

Ultragenyx Pharmaceutical Inc.  
Form DEF 14A  
May 23, 2014  
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**UNITED STATES**  
**SECURITIES AND EXCHANGE COMMISSION**  
**Washington, D.C. 20549**

**SCHEDULE 14A INFORMATION**  
**Proxy Statement Pursuant to Section 14(a) of the**  
**Securities Exchange Act of 1934**

Filed by the Registrant ☒ x

Filed by a party other than the Registrant ☐ ..

Check the appropriate box:

- ☐ .. Preliminary Proxy Statement  
☐ .. **Confidential, for Use of the Commission Only (as permitted by Rule 14a-6(e)(2))**  
☒ x Definitive Proxy Statement  
☐ .. Definitive Additional Materials  
☐ .. Soliciting Material Pursuant to §240.14a-12

**ULTRAGENYX PHARMACEUTICAL INC.**

**(Name of Registrant as Specified In Its Charter)**

**N/A**

**(Name of Person(s) Filing Proxy Statement, if Other Than The Registrant)**

Payment of Filing Fee (Check the appropriate box):

x No fee required.

.. Fee computed on table below per Exchange Act Rules 14a-6(i)(1) and 0-11.

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(1) Amount Previously Paid:

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(3) Filing Party:

(4) Date Filed:

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**60 Leveroni Court**

**Novato, California 94949**

**NOTICE OF ANNUAL MEETING OF STOCKHOLDERS**

**To Be Held on July 10, 2014**

Dear Stockholder:

You are cordially invited to attend the Annual Meeting of Stockholders of Ultragenyx Pharmaceutical Inc., a Delaware corporation (the **Company**), which will be held on July 10, 2014, at 4:00 p.m. Pacific Time virtually via the Internet at [www.virtualshareholdermeeting.com/RARE14](http://www.virtualshareholdermeeting.com/RARE14) (the **Annual Meeting**). Instructions on how to participate in the Annual Meeting and demonstrate proof of stock ownership are posted at [www.virtualshareholdermeeting.com/RARE14](http://www.virtualshareholdermeeting.com/RARE14). The webcast of the Annual Meeting will be archived for one year after the date of the Annual Meeting at [www.virtualshareholdermeeting.com/RARE14](http://www.virtualshareholdermeeting.com/RARE14). Only stockholders who held stock at the close of business on the record date, May 16, 2014, may vote at the Annual Meeting, including any adjournment or postponement thereof.

At the Annual Meeting, you will be asked to consider and vote upon: (1) the reelection of the one director named in the Proxy Statement as a Class I director, (2) the ratification of the selection of Ernst & Young LLP as our independent registered public accounting firm for the fiscal year ending December 31, 2014, (3) the reapproval of the Internal Revenue Code Section 162(m) provisions of the 2014 Incentive Plan, (4) the reapproval of the Internal Revenue Code Section 162(m) provisions of the Corporate Bonus Plan and (5) any other business that may properly come before the Annual Meeting or any adjournment thereof. No other items of business are expected to be considered, and no other director nominees will be entertained, at the Annual Meeting.

The accompanying Proxy Statement more fully describes the details of the business to be conducted at the Annual Meeting. Proposal 1 relates solely to the reelection of the director nominated by the Board of Directors and does not include any other matters relating to the election of directors, including, without limitation, the election of directors nominated by any stockholder of the Company. After careful consideration, our Board of Directors has unanimously approved the proposals and recommends that you vote FOR the director nominee and FOR each other proposal described in the Proxy Statement. In accordance with Delaware law, a list of stockholders entitled to vote at the meeting will be available in electronic form during the annual meeting at the following URL: [www.virtualshareholdermeeting.com/RARE14](http://www.virtualshareholdermeeting.com/RARE14), and will be accessible for ten days prior to the meeting at our principal place of business, 60 Leveroni Court, Novato, California 94949.

We are pleased to make use of the Securities and Exchange Commission rules that allow companies to furnish proxy materials to their stockholders via the Internet. We believe the ability to deliver proxy materials electronically allows us to provide our stockholders with the information they need, while lowering the costs of delivery and reducing the environmental impact from the distribution of our Annual Meeting materials.

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We look forward to speaking with you at the Annual Meeting.

Sincerely,

Emil D. Kakkis, M.D., Ph.D.

President and Chief Executive Officer

May 23, 2014

**WHETHER OR NOT YOU EXPECT TO ATTEND THE ANNUAL MEETING, PLEASE VOTE VIA THE INTERNET OR OVER THE TELEPHONE AS INSTRUCTED IN THE *NOTICE REGARDING THE AVAILABILITY OF PROXY MATERIALS* AND ON THE ENCLOSED PROXY CARD OR, IF YOU REQUESTED AND RECEIVED A PRINTED COPY OF THE PROXY STATEMENT, COMPLETE, DATE, SIGN AND RETURN THE ENCLOSED PROXY CARD USING THE ENCLOSED RETURN ENVELOPE, AS PROMPTLY AS POSSIBLE SO THAT YOUR SHARES MAY BE REPRESENTED AT THE ANNUAL MEETING. EVEN IF YOU HAVE VOTED BY PROXY, YOU MAY STILL VOTE THROUGH OUR VIRTUAL WEB CONFERENCE IF YOU ATTEND THE ANNUAL MEETING.**

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**60 Leveroni Court**

**Novato, California 94949**

**PROXY STATEMENT FOR**

**2014 ANNUAL MEETING OF STOCKHOLDERS**

**TO BE HELD ON JULY 10, 2014**

**GENERAL INFORMATION**

This Proxy Statement is furnished in connection with the solicitation of proxies by the Board of Directors (the **Board**) of Ultragenyx Pharmaceutical Inc. (**Ultragenyx** or the **Company**) for use at the Company's 2014 Annual Meeting of Stockholders, to be held virtually via the Internet at [www.virtualshareholdermeeting.com/RARE14](http://www.virtualshareholdermeeting.com/RARE14) on July 10, 2014, at 4:00 p.m. Pacific Time. The Notice Regarding the Availability of Proxy Materials (the **Notice**) containing instructions on how to access this proxy statement and our annual report is first being mailed on or about May 30, 2014 to all stockholders entitled to vote at the Annual Meeting. Pursuant to the rules promulgated by the Securities and Exchange Commission (the **SEC**), we have elected to provide access to our materials by notifying you of the availability of our proxy materials on the Internet. The proxy statement and 2013 Annual Report are available at [www.ultragenyx.com](http://www.ultragenyx.com) in the SEC Filings subsection of the Investors tab.

The Notice instructs you as to how you may access and review important information contained in the proxy materials. The Notice also instructs you on how you may submit your proxy via the Internet. If you receive a Notice by mail and would like to receive a printed copy of our proxy materials, you should follow the instructions for requesting such materials included in the Notice.

For a proxy to be effective, it must be properly executed and received prior to the Annual Meeting. Each proxy properly tendered will, unless otherwise directed by the stockholder, be voted for the proposals and nominee described in this Proxy Statement and at the discretion of the proxy holder(s) with regard to all other matters that may properly come before the Annual Meeting.

The Company will pay all of the costs of soliciting proxies. We will provide copies of our proxy materials to brokerage firms, fiduciaries and custodians for forwarding to beneficial owners who request printed copies of these materials and will reimburse these persons for their costs of forwarding these materials. Our directors, officers and employees may also solicit proxies by telephone, facsimile, or personal solicitation; however, we will not pay them additional compensation for any of these services.

**Shares Outstanding and Voting Rights**

Only holders of record of our common stock at the close of business on May 16, 2014 (the **Record Date**), are entitled to notice of and to vote at the Annual Meeting. On the Record Date, 30,059,288 shares of common stock were issued and outstanding. Each share of common stock is entitled to one vote on all matters to



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be voted upon at the Annual Meeting. Holders of common stock do not have the right to cumulative voting in the election of directors. The presence, in person or by proxy, of the holders of a majority of the outstanding shares on the Record Date will constitute a quorum for the transaction of business at the Annual Meeting and any postponement or adjournment thereof, though the Board may fix a new record date for purposes of a postponed or adjourned meeting.

Persons who hold shares of Ultragenyx common stock directly on the Record Date and not through a broker, bank or other financial institution ( **registered holders** ) must vote over the telephone (by calling 1-800-690-6903 until 11:59 p.m. Eastern Time the day before the Annual Meeting), through the Internet (you may vote before the meeting by going to [www.proxyvote.com](http://www.proxyvote.com) until 11:59 p.m. Eastern Time the day before the Annual Meeting or you may vote during the meeting by going to [www.virtualshareholdermeeting.com/RARE14](http://www.virtualshareholdermeeting.com/RARE14)), return an executed proxy card (that we must receive before the Annual Meeting) or attend the Annual Meeting in order to vote on the proposals. Persons who hold shares of Ultragenyx common stock indirectly on the Record Date through a brokerage firm, bank or other financial institution ( **beneficial holders** ) must return a voting instruction form to have their shares voted on their behalf. Brokerage firms, banks or other financial institutions that do not receive voting instructions from beneficial holders may either vote these shares on behalf of the beneficial holders with respect to routine matters or return a proxy leaving these shares un-voted (a **broker non-vote** ). The ratification of the selection of the independent registered public accounting firm in Proposal No. 2 is the only item on the agenda for the Annual Meeting that is considered routine. Abstentions and broker non-votes will be counted for the purpose of determining the presence or absence of a quorum. The required vote for each of the proposals expected to be acted upon at the Annual Meeting and the treatment of abstentions and broker non-votes under each proposal are described below:

*Proposal No. 1 Reelection of director.* Directors are elected by a plurality of the votes cast, with the nominee obtaining the most votes being elected. Because there is no minimum vote required, abstentions and broker non-votes will have no effect on the outcome.

*Proposal No. 2 Ratification of selection of independent registered public accounting firm.* This proposal must be approved by a majority of votes cast on the proposal. As a result, abstentions and broker non-votes will have no effect on the vote outcome.

*Proposal No. 3 Reapproval of the Internal Revenue Code Section 162(m) provisions of the Company's 2014 Incentive Plan.* This proposal must be approved by a majority of votes cast on the proposal. As a result, abstentions and broker non-votes will have no effect on the vote outcome.

*Proposal No. 4 Reapproval of the Internal Revenue Code Section 162(m) provisions of the Company's Corporate Bonus Plan.* This proposal must be approved by a majority of votes cast on the proposal. As a result, abstentions and broker non-votes will have no effect on the vote outcome.

We encourage you to vote by proxy, whether via telephone (you may vote by calling 1-800-690-6903 until 11:59 p.m. Eastern Time the day before the Annual Meeting), through the Internet (you may vote before the meeting by going to [www.proxyvote.com](http://www.proxyvote.com) until 11:59 p.m. Eastern Time the day before the Annual Meeting or you may vote during the meeting by going to [www.virtualshareholdermeeting.com/RARE14](http://www.virtualshareholdermeeting.com/RARE14)) or by mailing an executed proxy card (that we must receive before the Annual Meeting). Voting in advance of the Annual Meeting reduces the likelihood that the Company will be forced to incur additional expenses soliciting proxies for the Annual Meeting. Any registered holder of our common stock may attend the Annual Meeting and may change or revoke a delivered proxy by:

executing a new proxy card, and submitting it via telephone, through the Internet or by mail as instructed above in advance of the applicable deadline;

delivering a written revocation to the corporate secretary before the Annual Meeting; or

voting at the Annual Meeting.

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Beneficial holders of our common stock who wish to change or revoke their voting instructions should contact their brokerage firm, bank or other financial institution for information on how to do so. Beneficial holders who wish to vote at the Annual Meeting may go to [www.virtualshareholdermeeting.com/RARE14](http://www.virtualshareholdermeeting.com/RARE14) and follow the instructions regarding voting.

## **Explanatory Note**

We are an emerging growth company as defined in the Jumpstart Our Business Startups Act of 2012 (the JOBS Act). For as long as we are an emerging growth company, we will not be required to provide an auditor's attestation report on management's assessment of the effectiveness of our system of internal control over financial reporting pursuant to Section 404(b) of the Sarbanes-Oxley Act of 2002, or obtain stockholder approval of any golden parachute payments not previously approved. In addition, because we are an emerging growth company, we are not required to include a Compensation Discussion and Analysis section in this proxy statement and have elected to comply with the scaled-down executive compensation disclosure requirements applicable to emerging growth companies.

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**PROPOSAL NO. 1 ELECTION OF DIRECTORS**

The Amended and Restated Certificate of Incorporation of the Company provides that the Board is to be divided into three classes as nearly equal in number as reasonably possible, with directors in each class generally serving three-year terms. The total Board size is currently fixed at six directors. Currently, the Class I directors (whose terms expire at the Annual Meeting) are Emil D. Kakkis, M.D., Ph.D. and Mårten Steen, M.D., Ph.D.; Dr. Steen is not re-nominated for service on the Board. The Class II directors (whose terms expire at the 2015 annual meeting of stockholders) are Eran Nadav, Ph.D. and Clay B. Siegall, Ph.D. The Class III directors (whose terms expire at the 2016 annual meeting of stockholders) are William Aliski and Matthew K. Fust. The Class I director elected at the Annual Meeting will hold office until the 2017 annual meeting of stockholders and until his successor is elected and qualified, unless he resigns or his seat becomes vacant due to death, removal, or other cause in accordance with the Amended and Restated Bylaws of the Company.

As described below, the Board has nominated Dr. Kakkis for reelection as a Class I director at the Annual Meeting. Concurrent with the expiration of Dr. Steen's term at the Annual Meeting, the size of the Board will be fixed at five directors. Dr. Kakkis has indicated his willingness to serve if reelected. Should Dr. Kakkis become unavailable for reelection at the Annual Meeting, the persons named on the enclosed proxy as proxy holders may vote all proxies given in response to this solicitation for the election of a substitute nominee chosen by the Board or the Board may reduce the total size of the Board.

**Nomination of Directors**

The Nominating and Corporate Governance Committee reviews and recommends to the Board potential nominees for election to the Board. In reviewing potential nominees, the Nominating and Corporate Governance Committee considers the qualifications of each potential nominee in light of the Board's existing and desired mix of experience and expertise. Specifically, the Nominating and Corporate Governance Committee considers each potential nominee's personal and professional ethics, integrity and values, experience and interest in the Company, and commitment to the representation of the long-term interests of the stockholders. The Board membership criteria are set forth in our Corporate Governance Guidelines, a copy of which is available on our website at [www.ultragenyx.com](http://www.ultragenyx.com) in the Corporate Governance subsection of the Investors tab.

After reviewing the qualifications of potential Board candidates, the Nominating and Corporate Governance Committee presents its recommendations to the Board, which selects the final director nominees. Upon the recommendation of the Nominating and Corporate Governance Committee, the Board nominated Dr. Kakkis for reelection as a Class I director. The Company did not pay any fees to any third parties to identify or assist in identifying or evaluating nominees for the Annual Meeting.

The Nominating and Corporate Governance Committee considers stockholder nominees using the same criteria set forth above. Stockholders who wish to present a potential nominee to the Nominating and Corporate Governance Committee for consideration for election at a future annual meeting of stockholders must provide the Nominating and Corporate Governance Committee with notice of the recommendation and certain information regarding the candidate within the time periods set forth on page 35 under the caption Stockholder Proposals.

Although the Nominating and Corporate Governance Committee may consider whether nominees assist in achieving a mix of Board members that represents a diversity of background and experience, which is not only limited to race, gender or national origin, we have no formal policy regarding board diversity.

**Nominees and Incumbent Directors**

The Nominating and Corporate Governance Committee has recommended, and the Board has nominated, Dr. Kakkis to be reelected as a Class I director at the Annual Meeting. The following table sets forth the following information for Dr. Kakkis and the Company's continuing directors: the year each was first elected

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a director of the Company; their respective ages as of the Record Date; the positions currently held with the Company; the year their current term will expire; and their current class.

Nominee/Director Name	Age	Position	Director Since	Year Current Term Expires	Current Director Class
<b><i>Nominee for Class I Director</i></b>					
Emil D. Kakkis, M.D., Ph.D.	54	President and Chief Executive Officer	2010	2014	I
<b><i>Continuing Directors</i></b>					
Eran Nadav, Ph.D.	44	Chairman of the Board	2011	2015	II
Clay B. Siegall, Ph.D.	53	Director	2014	2015	II
William Aliski	66	Director	2011	2016	III
Matthew K. Fust	49	Director	2014	2016	III

**Class I Director Nominated for Reelection**

The following person has been nominated by the Board to be reelected as a Class I director at the 2014 Annual Meeting.

**Emil D. Kakkis, M.D., Ph.D.** is our founder and has served as our President and Chief Executive Officer and as a member of our Board since the Company's inception in April 2010. Prior to Ultragenyx, Dr. Kakkis served from September 1998 to February 2009 in various executive capacities, and ultimately as Chief Medical Officer, at BioMarin Pharmaceutical Inc., a biopharmaceutical company. Dr. Kakkis also serves as President and Founder of EveryLife Foundation for Rare Diseases, a non-profit organization he started in 2009 to accelerate biotechnology innovation for rare diseases. Dr. Kakkis is board certified in both Pediatrics and Medical Genetics. He holds a B.A. in Biology from Pomona College and received combined M.D. and Ph.D. degrees from the UCLA School of Medicine's Medical Scientist Training Program where he received the Bogen prize for his research. We believe that Dr. Kakkis possesses specific expert knowledge of genetics and rare diseases that qualifies him to serve on our Board, as well as his role as a founder of the Company and his operational experience in the life sciences sector.

**Class II Directors Continuing in Office Until 2015**

**Eran Nadav, Ph.D.** has served as a member of our Board since June 2011 and has served as Chairman of the Board since January 2012. Dr. Nadav is a Managing Director at TPG Biotech®, the life science venture investment arm of TPG, a global private investment firm. Dr. Nadav joined TPG in 2007 with a focus on global pharmaceuticals and biotechnology investments. Prior to TPG, Dr. Nadav served as Business Development Director at Eisai, a pharmaceutical company, from September 2003 to August 2007 and also as a manager at Johnson & Johnson Development Corporation, the venture capital arm of Johnson & Johnson, a healthcare company, from November 1999 until July 2002. Dr. Nadav served on the board of directors of Eden Springs Ltd., a European provider of drinking water solutions for the workplace, from July 2010 until August 2011. Since June 2013 he has been serving on the board of directors of MacroGenics, Inc., a biopharmaceutical company. Dr. Nadav received a B.Sc. magna cum laude in Life Sciences, an M.Sc. magna cum laude and Ph.D. in Biochemistry, as well as an M.B.A., from Tel Aviv University. We believe that Dr. Nadav is qualified to serve on our Board due to his experience in the venture capital industry and his years of analyzing development opportunities in the life sciences sector.

**Clay B. Siegall, Ph.D.** has served as a member of our Board since January 2014. Dr. Siegall currently serves as President and Chief Executive Officer and Chairman of the Board of Seattle Genetics, Inc., a biotechnology company. Dr. Siegall co-founded Seattle Genetics in 1998. Prior to Seattle Genetics, Dr. Siegall worked for the Bristol-Myers Squibb Pharmaceutical Research Institute from 1991 to 1997 and the National

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Cancer Institute, National Institutes of Health from 1988 to 1991. In addition to Seattle Genetics, Dr. Siegall serves as a director of Alder BioPharmaceuticals, Inc., a privately held biotechnology company, and Mirna Therapeutics, Inc., a privately held biotechnology company. Dr. Siegall received a B.S. in Zoology from the University of Maryland and a Ph.D. in Genetics from George Washington University. We believe that Dr. Siegall is qualified to serve on our Board due to his extensive experience in the life sciences industry and his role as Chief Executive Officer of a publicly-traded biotechnology company.

## **Class III Directors Continuing in Office Until 2016**

**William Aliski** has served as a member of our Board since January 2011. Mr. Aliski currently serves as a director of Edimer Pharmaceuticals, Inc., a privately held biotechnology company, and Scioderm, Inc., a privately held biopharmaceutical company. Mr. Aliski previously served as a commercial consultant for early stage orphan disease companies, including Enobia Pharma, from September 2011 until March 2012. Before that, Mr. Aliski served as Senior Vice President and Chief Commercial Officer of FoldRx Pharmaceuticals, a rare disease company that is now a wholly owned subsidiary of Pfizer Inc., from June 2009 until March 2011, as Director of Simon Kucher Partners, a global consulting firm, from January 2008 until June 2009 and as General Manager of BioMarin Europe at BioMarin Pharmaceuticals Inc. from December 2005 until January 2008. Mr. Aliski received a B.S. in Economics and a Master of Social Planning from Boston College and an M.P.A. from the Kennedy School of Government at Harvard University. We believe that Mr. Aliski is qualified to serve on our Board due to his extensive experience in the life sciences industry, membership on various boards of directors, and his leadership and management experience.

**Matthew K. Fust** has served as a member of our Board since January 2014. Mr. Fust served as Executive Vice President of Onyx Pharmaceuticals, Inc., a biopharmaceutical company, from January 2009 until January 2014. From May 2003 to December 2008, Mr. Fust served as Chief Financial Officer at Jazz Pharmaceuticals, Inc., a specialty pharmaceutical company. From 2002 to 2003, Mr. Fust served as Chief Financial Officer at Perlegen Sciences, a biopharmaceutical company. Previously, he was Senior Vice President and Chief Financial Officer at ALZA Corporation, a pharmaceutical company, where he was an executive from 1996 until 2002. From 1991 until 1996, Mr. Fust was a manager in the healthcare strategy practice at Andersen Consulting. Mr. Fust serves on the board of directors of Sunesis Pharmaceuticals, Inc. and MacroGenics, Inc., both of which are biopharmaceutical companies. Mr. Fust received a B.A. from the University of Minnesota and an M.B.A. from the Stanford Graduate School of Business. We believe that Mr. Fust is qualified to serve on our Board due to his extensive experience in the life sciences industry, his financial experience and ability to be our audit committee financial expert, and his service as a director of other public biopharmaceutical companies.

## **Vote Required**

The nominee who receives the greatest number of affirmative votes will be elected as a Class I director. Any shares that are not voted, whether by abstention, broker non-votes or otherwise, will not affect the election of directors.

Holders of proxies solicited by this Proxy Statement will vote the proxies received by them as directed on the proxy card or, if no direction is given, then FOR the election of the nominee named in this Proxy Statement.

**THE BOARD OF DIRECTORS RECOMMENDS A VOTE FOR  
THE DIRECTOR NOMINEE IDENTIFIED ABOVE.**





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**PROPOSAL NO. 2   RATIFICATION OF SELECTION OF INDEPENDENT REGISTERED  
PUBLIC ACCOUNTING FIRM**

Our Audit Committee has selected Ernst & Young LLP ( *EY* ) as our independent registered public accounting firm for the fiscal year ending December 31, 2014, and has further directed that we submit the selection of EY for ratification by our stockholders at the Annual Meeting.

The Company is not required to submit the selection of our independent registered public accounting firm for stockholder approval, but is submitting its selection of EY for stockholder ratification as a matter of good corporate governance. However, if the stockholders do not ratify this selection, the Audit Committee will reconsider its selection of EY. Even if the selection is ratified, our Audit Committee may direct the appointment of a different independent registered public accounting firm at any time during the year if the Audit Committee determines that the change would be in the best interests of the Company.

The Audit Committee reviews and pre-approves all audit and non-audit services performed by its independent registered public accounting firm. All services rendered by EY in fiscal 2013 were approved in accordance with these policies. In its review of non-audit services, the Audit Committee considers, among other things, the possible impact of the performance of such services on the independent registered public accounting firm's independence. The Audit Committee has determined that the non-audit services performed by EY in the fiscal year ended December 31, 2013 were compatible with maintaining the independent registered public accounting firm's independence. Additional information concerning the Audit Committee and its activities can be found in the following sections of this Proxy Statement: Board Committees and Report of the Audit Committee.

EY has audited our financial statements since inception. Representatives of EY are expected to be present at the Annual Meeting, will have the opportunity to make a statement if they desire to do so and will be available to respond to appropriate stockholder questions.

**Fees for Independent Registered Public Accounting Firm**

The following is a summary of the aggregate fees paid or accrued by the Company for the audit and other services rendered for the fiscal years ended December 31, 2013 and 2012.

	<b>2012</b>	<b>2013</b>
Audit fees <sup>(1)</sup>	\$ 18,000	\$ 1,264,000
Audit-related fees		
Tax fees <sup>(2)</sup>		8,000
All other fees <sup>(3)</sup>		2,000
<b>Total</b>	<b>\$ 18,000</b>	<b>\$ 1,274,000</b>

- (1) Audit fees consist of the aggregate fees billed for professional services rendered for (i) the audit of our annual financial statements, including for our Annual Report on Form 10-K for fiscal 2013 and (ii) the filing of our registration statements, including our Registration Statement on Form S-1 related to our initial public offering.

- (2) Tax fees principally include fees for tax compliance and tax advice.
- (3) All other fees consist of fees for accessing Ernst & Young LLP's online research database.

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**Vote Required**

Ratification of the selection of the independent registered public accounting firm requires the affirmative vote of a majority of the votes cast. Because abstentions and broker non-votes are not counted as votes for or against this proposal, they will have no effect on the outcome of the vote.

**THE BOARD OF DIRECTORS RECOMMENDS A VOTE FOR PROPOSAL NO. 2.**

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**PROPOSAL NO. 3 REAPPROVAL OF THE INTERNAL REVENUE CODE SECTION 162(m)**

**PROVISIONS OF THE 2014 INCENTIVE PLAN**

Prior to our initial public offering, our Board and stockholders approved the 2014 Incentive Plan (the ***2014 Plan***), including the list of performance goals and related provisions set forth in such plan for awards that are intended to qualify for the performance-based compensation exception under Section 162(m) ( ***Section 162(m)*** ) of the Internal Revenue Code of 1986, as amended (the ***Code*** ). **This proposal does not seek any amendment of the existing provisions of, or performance goals contained within, the 2014 Plan. Rather, this proposal is being presented to stockholders solely to address the periodic approval requirements of Section 162(m) described below and not to approve the 2014 Plan itself. Importantly, approval of this proposal would not increase or otherwise affect the number of shares available for grant under the 2014 Plan or the potential dilution to stockholders from awards granted under the 2014 Plan.**

Section 162(m) generally does not allow a publicly held corporation to deduct from its U.S. federal taxable income compensation above \$1,000,000 that is paid in any taxable year to its chief executive officer or other named executive officers. Compensation above \$1,000,000 may be deducted if, among other things, it is payable upon the attainment of performance goals whose material terms are approved by the company's stockholders. If the company's compensation committee retains discretion to select which performance goals will apply to a particular performance period, Section 162(m) requires that the material terms of such performance goals be reapproved by the company's stockholders at least once every five years. For purposes of Section 162(m), the material terms include (a) the individuals eligible to receive compensation, (b) a description of the business criteria on which the performance goal may be based, and (c) the maximum amount of compensation that can be paid to an individual under the performance goal. Each of these terms is discussed below and stockholder approval of this Proposal No. 3 will be deemed to constitute approval of each of these aspects of the 2014 Plan for purposes of the approval requirements of Section 162(m).

The performance goals set forth in the 2014 Plan that were approved by our stockholders prior to our initial public offering have not changed since such prior approval by the stockholders. In order to continue to provide us with the ability to deduct the performance-based compensation that we structure to comply with Section 162(m) and we pay to our Chief Executive Officer and our other named executive officers, we are submitting the list of performance goals and related provisions under our 2014 Plan to our stockholders for reapproval.

**Plan Information and Performance Goals**

Under the 2014 Plan, the Compensation Committee of our Board of Directors (the ***Compensation Committee*** ) may grant stock options, stock appreciation rights, restricted stock, unrestricted stock, stock units, performance awards and cash awards to participants that vest based on the achievement of performance goals over a performance period, a specified period of continuous employment, or a combination of both. All awards under the 2014 Plan are either performance-based and intended to comply with Section 162(m) or are awards that are not intended to comply with Section 162(m). The rules and regulations promulgated under Section 162(m) are complicated, however, and may change from time to time, sometimes with retroactive effect. As such, there can be no guarantee that any award intended to qualify as performance-based compensation within the meaning of Section 162(m) will so qualify. The Compensation Committee has the discretionary authority to determine the size of an award, whether it will be tied to meeting performance criteria and whether it will be settled in the form of stock and/or cash.

Participation in the 2014 Plan is limited to our employees, directors, consultants and advisors who are approved by the Compensation Committee. As of the Record Date, approximately 85 employees, directors and consultants are

currently eligible to participate in the 2014 Plan, which includes three executive officers, 76 employees who are not executive officers, one consultant and five non-employee directors. Subject to adjustments

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for stock splits and similar events, the maximum award granted to any one individual that is intended to qualify as performance-based compensation under Section 162(m) of the Code will not exceed 1,000,000 shares of Common Stock for any performance cycle and options or stock appreciation rights with respect to no more than 1,000,000 shares of Common Stock may be granted to any one individual during any calendar year period. If a performance-based award is payable in cash, it cannot exceed \$430,000 for any performance cycle.

To allow for awards granted under the 2014 Plan to our Chief Executive Officer and other named executive officers to qualify as performance-based compensation under Section 162(m), the 2014 Plan provides that the Compensation Committee may require that the vesting of such awards be conditioned on the satisfaction of an objectively determinable measure of performance ( *Performance Criterion* ) that may include any or all of the following: sales; revenues; assets; expenses; earnings from operations; earnings before or after deduction for all or any portion of interest, taxes, depreciation, amortization, incentives, service fees or extraordinary or special items, whether or not on a continuing operations or an aggregate or per share basis; net income or net income per common share (basic or diluted); return on equity, investment, capital or assets; one or more operating ratios; borrowing levels, leverage ratios or credit rating; market share; capital expenditures; cash flow, free cash flow, cash flow return on investment, or net cash provided by operations; stock price, dividends or total stockholder return; development of new technologies or products; sales of particular products or services; economic value created or added; operating margin or profit margin; customer acquisition or retention; raising or refinancing of capital; successful hiring of key individuals; resolution of significant litigation; acquisitions and divestitures (in whole or in part); joint ventures and strategic alliances; spin-offs, split-ups and the like; reorganizations; recapitalizations, restructurings, financings (issuance of debt or equity) or refinancings; or strategic business criteria, consisting of one or more objectives based on the following goals: meeting specified market penetration or value added, product development or introduction (including, without limitation, any clinical trial accomplishments, regulatory or other filings or approvals, or other product development milestones), geographic business expansion, cost targets, cost reductions or savings, customer satisfaction, operating efficiency, acquisition or retention, employee satisfaction, information technology, corporate development (including, without limitation, licenses, innovation, research or establishment of third party collaborations), manufacturing or process development, legal compliance or risk reduction, patent application or issuance goals, or goals relating to acquisitions or divestitures (in whole or in part), joint ventures or strategic alliances. A Performance Criterion and any targets with respect thereto determined by the Compensation Committee need not be based upon an increase, a positive or improved result or avoidance of loss. To the extent consistent with the requirements for satisfying the performance-based compensation exception under Section 162(m), the Compensation Committee may provide in the case of any award intended to qualify for such exception that one or more of the Performance Criteria applicable to such award will be adjusted in an objectively determinable manner to reflect events (for example, but without limitation, acquisitions or dispositions) occurring during the performance period that affect the applicable Performance Criterion or Criteria. The Compensation Committee will select the particular performance criteria in writing within 90 days following the commencement of a performance cycle and, prior to the event or occurrence (grant, vesting or payment, as the case may be) that is conditioned on the attainment of such Performance Criterion or Criteria, will certify whether it or they have been attained.

The material features of the 2014 Plan are:

2,250,000 shares of Common Stock have been initially reserved for the issuance of awards under the 2014 Plan. The 2014 Plan provides that the number of shares reserved and available for issuance thereunder will automatically increase each January 1, beginning in 2015, by an amount equal to the least of (i) 2,500,000 shares of common stock, (ii) 4% of the number of shares of Common Stock outstanding on a fully diluted basis as of the close of business on the immediately preceding

December 31 (calculated by adding to the number of shares of common stock outstanding, all outstanding securities convertible into common stock on such date on an as-converted basis), and (iii) a lesser amount determined by the Compensation Committee on or prior to January 1 of a given year;



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the shares issued by us under the 2014 Plan may be authorized but unissued shares or previously issued shares acquired by us. Any shares of our common stock underlying awards that are settled in cash or otherwise expire, terminate, or are forfeited prior to the issuance of stock will again be available for issuance under the 2014 Plan;

the award of stock options (both incentive and non-qualified options), stock appreciation rights, restricted stock, unrestricted stock, stock units, performance awards and cash awards is permitted. Dividend equivalents may also be provided in connection with an award under the 2014 Plan;

our Compensation Committee has the authority to determine the vesting schedule applicable to each award, and to accelerate the vesting or exercisability of any award;

the 2014 Plan expires on January 11, 2024;

any material amendment is subject to approval by our stockholders; and

the 2014 Plan is administered by the Compensation Committee. The Compensation Committee has full power and authority to: interpret the 2014 Plan; determine eligibility for and grant awards; determine, modify or waive the terms and conditions of any award; and prescribe forms, rules and procedures relating to the 2014 Plan.

## **Adjustments**

In the event of a stock dividend, stock split or combination of shares including a reverse stock split, recapitalization or other change in our capital structure that constitutes an equity restructuring within the meaning of the Financial Accounting Standards Board, Accounting Standards Codification Topic 718, *Compensation – Stock Compensation*, our Compensation Committee will make appropriate adjustments to the maximum number of shares that may be delivered under, and the individual share limits included in, the 2014 Plan, and will also make appropriate adjustments to the number and kind of shares of stock or securities subject to awards, the exercise prices of such awards or any other terms of awards affected by such change. Our Compensation Committee will also make the types of adjustments described above to take into account distributions and other events other than those listed above if it determines that such adjustments are appropriate to avoid distortion and preserve the value of awards.

## **Exercise of Stock Options and Stock Appreciation Rights**

The exercise price of stock options and base value of stock appreciation rights ( **SARs** ) granted under the 2014 Plan may not be less than 100% of the fair market value of our common stock on the date of grant (or in the case of an incentive stock option granted to a 10% stockholder within the meaning of subsection (b)(6) of Section 422 of the Code, 110%), and the term may not be longer than 10 years (or in the case of an incentive stock option granted to a 10% stockholder within the meaning of subsection (b)(6) of Section 422 of the Code, 5 years). The Compensation Committee determines at the time of grant when each award becomes exercisable. Payment of the exercise price of a stock option may be in cash or check or by such other legally permissible means, if any, as may be acceptable to the Compensation Committee. Similar provisions govern awards of SARs, which entitle the holder upon exercise to receive common stock or cash equal to the excess of the fair market value of the underlying shares on the date of

exercise over the base value of the SAR.

### **Vesting of Restricted Stock**

The Compensation Committee determines at the time of grant when awards of restricted stock and RSUs vest, and the related restrictions lapse.

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### **Effect of Certain Corporate Transactions**

In the event of a consolidation, merger or similar transaction, a sale or transfer of all or substantially all of our assets or our dissolution or liquidation, our Compensation Committee may, among other things, provide for continuation or assumption of outstanding awards, for new grants in substitution of outstanding awards, for the accelerated vesting or delivery of shares under awards or for a cash-out of outstanding awards, in each case on such terms and with such restrictions as it deems appropriate. Except as our Compensation Committee may otherwise determine, awards not assumed or continued will automatically terminate and in the case of outstanding shares of restricted stock, will automatically be forfeited upon the consummation of such covered transaction.

### **Termination of Employment**

Our Compensation Committee will determine the effect of termination of employment or service on an award. Unless otherwise provided by our Compensation Committee or in an award agreement, upon a termination of a participant's employment all unvested options then held by the participant and other awards requiring exercise will terminate and all other unvested awards will be forfeited. Unless otherwise provided for by our Compensation Committee, all vested stock options and stock appreciation rights then held by the participant will remain outstanding for three months, or one year in the case of death, or, in each case, until the applicable expiration date, if earlier. All stock options and stock appreciation rights held by a participant immediately prior to the participant's termination of employment will immediately terminate upon termination of employment if the termination is for cause as defined in the 2014 Plan or occurs in circumstances that would have constituted grounds for the participant's employment to be terminated for cause, in the determination of the Compensation Committee.

### **Transferability**

Awards under the 2014 Plan may not be transferred except by will or by the laws of descent and distribution, unless (for awards other than incentive stock options) otherwise provided by our Compensation Committee.

### **Administration**

The 2014 Plan is administered by our Compensation Committee. Our Compensation Committee has the authority to, among other things, interpret the 2014 Plan, determine eligibility for, grant and determine the terms of awards under the 2014 Plan, and to do all things necessary to carry out the purposes of the 2014 Plan. Our Compensation Committee's determinations under the 2014 Plan are conclusive and binding.

### **Amendment and Termination**

Our Compensation Committee will be able to amend the 2014 Plan or outstanding awards, or terminate the 2014 Plan as to future grants of awards, except that our Compensation Committee will not be able to alter the terms of an award if it would affect materially and adversely a participant's rights under the award without the participant's consent (unless expressly provided in the 2014 Plan or the right to alter the terms of an award was expressly reserved by our Compensation Committee at the time the award was granted). Stockholder approval will be required for any amendment to the 2014 Plan to the extent such approval is required by law, including the Code or applicable stock exchange requirements.

### **Federal Income Tax Consequences Under the 2014 Plan**

The following is a summary of some of the material federal income tax consequences associated with the grant and exercise of awards under the 2014 Plan under current federal tax laws and certain other tax considerations associated with awards under the 2014 Plan. The summary does not address tax rates or non-U.S., state or local tax consequences, nor does it address employment-tax or other federal tax consequences except as noted.

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*Restricted Stock.* A participant who is awarded or purchases shares subject to a substantial risk of forfeiture generally does not have income until the risk of forfeiture lapses. When the risk of forfeiture lapses, the participant has ordinary income equal to the excess of the fair market value of the shares at that time over the purchase price, if any, and a corresponding deduction is generally available to the Company. However, a participant may make an election under Section 83(b) of the Code to be taxed on restricted stock when it is acquired rather than later, when the substantial risk of forfeiture lapses. An 83(b) election must be made not later than thirty (30) days after the transfer of the shares to the participant and must satisfy certain other requirements. A participant who makes an effective 83(b) election will realize ordinary income equal to the fair market value of the shares as of the time of acquisition less any price paid for the shares. A corresponding deduction will generally be available to the Company. Fair market value for this purpose is determined without regard to the forfeiture restrictions. If a participant makes an effective 83(b) election, no additional income results by reason of the lapsing of the restrictions.

For purposes of determining capital gain or loss on a sale of shares awarded under the 2014 Plan, the holding period in the shares begins when the participant realizes taxable income with respect to the transfer. The participant's tax basis in the shares equals the amount paid for the shares plus any income realized with respect to the transfer. However, if a participant makes an effective 83(b) election and later forfeits the shares, the tax loss realized as a result of the forfeiture is limited to the excess of what the participant paid for the shares (if anything) over the amount (if any) realized in connection with the forfeiture.

*ISOs.* In general, a participant realizes no taxable income upon the grant or exercise of an incentive stock option ( *ISO* ). However, the exercise of an ISO may result in an alternative minimum tax liability to the participant. With some exceptions, a disposition of shares purchased under an ISO within two years from the date of grant or within one year after exercise produces ordinary income to the participant (and generally a deduction to the Company) equal to the value of the shares at the time of exercise less the exercise price. Any additional gain recognized in the disposition is treated as a capital gain for which the Company is not entitled to a deduction. If the participant does not dispose of the shares until after the expiration of these one and two-year holding periods, any gain or loss recognized upon a subsequent sale is treated as a long-term capital gain or loss for which the Company is not entitled to a deduction.

*NSOs.* In general, a participant has no taxable income upon the grant of a non-qualified stock option ( *NSO* ) but realizes income in connection with exercise of the option in an amount equal to the excess (at time of exercise) of the fair market value of the shares acquired upon exercise over the exercise price. A corresponding deduction is generally available to the Company. Upon a subsequent sale or exchange of the shares, any recognized gain or loss is treated as a capital gain or loss for which the Company is not entitled to a deduction. An ISO that is exercised more than three months after termination of employment (other than termination by reason of death) is generally treated as an NSO. ISOs are also treated as NSOs to the extent they first become exercisable by an individual in any calendar year for shares having a fair market value (determined as of the date of grant) in excess of \$100,000.

*SARs.* The grant of a SAR does not itself result in taxable income, nor does taxable income result merely because a SAR becomes exercisable. In general, a participant who exercises a SAR for shares of stock or receives payment in cancellation of a SAR will have ordinary income equal to the amount of any cash and the fair market value of any stock received. A corresponding deduction is generally available to the Company.

*Restricted Stock Units.* The grant of a restricted stock unit does not itself generally result in taxable income. Instead, the participant is taxed upon vesting (and a corresponding deduction is generally available to the Company), unless he or she has made a proper election to defer receipt of the shares (or cash if the award is cash settled) under Section 409A of the Code ( *Section 409A* ). If the shares delivered are restricted for tax purposes, the participant will instead be subject to the rules described above for restricted stock.

*Section 162(m)*. Stock options, SARs and certain performance awards under the 2014 Plan are generally intended to be exempt or eligible for exemption from the deductibility limits of Section 162(m). However, the

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Compensation Committee will have discretionary authority to provide compensation that is not exempt from the limits on deductibility under Section 162(m). In addition, the rules and regulations promulgated under Section 162(m) are complicated and may change from time to time, sometimes with retroactive effect. As such, there can be no guarantee that any award intended to qualify as performance-based compensation within the meaning of Section 162(m) will so qualify.

*Section 409A.* Section 409A imposes an additional 20% income tax, plus, in some cases, a further income tax in the nature of interest, on nonqualified deferred compensation that does not comply with deferral, payment-timing and other formal and operational requirements specified in Section 409A and related regulations and that is not exempt from those requirements. Stock options and SARs granted under the 2014 Plan are intended to be exempt from Section 409A. The 2014 Plan gives the Compensation Committee the flexibility to prescribe terms for other awards that are consistent with the requirements of, or an exemption from, Section 409A.

*Certain Change of Control Payments.* Under Section 280G of the Code, the vesting or accelerated exercisability of options or the vesting and payments of other awards in connection with a change of control of a corporation may be required to be valued and taken into account in determining whether participants have received compensatory payments, contingent on the change in control, in excess of certain limits. If these limits are exceeded, a substantial portion of amounts payable to the participant, including income recognized by reason of the grant, vesting or exercise of awards, may be subject to an additional 20% federal tax and may be non-deductible to the Company.

**New Plan Benefits**

Future awards under the 2014 Plan are discretionary and, therefore, are undeterminable at this time.

**Existing Plan Benefits**

Because the grant of awards under the 2014 Plan are undeterminable at this time, the following table provides information concerning the benefits that were received by the following persons and groups during 2013: each named executive officer; all current executive officers, as a group; all current directors who are not executive officers, as a group; and all current employees who are not executive officers, as a group.

Name and Position	Options	
	Average Exercise Price Per Share	Number (#)
Emil D. Kakkis, M.D., Ph.D.		
<i>President, Chief Executive Officer and Director</i>	\$ 6.86	47,854
Thomas Kassberg		
<i>Chief Business Officer and Senior Vice President</i>	\$ 6.86	39,878
Shalini Sharp	\$ 6.86	39,878

*Chief Financial Officer and Senior Vice President*

All current executive officers, as a group	\$	6.86	127,610
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All current directors who are not executive officers, as a group			
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All current employees who are not executive officers, as a group	\$	5.68	1,140,188
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**For additional information about the 2014 Plan, we encourage you to review the entire text of the plan, a copy of which is attached as Exhibit A to this Proxy Statement.**



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**Vote Required**

The affirmative vote of a majority of votes cast is required for the reapproval of the Internal Revenue Code Section 162(m) provisions of the 2014 Plan. Because abstentions and broker non-votes are not counted as votes for or against this proposal, they will have no effect on the outcome of the vote.

**THE BOARD OF DIRECTORS RECOMMENDS A VOTE FOR THE REAPPROVAL OF THE INTERNAL REVENUE CODE SECTION 162(m) PROVISIONS OF THE 2014 INCENTIVE PLAN.**

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**PROPOSAL NO. 4 REAPPROVAL OF THE INTERNAL REVENUE CODE SECTION 162(m) PROVISIONS OF THE CORPORATE BONUS PLAN**

Prior to our initial public offering, our Board and stockholders approved the Corporate Bonus Plan (the *Bonus Plan*), including the list of performance goals and related provisions set forth in such plan for awards that are intended to qualify for the performance-based compensation exception under Section 162(m). **This proposal does not seek any amendment of the existing provisions of, or performance goals contained within, the Bonus Plan. Rather, this proposal is being presented to stockholders solely to address the periodic approval requirements of Section 162(m) and not to approve the Bonus Plan itself.**

Section 162(m) generally does not allow a publicly held corporation to deduct from its U.S. federal taxable income compensation above \$1,000,000 that is paid in any taxable year to its chief executive officer or other named executive officers. Compensation above \$1,000,000 may be deducted if, among other things, it is payable upon the attainment of performance goals whose material terms are approved by the company's stockholders. If the company's compensation committee retains discretion to select which performance goals will apply to a particular performance period, Section 162(m) requires that the material terms of such performance goals be reapproved by the company's stockholders at least once every five years. For purposes of Section 162(m), the material terms include (a) the individuals eligible to receive compensation, (b) a description of the business criteria on which the performance goal may be based, and (c) the maximum amount of compensation that can be paid to an individual under the performance goal. Each of these terms is discussed below and stockholder approval of this Proposal No. 4 will be deemed to constitute approval of each of these aspects of the Corporate Bonus Plan for purposes of the approval requirements of Section 162(m).

The performance goals set forth in the Bonus Plan that were approved by our stockholders prior to our initial public offering have not changed since such prior approval by the stockholders. In order to continue to provide us with the ability to deduct the performance-based compensation that we structure to comply with Section 162(m) and we pay to our Chief Executive Officer and our other named executive officers, we are submitting the list of performance goals and related provisions under our Bonus Plan to our stockholders for reapproval.

**Plan Information and Performance Goals**

Under the Bonus Plan, the Compensation Committee may grant performance awards to eligible executives, the payment of which is determined by the achievement of one or more performance goals over a performance period. Participation in the Bonus Plan is limited to our employees who are approved by the Compensation Committee (the *Covered Executives*). As of the Record Date, approximately 79 employees are currently eligible to participate in the Bonus Plan. Each corporate performance goal under the Bonus Plan shall have a target and may also have a minimum hurdle and/or a maximum amount.

All awards under the Bonus Plan are intended to qualify as performance-based compensation under Section 162(m), and performance goals must be based on one or more objectively determinable measures of performance established by the Compensation Committee and relating to financial and/or operational metrics with respect to the Company (the *Corporate Performance Goals*), including the following: sales; revenue; assets; expenses; earnings from operations, earnings before or after deduction for all or any portion of interest, taxes, depreciation, amortization, incentives, service fees or extraordinary or special items, whether or not on a continuing operations or an aggregate or per share basis; net income or net income per common share (basic or diluted); return on equity, investment, capital or assets; one or more operating ratios; borrowing levels, leverage ratios or credit rating; market share; capital expenditures; cash flow, free cash flow, cash flow return on investment, or net cash provided by operations; stock price, dividends or total stockholder return; development of new technologies or products; sales of particular products or services;

economic value created or added; operating margin or profit margin; customer acquisition or retention; raising or refinancing of capital; successful hiring of key individuals; resolution of significant litigation; acquisitions and divestitures (in whole or in part); joint ventures and strategic alliances; spin-offs, split-ups and the like; reorganizations; recapitalizations,

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restructurings, financings (issuance of debt or equity) or refinancings; or strategic business criteria, consisting of one or more objectives based on the following goals: meeting specified market penetration or value added, product development or introduction (including, without limitation, any clinical trial accomplishments, regulatory or other filings or approvals, or other product development milestones), geographic business expansion, cost targets, cost reductions or savings, customer satisfaction, operating efficiency, acquisition or retention, employee satisfaction, information technology, corporate development (including, without limitation, licenses, innovation, research or establishment of third party collaborations), manufacturing or process development, legal compliance or risk reduction, patent application or issuance goals, or goals relating to acquisitions or divestitures (in whole or in part), joint ventures or strategic alliances, any of which may be (A) measured in absolute terms or compared to any incremental increase, (B) measured in terms of growth, (C) compared to another company or companies or to results of a peer group, (D) measured against the market as a whole and/or as compared to applicable market indices and/or (E) measured on a pre-tax or post-tax basis (if applicable). The rules and regulations promulgated under Section 162(m) are complicated, however, and may change from time to time, sometimes with retroactive effect. As such, there can be no guarantee that any award intended to qualify as performance-based compensation within the meaning of Section 162(m) will so qualify.

Corporate Performance Goals will be calculated in accordance with the Company's financial statements, generally accepted accounting principles, or under a methodology established by the Compensation Committee at the beginning of the performance period and which is consistently applied with respect to a Corporate Performance Goal in the relevant performance period.

Any bonuses paid to Covered Executives under the Bonus Plan shall be based upon objectively determinable bonus formulas that tie such bonuses to one or more performance targets relating to the Corporate Performance Goals, and any bonus formulas for Covered Executives shall be adopted in each performance period by the Compensation Committee and communicated to each Covered Executive at the beginning of each performance period. The Compensation Committee shall establish an individual target bonus opportunity for each Covered Executive for each performance period and the Compensation Committee has the authority to apportion the target award so that a portion of the award shall be tied to attainment of Corporate Performance Goals and a portion of the target award shall be tied to attainment of individual performance objectives.

No bonuses shall be paid to Covered Executives unless and until the Compensation Committee makes a determination with respect to the attainment of the performance targets relating to the Corporate Performance Goals. Notwithstanding the foregoing, the Compensation Committee may adjust bonuses payable under the Bonus Plan based on achievement of one or more individual performance objectives or pay bonuses (including, without limitation, discretionary bonuses) to Covered Executives under the Bonus Plan based on individual performance goals and/or upon such other terms and conditions as the Compensation Committee may in its discretion determine. The Compensation Committee has discretion to adjust bonuses payable under the Bonus Plan based on achievement of one or more individual performance objectives or pay bonuses (including, without limitation, discretionary bonuses) to participants based on individual performance goals and/or upon such other terms and conditions as the Compensation Committee may determine in its discretion. Actual bonuses are paid in cash no later than 74 days after the performance period ends.

**For additional information about the Bonus Plan, we encourage you to review the entire text of the plan, a copy of which is attached as Exhibit B to this Proxy Statement.**

## **Vote Required**

The affirmative vote of a majority of votes cast is required for the reapproval of the Internal Revenue Code Section 162(m) provisions of the Bonus Plan. Because abstentions and broker non-votes are not counted as votes for or against this proposal, they will have no effect on the outcome of the vote.

**THE BOARD OF DIRECTORS RECOMMENDS A VOTE FOR THE REAPPROVAL OF THE INTERNAL REVENUE CODE SECTION 162(m) PROVISIONS OF THE CORPORATE BONUS PLAN.**

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### **CORPORATE GOVERNANCE**

#### **Director Independence**

Our Board currently consists of six members and, concurrent with the expiration of Dr. Steen's term at the Annual Meeting, will consist of five members. Our Board has determined Dr. Nadav, Mr. Aliski, Mr. Fust, Dr. Siegall and Dr. Steen qualify as independent directors in accordance with the NASDAQ listing requirements and rules. Dr. Kakkis is not considered independent because he is an employee of Ultragenyx. Under NASDAQ rules, the Board's determination of a director's independence considers objective tests, such as whether the director is or has been within the last three years an employee of the Company and whether the director or any of his family members has engaged in certain types of business dealings with the Company. Under NASDAQ rules, our Board also evaluates whether any relationships exist that, in the opinion of our Board, would interfere with the exercise of independent judgment in carrying out the responsibilities of a director. In making these independence determinations, our Board reviewed and discussed information provided by the directors and us with regard to each director's business and personal activities and relationships as they may relate to us and our management, including any significant stock holdings by funds affiliated with the directors. There are no family relationships among any of our directors or executive officers.

#### **Code of Business Conduct and Ethics**

We have adopted a Code of Business Conduct and Ethics that applies to all of our employees, officers, and directors, including those officers responsible for financial reporting. Our Code of Business Conduct and Ethics is available on our website, [www.ultragenyx.com](http://www.ultragenyx.com), under the Corporate Governance subsection of the Investors tab. We will promptly disclose on our website any future changes or amendments to the Code of Business Conduct and Ethics, and any waivers of our Code of Business Conduct and Ethics that applies to our Chairman of the Board, any of our executive officers, or any member of our Board.

#### **Stockholder Communications**

Generally, stockholders who have questions or concerns regarding the Company should contact our Investor Relations department at (844) 758-7273. However, any stockholders who wish to address questions regarding the business or affairs of the Company directly with the Board, or any individual director, should direct his or her questions in writing to the Chairman of the Board, c/o Ultragenyx Pharmaceutical Inc., 60 Leveroni Court, Novato, California 94949. Upon receipt of any such communications, the correspondence will be directed to the appropriate person, including individual directors.

### **BOARD OF DIRECTORS AND COMMITTEES**

During fiscal 2013, our Board met 13 times. Each director attended at least 75% of the aggregate of the meetings of the Board and meetings of the committees of which he was a member in our last fiscal year.

The Board has a standing Audit Committee, Compensation Committee and Nominating and Corporate Governance Committee. All members of the Audit, Compensation and Nominating and Corporate Governance Committees are non-employee directors whom the Board has determined are independent.

Although the Company has no formal policies regarding director attendance at annual meetings, all members of the Board are expected to attend the 2014 Annual Meeting.

#### **Board Leadership Structure and Risk Oversight**

Our Corporate Governance Guidelines provide that the positions of Chairman of the Board and Chief Executive Officer are to be separated, which allows our Chief Executive Officer to focus on our day-to-day business, while allowing the Chairman of the Board to lead the Board in its fundamental role of providing advice to and independent oversight of management. Independent oversight of management is an important goal of the

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Board, which is why our Corporate Governance Guidelines provide that a lead independent director will be appointed by the Board if the Chairman is not independent. Additionally, our Board recognizes the time, effort, and energy that the Chief Executive Officer is required to devote to his position in the current business environment, as well as the commitment required to serve as our Chairman. Our Board also believes that the separation of the Chairman and Chief Executive Officer positions fosters a greater role for the independent directors in the oversight of our Company and active participation of the independent directors in setting agendas and establishing priorities and procedures for the work of our Board. The benefits of the separated Chairman and Chief Executive Officer positions are augmented by the independence of five of our six current directors, including our Chairman, and our independent Board committees that provide appropriate oversight in the areas described below. At executive sessions of independent directors, these directors can speak candidly on any matter of interest. The Board met in executive session twice in 2013. We believe this structure provides effective oversight of our management and the Company.

The Board has overall responsibility for the oversight of the Company's risk management process, which is designed to support the achievement of organizational objectives, including strategic objectives, to improve long-term organizational performance and enhance stockholder value. Risk management includes not only understanding company-specific risks and the steps management implements to manage those risks, but also what level of risk is acceptable and appropriate for the Company. Management is responsible for establishing our business strategy, identifying and assessing the related risks and implementing appropriate risk management practices. The Board periodically reviews our business strategy and management's assessment of the related risk and discusses with management the appropriate level of risk for the Company. The Board also delegates to Board committees oversight of selected elements of risk as set forth below.

## **Board Committees**

*Audit Committee.* As of the Record Date, the Audit Committee was comprised of Mr. Fust (Chairperson) and Drs. Nadav and Steen and, effective upon the completion of the Annual Meeting, is expected to be comprised of Mr. Fust (Chairperson), Mr. Aliski and Dr. Nadav. The Audit Committee appoints, approves the compensation of, reviews the performance of and assesses the independence of our independent registered public accounting firm; approves audit and permissible non-audit services, and the terms of such services, to be provided by our independent registered public accounting firm; reviews the audit plan with the independent registered public accounting firm and members of management responsible for preparing our financial statements; reviews and discusses with management and the independent registered public accounting firm our annual and quarterly financial statements and related disclosures and critical accounting policies; reviews the adequacy of our internal control over financial reporting; establishes policies and procedures for the receipt and retention of accounting-related complaints and concerns; recommends whether our audited financial statements shall be included in our Annual Report on Form 10-K; prepares the audit committee report to be included in our annual proxy statements; reviews all related-party transactions; reviews policies related to risk assessment and risk management; establishes, maintains and oversees our Code of Business Conduct and Ethics; and performs other duties, as specified in the Audit Committee Charter, a copy of which is available on the Company's website at [www.ultragenyx.com](http://www.ultragenyx.com) under the Corporate Governance subsection of the Investors tab. The Audit Committee met six times in fiscal 2013. All members of the Audit Committee satisfy the current independence and financial literacy standards promulgated by NASDAQ and the SEC, and the Board has determined that Mr. Fust qualifies as an audit committee financial expert, as the SEC has defined that term in Item 407 of Regulation S-K.

*Compensation Committee.* As of the Record Date, the Compensation Committee was comprised of, and effective upon completion of the Annual Meeting is expected to be comprised of, Dr. Nadav (Chairperson), Mr. Aliski and Dr. Siegall. The Compensation Committee annually reviews and approves individual and corporate goals and objectives relevant to the compensation of our executive officers; evaluates the performance of our executive officers



in light of such individual and corporate goals and objectives and determines the compensation of our executive officers; appoints, compensates and oversees the work of any compensation consultant, legal counsel or other advisor retained by the Compensation Committee; conducts the independence

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assessment outlined in NASDAQ rules with respect to any compensation consultant, legal counsel, or other advisor retained by the Compensation Committee; annually reviews and reassesses the adequacy of the Compensation Committee charter; oversees and, has the authority to administer, our compensation and similar plans; reviews and approves our policies and procedures for the grant of equity-based awards; reviews and makes recommendations to the Board with respect to director compensation; reviews and discusses with management the compensation discussion and analysis, if any, to be included in our annual proxy statements or Annual Reports on Form 10-K; reviews and discusses with the Board corporate succession plans for the Chief Executive Officer and other senior management positions; and performs other duties, as specified in the Compensation Committee Charter, a copy of which is available on the Company's website at [www.ultragenyx.com](http://www.ultragenyx.com) under the Corporate Governance subsection of the Investors tab. The Compensation Committee may form and delegate authority to subcommittees, each consisting of one or more members of the Compensation Committee, with such powers as the Compensation Committee shall from time to time confer. Our Chief Executive Officer may recommend the amount and form of compensation of other executive officers to the Compensation Committee. The Compensation Committee met four times in fiscal 2013. All members of the Compensation Committee satisfy the current NASDAQ and SEC independence standards.

*Nominating and Corporate Governance Committee.* As of the Record Date, the Nominating and Corporate Governance Committee was comprised of Mr. Fust (Chairperson) and Drs. Nadav and Steen and, effective upon completion of the Annual Meeting, is expected to be comprised of Mr. Fust (Chairperson) and Dr. Nadav. The Nominating and Corporate Governance Committee develops and recommends to the Board criteria for Board and committee membership; establishes procedures for identifying and evaluating Board candidates, including nominees recommended by stockholders; identifies individuals qualified to become members of the Board; recommends to the Board the persons to be nominated for election as directors and to each of the Board's committees; develops and recommends to the Board a set of corporate governance guidelines; and performs other duties, as specified in the Nominating and Corporate Governance Committee Charter, a copy of which is available on the Company's website at [www.ultragenyx.com](http://www.ultragenyx.com) under the Corporate Governance subsection of the Investors tab. The Nominating and Corporate Governance Committee was established in earlier this year, and therefore did not meet in fiscal 2013. All members of the Nominating and Corporate Governance Committee satisfy the current NASDAQ independence standards.

**Compensation Committee Interlocks and Insider Participation**

None of the members of our Compensation Committee has at any time during the prior three years been one of our officers or employees. None of our executive officers currently serves, or in the past fiscal year has served, as a member of the board of directors or compensation committee of any entity that has one or more executive officers serving on our Board or Compensation Committee. For a description of transactions between us and members of our Compensation Committee and affiliates of such members, please see Certain Relationships and Related Party Transactions.

**Table of Contents****EXECUTIVE OFFICERS AND KEY EMPLOYEES**

Our current executive officers and key employees and their respective ages and positions as of the Record Date are set forth in the following table. Biographical information regarding each executive officer (other than Dr. Kakkis) and key employee is set forth following the table. Biographical information for Dr. Kakkis is set forth above under Proposal No. 1 (Election of Directors).

<b>Name</b>	<b>Age</b>	<b>Position</b>
Emil D. Kakkis, M.D., Ph.D.	54	President and Chief Executive Officer, Director (principal executive officer)
Thomas Kassberg	53	Chief Business Officer and Senior Vice President
Shalini Sharp	39	Chief Financial Officer and Senior Vice President (principal financial officer)
Steven Jungles	48	Senior Vice President, Technical Operations
Michael P. Cohrs, Ph.D.	53	Vice President of Quality
John Ditton	49	Vice President, Commercial Planning
Tony Koutsoukos, Ph.D.	54	Vice President, Biometrics
Cordelia Leonard, RAC	53	Vice President, Regulatory Affairs and Quality Assurance
Javier San Martin, M.D.	49	Vice President, Clinical Development
Vimal Srivastava	48	Vice President, Program Development
Michael Vellard, Ph.D.	53	Vice President, Research
Spencer Guthrie	38	Senior Director, Clinical Operations
Alison Skrinar, Ph.D.	44	Senior Director, Clinical Sciences
Theodore A. Huizenga	43	Corporate Controller (principal accounting officer)

**Thomas Kassberg** has served as our Chief Business Officer and Senior Vice President since November 2011. Prior to Ultragenyx, Mr. Kassberg worked as Vice President of Business Development at Corium International, Inc., a biotechnology company, from July 2010 until October 2011. Prior to his work at Corium International, Inc., Mr. Kassberg worked as an independent consultant in corporate development and business strategy and consulted with a number of companies from March 2009 to June 2010, including Corium International, Inc. and Rib-X Pharmaceuticals, Inc., a pharmaceutical company focused on the development of novel antibiotics. Before becoming a consultant, Mr. Kassberg worked at Proteolix, Inc., a biotechnology company subsequently acquired by Onyx Pharmaceuticals, from January 2008 until February 2009, where he served as Senior Vice President of Corporate Development. Mr. Kassberg holds a B.A. in Business Administration from Gustavus Adolphus College and an M.B.A. from Northwestern University.

**Shalini Sharp** has served as our Chief Financial Officer and Senior Vice President since May 2012. Prior to Ultragenyx, Ms. Sharp served in various executive capacities, and ultimately as Chief Financial Officer, of Agenus Inc., a biotechnology company, from August 2003 until May 2012. Prior to Agenus, Ms. Sharp held strategic planning and corporate finance roles and ultimately served as chief of staff to the chairman of the board at Elan Pharmaceuticals, a biotechnology company, from August 1998 to August 1999 and September 2001 to August 2003. Prior to Elan, Ms. Sharp was a management consultant at McKinsey & Company and an investment banker at Goldman Sachs, specializing in pharmaceuticals and medical devices. Ms. Sharp has also served as a board member of Agenus since May 2012. Ms. Sharp holds a B.A. and an M.B.A. from Harvard University.

**Steven Jungles** has served as our Senior Vice President, Technical Operations since August 2011. Prior to Ultragenyx, Mr. Jungles worked as Vice President, Supply Chain at BioMarin Pharmaceutical Inc., a biopharmaceutical company, from June 1999 to July 2011, was Associate Director of Operations at Harvard Gene

Therapy Initiative from June 1997 until June 1999, and worked at Somatix Therapy Corporation, a research and development company in the field of gene therapy that was acquired by Cell Genesys, Inc., from March 1993 to May 1997. Mr. Jungles holds a B.S. in Biology from the University of Iowa.

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**Michael P. Cohrs, Ph.D.** has served as our Vice President of Quality since December 2013 and is responsible for overseeing our GMP, GLP, GCP, and Clinical Assay departments. Prior to Ultragenyx, Dr. Cohrs served as Senior Director of Quality Assurance/Quality Control at Medicines 360, a pharmaceutical company, from October 2012 through November 2013 and previously served as the Director of Quality Assurance at Medicines 360 from September 2009 through October 2012. Prior to Medicines 360, Dr. Cohrs served as the Director of Quality Assurance at Sunesis Pharmaceuticals Inc., a pharmaceutical company, from June 2007 to June 2008. Dr. Cohrs has also held increasingly responsible positions at a number of large pharmaceutical and small start-up companies, first in quality control and then in quality assurance, including Aventis Behring, Genetics Institute, Chiron, and InterMune. Dr. Cohrs has also contributed to numerous regulatory submissions resulting in several product approvals in both the United States and Europe. Dr. Cohrs received his Ph.D. in Organic Chemistry from Washington University and an M.B.A. in International Business from the University of Chicago.

**John Ditton** has served as our Vice President, Commercial Planning since April 2011. Prior to Ultragenyx, Mr. Ditton was the Chief Operating Officer at EveryLife Foundation for Rare Diseases, from January 2009 to April 2011. Prior to working at the EveryLife Foundation, Mr. Ditton served as the Vice President of Marketing at Diamics, Inc., a maker of cancer diagnostics, from October 2006 to December 2008 and Director of Global Marketing at BioMarin Pharmaceutical Inc., a biopharmaceutical company, from March 2004 to March 2006. Mr. Ditton holds an M.B.A. from the University of Tasmania.

**Tony Koutsoukos, Ph.D.** has served as our Vice President, Biometrics since October 2013. Prior to Ultragenyx, Mr. Koutsoukos worked as Vice President of Biometrics at Allos Therapeutics, a biopharmaceutical company, from September 2007 to March 2013, which was acquired by Spectrum Pharmaceuticals. He was also Director of Biostatistics at Amgen Inc., a biotechnology company, from May 2002 to September 2007. Prior to Amgen, Mr. Koutsoukos spent three years at Quintiles, a contract research company, as a Director of Biostatistics. His experience also includes five years at the FDA, Center for Drugs Evaluation and Research (CDER) division and approximately four years at the National Cancer Institute, Biometric Research Branch, CTEP, DCT. Dr. Koutsoukos received his Ph.D. and M.A., both in Mathematical Statistics from the University of Maryland, College Park.

**Cordelia Leonard** has served as our Vice President, Regulatory Affairs and Quality Assurance since July 2011. Prior to Ultragenyx, Ms. Leonard was Senior Director, Regulatory Affairs at BioMarin Pharmaceutical Inc., a biopharmaceutical company, from October 2003 until July 2011. Prior to BioMarin, Ms. Leonard was the Manager, Regulatory Affairs at Cerus Corporation, a biomedical products company, from May 1999 until October 2003. Ms. Leonard received bachelor degrees in Chemistry and Biological Science from the University of California, Irvine and holds both U.S. and EU Regulatory Affairs Certifications.

**Javier San Martin, M.D.** has served as our Vice President, Clinical Development since December 2013. Prior to Ultragenyx, Dr. San Martin served as Senior Vice President of Clinical Development at Alder Biopharmaceuticals Inc., a pharmaceutical company, from January 2012 through May 2013. Prior to Alder, Dr. San Martin served as Executive Director and Global Development Leader at Amgen, Inc., a biotechnology company, from March 2006 to September 2011. Prior to Amgen, Dr. San Martin served as Medical Advisor at Eli Lilly and Company, a pharmaceutical company, from June 1998 to February 2006. Dr. San Martin received his M.D. from the University of Buenos Aires Medical School and completed his residence in internal medicine at CEMIC University of Buenos Aires.

**Vimal Srivastava** has served as our Vice President, Program Development since August 2011. Before joining Ultragenyx, Mr. Srivastava was Senior Director, Portfolio and Project Management at Elan/Janssen Alzheimer Immunotherapy, a biotechnology company, from January 2008 until August 2011. He was also Director, Global Program Manager, Diabetes at Amgen Inc., a biotechnology company, from September 2005 to January 2008 and

Director, Program Management at BioMarin Pharmaceutical Inc., a biopharmaceutical company, from March 2003 to September 2005. Mr. Srivastava holds a B.S. in Pharmacy from Banaras Hindu University, an M.S. in Medicinal Chemistry from St. John's University and an M.A.S. in Management from Johns Hopkins University.

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**Michael Vellard, Ph.D.** has served as our Vice President, Research since May 2013. Prior to joining Ultragenyx, Dr. Vellard worked as Head of Lysosomal Biology at BioMarin Pharmaceutical Inc., a biopharmaceutical company, from October 1999 to May 2013. He was a postdoctoral fellow in the pediatric department at UCLA Harbor Medical Center from September 1992 to June 1995. Dr. Vellard received his B.S. in Natural and Life Sciences and M.S. in Molecular and Cellular Genetics from the University of Lyon I, France. He obtained his Ph.D. in Virology from the Pasteur and Curie Institutes (Universities Paris VI, VII and XI), France.

**Spencer Guthrie** has served as our Senior Director, Clinical Operations since June 2012. Prior to Ultragenyx, Mr. Guthrie worked as Director of Clinical Operations and Project Team Leader at Elan Pharmaceuticals, a biotechnology company, and Janssen Alzheimer's Immunotherapy from September 2007 to June 2012. Prior to that, Mr. Guthrie spent nine years with increasing responsibilities at Genentech in clinical operations and market planning. At Genentech, he worked on several innovative clinical programs, investigational new drug and biologics license application filings with Rituxan, Avastin, and Lucentis, including work on orphan indications. Mr. Guthrie also spent two years at ICON Clinical Research and a year at NASA's space science lab. Mr. Guthrie received his B.A. in Neuroscience from Vanderbilt University, an M.B.A. from the University of California, Irvine and he is certified as a Project Management Professional.

**Alison Skrinar, Ph.D.** has served as our Senior Director, Clinical Sciences since March 2012. Prior to joining Ultragenyx, Dr. Skrinar worked as the Senior Director of Clinical Outcomes and Regulatory Affairs from February 2009 to February 2012 at Enobia Pharma, Inc., a private clinical stage orphan company focused on the development of an enzyme replacement therapy for hypophosphastasia, which was acquired by Alexion in 2012. Prior to Enobia Pharma, Dr. Skrinar was the Senior Director of Clinical Outcomes at Genzyme Corporation, a biotechnology company, from May 2001 until January 2009. In her nearly 15 years in the biotechnology industry, Dr. Skrinar has worked exclusively on the clinical development and regulatory approval of ultra-orphan drugs. Dr. Skrinar received a B.B.A. from Emory University and a Ph.D. and a Master of Public Health degree from the University of Alabama.

**Theodore A. Huizenga** has served as our Corporate Controller since January 2014 and was appointed our Principal Accounting Officer in March 2014. Prior to joining us, Mr. Huizenga served as Controller of Crescendo Bioscience from August 2013 until January 2014. Prior to Crescendo Bioscience, Mr. Huizenga was employed by RDV Corporation, where he served as Director of Finance – Investment Group from January 2008 through December 2012 and as Corporate Controller from September 2003 through December 2007. Mr. Huizenga previously was a Manager in the Audit and Assurance Practice with PricewaterhouseCoopers LLP from January 1998 to September 2003. Mr. Huizenga has a B.S. in Accounting from Calvin College and is a Certified Public Accountant.

## **CERTAIN RELATIONSHIPS AND RELATED-PARTY TRANSACTIONS**

### **Related-Party Transactions**

Since January 1, 2013, other than participation in our initial public offering ( *IPO* ) by certain greater than 5% holders and entering into indemnification agreements with each of our executive officers and directors (as described below), we have not become, and are not currently proposed to be, a party to any transactions with any related parties, which are generally considered to be our directors and executive officers, nominees for director, holders of 5% or more of our outstanding common stock and members of their immediate families.

### **Participation in our Initial Public Offering**

In connection with our IPO, which closed in February 2014, the underwriters allocated shares of our common stock in the offering to certain of our greater than 5% holders on the same terms as the other shares that were offered and sold

in our IPO. These allocations included allocations of 240,000 shares to Adage Capital Partners, L.P. (who owned approximately 7.4% of our common stock immediately prior to the IPO) and 175,000



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shares to Beacon Bioventures Fund II Limited Partnership (who owned approximately 13.2% of our common stock immediately prior to the IPO). All of these shares were sold at \$21.00, which was the IPO price.

## **Indemnification Agreements and Directors and Officers Liability Insurance**

We have entered into indemnification agreements with each of our directors and executive officers. These agreements, among other things, require us to indemnify each director and executive officer to the fullest extent permitted by Delaware law, including indemnification of expenses such as attorneys' fees, judgments, penalties, fines and settlement amounts incurred by the director or executive officer in any action or proceeding, including any action or proceeding by or in right of us, arising out of the person's services as a director or executive officer.

## **Procedures for Related Party Transactions**

We have adopted a written related-party transaction approval policy that governs the review, approval and/or ratification of related-party transactions. Pursuant to this policy, if we want to enter into a transaction with a related party or an affiliate or immediate family member of a related party, our Chief Financial Officer will review the proposed transaction to determine, based on applicable NASDAQ and SEC rules, if such transaction qualifies as a related-party transaction. If the Chief Financial Officer determines that the proposed transaction is a related-party transaction, then the proposed transaction shall be submitted to the Audit Committee for pre-approval at the next regular or special Audit Committee meeting; if the Chief Financial Officer, in consultation with the Chief Executive Officer, determines that it is not practicable to wait until the next meeting of the Audit Committee, then the Chief Financial Officer shall submit the proposed transaction to the chairperson of the Audit Committee. In the event that our Chief Executive Officer or Chief Financial Officer becomes aware of a related-party transaction that has not been previously approved or previously ratified under our related-party transaction approval policy, the transaction, if pending or ongoing, will be promptly submitted to the Audit Committee or the chairperson of the Audit Committee for consideration. If the transaction is already completed, the Audit Committee or the chairperson of the Audit Committee shall evaluate the transaction to determine if rescission of the transaction and/or any disciplinary action is appropriate.

## **SECTION 16(A) BENEFICIAL OWNERSHIP REPORTING COMPLIANCE**

Under Section 16(a) of the Exchange Act and SEC rules, our directors, executive officers and beneficial owners of more than 10% of any class of equity security are required to file periodic reports of their ownership, and changes in that ownership, with the SEC. Because we did not become subject to the reporting requirements of Section 16(a) of the Exchange Act until January 2014, there were no such applicable SEC filings for the fiscal year ended December 31, 2013.

## **SECURITY OWNERSHIP OF CERTAIN BENEFICIAL OWNERS AND MANAGEMENT**

The following table sets forth information relating to the beneficial ownership of our common stock as of May 16, 2014, 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 current 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

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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 May 16, 2014 through the exercise of any stock option, warrants or other rights. We believe, based on the information furnished to us, that 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 30,059,288 shares of our common stock outstanding as of May 16, 2014. Shares of our common stock that a person has the right to acquire within 60 days of May 16, 2014 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 Ultragenyx Pharmaceutical Inc., at 60 Leveroni Court, Novato, California 94949.

Name and Address of Beneficial Owner	Beneficial Ownership	
	Number of Shares	% of Total
<b>Stockholders Owning Greater than 5%:</b>		
TPG Biotechnology Partners III, L.P. <sup>(1)</sup>	3,085,240	10.3%
Beacon Bioventures Fund II Limited Partnership <sup>(2)</sup>	3,260,240	10.9%
HealthCap VI, L.P. <sup>(3)</sup>	2,742,436	9.1%
Adage Capital Partners, L.P. <sup>(4)</sup>	1,867,713	6.2%
<b>Directors and Named Executive Officers:</b>		
Eran Nadav, Ph.D. <sup>(5)</sup>	2,431	*
Mårten Steen, M.D., Ph.D. <sup>(6)</sup>	2,431	*
William Aliski <sup>(7)</sup>	68,442	*
Matthew K. Fust <sup>(8)</sup>	7,431	*
Clay B. Siegall, Ph.D. <sup>(9)</sup>	2,431	*
Emil D. Kakkis, M.D., Ph.D. <sup>(10)</sup>	3,326,181	11.0%
Thomas Kassberg <sup>(11)</sup>	168,010	*
Shalini Sharp <sup>(12)</sup>	104,197	*
All executive officers and directors as a group <sup>(13)</sup> (8 persons)	3,681,554	12.1%

\* Indicates beneficial ownership of less than 1% of the total outstanding common stock.

- (1) Based on information set forth in a Form 4 filed with the SEC by TPG Group Holdings (SBS) Advisors, Inc. on February 7, 2014. TPG Biotechnology Partners III, L.P. is a Delaware limited partnership, whose general partner is TPG Biotechnology GenPar III, L.P., a Delaware limited partnership, whose general partner is TPG Biotechnology GenPar III Advisors, LLC, a Delaware limited liability company, whose sole member is TPG Holdings I, L.P., a Delaware limited partnership, whose general partner is TPG Holdings I-A, LLC, a Delaware

limited liability company, whose sole member is TPG Group Holdings (SBS), L.P., a Delaware limited partnership, whose general partner is TPG Group Holdings (SBS) Advisors, Inc., or Group Advisors, a Delaware corporation. Messrs. David Bonderman and James G. Coulter are officers and sole shareholders of Group Advisors and may therefore be deemed to be the beneficial owners of the shares held by TPG Biotechnology Partners III, L.P. Each of Messrs. Bonderman and Coulter and TPG Biotechnology Partners III, L.P. expressly disclaims beneficial ownership of the securities listed above except to the extent of any pecuniary interest therein. The address for Messrs. Bonderman and Coulter and TPG Biotechnology Partners III, L.P. is c/o TPG Global, LLC, 301 Commerce Street, Suite 3300, Fort Worth, TX 76102.

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- (2) Based on information set forth in a Schedule 13G filed with the SEC by FMR LLC on February 14, 2014. Beacon Bioventures Advisors Fund II Limited Partnership is the general partner of Beacon Bioventures Fund II Limited Partnership. Beacon Bioventures Advisors Fund II Limited Partnership is solely managed by Northern Neck Investors LLC, its general partner and investment manager. Northern Neck Investors LLC is owned by the shareholders and certain employees of FMR LLC, including certain members of the family of Edward C. Johnson 3d. Members of the family of Edward C. Johnson 3d, Chairman of FMR LLC, are the predominant owners, directly or through trusts, of Series B voting common shares of FMR LLC, representing 49% of the voting power of FMR LLC. The Johnson family group and all other Series B shareholders have entered into a shareholders voting agreement under which all Series B voting common shares will be voted in accordance with the majority vote of Series B voting common shares. Accordingly, through their ownership of voting common shares and the execution of the shareholders voting agreement, members of the Johnson family may be deemed, under the Investment Company Act of 1940, to form a controlling group with respect to FMR LLC. Each of the individuals and entities listed above expressly disclaims beneficial ownership of the securities listed above except to the extent of any pecuniary interest therein. The address for each of the individuals and entities listed above is 82 Devonshire Street, Boston, Massachusetts 02109.
- (3) HealthCap VI GP SA, L.L.C. ( **HCSA** ) is the sole general partner of HealthCap VI, L.P. HCSA has voting and dispositive power over the shares held by HealthCap VI, L.P. HCSA disclaims beneficial ownership of such shares, except to the extent of its pecuniary interest therein. Francois Kaiser, Dag Richter, and Daniel Schafer, the members of the board of HCSA, share voting and dispositive power over the shares held by HealthCap VI, L.P. and may be deemed to have indirect beneficial ownership of the shares held by such entities. The members of the board of HCSA disclaim beneficial ownership of shares held by HealthCap VI, L.P. except to the extent of any pecuniary interest therein. The address of HealthCap VI, L.P. is c/o HealthCap VI GP SA, 18, Avenue d Ouchy, 1006 Lausanne, Switzerland.
- (4) Adage Capital Partners, GP, LLC ( **ACPGP** ), serves as the general partner of Adage Capital Partners, L.P., a Delaware limited partnership (the **Fund** ) and as such has discretion over the portfolio of securities beneficially owned by the Fund. Adage Capital Advisors, LLC, a Delaware limited liability company ( **ACA** ), is managing member of ACPGP and directs ACPGP's operations. Robert Atchinson and Phillip Gross are the managing members of ACPGP and ACA and general partners of the Fund. Robert Atchinson and Phillip Gross disclaim beneficial ownership of the reported securities except to the extent of their pecuniary interest therein. The address of Adage Capital Partners, L.P. is 200 Clarendon Street, 52<sup>nd</sup> Floor, Boston, MA 02116.
- (5) Consists of 2,431 shares of common stock issuable pursuant to stock options exercisable within 60 days of May 16, 2014.
- (6) Consists of 2,431 shares of common stock issuable pursuant to stock options exercisable within 60 days of May 16, 2014.
- (7) Consists of (a) 36,903 shares of common stock, (b) 29,108 shares of common stock that may be acquired pursuant to the exercise of a warrant held by Mr. Aliski and (c) 2,431 shares of common stock issuable pursuant to stock options exercisable within 60 days of May 16, 2014.

- (8) Consists of (a) 5,000 shares of common stock and (b) 2,431 shares of common stock issuable pursuant to stock options exercisable within 60 days of May 16, 2014.
- (9) Consists of 2,431 shares of common stock issuable pursuant to stock options exercisable within 60 days of May 16, 2014.
- (10) Consists of (a) 2,552,241 shares of common stock held by the Emil Kakkis and Jenny Soriano Living Trust, dated June 18, 2009, (b) 624,240 shares of common stock held by Dr. Kakkis and (c) 149,700 shares of common stock that may be acquired pursuant to the exercise of warrants held by Dr. Kakkis. Dr. Kakkis shares voting and dispositive power over the 2,552,241 shares of common stock held by the Emil Kakkis and Jenny Soriano Living Trust, dated June 18, 2009; each of Dr. Kakkis and Dr. Soriano is a trustee of such trust. Dr. Kakkis has sole voting and dispositive power over the 624,240 shares of common stock held by him and the 149,700 shares of common stock that may be acquired pursuant to the exercise of warrants held by Dr. Kakkis.

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(11) Consists of (a) 125,804 shares of common stock and (b) 42,206 shares of common stock issuable pursuant to stock options exercisable within 60 days of May 16, 2014.

(12) Consists of (a) 84,255 shares of common stock and (b) 19,942 shares of common stock issuable pursuant to stock options exercisable within 60 days of May 16, 2014.

(13) Consists of (a) 3,428,443 shares held by our directors and officers, (b) 178,808 shares of common stock that may be acquired pursuant to the exercise of warrants by our certain of our directors and officers, and (c) 74,303 shares of common stock issuable pursuant to stock options held by our directors and officers that are exercisable within 60 days of May 16, 2014.

**EXECUTIVE COMPENSATION**

The following is a summary of the compensation arrangements of our named executive officers. Actual compensation programs that we may adopt may differ materially from current programs as summarized in this discussion. As an emerging growth company as defined in the JOBS Act, we are not required to include a Compensation Discussion and Analysis section and have elected to comply with the scaled disclosure requirements applicable to emerging growth companies.

**Summary Compensation Table**

The following table sets forth the compensation earned during the years ended December 31, 2013 and 2012 to our chief executive officer and our next two highest-paid executive officers. We refer to these officers as our named executive officers.

Name and Principal Position	Year	Salary	Bonus	Option Awards <sup>(1)</sup>	Non-equity	All	Total
					Incentive Plan Compensation <sup>(2)</sup>	Other Compensation <sup>(3)</sup>	
Emil D. Kakkis, M.D., Ph.D. <i>President and Chief Executive Officer</i>	2013	\$ 306,034	\$	\$ 218,873	\$ 140,910	\$ 19,721	\$ 685,538
	2012	\$ 300,172	\$	\$	\$ 92,125	\$ 9,056	\$ 401,353
Thomas Kassberg <i>Chief Business Officer and Senior Vice President</i>	2013	\$ 289,419	\$	\$ 182,394	\$ 114,345	\$ 20,352	\$ 606,510
	2012	\$ 272,041	\$	\$	\$ 81,813	\$ 15,640	\$ 369,494
Shalini Sharp <sup>(4)</sup> <i>Chief Financial Officer and Senior Vice President</i>	2013	\$ 278,440	\$ 30,000	\$ 182,394	\$ 110,206	\$ 24,193	\$ 625,233
	2012	\$ 148,669	\$	\$ 90,042	\$ 51,560	\$ 13,228	\$ 303,499

(1) The amounts reported in this column represent the grant date fair value of the stock options granted to our named executive officers during 2013 and 2012, respectively, 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 this column are set forth in the notes to our financial statements included in our Annual Report on Form 10-K for the fiscal year ended December 31, 2013. 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.

- (2) Amounts for fiscal 2013 represent cash bonuses earned in fiscal 2013, and paid during 2014, based on achievement of performance goals and other factors deemed relevant by our Board. Amounts for fiscal 2012 represent cash bonuses earned in 2012, and paid during 2013, based on achievement of performance goals and other factors deemed relevant by our Board.
- (3) Amounts reported in this column for Dr. Kakkis consist of medical, dental, vision and life/accidental death & dismemberment and key person life insurance premiums paid by us. Amounts reported in this column for Mr. Kassberg and Ms. Sharp consist of medical, dental, vision and life/accidental death & dismemberment premiums paid by us.
- (4) Ms. Sharp commenced employment with us in May 2012.



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**Narrative Disclosure to Summary Compensation Table**

***Employment Arrangements with Our Named Executive Officers***

***Emil D. Kakkis, M.D., Ph.D., President and Chief Executive Officer.*** We entered into an executive employment agreement with Dr. Kakkis in June 2011 for the position of President and Chief Executive Officer. Dr. Kakkis currently receives a base salary of \$500,000, which is subject to adjustment at the discretion of the Board or the Compensation Committee. Dr. Kakkis is also eligible for an annual performance bonus of up to 35% of his base salary, payable based on his individual performance evaluated against certain goals mutually agreed upon and our overall performance, as determined by the Board. Dr. Kakkis is also eligible to participate in our employee benefit plans, subject to the terms of those plans. Pursuant to the terms of the executive employment agreement, the employment of Dr. Kakkis is at will; we may terminate his employment at any time, without advance notice, for any reason or for no reason at all and Dr. Kakkis may terminate his employment at any time, upon four weeks' prior written notice, for any reason or for no reason at all.

***Thomas Kassberg, Chief Business Officer and Senior Vice President.*** We entered into an offer letter in October 2011 with Thomas Kassberg for the position of Chief Business Officer and Senior Vice President. Mr. Kassberg currently receives a base salary of \$350,000, which is subject to adjustment at the discretion of the Board or the Compensation Committee. Mr. Kassberg is also eligible for an annual performance bonus of up to 30% of his base salary, payable based on his individual performance evaluated against certain goals mutually agreed upon and our overall performance, as determined by the Chief Executive Officer in consultation with the Board. Additionally, pursuant to the terms of the offer letter, Mr. Kassberg received an option to purchase 202,584 shares of our common stock in connection with his hiring. Mr. Kassberg is eligible to participate in our employee benefit plans, subject to the terms of those plans. Pursuant to the terms of the offer letter, Mr. Kassberg's employment is at will and may be terminated either by us or by him, with or without advance notice, for any reason or for no reason at all.

***Shalini Sharp, Chief Financial Officer and Senior Vice President.*** We entered into an offer letter in March 2012 with Shalini Sharp for the position of Chief Financial Officer and Senior Vice President. Ms. Sharp currently receives a base salary of \$350,000, which is subject to adjustment at the discretion of the Board or the Compensation Committee. Ms. Sharp is also eligible for an annual performance bonus of up to 30% of her base salary, payable based on her individual performance evaluated against certain goals mutually agreed upon and our overall performance, as determined by the Chief Executive Officer in consultation with the Board. Additionally, pursuant to the terms of the offer letter, Ms. Sharp received an option to purchase 191,418 shares of our common stock in connection with her hiring. Ms. Sharp is eligible to participate in our employee benefit plans, subject to the terms of those plans. Pursuant to the terms of the offer letter, Ms. Sharp's employment is at will and may be terminated either by us or by her, with or without advance notice, for any reason or for no reason at all.

Each of these employment arrangements also contain provisions that provide for certain payments and benefits in the event of an involuntary termination of employment. In addition, the named executive officers may be entitled to accelerated vesting of their outstanding and unvested awards in certain circumstances. The information below describes certain compensation that may become due and payable as a result of certain events.

***Involuntary Termination of Employment***

Pursuant to their employment arrangements, each named executive officer is eligible to receive certain payments and benefits in the event of certain qualifying terminations, including termination of his or her employment by us without cause (as defined below) or resignation of his or her employment with good reason or because of a constructive termination (each, as defined below). Upon the timely execution of a general release of claims, each named executive

officer is eligible to receive the following payments and benefits:

if Dr. Kakkis is terminated by us other than for cause or because of death or disability, he shall be entitled to receive six months of base salary continuation;

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if Dr. Kakkis resigns his employment with us for good reason following a change in control (as defined below) within six months of the event constituting good reason and after providing us with 20 days to cure the good reason, then he shall be entitled to receive 12 months of base salary continuation; and

if Mr. Kassberg or Ms. Sharp is terminated by us without cause or resigns employment with us due to a constructive termination, each executive will be entitled to: (i) extend the exercise period applicable to any options then held such that the executive has 12 months from termination to exercise any of the vested shares, provided that in no event shall the exercise period be extended beyond the expiration date of any options then held; and (ii) six months of base salary continuation.

### ***Deemed Liquidation Event***

Pursuant to the offer letter with Mr. Kassberg, in addition to the severance benefits described above, in the event (i) we consummate a deemed liquidation event (as defined in our certificate of incorporation), which includes certain mergers or material asset sales, as well as any dissolution, liquidation, or winding down of the Company, (ii) Mr. Kassberg is employed by us on the date of the deemed liquidation event, and (iii) Mr. Kassberg is terminated by us without cause or resigns his employment with us due to a constructive termination within 12 months after the deemed liquidation event, the vesting of Mr. Kassberg's November 17, 2011 option to purchase 202,584 shares of our common stock shall accelerate with respect to 50% of the then-unvested shares subject to such option and any other equity held by Mr. Kassberg shall accelerate with respect to 100% of the then-unvested shares.

Pursuant to the offer letter with Ms. Sharp, in the event (i) we consummate a deemed liquidation event, (ii) Ms. Sharp is employed by us on the date of the deemed liquidation event, and (iii) Ms. Sharp is terminated without cause or resigns due to a constructive termination within 12 months of the deemed liquidation event, the vesting of all options held by Ms. Sharp as of the date of the deemed liquidation event shall accelerate with respect to 50% of the then-unvested shares.

### ***Definitions***

For purposes of Dr. Kakkis's employment agreement, cause means his:

commission of a felony or any crime involving dishonesty, breach of trust, or physical harm to any person;

willful engagement in conduct that is in bad faith and materially injurious to us, including but not limited to misappropriation of trade secrets, fraud, or embezzlement;

material breach of his employment agreement that is not cured within 10 days after written notice to him from us; or

willful refusal to implement or follow a lawful policy or directive of ours, which breach is not cured within 10 days after written notice to him from us.

For purposes of each of the offer letters with Mr. Kassberg and Ms. Sharp, cause means the named executive officer s:

gross negligence in carrying out, or material failure to carry out, his or her duties for us (including, without limitation, failure to cooperate in any company investigation), after notice from the Board and a reasonable opportunity to cure (if deemed curable);

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breach of his or her fiduciary duties to us, after notice from the Board and a reasonable opportunity to cure (if deemed curable);

conviction of, or plea of guilty or no contest to, any felony;

any act of fraud or embezzlement with respect to his or her obligations to us or otherwise relating to our business;

material violation of any of our policies;

material breach of any agreement entered into with us; or

unauthorized use or disclosure of confidential information or trade secrets of ours or of our affiliates.

For purposes of Dr. Kakkis's employment agreement, "good reason" means any of the following events if (i) we effect the event without the consent of Dr. Kakkis and (ii) such event occurs after a change in control:

a change in his position with us that materially reduces his level of responsibility;

a material reduction in his base salary, except for reductions that are comparable to reductions generally applicable to similarly situated executives of ours; or

a relocation of his principal place of employment by more than 50 miles.

For purposes of Dr. Kakkis's employment agreement, "change in control" means a change in ownership or control of us effected through a merger, consolidation, or acquisition by any person or related group of persons (other than an acquisition by us or by an employee benefit plan sponsored by us or by a person or persons that directly or indirectly control, is controlled by, or is under common control with, us) of beneficial ownership of securities possessing more than 50% of the total combined voting power of our outstanding securities.

For purposes of each of the offer letters with Mr. Kassberg and Ms. Sharp, "constructive termination" means the occurrence of any of the following events without the named executive officer's consent if (i) the executive provides us with written objection (or notice) to the event or condition within 30 days following the occurrence of the event or condition, (ii) we do not reverse or otherwise cure the event within 30 days of receiving such written objection, and (iii) the executive resigns his or her employment with us within 30 days following the expiration of that cure period:

a material reduction or change in the executive's job duties, responsibilities and requirements from the executive's job duties, responsibilities and requirements immediately prior to such reduction or change, taking into account the differences in job title and duties that are normally occasioned by

reason of an acquisition of one company by another;

a material reduction of the executive's base salary (other than an equal, across-the-board reduction in the compensation of all similarly-situated employees of ours or the surviving entity that is approved by the Board); or

a requirement that the executive relocate to a principal office that increases his or her one-way commute by more than 50 miles relative to the executive's immediately preceding principal office.

***Terms and Conditions of Annual Bonuses***

Our Board has adopted a corporate bonus plan, or the bonus plan, that provides for cash bonus payments based upon the attainment of performance targets established by our Compensation Committee. The payment targets relate to corporate, financial, and operational measures or objectives, or corporate performance goals, as well as individual performance objectives.

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Our Compensation Committee may select corporate performance goals from among the following: sales; revenue; assets; expenses; earnings from operations, earnings before or after deduction for all or any portion of interest, taxes, depreciation, amortization, incentives, service fees or extraordinary or special items, whether or not on a continuing operations or an aggregate or per share basis; net income or net income per common share (basic or diluted); return on equity, investment, capital or assets; one or more operating ratios; borrowing levels, leverage ratios or credit rating; market share; capital expenditures; cash flow, free cash flow, cash flow return on investment, or net cash provided by operations; stock price, dividends or total stockholder return; development of new technologies or products; sales of particular products or services; economic value created or added; operating margin or profit margin; customer acquisition or retention; raising or refinancing of capital; successful hiring of key individuals; resolution of significant litigation; acquisitions and divestitures (in whole or in part); joint ventures and strategic alliances; spin-offs, split-ups and the like; reorganizations; recapitalizations, restructurings, financings (issuance of debt or equity) or refinancings; or strategic business criteria, consisting of one or more objectives based on the following goals: meeting specified market penetration or value added, product development or introduction (including, without limitation, any clinical trial accomplishments, regulatory or other filings or approvals, or other product development milestones), geographic business expansion, cost targets, cost reductions or savings, customer satisfaction, operating efficiency, acquisition or retention, employee satisfaction, information technology, corporate development (including, without limitation, licenses, innovation, research or establishment of third party collaborations), manufacturing or process development, legal compliance or risk reduction, patent application or issuance goals, or goals relating to acquisitions or divestitures (in whole or in part), joint ventures or strategic alliances, any of which may be measured in absolute terms, as compared to any incremental increase, in terms of growth, as compared to results of a peer group, against the market as a whole, compared to applicable market indices and/or measured on a pre-tax or post-tax basis.

Each executive officer who is selected to participate in the bonus plan will have a target bonus opportunity set for each performance period. The bonus formulas will be adopted in each performance period by the Compensation Committee and communicated to each executive. The corporate performance goals will be measured at the end of each performance period after our financial reports have been published or such other appropriate time as the Compensation Committee determines. If the corporate performance goals and individual performance objectives are met, payments will be made following the end of each performance period. Subject to the rights contained in any agreement between the executive officer and the company, an executive officer must be employed by the company on the bonus payment date to be eligible to receive a bonus payment. The bonus plan also permits the compensation committee to approve additional bonuses in its sole discretion.

***Terms and Conditions of Equity Award Grants***

All of our named executive officers received options to purchase our common stock in fiscal year 2013. The table below entitled Outstanding Equity Awards at December 31, 2013 describes the material terms of option awards made to our named executive officers.

**Table of Contents****Equity Compensation*****Outstanding Equity Awards at December 31, 2013***

The following table sets forth information concerning the outstanding equity awards held by each of the named executive officers as of December 31, 2013.

Name	Option awards			
	Number of Securities Underlying Unexercised Options (#) Exercisable	Number of Securities Underlying Unexercised Options (#) Unexercisable	Option Exercise Price	Option Expiration Date
Emil D. Kakkis, M.D., Ph.D. <sup>(1)</sup>		47,854	\$ 6.86	10/31/2023
Thomas Kassberg <sup>(2)</sup>	12,661	97,071	\$ 0.31	11/16/2021
		39,878	\$ 6.86	10/31/2023
Shalini Sharp <sup>(3)</sup>	11,963	115,648	\$ 0.81	8/1/2022
		39,878	\$ 6.86	10/31/2023

(1) Represents an option to purchase 47,854 shares of our common stock granted on November 1, 2013. The shares underlying this option vest as follows: 25% vest on November 1, 2014, with the remainder of the shares vesting in equal monthly installments over the following three years through November 1, 2017, subject to the holder's continued service to us through each such vesting date.

(2) Represents an option to purchase 202,584 shares of our common stock granted on November 17, 2011. The shares underlying this option vest as follows: 25% vested on November 15, 2012, with the remainder of the shares vesting in equal monthly installments over the following three years through November 15, 2015, subject to the holder's continued service to us through each such vesting date. Vesting of 50% of the unvested shares shall accelerate in connection with a deemed liquidation event pursuant to the terms of Mr. Kassberg's offer letter dated October 31, 2011, as more fully described above under the section entitled "Narrative Disclosure to Summary Compensation Table Deemed Liquidation Event." Mr. Kassberg exercised 50,646 options on November 28, 2012 and 42,204 options on September 27, 2013.

Represents an option to purchase 39,878 shares of our common stock granted on November 1, 2013. The shares underlying this option vest as follows: 25% vest on November 1, 2014, with the remainder of the shares vesting in equal monthly installments over the following three years through November 1, 2017, subject to the holder's continued service to us through each such vesting date.

(3) Represents an option to purchase 191,418 shares of our common stock granted on August 2, 2012. The shares underlying this option vest as follows: 25% vested on May 21, 2013, with the remainder of the shares vesting in



equal monthly installments over the following three years through May 21, 2016, subject to the holder's continued service to us through each such vesting date. Vesting of 50% of the unvested shares shall accelerate in connection with a deemed liquidation event pursuant to the terms of Ms. Sharp's offer letter dated March 12, 2012, as more fully described above under the section entitled "Narrative Disclosure to Summary Compensation Table Deemed Liquidation Event." Ms. Sharp exercised 47,854 options on May 30, 2013, 7,975 options on July 24, 2013, and 7,975 options on September 26, 2013.

Represents an option to purchase 39,878 shares of our common stock granted on November 1, 2013. The shares underlying this option vest as follows: 25% vest on November 1, 2014, with the remainder of the shares vesting in equal monthly installments over the following three years through November 1, 2017, subject to the holder's continued service to us through each such vesting date.

### **Compensation Committee Report**

As an emerging growth company, the Company is not required to include a Compensation Discussion and Analysis section in this proxy statement.

**Table of Contents****Equity Compensation Plan Information**

The table below discloses information as of December 31, 2013 with respect to our equity compensation plans that have been approved by stockholders and equity compensation plans that have not been approved by stockholders.

Plan Category	Number of Securities to be Issued upon Exercise of Outstanding Options	Weighted-Average Exercise Price of Outstanding Options	Number of Securities Remaining Available for Future Issuance under Equity Compensation Plans (Excluding Securities Reflected in Column (a))
Equity compensation plans approved by security holders:			
2011 Equity Incentive Plan, as amended	2,222,839	\$ 3.41	799,963
Equity compensation plans not approved by security holders	-	-	-
Total	2,222,839	\$ 3.41	799,963

**DIRECTOR COMPENSATION**

Dr. Kakkis, our President and Chief Executive Officer, receives no compensation for his service as a director. None of our non-employee directors received compensation from us for their service on the Board or otherwise during fiscal 2013.

Our Board has adopted a non-employee director compensation policy that is designed to provide a total compensation package that enables us to attract and retain, on a long-term basis, high caliber non-employee directors. Under the policy, all non-employee directors will be paid cash compensation during fiscal 2014 and thereafter as set forth below:

	Annual Retainer
<b>Board of Directors:</b>	
All non-employee members	\$ 35,000
<b>Audit Committee:</b>	
Chairman	\$ 15,000
Non-Chairman members	\$ 7,500
<b>Compensation Committee:</b>	
Chairman	\$ 10,000
Non-Chairman members	\$ 5,000
<b>Nominating and Corporate Governance Committee:</b>	

Chairman	\$ 6,000
Non-Chairman members	\$ 3,000

Under the non-employee director compensation policy, each non-employee director who is initially appointed or elected to the Board will receive an option grant to purchase up to 17,500 shares of our common stock under our stock option plan on the date he or she first becomes a non-employee director, which will vest monthly over a three-year period, subject to the holder's continued service to us through each such vesting date. In addition, on the date of the annual meeting of stockholders, each continuing non-employee director will be

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eligible to receive an annual option grant to purchase up to 7,500 shares of our common stock, which will vest in full upon the first anniversary of the date of grant, subject to the holder's continued service to us through such vesting date. All of the foregoing options will be granted at fair market value as of the date of grant.

## **COMPENSATION RISK ASSESSMENT**

We believe that our executive compensation program does not encourage excessive or unnecessary risk taking. This is primarily due to the fact that our compensation programs are designed to encourage our executive officers and other employees to remain focused on both short-term and long-term strategic goals, particularly in connection with our pay-for-performance compensation philosophy. As a result, we do not believe that our compensation programs are reasonably likely to have a material adverse effect on us.

## **REPORT OF THE AUDIT COMMITTEE**

The Audit Committee evaluates auditor performance, manages relations with the Company's independent registered public accounting firm, and evaluates policies and procedures relating to internal control systems. The Audit Committee operates under a written Audit Committee Charter that has been adopted by the Board, a copy of which is available on the Company's website at [www.ultragenyx.com](http://www.ultragenyx.com). All members of the Audit Committee currently meet the independence and qualification standards for Audit Committee membership set forth in the listing standards and rules of NASDAQ and the SEC.

No member of the Audit Committee is a professional accountant or auditor. The members' functions are not intended to duplicate or to certify the activities of management and the independent registered public accounting firm. The Audit Committee serves a board-level oversight role in which it provides advice, counsel and direction to management and the auditors on the basis of the information it receives, discussions with management and the auditors, and the experience of the Audit Committee's members in business, financial and accounting matters.

The Audit Committee oversees the Company's financial reporting process on behalf of the Board. The Company's management has the primary responsibility for the financial statements and reporting process, including the Company's system of internal controls over financial reporting. In fulfilling its oversight responsibilities, the Audit Committee reviewed with management the audited financial statements included in the Annual Report on Form 10-K for the fiscal year ended December 31, 2013. This review included a discussion of the quality and the acceptability of the Company's financial reporting, including the nature and extent of disclosures in the financial statements and the accompanying notes.

The Audit Committee also reviewed with the Company's independent registered public accounting firm, which is responsible for expressing an opinion on the conformity of the audited financial statements with accounting principles generally accepted in the United States of America, their judgments as to the quality and the acceptability of the Company's financial reporting and such other matters as are required to be discussed with the Committee pursuant to Public Company Accounting Oversight Board Auditing Standard No. 16, Communication with Audit Committees. The Audit Committee has received the written disclosures and the letter from the independent registered public accounting firm required by the Public Company Accounting Oversight Board regarding the independent accountant's communications with the Audit Committee concerning independence. The Audit Committee discussed with the independent registered public accounting firm their independence from management and the Company, including the matters required by the applicable rules of the Public Company Accounting Oversight Board.

In addition to the matters specified above, the Audit Committee discussed with the Company's independent registered public accounting firm the overall scope, plans and estimated costs of their audit. The Committee met with the

independent registered public accounting firm periodically, with and without management present, to discuss the results of the independent registered public accounting firm's examinations,

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the overall quality of the Company's financial reporting and the independent registered public accounting firm's reviews of the quarterly financial statements, and drafts of the quarterly and annual reports.

In reliance on the reviews and discussions referred to above, the Audit Committee recommended to the Board that the Company's audited financial statements should be included in the Company's Annual Report on Form 10-K for the fiscal year ended December 31, 2013.

Submitted by the Audit Committee of the Board of

Directors

Matthew Fust, Chairperson

Eran Nadav

Mårten Steen

## **OTHER BUSINESS**

We know of no other matters to be submitted to a vote of stockholders at the Annual Meeting. If any other matter is properly brought before the Annual Meeting or any adjournment thereof, it is the intention of the persons named in the enclosed proxy to vote the shares they represent in accordance with their judgment. In order for any stockholder to nominate a candidate or to submit a proposal for other business to be acted upon at a given annual meeting, he or she must provide timely written notice to our corporate secretary in the form prescribed by our Bylaws, as described below.

## **STOCKHOLDER PROPOSALS**

Stockholder proposals intended to be included in the proxy materials for the 2015 annual meeting of stockholders must be received by the Secretary of the Company no later than January 30, 2015, or otherwise as permitted by applicable law. The form and substance of these proposals must satisfy the requirements established by the Company's Amended and Restated Bylaws and the SEC, and the timing for the submission of any such proposals may be subject to change as a result of changes in SEC rules and regulations.

Additionally, stockholders who intend to present a stockholder proposal at the 2015 annual meeting of stockholders must provide the Secretary of the Company with written notice of the proposal between 90 and 120 days prior to the one-year anniversary date of the 2014 annual meeting; *provided, however*, that if the 2015 annual meeting date is advanced by more than 30 days before or delayed by more than 60 days after the anniversary date of the 2014 annual meeting, then stockholders must provide notice on or before 10 days after the day on which the date of the 2015 annual meeting is first disclosed in a public announcement. Notice must be tendered in the proper form prescribed by our Amended and Restated Bylaws. Proposals not meeting the requirements set forth in our Amended and Restated Bylaws will not be entertained at the annual meeting.

Any stockholder seeking to recommend a director candidate or any director candidate who wishes to be considered by the Nominating and Corporate Governance Committee, the committee that recommends nominees to the Board for election at each annual meeting, must provide the Secretary of the Company with (a) all information relating to such nominee that would be required to be disclosed in a proxy statement for the election of such nominee as a director and such nominee's written consent to serve as a director if elected and (b) a description of all direct and indirect

compensation and other material monetary arrangements, agreements or understandings during the past three years, and any other material relationship, if any, between or concerning such stockholder and its respective affiliates or associates, on the one hand, and the proposed nominee, and his or her respective affiliates or associates, on the other hand. The Nominating and Corporate Governance Committee is not required to consider director candidates received after the applicable date or without the required information. The Nominating and Corporate Governance Committee will consider all director candidates who comply with these requirements and will evaluate these candidates using the criteria described above under the caption, Nomination of Directors. Director candidates who are then nominated by the Board will be included in the Company's proxy statement for that annual meeting.

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**DELIVERY OF PROXY MATERIALS**

**Our annual report to stockholders for the fiscal year ended December 31, 2013, including audited financial statements, accompanies this Proxy Statement. Copies of our Annual Report on Form 10-K for fiscal 2013 and the exhibits thereto are available from the Company without charge upon written request of a stockholder to our investor relations department at 60 Leveroni Court, Novato, California 94949.** Copies of these materials are also available online through the SEC at [www.sec.gov](http://www.sec.gov). The Company may satisfy SEC rules regarding delivery of proxy materials, including the proxy statement, annual report and Notice, by delivering a single Notice and, if applicable, a single set of proxy materials to an address shared by two or more Company stockholders. This delivery method can result in meaningful cost savings for the Company. In order to take advantage of this opportunity, the Company may deliver only one Notice and, if applicable, a single set of proxy materials to multiple stockholders who share an address, unless contrary instructions are received prior to the mailing date. Similarly, if you share an address with another stockholder and have received multiple copies of our Notice and/or other proxy materials, you may write or call us at the address and phone number below to request delivery of a single copy of the Notice and, if applicable, other proxy materials in the future. We undertake to deliver promptly upon written or oral request a separate copy of the Notice and, if applicable, other proxy materials, as requested, to a stockholder at a shared address to which a single copy of the Notice and/or other proxy materials was delivered. If you hold stock as a registered holder and prefer to receive separate copies of a Notice and, if applicable, other proxy materials either now or in the future, please contact the Company's investor relations department at 60 Leveroni Court, Novato, California 94949 or by telephone at (415) 483-8800. If your stock is held through a brokerage firm, bank or other financial institution and you prefer to receive separate copies of a Notice and, if applicable, other proxy materials either now or in the future, please contact your brokerage firm, bank or other financial institution.

**EACH STOCKHOLDER IS URGED TO COMPLETE, DATE, SIGN AND PROMPTLY RETURN  
THE ENCLOSED PROXY.**



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**Exhibit A**

**ULTRAGENYX PHARMACEUTICAL INC.**

**2014 INCENTIVE PLAN**

**1. DEFINED TERMS**

Exhibit A, which is incorporated by reference, defines the terms used in the Plan and sets forth certain operational rules related to those terms.

**2. PURPOSE**

The Plan has been established to advance the interests of the Company by providing for the grant to Participants of Stock-based and other incentive Awards.

**3. ADMINISTRATION**

The Administrator has discretionary authority, subject only to the express provisions of the Plan, to interpret the Plan; determine eligibility for and grant Awards; determine, modify or waive the terms and conditions of any Award; prescribe forms, rules and procedures relating to the Plan; and otherwise do all things necessary or appropriate to carry out the purposes of the Plan. Determinations of the Administrator made under the Plan will be conclusive and will bind all parties.

**4. LIMITS ON AWARDS UNDER THE PLAN**

(a) **Number of Shares.** The maximum number of shares of Stock that may be delivered in satisfaction of Awards under the Plan is 2,250,000, which includes the number of shares of Stock that were available for grant under the Ultragenyx Pharmaceutical Inc. 2011 Equity Incentive Plan as of the Date of Adoption that are hereby transferred to the Plan or become available for grant under the Ultragenyx Pharmaceutical Inc. 2011 Equity Incentive Plan following the Date of Adoption as a result of the termination, cancellation or forfeiture of awards, plus an annual increase to be added on January 1 of each year, beginning January 1, 2015 through January 1, 2024, equal to the least of (i) 2,500,000 shares of Stock, (ii) 4 percent (4%) of the number of shares of Stock outstanding on a fully diluted basis as of the close of business on the immediately preceding day (calculated by adding to the number of shares of Stock outstanding, all outstanding securities convertible into Stock on such date on an as converted basis), and (iii) a lesser amount determined by the Administrator on or prior to January 1 of a given year. Up to the total number of shares set forth in this Section 4(a) as increased by the provisions of this Section 4(a) above may be issued in satisfaction of ISOs, but nothing in this Section 4(a) will be construed as requiring that any, or any fixed number of, ISOs be awarded under the Plan. The limits set forth in this Section 4(a) shall be construed to comply with Section 422 of the Code. For purposes of this Section 4(a), the number of shares of Stock delivered in satisfaction of Awards will be determined (i) net of shares of Stock underlying the portion of any Award that is settled in cash or the portion of any Stock Option or SAR that expires, terminates or is forfeited prior to the issuance of Stock thereunder, and (ii) by treating as having been delivered the full number of shares covered by any portion of a SAR that is settled in Stock (and not only the number of shares of Stock delivered in settlement). If Awards are forfeited or are terminated for any

reason before being exercised or becoming vested, then the shares of Stock underlying such Awards shall again become available for issuance under the Plan. Any shares withheld from an Award to satisfy the tax withholding obligations with respect to such Award or in payment of the exercise price of an Award requiring exercise shall not again be available for issuance under the Plan.

**(b) Type of Shares.** Stock delivered by the Company under the Plan may be authorized but unissued Stock or previously issued Stock acquired by the Company. No fractional shares of Stock will be delivered under the Plan.

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(c) **Section 162(m) Limits.** The following additional limits will apply to Awards of the specified type granted, or in the case of Cash Awards, payable to any person in any calendar year, to the extent that such Awards are to be treated as performance-based compensation under Section 162(m) of the Code:

(1) Stock Options: 1,000,000 shares of Stock.

(2) SARs: 1,000,000 shares of Stock.

(3) Awards other than Stock Options, SARs or Cash Awards: 1,000,000 shares of Stock.

(4) Cash Awards: \$430,000.

In applying the foregoing limits, (i) all Awards of the specified type granted to the same person in the same calendar year will be aggregated and made subject to one limit; (ii) the limits applicable to Stock Options and SARs refer to the number of shares of Stock subject to those Awards; (iii) the share limit under clause (3) refers to the maximum number of shares of Stock that may be delivered, or the value of which could be paid in cash or other property, under an Award or Awards of the type specified in clause (3) assuming a maximum payout; and (iv) the dollar limit under clause (4) refers to the maximum dollar amount payable under an Award or Awards of the type specified in clause (4) assuming a maximum payout. The foregoing provisions will be construed in a manner consistent with Section 162(m), including, without limitation, where applicable, the rules under Section 162(m) pertaining to permissible deferrals of exempt awards. Any Awards granted to a person in any calendar year that are in excess of the foregoing limits shall not be deemed to be performance-based compensation under Section 162(m).

## **5. ELIGIBILITY AND PARTICIPATION**

The Administrator will select Participants from among key Employees and directors of, and consultants and advisors to, the Company and its Affiliates. Eligibility for ISOs is limited to individuals described in the first sentence of this Section 5 who are employees of the Company or of a parent corporation or subsidiary corporation of the Company as those terms are defined in Section 424 of the Code. Eligibility for Stock Options other than ISOs is limited to individuals described in the first sentence of this Section 5 who are providing direct services on the date of grant of the Stock Option to the Company or to a subsidiary of the Company that would be described in the first sentence of Treas. Regs. §1.409A-1(b)(5)(iii)(E).

## **6. RULES APPLICABLE TO AWARDS**

### **(a) All Awards.**

(1) **Award Provisions.** The Administrator will determine the terms of all Awards, subject to the limitations provided herein. By accepting (or, under such rules as the Administrator may prescribe, being deemed to have accepted) an Award, the Participant will be deemed to have agreed to the terms of the Award and the Plan. Notwithstanding any provision of this Plan to the contrary, awards of an acquired company that are converted, replaced or adjusted in connection with the acquisition may contain terms and conditions that are inconsistent with the terms and conditions specified herein, as determined by the Administrator.

(2) **Term of Plan.** No Awards may be made after ten years from the Date of Adoption, but previously granted Awards may continue beyond that date in accordance with their terms.

**(3) Transferability.** Neither ISOs nor, except as the Administrator otherwise expressly provides in accordance with the second sentence of this Section 6(a)(3), other Awards may be transferred other than by will or by the laws of descent and distribution. During a Participant's lifetime, ISOs (and, except as the Administrator otherwise expressly provides in accordance with the second sentence of this Section 6(a)(3), SARs and NSOs) may be exercised only by the Participant. The Administrator may permit the gratuitous transfer (*i.e.*, transfer not for value) of Awards other than ISOs to any transferee eligible to be covered by the provisions of Form S-8 (under the Securities Act of 1933), subject to such limitations as the Administrator may impose.

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**(4) Vesting, etc.** The Administrator will determine the time or times at which an Award will vest or become exercisable and the terms on which a Stock Option or SAR will remain exercisable. Without limiting the foregoing, the Administrator may at any time accelerate the vesting or exercisability of an Award, regardless of any adverse or potentially adverse tax or other consequences resulting from such acceleration. Unless the Administrator expressly provides otherwise, however, the following rules will apply if a Participant's Employment ceases:

**(A)** Immediately upon the cessation of the Participant's Employment and except as provided in (B) and (C) below, each Stock Option and SAR that is then held by the Participant or by the Participant's permitted transferees, if any, will cease to be exercisable and will terminate and all other Awards that are then held by the Participant or by the Participant's permitted transferees, if any, to the extent not already vested will be forfeited.

**(B)** Subject to (C) and (D) below, all Stock Options and SARs held by the Participant or the Participant's permitted transferees, if any, immediately prior to the cessation of the Participant's Employment, to the extent then exercisable, will remain exercisable for the lesser of (i) a period of three months or (ii) the period ending on the latest date on which such Stock Option or SAR could have been exercised without regard to this Section 6(a)(4), and will thereupon immediately terminate.

**(C)** All Stock Options and SARs held by a Participant or the Participant's permitted transferees, if any, immediately prior to the Participant's death, to the extent then exercisable, will remain exercisable for the lesser of (i) the one year period ending with the first anniversary of the Participant's death or (ii) the period ending on the latest date on which such Stock Option or SAR could have been exercised without regard to this Section 6(a)(4), and will thereupon immediately terminate.

**(D)** All Stock Options and SARs (whether or not exercisable) held by a Participant or the Participant's permitted transferees, if any, immediately prior to the cessation of the Participant's Employment will immediately terminate upon such cessation of Employment if the termination is for Cause or occurs in circumstances that in the sole determination of the Administrator would have constituted grounds for the Participant's Employment to be terminated for Cause.

**(5) Additional Restrictions.** The Administrator may cancel, rescind, withhold or otherwise limit or restrict any Award at any time if the Participant is not in compliance with all applicable provisions of the Award agreement and the Plan, or if the Participant breaches any agreement with the Company or its Affiliates with respect to confidentiality. Without limiting the generality of the foregoing, the Administrator may recover Awards made under the Plan and payments under or gain in respect of any Award to the extent required to comply with Company policy, with Section 10D of the Securities Exchange Act of 1934, as amended, or any stock exchange or similar rule adopted under said Section.

**(6) Taxes.** The delivery, vesting and retention of Stock, cash or other property under an Award are conditioned upon full satisfaction by the Participant of all tax withholding requirements with respect to the Award. The Administrator will prescribe such rules for the withholding of taxes as it deems necessary. The Administrator may, but need not, hold back shares of Stock from an Award or permit a Participant to tender previously owned shares of Stock in satisfaction of tax withholding requirements (but not in excess of the minimum withholding required by law).

**(7) Dividend Equivalents, Etc.** The Administrator may provide for the payment of amounts (on terms and subject to conditions established by the Administrator) in lieu of cash dividends or other cash distributions with respect to Stock subject to an Award whether or not the holder of such Award is otherwise entitled to share in the actual dividend or distribution in respect of such Award. Any entitlement to dividend equivalents or similar entitlements will be established and administered either consistent with an exemption from, or in compliance with, the requirements of Section 409A. Dividends or dividend equivalent amounts payable in respect of Awards that are subject to restrictions

may be subject to such limits or restrictions as the Administrator may impose.

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**(8) Rights Limited.** Nothing in the Plan will be construed as giving any person the right to continued employment or service with the Company or its Affiliates, or any rights as a stockholder except as to shares of Stock actually issued under the Plan. The loss of existing or potential profit in Awards will not constitute an element of damages in the event of termination of Employment for any reason, even if the termination is in violation of an obligation of the Company or any Affiliate to the Participant.

**(9) Section 162(m).** In the case of any Performance Award (other than a Stock Option or SAR) intended to qualify for the performance-based compensation exception under Section 162(m), the Administrator will establish the applicable Performance Criterion or Criteria in writing no later than ninety (90) days after the commencement of the period of service to which the performance relates (or at such earlier time as is required to qualify the Award as performance-based under Section 162(m)) and, prior to the event or occurrence (grant, vesting or payment, as the case may be) that is conditioned on the attainment of such Performance Criterion or Criteria, will certify whether it or they have been attained. The preceding sentence will not apply to an Award eligible (as determined by the Administrator) for exemption from the limitations of Section 162(m) by reason of the post-initial public offering transition relief in Section 1.162-27(f) of the Treasury Regulations.

**(10) Coordination with Other Plans.** Awards under the Plan may be granted in tandem with, or in satisfaction of or substitution for, other Awards under the Plan or awards made under other compensatory plans or programs of the Company or its Affiliates. For example, but without limiting the generality of the foregoing, awards under other compensatory plans or programs of the Company or its Affiliates may be settled in Stock (including, without limitation, Unrestricted Stock) if the Administrator so determines, in which case the shares delivered will be treated as awarded under the Plan (and will reduce the number of shares thereafter available under the Plan in accordance with the rules set forth in Section 4). In any case where an award is made under another plan or program of the Company or its Affiliates and such award is intended to qualify for the performance-based compensation exception under Section 162(m), and such award is settled by the delivery of Stock or another Award under the Plan, the applicable Section 162(m) limitations under both the other plan or program and under the Plan will be applied to the Plan as necessary (as determined by the Administrator) to preserve the availability of the Section 162(m) performance-based compensation exception with respect thereto.

**(11) Section 409A.** Each Award will contain such terms as the Administrator determines, and will be construed and administered, such that the Award either qualifies for an exemption from the requirements of Section 409A or satisfies such requirements.

**(12) Fair Market Value.** In determining the fair market value of any share of Stock under the Plan, the Administrator will make the determination in good faith consistent with the rules of Section 422 and Section 409A to the extent applicable.

**(b) Stock Options and SARs.**

**(1) Time and Manner of Exercise.** Unless the Administrator expressly provides otherwise, no Stock Option or SAR will be deemed to have been exercised until the Administrator receives a notice of exercise (in form acceptable to the Administrator), which may be an electronic notice, signed (including electronic signature in form acceptable to the Administrator) by the appropriate person and accompanied by any payment required under the Award. A Stock Option or SAR exercised by any person other than the Participant will not be deemed to have been exercised until the Administrator has received such evidence as it may require that the person exercising the Award has the right to do so.

**(2) Exercise Price.** The exercise price (or the base value from which appreciation is to be measured) of each Award requiring exercise will be no less than 100% (or in the case of an ISO granted to a ten-percent shareholder within the

meaning of subsection (b)(6) of Section 422, 110%) of the fair market value of the Stock subject to the Award, determined as of the date of grant, or such higher amount as the Administrator may

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determine in connection with the grant. No such Award, once granted, may be repriced other than with stockholder approval. Fair market value will be determined by the Administrator consistent with the applicable requirements of Section 422 and Section 409A.

**(3) Payment of Exercise Price.** Where the exercise of an Award is to be accompanied by payment, payment of the exercise price will be by cash or check acceptable to the Administrator or by such other legally permissible means, if any, as may be acceptable to the Administrator.

**(4) Maximum Term.** Stock Options and SARs will have a maximum term not to exceed ten (10) years from the date of grant (or five (5) years from the date of grant in the case of an ISO granted to a ten-percent shareholder described in Section 6(b)(2) above); provided, however, that, if a Participant still holding an outstanding but unexercised NSO or SAR ten (10) years from the date of grant (or, in the case of an NSO or SAR with a maximum term of less than ten (10) years, such maximum term) is prohibited by applicable law or a written policy of the Company applicable to similarly situated employees from engaging in any open-market sales of Stock, and if at such time the Stock is publicly traded (as determined by the Administrator), the maximum term of such Award will instead be deemed to expire on the thirtieth (30<sup>th</sup>) day following the date the Participant is no longer prohibited from engaging in such open market sales.

## **7. EFFECT OF CERTAIN TRANSACTIONS**

**(a) Mergers, etc.** Except as otherwise provided in an Award agreement, the following provisions will apply in the event of a Covered Transaction:

**(1) Assumption or Substitution.** If the Covered Transaction is one in which there is an acquiring or surviving entity, the Administrator may (but, for the avoidance of doubt, need not) provide (i) for the assumption or continuation of some or all outstanding Awards or any portion thereof or (ii) for the grant of new awards in substitution therefor by the acquiror or survivor or an affiliate of the acquiror or survivor.

**(2) Cash-Out of Awards.** Subject to Section 7(a)(5) below the Administrator may (but, for the avoidance of doubt, need not) provide for payment (a cash-out ), with respect to some or all Awards or any portion thereof, equal in the case of each affected Award or portion thereof to the excess, if any, of (A) the fair market value of one share of Stock (as determined by the Administrator in its reasonable discretion) times the number of shares of Stock subject to the Award or such portion, over (B) the aggregate exercise or purchase price, if any, under the Award or such portion (in the case of an SAR, the aggregate base value above which appreciation is measured), in each case on such payment terms (which need not be the same as the terms of payment to holders of Stock) and other terms, and subject to such conditions, as the Administrator determines.

**(3) Acceleration of Certain Awards.** Subject to Section 7(a)(5) below, the Administrator may (but, for the avoidance of doubt, need not) provide that any Award requiring exercise will become exercisable, in full or in part and/or that the delivery of any shares of Stock remaining deliverable under any outstanding Award of Stock Units (including Restricted Stock Units and Performance Awards to the extent consisting of Stock Units) will be accelerated in full or in part, in each case on a basis that gives the holder of the Award a reasonable opportunity, as determined by the Administrator, following exercise of the Award or the delivery of the shares, as the case may be, to participate as a stockholder in the Covered Transaction.

**(4) Termination of Awards Upon Consummation of Covered Transaction.** Except as the Administrator may otherwise determine in any case, each Award will automatically terminate (and in the case of outstanding shares of

Restricted Stock, will automatically be forfeited) upon consummation of the Covered Transaction, other than Awards assumed or continued pursuant to Section 7(a)(1) above.

**(5) Additional Limitations.** Any share of Stock and any cash or other property delivered pursuant to Section 7(a)(2) or Section 7(a)(3) above with respect to an Award may, in the discretion of the Administrator, contain such restrictions, if any, as the Administrator deems appropriate to reflect any performance or other

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vesting conditions to which the Award was subject and that did not lapse (and were not satisfied) in connection with the Covered Transaction. For purposes of the immediately preceding sentence, a cash-out under Section 7(a)(2) above or acceleration under Section 7(a)(3) above will not, in and of itself, be treated as the lapsing (or satisfaction) of a performance or other vesting condition. In the case of Restricted Stock that does not vest and is not forfeited in connection with the Covered Transaction, the Administrator may require that any amounts delivered, exchanged or otherwise paid in respect of such Stock in connection with the Covered Transaction be placed in escrow or otherwise made subject to such restrictions as the Administrator deems appropriate to carry out the intent of the Plan.

**(b) Changes in and Distributions With Respect to Stock.**

**(1) Basic Adjustment Provisions.** In the event of a stock dividend, stock split or combination of shares (including a reverse stock split), recapitalization or other change in the Company's capital structure that constitutes an equity restructuring within the meaning of FASB ASC 718, the Administrator will make appropriate adjustments to the maximum number of shares specified in Section 4(a) that may be delivered under the Plan and to the maximum share limits described in Section 4(c), and will also make appropriate adjustments to the number and kind of shares of stock or securities subject to Awards then outstanding or subsequently granted, any exercise prices relating to Awards and any other provision of Awards affected by such change.

**(2) Certain Other Adjustments.** The Administrator may also make adjustments of the type described in Section 7(b)(1) above to take into account distributions to stockholders other than those provided for in Section 7(a) and 7(b)(1), or any other event, if the Administrator determines that adjustments are appropriate to avoid distortion in the operation of the Plan, having due regard for the qualification of ISOs under Section 422, the requirements of Section 409A, and for the performance-based compensation rules of Section 162(m), where applicable.

**(3) Continuing Application of Plan Terms.** References in the Plan to shares of Stock will be construed to include any stock or securities resulting from an adjustment pursuant to this Section 7.

**8. LEGAL CONDITIONS ON DELIVERY OF STOCK**

The Company will not be obligated to deliver any shares of Stock pursuant to the Plan or to remove any restriction from shares of Stock previously delivered under the Plan until: (i) the Company is satisfied that all legal matters in connection with the issuance and delivery of such shares have been addressed and resolved; (ii) if the outstanding Stock is at the time of delivery listed on any stock exchange or national market system, the shares to be delivered have been listed or authorized to be listed on such exchange or system upon official notice of issuance; and (iii) all conditions of the Award have been satisfied or waived. The Company may require, as a condition to exercise of the Award, such representations or agreements as counsel for the Company may consider appropriate to avoid violation of the Securities Act of 1933 or any applicable state or non-U.S. securities law. Any Stock required to be issued to Participants under the Plan will be evidenced in such manner as the Administrator may deem appropriate, including book-entry registration or delivery of stock certificates. In the event that the Administrator determines that Stock certificates will be issued to Participants under the Plan, the Administrator may require that certificates evidencing Stock issued under the Plan bear an appropriate legend reflecting any restriction on transfer applicable to such Stock, and the Company may hold the certificates pending lapse of the applicable restrictions.

**9. AMENDMENT AND TERMINATION**

The Administrator may at any time or times amend the Plan or any outstanding Award for any purpose which may at the time be permitted by law, and may at any time terminate the Plan as to any future grants of Awards; provided, that except as otherwise expressly provided in the Plan the Administrator may not, without the Participant's consent, alter the terms of an Award so as to affect materially and adversely the Participant's rights

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under the Award, unless the Administrator expressly reserved the right to do so at the time the Award was granted. Any amendments to the Plan will be conditioned upon stockholder approval only to the extent, if any, such approval is required by law (including the Code and applicable stock exchange requirements), as determined by the Administrator.

## **10. OTHER COMPENSATION ARRANGEMENTS**

The existence of the Plan or the grant of any Award will not in any way affect the Company's right to Award a person bonuses or other compensation in addition to Awards under the Plan.

## **11. MISCELLANEOUS**

**(a) Waiver of Jury Trial.** By accepting an Award under the Plan and to the extent permitted under applicable law, each Participant waives any right to a trial by jury in any action, proceeding or counterclaim concerning any rights under the Plan and any Award, or under any amendment, waiver, consent, instrument, document or other agreement delivered or which in the future may be delivered in connection therewith, and agrees that any such action, proceedings or counterclaim will be tried before a court and not before a jury. By accepting an Award under the Plan, each Participant certifies that no officer, representative, or attorney of the Company has represented, expressly or otherwise, that the Company would not, in the event of any action, proceeding or counterclaim, seek to enforce the foregoing waivers. Notwithstanding anything to the contrary in the Plan, nothing herein is to be construed as limiting the ability of the Company and a Participant to agree to submit disputes arising under the terms of the Plan or any Award made hereunder to binding arbitration or as limiting the ability of the Company to require any eligible individual to agree to submit such disputes to binding arbitration as a condition of receiving an Award hereunder.

**(b) Limitation of Liability.** Notwithstanding anything to the contrary in the Plan, neither the Company, nor any Affiliate, nor the Administrator, nor any person acting on behalf of the Company, any Affiliate, or the Administrator, will be liable to any Participant or to the estate or beneficiary of any Participant or to any other holder of an Award by reason of any acceleration of income, or any additional tax (including any interest and penalties), asserted by reason of the failure of an Award to satisfy the requirements of Section 422 or Section 409A or by reason of Section 4999 of the Code, or otherwise asserted with respect to the Award; provided, that nothing in this Section 11(b) will limit the ability of the Administrator or the Company, in its discretion, to provide by separate express written agreement with a Participant for any payment in connection with any such acceleration of income or additional tax.

## **12. ESTABLISHMENT OF SUB-PLANS**

The Administrator may from time to time establish one or more sub-plans under the Plan for purposes of satisfying applicable blue sky, securities or tax laws of various jurisdictions. The Administrator will establish such sub-plans by adopting supplements to the Plan setting forth (i) such limitations on the Administrator's discretion under the Plan as it deems necessary or desirable and (ii) such additional terms and conditions not otherwise inconsistent with the Plan as it deems necessary or desirable. All supplements so established will be deemed to be part of the Plan, but each supplement will apply only to Participants within the affected jurisdiction (as determined by the Administrator).

## **13. GOVERNING LAW**

**(a) Certain Requirements of Corporate Law.** Awards will be granted and administered consistent with the requirements of applicable Delaware law relating to the issuance of stock and the consideration to be received therefor, and with the applicable requirements of the stock exchanges or other trading systems on which the Stock is listed or entered for trading, in each case as determined by the Administrator.

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**(b) Other Matters.** Except as otherwise provided by the express terms of an Award agreement, under a sub-plan described in Section 12 or as provided in Section 13(a) above, the provisions of the Plan and of Awards under the Plan and all claims or disputes arising out of or based upon the Plan or any Award under the Plan or relating to the subject matter hereof or thereof will be governed by and construed in accordance with the domestic substantive laws of the State of Delaware without giving effect to any choice or conflict of laws provision or rule that would cause the application of the domestic substantive laws of any other jurisdiction.

**(c) Jurisdiction.** By accepting an Award, each Participant will be deemed to (a) have submitted irrevocably and unconditionally to the jurisdiction of the federal and state courts located within the geographic boundaries of the United States District Court for the Northern District of California for the purpose of any suit, action or other proceeding arising out of or based upon the Plan or any Award; (b) agree not to commence any suit, action or other proceeding arising out of or based upon the Plan or an Award, except in the federal and state courts located within the geographic boundaries of the United States District Court for the Northern District of California; and (c) waive, and agree not to assert, by way of motion as a defense or otherwise, in any such suit, action or proceeding, any claim that it is not subject personally to the jurisdiction of the above-named courts that its property is exempt or immune from attachment or execution, that the suit, action or proceeding is brought in an inconvenient forum, that the venue of the suit, action or proceeding is improper or that the Plan or an Award or the subject matter thereof may not be enforced in or by such court.

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**EXHIBIT A**

**Definition of Terms**

The following terms, when used in the Plan, will have the meanings and be subject to the provisions set forth below:

**Administrator** : The Compensation Committee, except that the Compensation Committee may delegate (i) to one or more of its members (or one or more other members of the Board (including the full Board)) such of its duties, powers and responsibilities as it may determine; (ii) to one or more officers of the Company the power to grant Awards to the extent permitted by Section 157(c) of the Delaware General Corporation Law; and (iii) to such Employees or other persons as it determines such ministerial tasks as it deems appropriate. In the event of any delegation described in the preceding sentence, the term Administrator will include the person or persons so delegated to the extent of such delegation.

**Affiliate** : Any corporation or other entity that stands in a relationship to the Company that would result in the Company and such corporation or other entity being treated as one employer under Section 414(b) and Section 414(c) of the Code.

**Award** : Any or a combination of the following:

- (i) Stock Options.
- (ii) SARs.
- (iii) Restricted Stock.
- (iv) Unrestricted Stock.
- (v) Stock Units, including Restricted Stock Units.
- (vi) Performance Awards.
- (vii) Cash Awards.
- (viii) Awards (other than Awards described in (i) through (vii) above) that are convertible into or otherwise based on Stock.

**Board** : The Board of Directors of the Company.

**Cash Award** : An Award denominated in cash.

**Cause** : In the case of any Participant who is party to an employment or severance-benefit agreement that contains a definition of Cause, the definition set forth in such agreement will apply with respect to such Participant under the Plan. In the case of any other Participant, Cause will mean, as determined by the Administrator in its reasonable judgment, (i) a substantial failure of the Participant to perform the Participant's duties and responsibilities to the Company or subsidiaries or substantial negligence in the performance of such duties and responsibilities; (ii) the commission by the Participant of a felony or a crime involving moral turpitude; (iii) the commission by the Participant of theft, fraud, embezzlement, material breach of trust or any material act of dishonesty involving the Company or any



of its subsidiaries; (iv) a significant violation by the Participant of the code of conduct of the Company or its subsidiaries of any material policy of the Company or its subsidiaries, or of any statutory or common law duty of loyalty to the Company or its subsidiaries; (v) material breach of any of the terms of the Plan or any Award made under the Plan, or of the terms of any other agreement between the Company or subsidiaries and the Participant; or (vi) other conduct by the Participant that could be expected to be harmful to the business, interests or reputation of the Company.

**Code :** The U.S. Internal Revenue Code of 1986 as from time to time amended and in effect, or any successor statute as from time to time in effect.

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**Compensation Committee** : The Compensation Committee of the Board.

**Company** : Ultragenyx Pharmaceutical Inc.

**Covered Transaction** : Any of (i) a consolidation, merger, or similar transaction or series of related transactions, including a sale or other disposition of stock, in which the Company is not the surviving corporation or which results in the acquisition of all or substantially all of the Company's then outstanding common stock by a single person or entity or by a group of persons and/or entities acting in concert, (ii) a sale or transfer of all or substantially all the Company's assets, or (iii) a dissolution or liquidation of the Company. Where a Covered Transaction involves a tender offer that is reasonably expected to be followed by a merger described in clause (i) (as determined by the Administrator), the Covered Transaction will be deemed to have occurred upon consummation of the tender offer.

**Date of Adoption** : The earlier of the date the Plan was approved by the Company's stockholders or adopted by the Board, as determined by the Compensation Committee.

**Employee** : Any person who is employed by the Company or an Affiliate.

**Employment** : A Participant's employment or other service relationship with the Company and its Affiliates, which may include service as a director, consultant or independent contractor. Employment will be deemed to continue, unless the Administrator expressly provides otherwise, so long as the Participant is employed by, or otherwise is providing services in a capacity described in Section 5 to the Company or an Affiliate. If a Participant's employment or other service relationship is with an Affiliate and that entity ceases to be an Affiliate, the Participant's Employment will be deemed to have terminated when the entity ceases to be an Affiliate unless the Participant transfers Employment to the Company or its remaining Affiliates. Notwithstanding the foregoing and the definition of Affiliate above, in construing the provisions of any Award relating to the payment of nonqualified deferred compensation (subject to Section 409A) upon a termination or cessation of Employment, references to termination or cessation of employment, separation from service, retirement or similar or correlative terms will be construed to require a separation from service (as that term is defined in Section 1.409A-1(h) of the Treasury Regulations) from the Company and from all other corporations and trades or businesses, if any, that would be treated as a single service recipient with the Company under Section 1.409A-1(h)(3) of the Treasury Regulations. The Company may, but need not, elect in writing, subject to the applicable limitations under Section 409A, any of the special elective rules prescribed in Section 1.409A-1(h) of the Treasury Regulations for purposes of determining whether a separation from service has occurred. Any such written election will be deemed a part of the Plan.

**ISO** : A Stock Option intended to be an incentive stock option within the meaning of Section 422. Each Stock Option granted pursuant to the Plan will be treated as providing by its terms that it is to be an NSO unless, as of the date of grant, it is expressly designated as an ISO.

**NSO** : A Stock Option that is not intended to be an incentive stock option within the meaning of Section 422.

**Participant** : A person who is granted an Award under the Plan.

**Performance Award** : An Award subject to Performance Criteria. The Administrator in its discretion may grant Performance Awards that are intended to qualify for the performance-based compensation exception under Section 162(m) and Performance Awards that are not intended so to qualify.

**Performance Criteria** : Specified criteria, other than the mere continuation of Employment or the mere passage of time, the satisfaction of which is a condition for the grant, exercisability, vesting or full enjoyment of an Award. For

purposes of Awards that are intended to qualify for the performance-based compensation

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exception under Section 162(m), a Performance Criterion will mean an objectively determinable measure of performance relating to any, or any combination, of the following (measured either absolutely or by reference to an index or indices and determined either on a consolidated basis or, as the context permits, on a divisional, subsidiary, line of business, project or geographical basis or in combinations thereof): sales; revenues; assets; expenses; earnings from operations; earnings before or after deduction for all or any portion of interest, taxes, depreciation, amortization, incentives, service fees or extraordinary or special items, whether or not on a continuing operations or an aggregate or per share basis; net income or net income per common share (basic or diluted); return on equity, investment, capital or assets; one or more operating ratios; borrowing levels, leverage ratios or credit rating; market share; capital expenditures; cash flow, free cash flow, cash flow return on investment, or net cash provided by operations; stock price, dividends or total stockholder return; development of new technologies or products; sales of particular products or services; economic value created or added; operating margin or profit margin; customer acquisition or retention; raising or refinancing of capital; successful hiring of key individuals; resolution of significant litigation; acquisitions and divestitures (in whole or in part); joint ventures and strategic alliances; spin-offs, split-ups and the like; reorganizations; recapitalizations, restructurings, financings (issuance of debt or equity) or refinancings; or strategic business criteria, consisting of one or more objectives based on the following goals: meeting specified market penetration or value added, product development or introduction (including, without limitation, any clinical trial accomplishments, regulatory or other filings or approvals, or other product development milestones), geographic business expansion, cost targets, cost reductions or savings, customer satisfaction, operating efficiency, acquisition or retention, employee satisfaction, information technology, corporate development (including, without limitation, licenses, innovation, research or establishment of third party collaborations), manufacturing or process development, legal compliance or risk reduction, patent application or issuance goals, or goals relating to acquisitions or divestitures (in whole or in part), joint ventures or strategic alliances. A Performance Criterion and any targets with respect thereto determined by the Administrator need not be based upon an increase, a positive or improved result or avoidance of loss. To the extent consistent with the requirements for satisfying the performance-based compensation exception under Section 162(m), the Administrator may provide in the case of any Award intended to qualify for such exception that one or more of the Performance Criteria applicable to such Award will be adjusted in an objectively determinable manner to reflect events (for example, but without limitation, acquisitions or dispositions) occurring during the performance period that affect the applicable Performance Criterion or Criteria.

**Plan** : The Ultragenyx Pharmaceutical Inc. 2014 Incentive Plan as from time to time amended and in effect.

**Restricted Stock** : Stock subject to restrictions requiring that it be redelivered or offered for sale to the Company if specified conditions are not satisfied.

**Restricted Stock Unit** : A Stock Unit that is, or as to which the delivery of Stock or cash in lieu of Stock is, subject to the satisfaction of specified performance or other vesting conditions.

**SAR** : A right entitling the holder upon exercise to receive an amount (payable in cash or in shares of Stock of equivalent value) equal to the excess of the fair market value of the shares of Stock subject to the right over the base value from which appreciation under the SAR is to be measured.

**Section 409A** : Section 409A of the Code.

**Section 422** : Section 422 of the Code.

**Section 162(m)** : Section 162(m) of the Code.

**Stock** : Common stock of the Company, par value \$0.001 per share.

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**Stock Option** : An option entitling the holder to acquire shares of Stock upon payment of the exercise price.

**Stock Unit** : An unfunded and unsecured promise, denominated in shares of Stock, to deliver Stock or cash measured by the value of Stock in the future.

**Unrestricted Stock** : Stock not subject to any restrictions under the terms of the Award.

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**Exhibit B**

**ULTRAGENYX PHARMACEUTICAL INC.**

**CORPORATE BONUS PLAN**

1. Purpose

This Corporate Bonus Plan (the ***Bonus Plan***) is intended to provide an incentive for superior work and to motivate eligible executives of Ultragenyx Pharmaceutical Inc. (the ***Company***) toward even higher achievement and business results, to tie their goals and interests to those of the Company and its stockholders and to enable the Company to attract and retain highly qualified executives. The Bonus Plan is for the benefit of Covered Executives (as defined below).

2. Covered Executives

From time to time, the Compensation Committee of the Board of Directors of the Company (the ***Compensation Committee***) may select certain key executives (the ***Covered Executives***) to be eligible to receive bonuses hereunder. Participation in this Plan does not change the at will nature of a Covered Executive's employment with the Company.

3. Administration

The Compensation Committee shall have the sole discretion and authority to administer and interpret the Bonus Plan.

4. Bonus Determinations

(a) Corporate Performance Goals. A Covered Executive may receive a bonus payment under the Bonus Plan based upon the attainment of one or more performance objectives that are established by the Compensation Committee and relate to financial and/or operational metrics with respect to the Company or any of its subsidiaries (the ***Corporate Performance Goals***), including the following: sales; revenue; assets; expenses; earnings from operations, earnings before or after deduction for all or any portion of interest, taxes, depreciation, amortization, incentives, service fees or extraordinary or special items, whether or not on a continuing operations or an aggregate or per share basis; net income or net income per common share (basic or diluted); return on equity, investment, capital or assets; one or more operating ratios; borrowing levels, leverage ratios or credit rating; market share; capital expenditures; cash flow, free cash flow, cash flow return on investment, or net cash provided by operations; stock price, dividends or total stockholder return; development of new technologies or products; sales of particular products or services; economic value created or added; operating margin or profit margin; customer acquisition or retention; raising or refinancing of capital; successful hiring of key individuals; resolution of significant litigation; acquisitions and divestitures (in whole or in part); joint ventures and strategic alliances; spin-offs, split-ups and the like; reorganizations; recapitalizations, restructurings, financings (issuance of debt or equity) or refinancings; or strategic business criteria, consisting of one or more objectives based on the following goals: meeting specified market penetration or value added, product development or introduction (including, without limitation, any clinical trial accomplishments, regulatory or other filings or approvals, or other product development milestones), geographic business expansion, cost targets, cost reductions or savings, customer satisfaction, operating efficiency, acquisition or retention, employee satisfaction,

information technology, corporate development (including, without limitation, licenses, innovation, research or establishment of third party collaborations), manufacturing or process development, legal compliance or risk reduction, patent application or issuance goals, or goals relating to acquisitions or divestitures (in whole or in part), joint ventures or strategic alliances, any of which may be (A) measured in absolute terms or compared to any incremental increase, (B) measured in terms of growth, (C) compared to another company or companies or to results of a peer group, (D) measured against the market as a whole and/or as compared to applicable market indices and/or

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(E) measured on a pre-tax or post-tax basis (if applicable). Further, any Corporate Performance Goals may be used to measure the performance of the Company as a whole or a business unit or other segment of the Company, or one or more product lines or specific markets. The Corporate Performance Goals may differ from Covered Executive to Covered Executive.

(b) Calculation of Corporate Performance Goals. At the beginning of each applicable performance period, the Compensation Committee will determine whether any significant element(s) will be included in or excluded from the calculation of any Corporate Performance Goal with respect to any Covered Executive. In all other respects, Corporate Performance Goals will be calculated in accordance with the Company's financial statements, generally accepted accounting principles, or under a methodology established by the Compensation Committee at the beginning of the performance period and which is consistently applied with respect to a Corporate Performance Goal in the relevant performance period.

(c) Target; Minimum; Maximum. Each Corporate Performance Goal shall have a target (*e.g.*, 100 percent attainment of the Corporate Performance Goal) and may also have a minimum hurdle and/or a maximum amount.

(d) Bonus Requirements; Individual Goals. Except as otherwise set forth in this Section 4(d), to the extent practicable under the circumstances: (i) any bonuses paid to Covered Executives under the Bonus Plan shall be based upon objectively determinable bonus formulas that tie such bonuses to one or more performance targets relating to the Corporate Performance Goals, (ii) bonus formulas for Covered Executives shall be adopted in each performance period by the Compensation Committee and communicated to each Covered Executive at the beginning of each performance period and (iii) no bonuses shall be paid to Covered Executives unless and until the Compensation Committee makes a determination with respect to the attainment of the performance targets relating to the Corporate Performance Goals. Notwithstanding the foregoing, the Compensation Committee may adjust bonuses payable under the Bonus Plan based on achievement of one or more individual performance objectives or pay bonuses (including, without limitation, discretionary bonuses) to Covered Executives under the Bonus Plan based on individual performance goals and/or upon such other terms and conditions as the Compensation Committee may in its discretion determine.

(e) Individual Target Bonuses. The Compensation Committee shall establish a target bonus opportunity for each Covered Executive for each performance period. For each Covered Executive, the Compensation Committee shall have the authority to apportion the target award so that a portion of the target award shall be tied to attainment of Corporate Performance Goals and a portion of the target award shall be tied to attainment of individual performance objectives.

(f) Employment Requirement. Subject to any additional terms contained in a written agreement between the Covered Executive and the Company, the payment of a bonus to a Covered Executive with respect to a performance period shall be conditioned upon the Covered Executive's employment by the Company on the bonus payment date. If a Covered Executive was not employed for an entire performance period, the Compensation Committee may pro rate the bonus based on the number of days employed during such period.

**5. Timing of Payment**

(a) The Corporate Performance Goals will be measured at the end of each performance period after the Company's financial reports with respect to such period(s) have been published. If the Corporate Performance Goals and/or individual goals for such period are met, payments will be made as soon as practicable following the end of such period, but not later 74 days after the end of the fiscal year in which such performance period ends.

(b) With respect to Corporate Performance Goals established and measured on an annual or multi-year basis, Corporate Performance Goals will be measured as of the end of each such performance period (*e.g.*, the

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end of each fiscal year) after the Company's financial reports with respect to such period(s) have been published. If the Corporate Performance Goals and/ or individual goals for any such period are met, bonus payments will be made as soon as practicable, but not later than 74 days after the end of the relevant fiscal year.

(c) For the avoidance of doubt, bonuses earned at any time in a fiscal year must be paid no later than 74 days after the last day of such fiscal year.

6. Amendment and Termination

The Company reserves the right to amend or terminate the Bonus Plan at any time in its sole discretion.

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**ULTRAGENYX PHARMACEUTICAL INC.**

**60 LEVERONI COURT**

**NOVATO, CA 94949**

**VOTE BY INTERNET**

*Before The Meeting* Go to **[www.proxyvote.com](http://www.proxyvote.com)**

Use the Internet to transmit your voting instructions and for electronic delivery of information up until 11:59 P.M. Eastern Time the day before the cut-off date or meeting date. Have your proxy card in hand when you access the web site and follow the instructions to obtain your records and to create an electronic voting instruction form.

*During The Meeting* Go to **[www.virtualshareholdermeeting.com/RARE14](http://www.virtualshareholdermeeting.com/RARE14)**

You may attend the Meeting via the Internet and vote during the Meeting. Have the information that is printed in the box marked by the arrow available and follow the instructions.

**VOTE BY PHONE 1-800-690-6903**

Use any touch-tone telephone to transmit your voting instructions up until 11:59 P.M. Eastern Time the day before the cut-off date or meeting date. Have your proxy card in hand when you call and then follow the instructions.

**VOTE BY MAIL**

Mark, sign and date your proxy card and return it in the postage-paid envelope we have provided or return it to Vote Processing, c/o Broadridge, 51 Mercedes Way, Edgewood, NY 11717.

TO VOTE, MARK BLOCKS BELOW IN BLUE OR BLACK INK AS FOLLOWS:

KEEP THIS PORTION FOR YOUR RECORDS

DETACH AND RETURN THIS PORTION ONLY

**THIS PROXY CARD IS VALID ONLY WHEN SIGNED AND DATED.**

**ULTRAGENYX PHARMACEUTICAL INC.**

**The Board of Directors recommends that you vote FOR the election of the nominee for director named below:**

**Nominee:**

1a. Emil D. Kakkis, M.D., Ph.D.

**For**

**Withhold**

..

..

**The Board of Directors recommends you vote FOR proposals 2, 3 and 4:**

**For Against Abstain**

- |    |   |    |    |    |
|----|---|----|----|----|
| 2. | Ratification of the selection of Ernst & Young LLP as independent registered public accounting firm for the fiscal year ending December 31, 2014. | .. | .. | .. |
| 3. | Reapproval of the Internal Revenue Code Section 162(m) provisions of the 2014 Incentive Plan.   | .. | .. | .. |
| 4. | Reapproval of the Internal Revenue Code Section 162(m) provisions of the Corporate Bonus Plan.  | .. | .. | .. |

For address changes and/or comments, please check this box and write them on the back where indicated.

..

Please indicate if you plan to attend this meeting.

..

..

**Yes**

**No**

Please sign exactly as your name(s) appear(s) hereon. When signing as attorney, executor, administrator, or other fiduciary, please give full title as such. Joint owners should each sign personally. All holders must sign. If a corporation or partnership, please sign in full corporate or partnership name, by authorized officer.

Signature [PLEASE SIGN WITHIN BOX]

Date

Signature (Joint Owners)

Date

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**ULTRAGENYX PHARMACEUTICAL INC.**

**Annual Meeting of Stockholders**

**July 10, 2014 4:00 p.m., Pacific Time**

**This proxy is solicited by the Board of Directors**

The stockholder(s) hereby appoint(s) Emil D. Kakkis, M.D., Ph.D. and Shalini Sharp, or either of them, as proxies, each with the power to appoint his or her substitute, and hereby authorize(s) them to represent and to vote, as designated on the reverse side of this ballot, all of the shares of Common Stock of Ultragenyx Pharmaceutical Inc. that the stockholder(s) is/are entitled to vote at the 2014 Annual Meeting of Stockholders to be held at 4:00 p.m., Pacific Time, on July 10, 2014, and at any adjournment or postponement thereof.

**This proxy, when properly executed, will be voted in the manner directed herein. If no such direction is made, this proxy will be voted in accordance with the Board of Directors' recommendations and in the discretion of the proxy holders on all other matters that may come before the meeting.**

**Address Changes/Comments:**

(If you noted any Address Changes/Comments above, please mark corresponding box on the reverse side.)

**Continued and to be signed on reverse side**