June 2013, by and between Joseph Gardner ("**Pledgor**"), and Akebia Therapeutics, Inc., a Delaware corporation ("**Lender**").

WHEREAS, Lender has extended a loan to Pledgor in the principal amount of \$96,026.84 (the "**Loan**"), which Loan is evidenced by a promissory note in favor of Lender dated as of June 15, 2011 (the "**Prior Note**"); and

WHEREAS, the parties have agreed to amend and restate the Prior Note with the promissory note in favor of Lender dated as of June 15, 2013 (the "**Note**"); and

WHEREAS, to secure the payment and performance of all obligations under the terms and conditions of the Note, the Parties wish to amend and restate that certain Stock Pledge Agreement by and between Joseph Gardner and Lender dated June 15, 2011 (the "**Prior Stock Pledge Agreement**") with this Agreement, the terms of which amend, restate and supersede in all respects the Prior Stock Pledge Agreement, and under which Pledgor pledges to Lender all of Pledgor's right, title and interest in the capital stock of Lender owned by Pledgor and listed on **Exhibit A** hereto (the "**Stock**").

NOW, THEREFORE, the parties hereto agree as follows:

1. **Warranty.** Pledgor hereby represents and warrants to Lender that except for the security interest created hereby, Pledgor owns the Stock free and clear of all liens, charges and encumbrances, that the Stock is duly issued, fully paid and nonassessable, and that Pledgor has the unencumbered right to pledge the Stock.

2. **Security Interest.** Pledgor hereby unconditionally grants and assigns to Lender, its successors and assigns, a continuing security interest in the security title to the Stock. Pledgor has delivered to and deposited with Lender herewith all of Pledgor's right, title and interest in and to the Stock, together with certificates representing the Stock and stock powers endorsed in blank by Pledgor, as security for payment and performance of all obligations of Pledgor to Lender under the Note or any extension, renewal, amendment or modification of the Note, however created, acquired, arising or evidenced, whether direct or indirect, absolute or contingent, now or hereafter existing, or due or to become due. Beneficial ownership of the Stock, including, without limitation, all voting, consensual and dividend rights, shall remain in Pledgor until the occurrence of a Default under the terms hereof (as defined in Section 4 below).

3. **Additional Shares.** In the event that, during the term of this Agreement:

(a) any stock dividend, stock split, reclassification, readjustment or other change is declared or made in the capital structure of Lender, all new, substituted and additional shares, or other securities, issued by reason of any such change and received by Pledgor or to which Pledgor shall be entitled shall be immediately delivered to Lender, together with stock powers endorsed in blank by Pledgor, and shall thereupon constitute Stock to be held by Lender under the terms of this Agreement ; and

(b) subscriptions, warrants or any other rights or options are issued in connection with the Stock, all new stock or other securities acquired through such subscriptions, warrants, rights or options by Pledgor shall be immediately delivered to Lender and shall thereupon constitute Stock to be held by Lender under the terms of this Agreement.

4. **Default.** Failure of Pledgor to pay any amount of principal or interest when due pursuant to the terms of the Note or a default by Pledgor under this Agreement shall constitute a default under the terms of this Agreement (any of such occurrences being hereinafter referred to as a “**Default**”). Upon the occurrence of a Default, Lender may take the actions described in the following sentence and thereafter, or may elect, as its sole recourse hereunder and under the Note and full remedy hereunder and thereunder, in full settlement and repayment of all amounts due and owing under the Note (the “**Obligations**”), and without the requirement of Pledgor’s consent or approval, to redeem that number of shares of Stock equal to the amount of the Obligations (or, if the Obligations exceed the total value of the Stock, then all of the Stock), based upon a price per share of the Stock equal to the fair market value thereof as determined in the most recent third-party appraisal thereof. Alternatively, Lender may sell or make other commercially reasonable disposition of the Stock or any portion thereof after ten (10) business days’ written notice to Pledgor, and Lender may purchase the Stock or any portion thereof at any public sale. The proceeds of the public or private sale or other disposition shall be applied (i) to the costs incurred in connection with the sale; (ii) to any unpaid interest which may have accrued on any obligations secured hereby; (iii) to any unpaid principal; and (iv) to damages incurred by Lender by reason of any breach of the obligations secured against hereby, in such order as Lender may determine but in any event the proceeds shall be applied first to the Non-Recourse Portion of the Note (as defined in the Note) and then to the balance of the sums due under the Note, and any remaining proceeds shall be paid over to Pledgor or others as law provides. Pledgor shall not be liable to Lender for any deficiency in the Non-Recourse Portion of the Note in the event the proceeds of the sale or other disposition of the Stock are insufficient to pay such expenses, interest, principal, obligations and damages.

5. **Additional Rights of Secured Parties.** In addition to other rights and privileges under this Agreement, Lender shall have the rights, powers and privileges of secured parties under the Uniform Commercial Code.

6. **Return of Stock to Pledgor.** Upon payment in full of all principal and interest on the Note, Lender shall return to Pledgor all of the then remaining Stock and all rights received by Lender as agent for Pledgor as a result of its possessory interest in the Stock.

7. **Voting Rights.** Pledgor shall retain all rights to vote the Stock until such time as Lender either cancels or sells the Stock after a Default under the Note.

8. Notices. All notices and other communications required or permitted hereunder shall be in writing and, if mailed by prepaid certified mail, shall be deemed to have been received on the earlier of the date shown on the receipt or three (3) business days after the postmarked date thereof. In addition, notices hereunder may be delivered by hand, by facsimile or by email, in which event such notice shall be deemed effective when delivered. Notice of change of address for notice shall also be governed by this Section. Notices shall be addressed as follows:

If to Pledgor: Name: Joseph Gardner
Mailing Address: 4060 Boomer
Cincinnati, OH 45247

If to Lender: Akebia Therapeutics, Inc.
Attention: CFO
Mailing Address: 9987 Carver Road
Cincinnati, OH 45242
Email: afishman@akebia.com

With a copy to:
Thompson Hine LLP
Attention: David J. Willbrand
312 Walnut Street, Suite 1400
Cincinnati, Ohio 45202
Facsimile: (513) 241-4771
Email: David.Willbrand@ThompsonHine.com

9. Binding Agreement. The provisions of this Agreement shall be construed and interpreted, and all rights and obligations of the parties hereto determined, in accordance with the laws of the State of Delaware. This Agreement, together with all documents referred to herein, constitutes the entire agreement between Pledgor and Lender with respect to the matters addressed herein and may not be modified except by a writing executed by Lender and Pledgor. This Agreement may be executed in multiple counterparts and by facsimile or PDF, each of which shall be deemed an original but all of which, taken together, shall constitute one and the same instrument.

10. Severability. If any paragraph or part thereof shall for any reason be held or adjudged to be invalid, illegal or unenforceable by any court of competent jurisdiction, such paragraph or part thereof so adjudicated invalid, illegal or unenforceable shall be deemed separate, distinct and independent, and the remainder of this Agreement shall remain in full force and effect and shall not be affected by such holding or adjudication .

11. Assignability. This Agreement, and the rights and obligations of Lender hereunder, may be assigned by Lender to any person or entity to which the Note is transferred by Lender, and such transferee shall be deemed the "**Lender**" for purposes of this Agreement; provided that the transferee provides written notice of such assignment to Pledgor and agrees to be bound by the terms of this Agreement.

Signature Page Follows

IN WITNESS WHEREOF, the undersigned have hereunto set their hands, by and through their duly authorized officers, as of the day and year first above written.

Pledgor:

/s/ Joseph H. Gardner

Joseph Gardner, CEO

Lender:

Akebia Therapeutics, Inc.

By: /s/ Anupam Dalal

Its: **CFO**

Exhibit A
STOCK CERTIFICATE NUMBERS

<u>Number</u>	<u>Owner</u>	<u>Class of Shares</u>	<u>Number of Shares Represented</u>
12	Joseph Gardner	Common	2,080,557
13	Joseph Gardner	Common	1,485,714
14	Joseph Gardner	Common	2,835,518

*Akebia Therapeutics, Inc.***AMENDED AND RESTATED PARTIAL RECOURSE PROMISSORY NOTE**

\$14,571.54

Cincinnati, Ohio
May 9, 2013

FOR VALUE RECEIVED, Robert Shalwitz ("**Borrower**") promises to pay to Akebia Therapeutics, Inc., a Delaware corporation ("**Lender**"), or order, the principal sum of \$14,571.54 with interest as set forth below, both principal and interest payable in lawful money of the United States of America, at such place as Lender may designate in writing.

The principal and interest shall be due and payable as follows:

Interest shall accrue at the rate of six percent (3%) per annum from the date hereof up to and through the Maturity date (as defined herein). The entire aggregate unpaid principal balance interest shall be due and payable on the first to occur of (a) the consummation of Lender's first underwritten public offering of its Common Stock (other than a registration statement relating either to the sale of securities to employees of Lender pursuant to its stock option, stock purchase or similar plan or an SEC Rule 145 transaction); (b) the consummation of a "*Deemed Liquidation Event*" and distribution of proceeds to or escrow for the benefit of the stockholders of Lender in accordance with Lender's certificate of incorporation as in effect and amended from time to time; or (c) May 9, 2023 (the date of whichever such event occurs first being the "**Maturity Date**" of the Note).

The Note may be prepaid in full or in part at any time without penalty or premium; provided, however, that partial prepayments shall be applied first to the payment of interest accrued to the date of such prepayment and then to the payment of principal.

All parties to this Note, including maker and any sureties, endorsers or guarantors, hereby waive protest, presentment, notice of dishonor and notice of acceleration of maturity and agree to continue to remain bound for the payment of principal, interest and all other sums due under this Note, notwithstanding any change or changes by way of any extension or extensions of time for the payment of principal and interest; and all such parties waive all and every kind of notice of such change or changes and agree that the same may be made without notice or consent of any of them.

As an inducement for Lender to accept from Borrower this Note and as collateral security for the payment of any and all indebtedness and liabilities whatsoever of Borrower to Lender evidenced by this Note, the parties hereto have executed a certain Amended and Restated Stock Pledge Agreement of even date herewith (the "**Pledge Agreement**"), substantially in the form attached hereto as Exhibit A, pursuant to which Borrower has delivered, assigned and pledged to Lender and has granted to Lender a first priority security interest in 9,714.36 shares of Common Stock of Lender owned by Borrower (the "**Stock**").

Upon default of Borrower in the payment of any indebtedness under this Note, Lender's sole recourse with respect to fifty percent (50%) of the sum of (a) unpaid principal of this Note, (b)

accrued but unpaid interest on this Note, and (c) collection costs including attorneys' fees in connection therewith (the "**Non-Recourse Portion**") shall be to exercise its rights under the Pledge Agreement. Liability of Borrower under the Non-Recourse Portion of this Note is limited to the shares held by Lender pursuant to the Pledge Agreement, and in no event shall Borrower be liable on the Non-Recourse Portion of this Note for any deficiency resulting from any sale of shares pursuant to the Pledge Agreement, nor shall any action or proceeding be brought by Lender against Borrower to recover judgment against Borrower upon the Non-Recourse Portion of this Note or the Pledge Agreement. Upon default of Borrower in the payment of any indebtedness under this Note, Borrower shall be fully liable for all amounts due under this Note other than the Non-Recourse Portion.

At the sole and absolute discretion of Borrower, Borrower may elect to repay some or all of the amounts due and owing hereunder, at any time and from time to time, whether in the event of Default or otherwise, and without the requirement of Lender's consent or approval, by putting to Lender that number of shares of Stock equal to the amount of such repayment, based upon a price per share of the Stock equal to the fair market value thereof as determined in the most recent third-party appraisal thereof.

This Note amends, restates, and supersedes in all respects that certain Partial Recourse Promissory Note by Robert Shalwitz to Lender dated as of October 15, 2009 with respect to the time period beginning on May 9, 2013 and ending on the Maturity Date.

This Note is to be governed and construed in accordance with the laws of the State of Delaware.

IN TESTIMONY WHEREOF, the undersigned has executed this instrument the day and year first above written.

/s/ Robert Shalwitz

Borrower (Print Name): Robert Shalwitz

ACKNOWLEDGED AND ACCEPTED

AKEBIA THERAPEUTICS, INC.

By: /s/ Joseph H. Gardner
Name: Joseph H. Gardner
Title: President & CEO

Stock Pledge Agreement

Akebia Therapeutics, Inc.

AMENDED AND RESTATED STOCK PLEDGE AGREEMENT

THIS AMENDED AND RESTATED STOCK PLEDGE AGREEMENT (the "**Agreement**") is made as of this 9th day of May, 2013, by and between Robert Shalwitz ("**Pledgor**"), and Akebia Therapeutics, Inc., a Delaware corporation ("**Lender**").

WHEREAS, Lender has extended a loan to Pledgor in the principal amount of \$14,571.54 (the "**Loan**"), which Loan is evidenced by a promissory note in favor of Lender attached hereto as **Exhibit A** (the "**Note**"); and

WHEREAS, to secure the payment and performance of all obligations under the Note, Pledgor wishes to pledge to Lender all of Pledgor's right, title and interest in the capital stock of Lender owned by Pledgor and listed on **Exhibit B** hereto (the "**Stock**").

NOW, THEREFORE, the parties hereto agree as follows:

1. **Warranty.** Pledgor hereby represents and warrants to Lender that except for the security interest created hereby, Pledgor owns the Stock free and clear of all liens, charges and encumbrances, that the Stock is duly issued, fully paid and nonassessable, and that Pledgor has the unencumbered right to pledge the Stock.

2. **Security Interest.** Pledgor hereby unconditionally grants and assigns to Lender, its successors and assigns, a continuing security interest in the security title to the Stock. Pledgor has delivered to and deposited with Lender herewith all of Pledgor's right, title and interest in and to the Stock, together with certificates representing the Stock and stock powers endorsed in blank by Pledgor, as security for payment and performance of all obligations of Pledgor to Lender under the Note or any extension, renewal, amendment or modification of the Note, however created, acquired, arising or evidenced, whether direct or indirect, absolute or contingent, now or hereafter existing, or due or to become due. Beneficial ownership of the Stock, including, without limitation, all voting, consensual and dividend rights, shall remain in Pledgor until the occurrence of a Default under the terms hereof (as defined in Section 4 below).

3. **Additional Shares.** In the event that, during the term of this Agreement:

(a) any stock dividend, stock split, reclassification, readjustment or other change is declared or made in the capital structure of Lender, all new, substituted and additional shares, or other securities, issued by reason of any such change and received by Pledgor or to which Pledgor shall be entitled shall be immediately delivered to Lender, together with stock powers endorsed in blank by Pledgor, and shall thereupon constitute Stock to be held by Lender under the terms of this Agreement; and

(b) subscriptions, warrants or any other rights or options are issued in connection with the Stock, all new stock or other securities acquired through such subscriptions, warrants, rights or options by Pledgor shall be immediately delivered to Lender and shall thereupon constitute Stock to be held by Lender under the terms of this Agreement.

4. Default. Failure of Pledgor to pay any amount of principal or interest when due pursuant to the terms of the Note or a default by Pledgor under this Agreement shall constitute a default under the terms of this Agreement (any of such occurrences being hereinafter referred to as a “**Default**”). Upon the occurrence of a Default, Lender may take the actions described in the following sentence and thereafter, or may elect, as its sole recourse hereunder and under the Note and full remedy hereunder and thereunder, in full settlement and repayment of all amounts due and owing under the Note (the “**Obligations**”), and without the requirement of Pledgor’s consent or approval, to redeem that number of shares of Stock equal to the amount of the Obligations (or, if the Obligations exceed the total value of the Stock, then all of the Stock), based upon a price per share of the Stock equal to the fair market value thereof as determined in the most recent third-party appraisal thereof. Alternatively, Lender may sell or make other commercially reasonable disposition of the Stock or any portion thereof after ten (10) business days’ written notice to Pledgor, and Lender may purchase the Stock or any portion thereof at any public sale. The proceeds of the public or private sale or other disposition shall be applied (i) to the costs incurred in connection with the sale; (ii) to any unpaid interest which may have accrued on any obligations secured hereby; (iii) to any unpaid principal; and (iv) to damages incurred by Lender by reason of any breach of the obligations secured against hereby, in such order as Lender may determine but in any event the proceeds shall be applied first to the Non-Recourse Portion of the Note (as defined in the Note) and then to the balance of the sums due under the Note, and any remaining proceeds shall be paid over to Pledgor or others as law provides. Pledgor shall not be liable to Lender for any deficiency in the Non-Recourse Portion of the Note in the event the proceeds of the sale or other disposition of the Stock are insufficient to pay such expenses, interest, principal, obligations and damages.

5. Additional Rights of Secured Parties. In addition to other rights and privileges under this Agreement, Lender shall have the rights, powers and privileges of secured parties under the Uniform Commercial Code.

6. Return of Stock to Pledgor. Upon payment in full of all principal and interest on the Note, Lender shall return to Pledgor all of the then remaining Stock and all rights received by Lender as agent for Pledgor as a result of its possessory interest in the Stock.

7. Voting Rights. Pledgor shall retain all rights to vote the Stock until such time as Lender either cancels or sells the Stock after a Default under the Note.

8. Notices. All notices and other communications required or permitted hereunder shall be in writing and, if mailed by prepaid certified mail, shall be deemed to have been received on the earlier of the date shown on the receipt or three (3) business days after the postmarked date thereof. In addition, notices hereunder may be delivered by hand, by facsimile or by email, in which event such notice shall be deemed effective when delivered. Notice of change of address for notice shall also be governed by this Section. Notices shall be addressed as follows:

If to Pledgor: Robert Shalwitz
2549 Bryden Road
Bexley, Ohio 43209

If to Lender: Akebia Therapeutics, Inc.
Attention: Joseph Gardner
President and Chief Executive Officer
9987 Carver Road, Suite 420
Cincinnati, Ohio 45242
Email: JGardner@Akebia.com

With a copy to:
Thompson Hine LLP
Attention: David J. Willbrand
312 Walnut Street, Suite 1400
Cincinnati, Ohio 45202
Facsimile: (513) 241-4771
Email: David.Willbrand@ThompsonHine.com

9. Binding Agreement. This Agreement amends, restates and supersedes in all respects the Stock Pledge Agreement between Pledgor and Lender dated as of October 15, 2009. The provisions of this Agreement shall be construed and interpreted, and all rights and obligations of the parties hereto determined, in accordance with the laws of the State of Delaware. This Agreement, together with all documents referred to herein, constitutes the entire agreement between Pledgor and Lender with respect to the matters addressed herein and may not be modified except by a writing executed by Lender and Pledgor. This Agreement may be executed in multiple counterparts and by facsimile or PDF, each of which shall be deemed an original but all of which, taken together, shall constitute one and the same instrument.

10. Severability. If any paragraph or part thereof shall for any reason be held or adjudged to be invalid, illegal or unenforceable by any court of competent jurisdiction, such paragraph or part thereof so adjudicated invalid, illegal or unenforceable shall be deemed separate, distinct and independent, and the remainder of this Agreement shall remain in full force and effect and shall not be affected by such holding or adjudication.

11. Assignability. This Agreement, and the rights and obligations of Lender hereunder, may be assigned by Lender to any person or entity to which the Note is transferred by Lender, and such transferee shall be deemed the "Lender" for purposes of this Agreement; provided that the transferee provides written notice of such assignment to Pledgor and agrees to be bound by the terms of this Agreement.

Signature Page Follows

IN WITNESS WHEREOF, the undersigned have hereunto set their hands, by and through their duly authorized officers, as of the day and year first above written.

Pledgor:

/s/ Robert Shalwitz

Print Name: Robert Shalwitz

Lender:

Akebia Therapeutics, Inc.

By: /s/ Joseph H. Gardner

Name: Joseph H. Gardner

Title: CEO

NOTE

Exhibit B

STOCK CERTIFICATE NUMBERS

<u>Number</u>	<u>Owner</u>	<u>Class of Shares</u>	<u>Number of Shares Represented</u>
—	Robert Shalwitz	Common	9,714.36

Akebia Therapeutics, Inc.

AMENDED AND RESTATED PARTIAL RECOURSE PROMISSORY NOTE

\$71,834.85

Cincinnati, Ohio
June 15, 2013

FOR VALUE RECEIVED, Robert Shalwitz ("**Borrower**") promises to pay to Akebia Therapeutics, Inc., a Delaware corporation ("**Lender**"), or order, the principal sum of \$71,834.85 with interest as set forth below, both principal and interest payable in lawful money of the United States of America, at such place as Lender may designate in writing.

The principal and interest shall be due and payable as follows:

Interest shall accrue at the rate of six percent (3%) per annum from the date hereof up to and through the Maturity date (as defined herein). The entire aggregate unpaid principal balance interest shall be due and payable on the first to occur of (a) the consummation of Lender's first underwritten public offering of its Common Stock (other than a registration statement relating either to the sale of securities to employees of Lender pursuant to its stock option, stock purchase or similar plan or an SEC Rule 145 transaction); (b) the consummation of a "*Deemed Liquidation Event*" and distribution of proceeds to our escrow for the benefit of the stockholders of Lender in accordance with Lender's certificate of incorporation as in effect and amended from time to time; or (c) June 15, 2023 (the date of whichever such event occurs first being the "**Maturity Date**" of the Note).

The Note may be prepaid in full or in part at any time without penalty or premium; provided, however, that partial prepayments shall be applied first to the payment of interest accrued to the date of such prepayment and then to the payment of principal.

All parties to this Note, including maker and any sureties, endorsers or guarantors, hereby waive protest, presentment, notice of dishonor and notice of acceleration of maturity and agree to continue to remain bound for the payment of principal, interest and all other sums due under this Note, notwithstanding any change or changes by way of any extension or extensions of time for the payment of principal and interest; and all such parties waive all and every kind of notice of such change or changes and agree that the same may be made without notice or consent of any of them.

As an inducement for Lender to accept from Borrower this Note and as collateral security for the payment of any and all indebtedness and liabilities whatsoever of Borrower to Lender evidenced by this Note, the parties hereto have executed a certain Stock Pledge Agreement of even date herewith (the "**Pledge Agreement**"), pursuant to which Borrower has delivered, assigned and pledged to Lender and has granted to Lender a first priority security interest in 4,788,989 shares of Common Stock of Lender owned by Borrower (the "**Stock**").

Upon default of Borrower in the payment of any indebtedness under this Note, Lender's sole recourse with respect to fifty percent (50%) of the sum of (a) unpaid principal of this Note, (b)

accrued but unpaid interest on this Note, and (c) collection costs including attorneys' fees in connection therewith (the "**Non-Recourse Portion**") shall be to exercise its rights under the Pledge Agreement. Liability of Borrower under the Non-Recourse Portion of this Note is limited to the shares held by Lender pursuant to the Pledge Agreement, and in no event shall Borrower be liable on the Non-Recourse Portion of this Note for any deficiency resulting from any sale of shares pursuant to the Pledge Agreement, nor shall any action or proceeding be brought by Lender against Borrower to recover judgment against Borrower upon the Non-Recourse Portion of this Note or the Pledge Agreement. Upon default of Borrower in the payment of any indebtedness under this Note, Borrower shall be fully liable for all amounts due under this Note other than the Non-Recourse Portion.

At the sole and absolute discretion of Borrower, Borrower may elect to repay some or all of the amounts due and owing hereunder, at any time and from time to time, whether in the event of Default or otherwise, and without the requirement of Lender's consent or approval, by putting to Lender that number of shares of Stock equal to the amount of such repayment, based upon a price per share of the Stock equal to the fair market value thereof as determined in the most recent third-party appraisal thereof.

This Note amends, restates, and supersedes in all respects that certain Partial Recourse Promissory Note by Robert Shalwitz to Lender dated as of June 15, 2011 with respect to the time period beginning on June 15, 2013 and ending on the Maturity Date.

This Note is to be governed and construed in accordance with the Laws of the State of Delaware.

IN TESTIMONY WHEREOF, the undersigned has executed this instrument the day and year first above written.

/s/ Robert Shalwitz

Borrower (Print Name): Robert Shalwitz

Akebia Therapeutics, Inc.

AMENDED AND RESTATED STOCK PLEDGE AGREEMENT

THIS AMENDED AND RESTATED STOCK PLEDGE AGREEMENT (the "**Agreement**") is made as of this 15th day of June 2013, by and between Robert Shalwitz ("**Pledgor**"), and Akebia Therapeutics, Inc., a Delaware corporation ("**Lender**").

WHEREAS, Lender has extended a loan to Pledgor in the principal amount of \$71,834.85 (the "**Loan**"), which Loan is evidenced by a promissory note in favor of Lender dated as of June 15, 2011 (the "**Prior Note**"); and

WHEREAS, the Parties have agreed to amend and restate the Prior Note with the promissory note in favor of Lender dated as of June 15, 2013 (the "**Note**"); and

WHEREAS, to secure the payment and performance of all obligations under the terms and conditions of the Note, the Parties wish to amend and restate that certain Stock Pledge Agreement by and between Robert Shalwitz and Lender dated June 15, 2011 (the "**Prior Stock Pledge Agreement**") with this Agreement, the terms of which amend, restate and supersede in all respects the Prior Stock Pledge Agreement, and under which Pledgor pledges to Lender all of Pledgor's right, title and interest in the capital stock of Lender owned by Pledgor and listed on **Exhibit A** hereto (the "**Stock**").

NOW, THEREFORE, the parties hereto agree as follows:

1. Warranty. Pledgor hereby represents and warrants to Lender that except for the security interest created hereby, Pledgor owns the Stock free and clear of all liens, charges and encumbrances, that the Stock is duly issued, fully paid and nonassessable, and that Pledgor has the unencumbered right to pledge the Stock.

2. Security Interest. Pledgor hereby unconditionally grants and assigns to Lender, its successors and assigns, a continuing security interest in the security title to the Stock. Pledgor has delivered to and deposited with Lender herewith all of Pledgor's right, title and interest in and to the Stock, together with certificates representing the Stock and stock powers endorsed in blank by Pledgor, as security for payment and performance of all obligations of Pledgor to Lender under the Note or any extension, renewal, amendment or modification of the Note, however created, acquired, arising or evidenced, whether direct or indirect, absolute or contingent, now or hereafter existing, or due or to become due. Beneficial ownership of the Stock, including, without limitation, all voting, consensual and dividend rights, shall remain in Pledgor until the occurrence of a Default under the terms hereof (as defined in Section 4 below).

3. Additional Shares. In the event that, during the term of this Agreement:

(a) any stock dividend, stock split, reclassification, readjustment or other change is declared or made in the capital structure of Lender, all new, substituted and additional shares, or other securities, issued by reason of any such change and received by Pledgor or to which Pledgor shall be entitled shall be immediately delivered to Lender, together with stock powers endorsed in blank by Pledgor, and shall thereupon constitute Stock to be held by Lender under the terms of this Agreement; and

(b) subscriptions, warrants or any other rights or options are issued in connection with the Stock, all new stock or other securities acquired through such subscriptions, warrants, rights or options by Pledgor shall be immediately delivered to Lender and shall thereupon constitute Stock to be held by Lender under the terms of this Agreement.

4. **Default.** Failure of Pledgor to pay any amount of principal or interest when due pursuant to the terms of the Note or a default by Pledgor under this Agreement shall constitute a default under the terms of this Agreement (any of such occurrences being hereinafter referred to as a “**Default**”). Upon the occurrence of a Default, Lender may take the actions described in the following sentence and thereafter, or may elect, as its sole recourse hereunder and under the Note and full remedy hereunder and thereunder, in full settlement and repayment of all amounts due and owing under the Note (the “**Obligations**”), and without the requirement of Pledgor’s consent or approval, to redeem that number of shares of Stock equal to the amount of the Obligations (or, if the Obligations exceed the total value of the Stock, then all of the Stock), based upon a price per share of the Stock equal to the fair market value thereof as determined in the most recent third-party appraisal thereof. Alternatively, Lender may sell or make other commercially reasonable disposition of the Stock or any portion thereof after ten (10) business days’ written notice to Pledgor, and Lender may purchase the Stock or any portion thereof at any public sale. The proceeds of the public or private sale or other disposition shall be applied (i) to the costs incurred in connection with the sale; (ii) to any unpaid interest which may have accrued on any obligations secured hereby; (iii) to any unpaid principal; and (iv) to damages incurred by Lender by reason of any breach of the obligations secured against hereby, in such order as Lender may determine but in any event the proceeds shall be applied first to the Non-Recourse Portion of the Note (as defined in the Note) and then to the balance of the sums due under the Note, and any remaining proceeds shall be paid over to Pledgor or others as law provides. Pledgor shall not be liable to Lender for any deficiency in the Non-Recourse Portion of the Note in the event the proceeds of the sale or other disposition of the Stock are insufficient to pay such expenses, interest, principal, obligations and damages.

5. **Additional Rights of Secured Parties.** In addition to other rights and privileges under this Agreement, Lender shall have the rights, powers and privileges of secured parties under the Uniform Commercial Code.

6. **Return of Stock to Pledgor.** Upon payment in full of all principal and interest on the Note, Lender shall return to Pledgor all of the then remaining Stock and all rights received by Lender as agent for Pledgor as a result of its possessory interest in the Stock.

7. **Voting Rights.** Pledgor shall retain all rights to vote the Stock until such time as Lender either cancels or sells the Stock after a Default under the Note.

8. **Notices.** All notices and other communications required or permitted hereunder shall be in writing and, if mailed by prepaid certified mail, shall be deemed to have been received on the earlier of the date shown on the receipt or three (3) business days after the postmarked date thereof. In addition, notices hereunder may be delivered by hand, by facsimile

or by email, in which event such notice shall be deemed effective when delivered. Notice of change of address for notice shall also be governed by this Section. Notices shall be addressed as follows:

If to Pledgor: Name: Robert Shalwitz
Mailing Address: 2549 Bryden Road
Bexley, OH 43209
Email: rshalwitz@akebia.com

If to Lender: Akebia Therapeutics, Inc.
Attention: CEO
Mailing Address: 9987 Carver Road
Cincinnati, OH 45242
Facsimile: 513 985 0999
Email: afishman@akebia.com

With a copy to:

Thompson Hine LLP
Attention: David J. Willbrand
312 Walnut Street, Suite 1400
Cincinnati, Ohio 45202
Facsimile: (513) 241-4771
Email: David.Willbrand@Thompsonline.com

9. Binding Agreement. The provisions of this Agreement shall be construed and interpreted, and all rights and obligations of the parties hereto determined, in accordance with the laws of the State of Delaware. This Agreement, together with all documents referred to herein, constitutes the entire agreement between Pledgor and Lender with respect to the matters addressed herein and may not be modified except by a writing executed by Lender and Pledgor. This Agreement may be executed in multiple counterparts and by facsimile or PDF, each of which shall be deemed an original but all of which, taken together, shall constitute one and the same instrument.

10. Severability. If any paragraph or part thereof shall for any reason be held or adjudged to be invalid, illegal or unenforceable by any court of competent jurisdiction, such paragraph or part thereof so adjudicated invalid, illegal or unenforceable shall be deemed separate, distinct and independent, and the remainder of this Agreement shall remain in full force and effect and shall not be affected by such holding or adjudication.

11. Assignability. This Agreement, and the rights and obligations of Lender hereunder, may be assigned by Lender to any person or entity to which the Note is transferred by Lender, and such transferee shall be deemed the "Lender" for purposes of this Agreement; provided that the transferee provides written notice of such assignment to Pledgor and agrees to be bound by the terms of this Agreement.

Signature Page Follows

IN WITNESS WHEREOF, the undersigned have hereunto set their hands, by and through their duly authorized officers, as of the day and year first above written.

Pledgor:

/s/ Robert Shalwitz

Robert Shalwitz

Lender:

Akebia Therapeutics, Inc.

By: /s/ Joseph H. Gardner

Its: CEO

Joseph H. Gardner

President & CEO

Exhibit A

STOCK CERTIFICATE NUMBERS

<u>Number</u>	<u>Owner</u>	<u>Class of Shares</u>	<u>Number of Shares Represented</u>
15	Robert Shalwitz	Common	1,804,095
16	Robert Shalwitz	Common	2,242,038
17	Robert Shalwitz	Common	742,857