

AGIOS PHARMACEUTICALS INC

Form DEF 14A

April 18, 2019

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UNITED STATES

SECURITIES AND EXCHANGE COMMISSION

Washington, D.C. 20549

SCHEDULE 14A

(Rule 14a-101)

SCHEDULE 14A INFORMATION

Proxy Statement Pursuant to Section 14(a) of the

Securities Exchange Act of 1934

(Amendment No.)

Filed by the Registrant

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))

Definitive Proxy Statement

Definitive Additional Materials

Soliciting Material under § 240.14a-12

Agios Pharmaceuticals, Inc.

(Name of Registrant as Specified In Its Charter)

(Name of Person(s) Filing Proxy Statement, if other than the Registrant)

Payment of Filing Fee (Check the appropriate box):

No fee required.

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

- (1) Title of each class of securities to which transaction applies:

- (2) Aggregate number of securities to which transaction applies:

- (3) Per unit price or other underlying value of transaction computed pursuant to Exchange Act Rule 0-11 (set forth the amount on which the filing fee is calculated and state how it was determined):

- (4) Proposed maximum aggregate value of transaction:

- (5) Total fee paid:

Fee paid previously with preliminary materials.

Check box if any part of the fee is offset as provided by Exchange Act Rule 0-11(a)(2) and identify the filing for which the offsetting fee was paid previously. Identify the previous filing by registration statement number, or the Form or Schedule and the date of its filing.

- (1) Amount Previously Paid:

- (2) Form, Schedule or Registration Statement No.:

(3) Filing Party:

(4) Date Filed:

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88 Sidney Street, Cambridge, Massachusetts 02139

NOTICE OF ANNUAL MEETING OF STOCKHOLDERS

TO BE HELD ON WEDNESDAY, MAY 29, 2019

Dear Stockholder:

You are cordially invited to our Annual Meeting of Stockholders on Wednesday, May 29, 2019, beginning at 9:00 a.m., Eastern Time, at our offices located at 88 Sidney Street, Cambridge, Massachusetts 02139, for the following purposes:

1. To elect each of the three Class III director nominees set forth in the Proxy Statement, each to serve for a three-year term expiring at the 2022 annual meeting of stockholders and until his or her respective successor is duly elected and qualified;
2. To vote, on an advisory basis, to approve the compensation paid to our named executive officers;
3. To ratify the appointment of PricewaterhouseCoopers LLP as our independent registered public accounting firm for the fiscal year ending December 31, 2019; and
4. To transact such other business as may be properly brought before the meeting or any adjournment or postponement thereof.

Stockholders of record as of the close of business on April 8, 2019 are entitled to vote at the meeting.

At Agios we are keenly focused on the contribution we can make to environmental sustainability. Instead of mailing a paper copy of our proxy materials to all of our stockholders, this year we are providing access to our proxy materials over the internet under the U.S. Securities and Exchange Commission's notice and access rules. As a result, we are mailing to our stockholders a Notice of Internet Availability of Proxy Materials (the Notice) instead of a paper copy of this Proxy Statement and our Annual Report for the fiscal year ended December 31, 2018 (the 2018 Annual Report). We are mailing the Notice on or about April 18, 2019, and it contains instructions on how to access those documents over the internet. The Notice also contains instructions on how each of our stockholders can receive a paper copy of our proxy materials, including this Proxy Statement, our 2018 Annual Report, and a form of proxy card or voting instruction card. All stockholders who do not receive the Notice, including shareholders who have previously requested to receive paper copies of proxy materials, will receive a paper copy of the proxy materials by mail unless they have previously requested delivery of proxy materials electronically. We have chosen to employ this distribution process to conserve natural resources and reduce the costs of printing and distributing our proxy materials.

We encourage all stockholders to attend the Annual Meeting in person. Whether or not you plan to attend the Annual Meeting in person, we encourage you to read this Proxy Statement and submit your proxy or voting instructions as soon as possible by using the internet as described in the instructions included on your Notice, by calling the toll-free telephone number included on your Notice, or, if you received a paper copy of the proxy materials, by completing,

signing, dating and returning your proxy card or voting instruction form.

Thank you for your ongoing support and continued interest in Agios Pharmaceuticals, Inc.

By Order of the Board of Directors,

Jacquelyn A. Fouse, Ph.D.

Chief Executive Officer

Cambridge, Massachusetts

April 18, 2019

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Important Notice Regarding the Availability of Proxy Materials for the Annual Meeting of Stockholders to be Held on May 29, 2019: This Proxy Statement and our 2018 Annual Report to Stockholders are available at www.proxydocs.com/agio. These documents are also available to any stockholder who wishes to receive a paper copy by calling (866) 648-8133, visiting www.investorelections.com/agio or emailing paper@investorelections.com.

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88 Sidney Street, Cambridge, Massachusetts 02139

PROXY STATEMENT FOR THE 2019 ANNUAL MEETING OF STOCKHOLDERS

TO BE HELD ON WEDNESDAY, MAY 29, 2019

INFORMATION CONCERNING SOLICITATION AND VOTING

This Proxy Statement contains information about our 2019 annual meeting of stockholders, or the Annual Meeting. The meeting will be held on Wednesday, May 29, 2019, beginning at 9:00 a.m. Eastern Time, at our offices located at 88 Sidney Street, Cambridge, Massachusetts 02139. Except where the context otherwise requires, references to Agios Pharmaceuticals, Agios, we, us, our and similar terms refer to Agios Pharmaceuticals, Inc. and its consolidated subsidiaries. References to our website are inactive textual references only and the contents of our website are not incorporated by reference into this Proxy Statement.

This Proxy Statement and the enclosed proxy card are being furnished in connection with the solicitation of proxies by our board of directors for use at the Annual Meeting and at any adjournment of that meeting. All proxies will be voted in accordance with the instructions they contain. If you do not specify your voting instructions on your proxy, it will be voted in accordance with the recommendations of our board of directors. We are making this Proxy Statement, the related proxy card and our annual report to stockholders for the fiscal year ended December 31, 2018, or the 2018 Annual Report, available to stockholders for the first time on or about April 18, 2019.

GENERAL INFORMATION ABOUT THE ANNUAL MEETING AND VOTING

Why do I have access to these materials?

We have made these proxy materials available to you because our board of directors is soliciting your proxy to vote at the Annual Meeting to be held on May 29, 2019 at 9:00 a.m., Eastern Time, including at any adjournments or postponements of the meeting. As a holder of record of common stock as of the close of business on April 8, 2019, you are invited to attend the Annual Meeting and are requested to vote on the items of business described in this Proxy Statement. This Proxy Statement includes information that we are required to provide to you under the rules adopted by the U.S. Securities and Exchange Commission, or the SEC, and that is designed to assist you in voting your shares.

Why did I receive a notice in the mail regarding the internet availability of proxy materials instead of a full set of proxy materials?

Because we care about the sustainability of our environment, and in accordance with SEC rules, we have elected to provide access to our proxy materials, including this Proxy Statement and our 2018 Annual Report, over the internet. Accordingly, we have sent a Notice Regarding the Availability of Proxy Materials, or the Notice, to our stockholders of record entitled to vote at the Annual Meeting with instructions for accessing the proxy materials and voting over the internet or by telephone. We mailed the Notice on or about April 18, 2019 to all stockholders entitled to vote at the Annual Meeting.

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All stockholders entitled to vote at the Annual Meeting will have the ability to access the proxy materials by visiting the website referred to in the Notice, www.proxydocs.com/agio. This makes the proxy distribution process more efficient and less costly and helps conserve natural resources. The Notice also contains instructions to request to receive a printed set of the proxy materials. You may request the proxy materials over the internet at www.investorelections.com/agio, by emailing paper@investorelections.com, or by calling (866) 648-8133.

The Notice also identifies the date, time and location of the Annual Meeting; the matters to be acted upon at the Annual Meeting and our board of directors' recommendation with regard to each matter; a toll-free telephone number, an e-mail address, and a website where stockholders can request to receive, free of charge, a paper or e-mail copy of the Proxy Statement, our Annual Report on Form 10-K for the year ended December 31, 2018, and a form of proxy relating to the Annual Meeting; and information on how to access and vote the form of proxy.

Can I vote my shares by filling out and returning the Notice?

No. The Notice identifies the items to be voted on at the Annual Meeting, but you cannot vote by marking the Notice and returning it. The Notice provides instructions on how to vote over the internet or by telephone, by requesting and returning a printed proxy card, or by submitting a ballot in person at the Annual Meeting.

What does it mean if I receive more than one Notice?

If you receive more than one Notice, your shares may be registered in more than one name or in different accounts. Please follow the voting instructions on the Notices to ensure that all of your shares are voted.

What is the purpose of the Annual Meeting?

At the Annual Meeting, stockholders will consider and vote on the following matters:

- (1) To elect each of the three Class III director nominees set forth in the Proxy Statement, each to serve for a three-year term expiring at the 2022 annual meeting of stockholders and until his or her respective successor is duly elected and qualified.
- (2) To vote, on an advisory basis, to approve the compensation paid to our named executive officers.
- (3) To ratify the appointment of PricewaterhouseCoopers LLP as our independent registered public accounting firm for the fiscal year ending December 31, 2019.

Stockholders will also act on any other business that may properly come before the meeting, or any adjournment or postponement thereof.

Who can vote?

Only stockholders of record at the close of business on April 8, 2019, the record date for the Annual Meeting, are entitled to vote at the Annual Meeting. On this record date, there were 58,680,674 shares of our common stock outstanding. Common stock is our only class of stock outstanding.

How many votes do I have?

Each share of our common stock that you own as of the record date, April 8, 2019, entitles you to one vote on each matter that is voted on.

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Is my vote important?

Your vote is important no matter how many shares you own. Please take the time to vote. Take a moment to read the instructions, choose the way to vote that is the easiest and most convenient for you and cast your vote as soon as possible.

How do I vote?

If you are the record holder of your shares, meaning that you own your shares in your own name and not through a bank, brokerage firm or other nominee, you may vote:

- (1) **Over the Internet:** To vote over the internet, please go to the following website: www.proxypush.com/agio, and follow the instructions at that site for submitting your proxy electronically. If you vote over the internet, you do not need to complete and mail your proxy card or vote your proxy by telephone. Your vote must be received by 8:59 a.m., Eastern Time, on May 29, 2019 to be counted.
- (2) **By Telephone:** To vote by telephone, please call (866) 509-2148, and follow the instructions provided on the proxy card. If you vote by telephone, you do not need to complete and mail your proxy card or vote your proxy over the internet. Your vote must be received by 8:59 a.m., Eastern Time, on May 29, 2019 to be counted.
- (3) **By Mail:** To vote using the printed proxy card that may be delivered to you upon request, simply complete, sign and date the proxy card that may be delivered and return it promptly in the postage prepaid envelope provided to Proxy Tabulator for Agios Pharmaceuticals, Inc., c/o Mediant Communications, P.O. Box 8016, Cary, NC 27512-9903. If you vote by mail, you do not need to vote over the internet or by telephone. If we receive the proxy card no later than May 28, 2019, we will vote your shares as you direct.
- (4) **In Person at the Meeting:** If you attend the Annual Meeting, you may deliver your completed proxy card in person or you may vote by completing a ballot, which we will provide to you at the meeting.

If your shares are held in street name, meaning they are held for your account by a bank, brokerage firm, or other nominee, you may vote:

- (1) **Over the Internet or by Telephone:** You will receive instructions from your bank, brokerage firm, or other nominee if they permit internet or telephone voting. You should follow those instructions.
- (2) **By Mail:** You will receive instructions from your bank, brokerage firm, or other nominee explaining how you can vote your shares by mail. You should follow those instructions.

- (3) **In Person at the Meeting:** You must bring an account statement or letter from your bank, brokerage firm or other nominee showing that you are the beneficial owner of the shares as of the record date in order to vote your shares at the meeting. To be able to vote your shares held in street name at the meeting, you will need to obtain a proxy card from the holder of record.

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Can I change my vote?

If your shares are registered directly in your name, you may revoke your proxy and change your vote by following one of the below procedures:

- (1) Vote over the internet or by telephone as instructed above. Only your latest internet or telephone vote is counted. You may not change your vote over the internet or by telephone after 8:59 a.m., Eastern Time, on May 29, 2019.
- (2) Sign and complete a new proxy card and send it by mail to Proxy Tabulator for Agios Pharmaceuticals, Inc., c/o Mediant Communications, P.O. Box 8016, Cary, NC 27512-9903. Mediant must receive the proxy card no later than May 28, 2019. Only your latest dated proxy will be counted.
- (3) Attend the Annual Meeting and vote in person as instructed above. Attending the Annual Meeting alone, without voting in person, will not revoke your internet vote, telephone vote or proxy submitted by mail, as the case may be.
- (4) Give our corporate secretary written notice before or at the Annual Meeting that you want to revoke your proxy. Such written notice should be sent to Agios Pharmaceuticals, Inc., Attention: James Burns, Corporate Secretary, 88 Sidney Street, Cambridge, MA 02139.

If your shares are held in street name, you may submit new voting instructions with a later date by contacting your bank, brokerage firm, or other nominee. You may also vote in person at the Annual Meeting, which will have the effect of revoking any previously submitted voting instructions, if you obtain a broker's proxy as described in the answer to the question "How do I vote?" above.

Will my shares be voted if I do not return my proxy?

If your shares are registered directly in your name, your shares will not be voted if you do not vote over the internet, by telephone, by returning your proxy by mail or by ballot at the Annual Meeting.

If your shares are held in street name, your brokerage firm may under certain circumstances vote your shares if you do not return your voting instructions. Brokerage firms can vote customers' unvoted shares on routine matters but they will not be allowed to vote your shares with respect to certain non-routine items. If you do not return voting instructions to your brokerage firm to vote your shares, your brokerage firm may, on routine matters, either vote your shares or leave your shares unvoted.

Your brokerage firm cannot vote your shares on any matter that is not considered routine. Proposal 1, the election of three Class III directors, and Proposal 2, an advisory vote on the compensation paid to our named executive officers, are not considered routine matters. If you do not instruct your brokerage firm how to vote with respect to these items, your brokerage firm may not vote with respect to these proposals and those votes will be counted as broker non-votes. Broker non-votes are shares that are held in street name by a bank or brokerage firm that indicates on its proxy that it does not have or did not exercise discretionary authority to vote on a particular matter. Proposal 3, the ratification of the appointment of PricewaterhouseCoopers LLP as our independent registered public

accounting firm for the fiscal year ending December 31, 2019, is considered a routine matter, and your brokerage firm will be able to vote on that item even if it does not receive instructions from you, so long as it holds your shares in its name. We encourage you to provide voting instructions to your brokerage firm or other nominee. This ensures that your shares will be voted at the Annual Meeting according to your instructions. You should receive directions from your brokerage firm or other nominee about how to submit your voting instructions.

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How many shares must be represented to hold the Annual Meeting?

A majority of our shares of common stock outstanding at the record date must be present in person or represented by proxy to hold the Annual Meeting. This is called a quorum. For purposes of determining whether a quorum exists, we count as present any shares that are voted over the internet, by telephone, by completing and submitting a proxy by mail, or that are represented in person at the meeting. Further, for purposes of establishing a quorum, we will count as present shares that a stockholder holds even if the stockholder votes to abstain or only votes on one of the proposals. In addition, we will count as present shares held in street name by banks, brokerage firms or nominees who indicate on their proxies that they do not have authority to vote those shares on Proposals 1 or 2. If a quorum is not present, we expect to adjourn the Annual Meeting until we obtain a quorum.

The presence at the Annual Meeting, in person or by proxy, of holders representing a majority of our outstanding common stock as of the record date, April 8, 2019, or approximately 29,340,338 shares, constitutes a quorum at the meeting and permits us to conduct the business of the meeting.

What vote is required to approve each matter and how are votes counted?

Proposal 1 Election of Directors

The three nominees for director to receive the highest number of votes FOR election will be elected as directors. This is called a plurality. **Proposal 1 is not considered a routine matter.** Therefore, if your shares are held by your brokerage firm in street name and you do not provide voting instructions with respect to your shares, your brokerage firm cannot vote your shares on Proposal 1. Shares held in street name by banks, brokerage firms, or nominees who indicate on their proxies that they do not have authority to vote the shares on Proposal 1 will not be counted as votes FOR or WITHHELD from any nominee. As a result, such broker non-votes will have no effect on the voting on Proposal 1. You may:

vote FOR all nominees;

vote FOR a particular nominee or nominees and WITHHOLD your vote from the other nominees; or

WITHHOLD your vote from all nominees.

Votes that are withheld will not be included in the vote tally for the election of directors and will not affect the results of the vote.

Proposal 2 Advisory Vote on the Compensation Paid to Named Executive Officers

To approve Proposal 2, holders of a majority of the votes cast on the matter must vote FOR the proposal. **Proposal 2 is not considered a routine matter.** Shares which abstain and broker non-votes will not be counted as votes in favor of, or with respect to, these proposals and will also not be counted as votes cast. Accordingly, abstentions and broker non-votes will have no effect on the outcome of these proposals. Proposal 2 is non-binding. Because this vote is advisory and not binding on us or our board of directors in any way, our board may decide that it is in our and our stockholders' best interests to compensate our named executive officers in an amount or manner that differs from that which is approved by our stockholders.

Proposal 3 Ratification of Appointment of Independent Registered Public Accounting Firm

To approve Proposal 3, holders of a majority of the votes cast on the matter must vote FOR the proposal. **Proposal 3 is considered a routine matter.** If your shares are held by your brokerage firm in street name and

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you do not provide voting instructions with respect to your shares, your brokerage firm may vote your unvoted shares on Proposal 3. If you ABSTAIN from voting on Proposal 3, your shares will not be voted FOR or AGAINST the proposal and will also not be counted as votes cast or shares voting on the proposal. As a result, voting to ABSTAIN will have no effect on the outcome of Proposal 3.

Although stockholder approval of our audit committee's appointment of PricewaterhouseCoopers LLP as our independent registered public accounting firm for the fiscal year ending December 31, 2019 is not required, we believe that it is advisable to give stockholders an opportunity to ratify this appointment. If this proposal is not approved at the Annual Meeting, our audit committee will reconsider its appointment of PricewaterhouseCoopers LLP as our independent registered public accounting firm for the year ended December 31, 2019.

How does the board of directors recommend that I vote on the proposals?

Our board of directors recommends that you vote:

FOR the election of each of the three nominees to serve on our board of directors, each for a three-year term;

FOR the approval, on an advisory (non-binding) basis, of the compensation of our named executive officers; and

FOR the ratification of the appointment of PricewaterhouseCoopers LLP as our independent registered public accounting firm for the fiscal year ending December 31, 2019.

Are there other matters to be voted on at the Annual Meeting?

We do not know of any matters that may come before the Annual Meeting other than the election of our board of directors, the approval of the compensation of our named executive officers and the ratification of our independent registered public accounting firm. If any other matters are properly presented at the Annual Meeting, the persons named in the accompanying proxy intend to vote, or otherwise act, in accordance with their judgment on the matter.

Who will count the votes?

The votes will be counted, tabulated and certified by Mediant Communications Inc.

Will my vote be kept confidential?

Your vote will be kept confidential and we will not disclose your vote, unless (1) we are required to do so by law (including in connection with the pursuit or defense of a legal or administrative action or proceeding), or (2) there is a contested election for the board of directors. The inspector of election will forward any written comments that you make on the proxy card to management without providing your name, unless you expressly request disclosure on the proxy card.

How can I find out the results of the voting at the Annual Meeting?

Preliminary voting results will be announced at the Annual Meeting. Final voting results will be tallied by the inspector of election and published in a current report on Form 8-K to be filed with the SEC within four business days after the Annual Meeting.

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How and when may I submit a stockholder proposal, including a stockholder nomination for director for the 2020 annual meeting?

Stockholders wishing to suggest a candidate for director should write to our corporate secretary. In order to give the nominating and corporate governance committee sufficient time to evaluate a recommended candidate and/or include the candidate in our proxy statement for the 2020 annual meeting, the recommendation should be received by our corporate secretary at our principal executive offices in accordance with our procedures detailed in the section below entitled Stockholder Proposals. Such submissions must state the nominee's name, together with appropriate biographical information and background materials, and information with respect to the stockholder or group of stockholders making the recommendation, including the number of shares of common stock owned by such stockholder or group of stockholders, as well as other information required by our bylaws or SEC regulations. We may require any proposed nominee to furnish such other information as we may reasonably require in determining the eligibility of such proposed nominee to serve as an independent director or that could be material to a reasonable stockholder's understanding of the independence, or lack thereof, of such proposed nominee.

Who is paying the costs of soliciting these proxies?

We will pay all of the costs of soliciting proxies. Our directors, officers and other employees may solicit proxies in person or by mail, telephone, fax or email. We will pay our directors, officers and other employees no additional compensation for these services. We will ask banks, brokerage firms and other nominees to forward these proxy materials to their principals and to obtain authority to execute proxies. We may reimburse them for their expenses.

How do I obtain an Annual Report on Form 10-K?

If you would like a copy of our Annual Report on Form 10-K for the year ended December 31, 2018 that we filed with the SEC, we will send you one, without exhibits, free of charge. Please contact Kendra Adams, Vice President, External Communications and Investor Relations. She may be contacted at 88 Sidney Street, Cambridge, Massachusetts 02139; telephone: 617-649-8600; e-mail: kendra.adams@agios.com.

All of our SEC filings are also available free of charge in the Investors Financials SEC Filings section of our website at www.agios.com.

Whom should I contact if I have any questions?

If you have any questions about the Annual Meeting or your ownership of our common stock, please contact Kendra Adams in our Investor and Public Relations department. She may be contacted at 88 Sidney Street, Cambridge, Massachusetts 02139; telephone: 617-649-8600; e-mail: kendra.adams@agios.com.

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STOCKHOLDERS SHARING THE SAME ADDRESS

The SEC has adopted rules that permit companies and intermediaries (e.g., brokers) to satisfy the delivery requirements for Notices and, if applicable, our annual report and other proxy materials, with respect to two or more stockholders sharing the same address by delivering a single Notice and, if applicable, a single set of our annual report and proxy materials, addressed to those stockholders. This practice, which is commonly referred to as householding, potentially means extra convenience for stockholders and cost savings for companies.

This year, a number of brokers with account holders who are our stockholders will be householding our proxy materials. A single Notice and, if applicable, a single copy of our annual report and our proxy materials, will be delivered to multiple stockholders sharing an address unless contrary instructions have been received from the affected stockholders. Once you have received notice from your broker that they will be householding communications to your address, householding will continue until you are notified otherwise or until you revoke your consent.

If, at any time, you no longer wish to participate in householding and would prefer to receive a separate Notice and, if applicable, a separate set of proxy materials in the future, please notify your broker or contact us. If you wish to receive a separate set of proxy materials for this year's Annual Meeting, we will deliver them promptly upon written or oral request. Stockholders who currently receive multiple copies of the Notice, and, if applicable, our annual report and other proxy materials at their addresses and would like to request householding of their communications should contact their brokers or us. To contact us, direct your written request to: Agios Pharmaceuticals, Inc., 88 Sidney Street, Cambridge, MA 02139, Attention: Corporate Secretary, 617-649-8600 or contact Investor and Public Relations at 617-649-8600.

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The following table sets forth information relating to the beneficial ownership of our common stock as of March 31, 2019, by:

each person known by us to beneficially own more than 5% of our outstanding shares of common stock;

each of our directors and nominees for director;

our principal executive officer, our principal financial officer and our other executive officers named in the Summary Compensation Table below, whom we collectively refer to as our named executive officers; and

all directors and executive officers as a group.

The percentage of shares beneficially owned is computed on the basis of 58,659,821 shares of our common stock outstanding as of March 31, 2019. The number of shares beneficially owned by each stockholder is determined under rules of the SEC. Under these rules, beneficial ownership includes any shares as to which the individual or entity has sole or shared voting power or investment power. In computing the number of shares beneficially owned by an individual or entity and the percentage ownership of that person, shares of common stock subject to options or other rights held by such person that are currently exercisable or will become exercisable within 60 days of March 31, 2019 are considered outstanding, although these shares are not considered outstanding for purposes of computing the percentage ownership of any other person. Unless otherwise indicated, the address of all listed stockholders is c/o Agios Pharmaceuticals, Inc., 88 Sidney Street, Cambridge, MA 02139. Each of the stockholders listed has sole voting and investment power with respect to the shares beneficially owned by the stockholder unless noted otherwise, subject to community property laws where applicable.

| Name of Beneficial Owner | Shares of Common Stock Owned | + | Common Stock Underlying Options and Other Rights Acquirable Within 60 Days | = | Total Beneficial Ownership | |
|--|---------------------------------------|---|--|---|----------------------------|------------|
| | | | | | Number | Percentage |
| 5% Stockholders | | | | | | |
| Entities affiliated with Fidelity Management & Research Company(1) | 8,726,801 | | | | 8,726,801 | 14.88% |
| Wellington Management Group LLP(2) | 8,143,116 | | | | 8,143,116 | 13.88% |

| | | | | |
|---|-----------|--|-----------|--------|
| Entities affiliated with Celgene Corporation(3) | 6,718,433 | | 6,718,433 | 11.45% |
| The Vanguard Group(4) | 4,621,949 | | 4,621,949 | 7.88% |
| Vanguard Specialized Funds Vanguard Health Care Fund(5) | 4,402,124 | | 4,402,124 | 7.50% |
| Capital International Investors(6) | 4,037,544 | | 4,037,544 | 6.88% |
| BlackRock, Inc.(7) | 3,324,479 | | 3,324,479 | 5.67% |
| BB Biotech AG(8) | 2,978,134 | | 2,978,134 | 5.08% |

Named Executive Officers and Directors

| | | | | |
|--|---------|-----------|-----------|-------|
| Scott Biller, Ph.D.(9) | 79,570 | 157,818 | 237,388 | * |
| Christopher Bowden, M.D. | 7,238 | 171,818 | 179,056 | * |
| Jacquelyn A. Fouse, Ph.D. | | 5,665 | 5,665 | * |
| Andrew Hirsch | 12,374 | 120,409 | 132,783 | * |
| Steve Hoerter(10) | 10,941 | 23,892 | 34,833 | * |
| David P. Schenkein, M.D.(11) | 439,357 | 603,557 | 1,042,914 | 1.76% |
| Paul J. Clancy | | 67,400 | 67,400 | * |
| Ian T. Clark | | 17,661 | 17,661 | * |
| Kaye Foster | 2,200 | 36,899 | 39,099 | * |
| Maykin Ho, Ph.D. | | 36,807 | 36,807 | * |
| John M. Maraganore, Ph.D. | 36,209 | 49,125 | 85,334 | * |
| David Scadden, M.D. | 571 | 15,996 | 16,567 | * |
| All executive officers and directors as a group (12 persons) | 588,460 | 1,307,047 | 1,895,507 | 3.16% |

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* Less than 1%.

- (1) Based solely on a Schedule 13G/A filed with the SEC on February 13, 2019. FMR LLC and Abigail P. Johnson are each the beneficial owners of 8,726,801 shares of common stock. FMR LLC has sole voting power over 1,777,875 shares of common stock and sole dispositive power over 8,726,801 shares of common stock. The address of FMR LLC is 245 Summer Street, Boston, MA 02210.
- (2) Based solely on a Schedule 13G/A filed with the SEC on February 12, 2019. Wellington Management Group LLP (Wellington), Wellington Group Holdings LLP and Wellington Investment Advisors Holdings LLP are each deemed to be the beneficial owner of 8,143,116 shares of common stock, with respect to which each entity reported shared voting power over 3,372,894 shares and shared dispositive power over 8,143,116 shares. Includes 4,402,124 shares of common stock owned by Vanguard Health Care Fund that may be duplicative of shares described in footnote 5 of this table. Wellington Management Company LLP is deemed to be the beneficial owner of 8,039,836 shares of common stock, with respect to which it reported shared voting power over 3,361,282 shares and shared dispositive power over 8,039,836 shares. The shares are owned of record by clients of the following investment advisers (the Wellington Investment Advisers): Wellington Management Company LLP, Wellington Management Canada LLC, Wellington Management Singapore Pte Ltd, Wellington Management Hong Kong Ltd, Wellington Management International Ltd, Wellington Management Japan Pte Ltd and Wellington Management Australia Pty Ltd. Wellington Investment Advisors Holdings LLP controls directly, or indirectly through Wellington Management Global Holdings, Ltd., the Wellington Investment Advisers. Wellington Investment Advisors Holdings LLP is owned by Wellington Group Holdings LLP. Wellington Group Holdings LLP is owned by Wellington. The address of Wellington is 280 Congress Street, Boston, MA 02210.
- (3) Based solely on a Schedule 13D/A filed with the SEC on January 23, 2018. Consists of 4,010,926 shares of common stock held by Celgene European Investment Company LLC (Celgene LLC), 708,333 shares of common stock held by Celgene Alpine Investment Co., LLC (Celgene Alpine LLC), 624,575 shares of common stock held by Celgene Switzerland LLC and 1,374,599 shares of common stock held by Celgene Corporation (Celgene). Celgene LLC, Celgene Alpine LLC and Celgene Switzerland LLC are wholly-owned subsidiaries of Celgene. Celgene LLC has shared voting and dispositive power over 4,010,926 shares of common stock, Celgene Alpine LLC has shared voting and dispositive power over 708,333 shares of common stock, Celgene Switzerland LLC has shares voting and dispositive power over 624,575 shares of common stock and Celgene has sole voting and dispositive power over 1,374,599 shares of common stock and shared voting and dispositive power over 5,343,834 shares of common stock. The address of Celgene Corporation is 86 Morris Avenue, Summit, NJ 07901.
- (4) Based solely on a Schedule 13G/A filed with the SEC on February 11, 2019. The Vanguard Group (Vanguard) is deemed to be the beneficial owner of 4,621,949 shares of common stock, with respect to which it reported sole voting power over 28,470 shares, shared voting power over 12,853 shares, sole dispositive power over 4,586,328 shares and shared dispositive power over 35,621 shares. Includes 22,768 shares beneficially owned by Vanguard Fiduciary Trust Company, a wholly owned subsidiary of The Vanguard Group, Inc. as a result of Vanguard Fiduciary Trust Company serving as investment manager of collective trust accounts. Vanguard Investments Australia, Ltd., a wholly owned subsidiary of The Vanguard Group, Inc., is the beneficial owner of 18,555 shares as a result of Vanguard Investments Australia, Ltd. serving as investment manager of Australian

investment offerings. The address of Vanguard is 100 Vanguard Blvd., Malvern, PA 19355.

- (5) Based solely on a Schedule 13G/A filed with the SEC on January 31, 2019. Vanguard Specialized Funds Vanguard Health Care Fund (Vanguard Health Care Fund) is deemed to be the beneficial owner of 4,402,124 shares of common stock, with respect to which it reported sole voting power over 4,402,124 shares. The address of Vanguard Health Care Fund is 100 Vanguard Blvd., Malvern, PA 19355.

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- (6) Based solely on a Schedule 13G/A filed with the SEC on February 14, 2019. Capital International Investors has sole voting power over 3,617,401 shares of common stock and sole dispositive power over 4,037,544 shares of common stock. The address of Capital International Investors is 11100 Santa Monica Boulevard, 16th Floor, Los Angeles, CA 90025.

- (7) Based solely on a Schedule 13G/A filed with the SEC on February 4, 2019 by BlackRock, Inc. (BlackRock) and certain of its subsidiaries. BlackRock is deemed to be the beneficial owner of 3,324,479 shares of common stock, with respect to which it reported sole voting power over 3,116,544 shares and sole dispositive power over 3,324,479 shares. The address of BlackRock 55 East 52nd Street, New York, NY 10055.

- (8) Based solely on a Schedule 13G/A filed with the SEC on January 28, 2019. BB Biotech AG (BB Biotech) and its wholly-owned subsidiary Biotech Target N.V. (Biotech Target) share voting and dispositive power over 2,978,134 shares of common stock. The address of BB Biotech is Schwertstrasse 6, CH-8200 Schaffhausen, Switzerland and the address of Biotech Target is Snipweg 26, Curacao.

- (9) Includes shares of common stock held by a trust of which the reporting person is trustee and beneficiary.

- (10) Effective March 15, 2019, Mr. Hoerter resigned as our chief commercial officer.

- (11) Includes shares of common stock held by the David P. Schenkein 2004 Revocable Trust and shares of common stock held by the Amy P. Schenkein 2004 Revocable Trust.

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SECTION 16(a) BENEFICIAL OWNERSHIP REPORTING COMPLIANCE

Under Section 16(a) of the Exchange Act, directors, executive officers, our principal accounting officer and beneficial owners of 10% or more of our common stock, or reporting persons, are required to report to the SEC on a timely basis the initiation of their status as a reporting person and any changes with respect to their beneficial ownership of our common stock. Based solely on our review of copies of such forms that we have received, or written representations from reporting persons, we believe that during the fiscal year ended December 31, 2018, all reporting persons complied with all applicable filing requirements, except with respect to three Form 4 filings made on May 29, 2018 to report shares of common stock sold by each of Dr. Biller, Mr. Hoerter and Dr. Schenkein on May 23, 2018 and a Form 4 filing made on February 20, 2019 to report shares of common stock sold by Carman Alenson, our principal accounting officer, to report shares of common stock sold on September 28, 2018.

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PROPOSAL 1:

ELECTION OF DIRECTORS

Our board of directors is divided into three classes, with one class of our directors standing for election each year. Directors in each class are elected at the annual meeting of stockholders held in the year in which the term for their class expires and hold office for a three-year term and until their resignation or removal or their successors are duly elected and qualified. In accordance with our certificate of incorporation and bylaws, our directors may fill existing vacancies on the board of directors.

The term of office of our Class III directors, Jacquelyn A. Fouse, Ph.D., David Scadden, M.D., and David P. Schenkein, M.D., will expire at the Annual Meeting. The nominees for Class III directors for election at the Annual Meeting are Dr. Fouse, Dr. Scadden and Dr. Schenkein. If any of Dr. Fouse, Dr. Scadden and Dr. Schenkein is elected at the Annual Meeting, such individual will be elected to serve for a three-year term that will expire at our 2022 annual meeting of stockholders and until such individual's successor is elected and qualified.

If no contrary indication is made, proxies in the accompanying form will be voted for Dr. Fouse, Dr. Scadden and Dr. Schenkein or, in the event that any of Dr. Fouse, Dr. Scadden and Dr. Schenkein is not a candidate or is unable to serve as a director at the time of the election (which is not currently expected), for any nominee who is designated by our board of directors to fill the vacancy.

Our priority in selection of board members is identification of members who will further the interests of our stockholders through their established record of professional accomplishment, the ability to contribute positively to the collaborative culture among board members, knowledge of our business, understanding of the competitive landscape and adherence to high ethical standards. Certain individual qualifications and skills of our directors that contribute to the board of directors' effectiveness as a whole are described in the following paragraphs.

Information Regarding Directors

The following paragraphs provide information as of the date of this Proxy Statement about each director and nominee for director, as furnished to us by the directors and nominees for director. The information presented includes information each such individual has given us about his or her age, all positions he or she holds, his or her principal occupation and business experience for the past five years, and the names of other publicly-held companies of which he or she currently serves as a director or has served as a director during the past five years. In addition to the information presented below regarding each such individual's specific experience, qualifications, attributes and skills that led our board of directors to the conclusion that he or she should serve as a director, we also believe that each of our directors and director nominees has a reputation for integrity, honesty and adherence to high ethical standards. Each has demonstrated business acumen and an ability to exercise sound judgment, as well as a commitment of service to our company and our board of directors. Finally, we value their significant experience on other public company boards of directors and board committees.

Information about the number of shares of common stock beneficially owned by each of our directors and nominees for director appears above under the heading "Security Ownership of Certain Beneficial Owners and Management."

There are no family relationships between or among any of our executive officers, directors or nominees for director.

Table of Contents**Nominees for Election to the Board of Directors****Term Expiring at the 2022 Annual Meeting of Stockholders, if elected at the Annual Meeting (Class III)**

| Name | Age | Present Position with Agios Pharmaceuticals, Inc. |
|---------------------------|------------|--|
| Jacquelyn A. Fouse, Ph.D. | 57 | Chief Executive Officer and Director |
| David Scadden, M.D. | 66 | Director |
| David P. Schenkein, M.D. | 61 | Executive Chairman |

Jacquelyn A. Fouse, Ph.D. joined Agios as chief executive officer on February 1, 2019 and has served as a member of our board of directors since December 2017. Dr. Fouse served as executive chair of Dermavant Sciences, a biopharmaceutical company, from July 2017 until September 2018. From September 2010 until June 2017, Dr. Fouse served in various capacities at Celgene Corporation, a biopharmaceutical company, serving as strategic advisor to the management executive committee from April 2017 to June 2017, president and chief operating officer from March 2016 to March 2017, president, hematology and oncology from August 2014 to February 2016, executive vice president and chief financial officer from February 2012 to July 2014, and senior vice president and chief financial officer from September 2010 to February 2012. Prior to joining Celgene, Dr. Fouse served as chief financial officer of Bunge Limited, or Bunge, a global agribusiness and food company, from 2007 to 2010. Prior to joining Bunge, Dr. Fouse served as senior vice president, chief financial officer and corporate strategy at Alcon Laboratories, Inc., or Alcon, a global medical company, since 2006, and as its senior vice president and chief financial officer since 2002. Prior to her time with Alcon she held a variety of senior leadership roles with international companies. Dr. Fouse is also a director of Incyte Corporation, a public biopharmaceutical company, and Dick's Sporting Goods, Inc., a public sporting goods retailer, and was a director of Perrigo Company from November 2012 to April 2016 and a director of Celgene Corporation from February 2016 until June 2017. Dr. Fouse earned a B.A. and an M.A. in Economics and a Ph.D. in Finance from the University of Texas at Arlington. We believe Dr. Fouse is qualified to serve as a member of our board of directors due to her extensive experience in the biotechnology sector and her senior leadership experience.

David Scadden, M.D. has served as a member of our board of directors since May 2017. Dr. Scadden is a hematologist/oncologist and an expert on the medical application of stem cell biology with a particular emphasis on its use in the setting of cancer. He is the Gerald and Darlene Jordan Professor of Medicine at Harvard University, a position he has held since 2006. Since 1995, Dr. Scadden has practiced at the Massachusetts General Hospital, where he founded and directs the Center for Regenerative Medicine and directed the Hematologic Malignancies Center of the MGH Cancer Center for 10 years. Dr. Scadden co-founded and co-directs the Harvard Stem Cell Institute, and is Chairman emeritus and Professor of the Harvard University Department of Stem Cell and Regenerative Biology. He is a member of the National Academy of Medicine, the American Academy of Arts and Sciences, is an associate member of the Broad Institute of Harvard and MIT and is a former member of the Board of External Experts for the National Heart, Lung and Blood Institute, the Board of Scientific Counselors for the National Cancer Institute, Board of Directors of the International Society for Stem Cell Research and the Executive Committee of the American Society of Hematology. Dr. Scadden is on the board of the public biotechnology company, Editas Medicines, the boards of the private biotechnology companies LifeVault Bio, Clear Creek Bio, Inc. and Red Oak Medicines, is a scientific founder of Fate Therapeutics, a public biotechnology company, and a scientific founder and member of the board of Magenta Therapeutics, a public biotechnology company. He is the recipient of numerous honors including the E. Donnell Thomas and the Dameshek awards from the American Society of Hematology, awards from the Doris Duke Charitable Trust, the Ellison Medical Foundation, the Burroughs Welcome Fund, and the Leukemia and Lymphoma Society. Dr. Scadden holds degrees from Bucknell University, Case Western Reserve University and honorary degrees from Harvard University, Bucknell University and Lund University in Sweden. We believe

Dr. Scadden is qualified to serve on our board of directors due to his scientific expertise in the fields of hematology and oncology.

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David P. Schenkein, M.D. has served as a member of our board of directors since August 2009 and as our executive chairman since February 2019. Dr. Schenkein also served as our president and chief executive officer from August 2009 until February 2019. Dr. Schenkein has been a hematologist and medical oncologist for more than 20 years. He currently serves as a general partner of GV, the venture capital arm of Alphabet Inc. (formerly Google), and is an adjunct attending physician in hematology at Tufts Medical Center. Prior to joining Agios, from 2006 to 2009, Dr. Schenkein was the senior vice president, clinical hematology/oncology at Genentech, Inc., a pharmaceutical company (now a member of the Roche Group, a global healthcare company), or Genentech, where he was responsible for numerous successful oncology drug approvals and leading the medical and scientific strategies for its bio-oncology portfolio. While at Genentech, he served as an adjunct clinical professor of medical oncology at Stanford University School of Medicine. Prior to joining Genentech, he served as the senior vice president of clinical research at Millennium Pharmaceuticals, Inc., a biopharmaceutical company (now Takeda Oncology), or Millennium, overseeing the clinical development and worldwide approval of VELCADE®, a first-in-class cancer therapy now approved to treat multiple myeloma and non-Hodgkins lymphoma. Dr. Schenkein currently serves on the board of directors of bluebird bio, Inc. and Denali Therapeutics Inc., both public biotechnology companies, and was formerly on the board of Foundation Medicine, Inc., a public biotechnology company. Dr. Schenkein holds a B.A. in chemistry from Wesleyan University and an M.D. from the State University of New York Upstate Medical School. We believe that Dr. Schenkein's detailed knowledge of our company and his extensive background in the biotechnology industry, including his roles at Genentech and Millennium, provide a critical contribution to our board of directors.

Recommendation of the Board of Directors

OUR BOARD OF DIRECTORS UNANIMOUSLY RECOMMENDS THAT THE STOCKHOLDERS VOTE FOR THE ELECTION OF EACH OF DR. FOUSE, DR. SCADDEN AND DR. SCHENKEIN.

Members of the Board of Directors Continuing in Office**Term Expiring at the****2020 Annual Meeting of Stockholders (Class I)**

| Name | Age | Present Position with Agios Pharmaceuticals, Inc. |
|-------------|------------|--|
| Paul Clancy | 57 | Director |
| Ian Clark | 58 | Director |

Paul J. Clancy has served as a member of our board of directors since September 2013. Mr. Clancy has more than 20 years of experience in financial management and strategic business planning, and has served as chief financial officer of Alexion Pharmaceuticals, Inc., a public biopharmaceutical company, since July 2017. Prior to Alexion, Mr. Clancy served as the executive vice president and chief financial officer at Biogen Inc. (formerly known as Biogen Idec Pharmaceutical Corporation), or Biogen, since 2007. He also served as senior vice president of finance of Biogen Idec, with responsibilities for leading the treasury, tax, investor relations and business planning groups. Prior to the merger of Biogen and Idec Pharmaceutical Corporation, Mr. Clancy was the vice president of portfolio management at Biogen. He joined Biogen in 2001 as vice president of U.S. marketing. Before Biogen, Mr. Clancy spent 13 years at PepsiCo Inc., a publicly-traded food and beverage company, serving in a variety of financial and general management positions, including vice president and general manager of their Great West Business Unit. Mr. Clancy is a member of the board of directors and audit committee of Incyte Corporation, a biopharmaceutical company. Mr. Clancy has an M.B.A. from Columbia Business School and received his B.S. in business administration from Babson College in Wellesley, MA. We believe Mr. Clancy is qualified to serve as a member of our board of directors due to his

extensive financial and executive leadership experience at large multi-national companies.

Ian T. Clark has served as a member of our board of directors since December 2016. Mr. Clark has more than 30 years of experience in the biotechnology industry. Mr. Clark currently serves as an operating partner at

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Blackstone Life Sciences, a private investment firm. Prior to that, he was chief executive officer and head of North American Commercial Operations of Genentech (now a member of the Roche Group), a position he held from 2010 to 2016. Mr. Clark also led the Genentech executive committee and was a member of the Genentech board of directors. Mr. Clark joined Genentech in 2003 as senior vice president and general manager, bio-oncology. In 2005, he was named senior vice president, commercial operations and became a member of the executive committee, and in 2006, he was named executive vice president, commercial operations. In 2009, he was named head of global product strategy and chief marketing officer of Roche. Prior to joining Genentech, Mr. Clark held roles of increasing responsibility at Novartis, Sanofi-Synthelabo SA (Aventis SA), Ivax Pharmaceuticals, Inc. and G.D. Searle, LLC, a subsidiary of Monsanto Corporation in the United States, Canada, the United Kingdom, France and Eastern Europe. Mr. Clark is a member of the boards of directors of publicly-traded biopharmaceutical companies Takeda Pharmaceutical Company Limited, Guardant Health, Inc., Forty Seven, Inc. and Corvus Pharmaceuticals, Inc., and is on the board of AvroBio, Inc., a private biotechnology company. He was formerly on the boards of directors of Shire plc (prior to its acquisition by Takeda), Kite Pharma, Inc. (prior to its acquisition by Gilead), Dendreon Corporation, Vernalis plc, and TerraVia Holdings, Inc. Mr. Clark also serves on the BioFulcrum Board of the Gladstone Institute, an independent, nonprofit life science research organization. Mr. Clark was formerly on the board of the Biotechnology Innovation Organization (BIO) and on the Economic Advisory Council of the 12th District of the Federal Reserve Bank of San Francisco. Mr. Clark received a B.S. degree in biological sciences and an honorary doctorate from Southampton University in the United Kingdom. We believe Mr. Clark is qualified to serve as a member of our board of directors due to his extensive experience in and knowledge of the biotechnology sector and his leadership experience.

Term Expiring at the**2021 Annual Meeting of Stockholders (Class II)**

| Name | Age | Present Position with Agios Pharmaceuticals, Inc. |
|---------------------------|------------|--|
| Kaye Foster | 59 | Director |
| Maykin Ho, Ph.D. | 66 | Director |
| John M. Maraganore, Ph.D. | 56 | Director |

Kaye Foster has served as a member of our board of directors since December 2014. Ms. Foster has more than 25 years of experience in human resources in the pharmaceutical industry and has been a senior advisor at the Boston Consulting Group since 2014. Previously, she was senior vice president, global human resources at Onyx Pharmaceuticals, Inc., an Amgen, Inc. subsidiary and a biopharmaceutical company, or Onyx from 2010 to 2014. At Onyx, which was acquired by Amgen in 2013, she led all aspects of human resources for U.S. and global operations. Prior to joining Onyx, Ms. Foster was global vice president of human resources and an executive committee member at Johnson and Johnson Corporation, a publicly-traded healthcare company, from 2003 to 2010. Before Johnson and Johnson, Ms. Foster held several senior human resources executive positions with Pfizer Inc., a publicly-traded pharmaceuticals company, supporting its pharmaceuticals businesses in Japan, Asia, Africa, Middle East and Latin America and, notably, led the integration of both the Warner-Lambert and Pharmacia mergers for these regions. Prior to that, she gained 10 years of operational experience within The Yellow Pages. She currently serves on the board of directors of Grail, Inc., a private biotechnology company, and Stanford Health Care, a hospital and healthcare system, and also serves on the board of Spelman College and chairs the Glide Memorial Foundation Board of Trustees. She earned her undergraduate degree at Baruch College of the City University of New York and received her M.B.A. from Columbia University, Graduate School of Business. We believe Ms. Foster's qualifications to serve on our board of directors include her extensive experience as an executive in the pharmaceuticals industry.

Maykin Ho, Ph.D. has served as a member of our board of directors since June 2015. Dr. Ho has more than 30 years of experience in the healthcare and finance industries. She has been a venture partner at Qiming

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Venture Partners, a venture capital firm in China and Hong Kong, since July 2015. From July 1992 to February 2015, she held various positions at Goldman Sachs, a global investment bank, including: from 2010 to 2015, she served as advisory director of global healthcare investment banking; from 2002 to 2010, she served as partner and co-head of healthcare investment research; and from 1992 to 2010 she served as senior biotechnology research analyst. Prior to Goldman Sachs, Dr. Ho held various managerial positions in licensing, strategic planning, marketing and research at DuPont-Merck Pharmaceuticals, a global pharmaceutical company, and DuPont de Nemours & Company. She is a member of the board of directors of Fibrogen, Inc., a publicly-traded biotechnology company, and Parexel International, a privately-held, global pharmaceutical services company, and serves on the board of directors of two non-profit research institutes: the Aaron Diamond AIDS Research Center and the Institute for Protein Innovation. In addition, Dr. Ho is a member of the Biotech Advisory Panel of The Stock Exchange of Hong Kong. Previously, Dr. Ho served on the board of directors and audit committee of Parexel International when it was a publicly-traded company, and on the investment committee of the Society of Neuroscience. She was a postdoctoral fellow at the pathology department of Harvard Medical School and a graduate of the Advanced Management Program at The Fuqua School of Business, Duke University. Dr. Ho received a Ph.D. in microbiology and immunology and a B.S. from the State University of New York, Downstate Medical Center. We believe Dr. Ho is qualified to serve on our board of directors due to her extensive experience in healthcare investment research and banking.

John M. Maraganore, Ph.D. has served as a member of our board of directors since November 2011. Since 2002, Dr. Maraganore has served as the chief executive officer and as a director of Alnylam Pharmaceuticals, Inc., a publicly-traded biopharmaceutical company, or Alnylam. From 2002 to 2007, Dr. Maraganore served as president of Alnylam. From 2000 to 2002, Dr. Maraganore served as senior vice president, strategic product development with Millennium. Before Millennium, he served as director of molecular biology and director of market and business development at Biogen. Prior to Biogen, Dr. Maraganore was a scientist at ZymoGenetics, Inc., a biotechnology company, and The Upjohn Company, a pharmaceutical manufacturing company. Dr. Maraganore was formerly a director of bluebird bio, Inc., a publicly-traded biopharmaceutical company. In addition, he was formerly a venture partner at Third Rock Ventures, L.P., where he participated in a limited capacity focusing on guiding strategy for Third Rock and its portfolio companies, and was formerly chairman of the board of directors of Regulus Therapeutics, Inc., a publicly-traded company. He is also chair of the Biotechnology Innovation Organization. Dr. Maraganore holds an M.S. and a Ph.D. in Biochemistry and Molecular Biology from the University of Chicago and a B.A. in Biological Sciences also from the University of Chicago. We believe that Dr. Maraganore is qualified to serve on our board of directors because he has over 30 years of experience in the biotechnology industry, bringing to our board of directors critical scientific, research and development, and general management expertise.

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CORPORATE GOVERNANCE

General

We believe that good corporate governance is important to ensure that Agios Pharmaceuticals, Inc. is managed for the long-term benefit of our stockholders. This section describes key corporate governance practices that we have adopted. We have adopted a code of business conduct and ethics, which applies to all of our officers, directors and employees, corporate governance guidelines and charters for our audit committee, our compensation committee, our nominating and governance committee and our science and technology committee. We have posted copies of our code of business conduct and ethics and corporate governance guidelines, as well as each of our committee charters, on the Corporate Governance page of the Investors section of our website, *www.agios.com*, which you can access free of charge. Information contained on the website is not incorporated by reference in, or considered part of, this Proxy Statement. We intend to disclose on our website any amendments to, or waivers from, our code of business conduct and ethics that are required to be disclosed by law or Nasdaq listing standards. We will also provide copies of these documents, as well as our other corporate governance documents, free of charge, to any stockholder upon written request to Kendra Adams, Vice President, External Communications and Investor Relations, at 88 Sidney Street, Cambridge, MA 02139, telephone: 617-649-8600; e-mail: kendra.adams@agios.com.

Corporate Governance Guidelines

Our board of directors has adopted corporate governance guidelines to assist in the exercise of its duties and responsibilities and to serve the best interests of Agios and our stockholders. These guidelines, which provide a framework for the conduct of our board's business, provide that:

our board's principal responsibility is to oversee the management of Agios;

a majority of the members of our board shall be independent directors;

the independent directors meet regularly in executive session;

directors have full and free access to management and, as necessary and appropriate, independent advisors;

new directors participate in an orientation program and all directors are expected to participate in continuing director education on an ongoing basis; and

our board and its committees will conduct a self-evaluation periodically to determine whether they are functioning effectively.

Director Determination of Independence

Rule 5605 of the Nasdaq Listing Rules requires a majority of a listed company's board of directors to be comprised of independent directors within one year of listing. In addition, the Nasdaq Listing Rules require that, subject to specified exceptions, each member of a listed company's audit, compensation and nominating and corporate governance committees be independent, that audit committee members also satisfy independence criteria set forth in Rule 10A-3 under the Securities Exchange Act of 1934, as amended, or the Exchange Act and that compensation committee members also satisfy the independence criteria set forth in Rule 10C-1 under the Exchange Act.

Under Rule 5605(a)(2) of the Nasdaq Listing Rules, a director will only qualify as an independent director if, in the opinion of our board of directors, that person does not have a relationship that would interfere

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with the exercise of independent judgment in carrying out the responsibilities of a director. In order to be considered independent for purposes of Rule 10A-3 of the Exchange Act, a member of an audit committee of a listed company may not, other than in their capacity as a member of the audit committee, the board of directors, or any other board committee, accept, directly or indirectly, any consulting, advisory, or other compensatory fee from the listed company or any of its subsidiaries or otherwise be an affiliated person of the listed company or any of its subsidiaries.

In addition, in affirmatively determining the independence of any director who will serve on a company's compensation committee, Rule 10C-1 under the Exchange Act requires that a company's board of directors consider all factors specifically relevant to determining whether a director has a relationship to such company which is material to that director's ability to be independent from management in connection with the duties of a compensation committee member, including, but not limited to: (i) the source of compensation of the director, including any consulting advisory or other compensatory fee paid by such company to the director; and (ii) whether the director is affiliated with the company or any of its subsidiaries or affiliates.

Our board of directors undertook a review of the composition of our board of directors and its committees and the independence of each director. Based upon information requested from and provided by each director concerning his or her background, employment and affiliations, including family relationships, our board of directors has determined that each of our directors, with the exception of Dr. Fouse, our chief executive officer, and Dr. Schenkein, our executive chairman, is an independent director as defined under Rule 5605(a)(2) of the Nasdaq Listing Rules. In addition, our board of directors determined that Mr. Clancy, Dr. Ho and Dr. Maraganore, who comprise our audit committee, Ms. Foster, Dr. Maraganore and Mr. Clark, who comprise our compensation committee, and Mr. Clark, Ms. Foster and Dr. Ho, who are members of our nominating and corporate governance committee, satisfy the independence standards for such committees established by the SEC and the Nasdaq Listing Rules, as applicable. In making such determinations, our board of directors considered the relationships that each such non-employee director has with our company, including any relevant transactions described below in *Certain Relationships and Related Party Transactions* and the beneficial ownership of our capital stock by each non-employee director, as well as all other facts and circumstances our board of directors deemed relevant in determining independence.

Board Leadership Structure

Dr. Schenkein currently serves as executive chairman of our board of directors. Effective February 1, 2019, Dr. Schenkein transitioned from his role of president and chief executive officer to executive chairman, and Dr. Fouse assumed the position of chief executive officer. Our board of directors believes this separate governance structure benefits our company because it enables Dr. Fouse to focus her entire energy on running our company while providing for the continued leadership and other contributions from Dr. Schenkein.

Our board of directors, upon the recommendation of our nominating and corporate governance committee, has determined that at this point in time having an executive chairman of our board of directors is in the best interests of our company and our stockholders and is consistent with good corporate governance for the following reasons:

Our executive chairman is more familiar with our business and strategy than an independent, non-employee Chairman would be and is thus better positioned to focus our board of directors' agenda on the key issues facing our company.

Oversight of our company is the responsibility of our board of directors as a whole, and this responsibility can be properly discharged without an independent chairman.

Our lead independent director can provide similar benefits to those associated with an independent chairman.

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Our board of directors, upon the recommendation of our nominating and corporate governance committee, has appointed Dr. Maraganore as lead independent director. Dr. Maraganore is an independent director within the meaning of the Nasdaq Listing Rules (see [Director Determination of Independence](#) above). His duties as lead director include the following:

Chairing meetings of the independent directors in executive session;

Meeting with any director who is not adequately performing his or her duties as a member of our board of directors or any committee;

Facilitating communications between other members of our board of directors, our executive chairman and our chief executive officer;

Working with our executive chairman and our chief executive officer in the preparation of the agenda for each board meeting and in determining the need for special meetings of our board of directors;

Reviewing and, if appropriate, recommending action to be taken with respect to written communications from stockholders submitted to our board of directors (see [Communications with our Board of Directors](#) below); and

Consulting with our executive chairman and our chief executive officer on matters relating to corporate governance and board performance.

Our nominating and corporate governance committee evaluates our board leadership structure from time to time and may recommend further alterations of this structure in the future.

Director Nomination Process

Director Qualifications

In evaluating director nominees the nominating and corporate governance committee will consider among other things the following factors:

reputation for personal and professional integrity, honesty and adherence to high ethical standards;

demonstrated business acumen, experience and ability to exercise sound judgments in matters that relate to the current and long-term objectives of our company;

strong finance experience;

commitment to understand our company and its industry;

interest and ability to understand the sometimes conflicting interests of the various constituencies of our company, which include stockholders, employees, customers, governmental units, creditors and the general public, and to act in the interests of all stockholders;

diversity of expertise and experience in substantive matters pertaining to our business relative to other board members;

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diversity of background and perspective, including with respect to age, gender, race, place of residence and specialized experience; and

practical and mature business judgment, including the ability to make independent analytical inquiries. The nominating and corporate governance committee's goal is to assemble a board of directors that brings to the company a variety of perspectives and skills derived from high quality business and professional experience. Moreover, the nominating and corporate governance committee believes that the background and qualifications of the board of directors, considered as a group, should provide a significant mix of experience, knowledge and abilities that will allow the board of directors to fulfill its responsibilities. Nominees are not discriminated against on the basis of race, religion, national origin, sex, sexual orientation, disability or any other basis proscribed by law.

The nominating and corporate governance committee has not adopted a formal policy with respect to a fixed set of specific minimum qualifications, including diversity, for its candidates for membership on the board of directors. The committee may consider such other facts, including, without limitation, diversity, as it may deem are in the best interests of our company and its stockholders. Our directors' performance and qualification criteria are reviewed periodically by the nominating and corporate governance committee.

Identification and Evaluation of Nominees for Directors

The nominating and corporate governance committee identifies nominees for director by first evaluating the current members of our board of directors willing to continue in service. Current members with qualifications and skills that are consistent with the nominating and corporate governance committee's criteria for board of director service and who are willing to continue in service are considered for re-nomination, balancing the value of continuity of service by existing members of our board of directors with that of obtaining a new perspective or expertise.

If any member of our board of directors does not wish to continue in service or if our board of directors decides not to re-nominate a member for re-election, the nominating and corporate governance committee identifies a new nominee that meets the criteria above. The committee generally inquires of our board of directors and members of management for their recommendations. The committee may also review the composition and qualification of the boards of directors of our competitors, and may seek input from industry experts or analysts. The nominating and corporate governance committee reviews the qualifications, experience and background of candidates. Final candidates, if other than our current directors, would be interviewed by the members of the nominating and corporate governance committee and by certain of our other independent directors and executive management. In making its determinations, the nominating and corporate governance committee evaluates each individual in the context of our board of directors as a whole, with the objective of assembling a group that can best contribute to the success of our company and represent stockholder interests through the exercise of sound judgment. After review and deliberation of all feedback and data, the nominating and corporate governance committee makes its recommendation to our board of directors. To date, the nominating and corporate governance committee has not utilized third-party search firms to identify board of director candidates. The nominating and corporate governance committee may in the future choose to do so in those situations where particular qualifications are required or where existing contacts are not sufficient to identify an appropriate candidate.

Stockholders may recommend individuals to our nominating and corporate governance committee for consideration as potential director candidates by submitting their names, together with appropriate biographical information and background materials and a statement as to whether the stockholder or group of stockholders making the recommendation has beneficially owned more than 5% of our common stock for at least a year as of

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the date such recommendation is made, to Nominating and Corporate Governance Committee, c/o Corporate Secretary, Agios Pharmaceuticals, Inc. 88 Sidney Street, Cambridge, Massachusetts 02139. Assuming that appropriate biographical and background material has been provided on or before the dates set forth in the section below entitled Stockholder Proposals, the committee will evaluate stockholder-recommended candidates by following substantially the same process, and applying substantially the same criteria, as it follows for candidates submitted by others, as described above.

Communications with Our Board of Directors

Our board of directors will give appropriate attention to written communications that are submitted by stockholders, and will respond if and as appropriate. The chairman of the board of directors (if an independent director), or the lead director (if one is appointed), or otherwise the chair of our Nominating and Governance Committee, with the assistance of our Corporate Secretary or his or her designee, is primarily responsible for monitoring communications from stockholders and for providing copies or summaries to the other directors as he or she considers appropriate.

Communications are forwarded to all directors if they relate to important substantive matters and include suggestions or comments that the chairman of the board considers to be important for the directors to know. In general, communications relating to corporate governance and corporate strategy are more likely to be forwarded than communications relating to ordinary business affairs, personal grievances and matters as to which we receive repetitive or duplicative communications.

Stockholders who wish to send communications on any topic to our board of directors should address such communications to Agios Pharmaceuticals, Inc., c/o Corporate Secretary, 88 Sidney Street, Cambridge, Massachusetts 02139.

Board Meetings and Attendance

Our board of directors met seven times during our fiscal year 2018, including telephonic meetings. During the year, each of our directors attended 75% or more of the aggregate number of meetings of the board of directors and the committees on which they served, other than Dr. Fouse, who attended 50% of the aggregate number of meetings of the board of directors and the committees on which she served.

In fiscal year 2018, in order to avoid the appearance of a conflict of interest and at our request, Dr. Fouse recused herself from attendance at three meetings of the board of directors that were called to exclusively discuss Dr. Fouse's potential transition to the role of chief executive officer of our company upon Dr. Schenkein's resignation from such position.

Had Dr. Fouse attended those three meetings, she would have attended more than 75% of the aggregate number of meetings of the board of directors and the committees on which she served in fiscal year 2018.

Since Dr. Fouse joined our board of directors, she has provided invaluable service to our board of directors and has served on the science and technology committee. Her extensive experience in the biotechnology sector makes her particularly well-suited to help our board of directors address the types of challenges commonly faced by biopharmaceutical companies, and she has brought that experience to bear as a participant in every major decision, other than the aforementioned executive transition, that our board of directors has made during her tenure both formally at regularly scheduled meetings and informally outside of regularly scheduled board meetings. Moreover, as chief executive officer of our company, Dr. Fouse's extensive knowledge of our company and our business is of significant value to our board of directors.

In discussions with Dr. Fouse, she has re-affirmed her commitment to attend at least 75% of all board and committee meetings on which she is then-serving, absent an illness or family emergency, in the future, and she

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has attended 100% of the meetings of the board of directors and the committees on which she was then-serving during the current fiscal year.

Director Attendance at Annual Meetings

Although our company does not have a formal policy regarding attendance by members of our board of directors at our annual meeting, we encourage all of our directors to attend. Seven of our eight then-serving directors attended our 2018 annual meeting of stockholders.

Board Committees

We have four standing committees: the audit committee, the compensation committee, the nominating and corporate governance committee and the science and technology committee. Each of these committees has a written charter approved by our board of directors. A copy of each charter can be found on the Corporate Governance page of the Investors section of our website, www.agios.com.

Audit Committee

The members of our audit committee are Mr. Clancy, Dr. Ho and Dr. Maraganore. Mr. Clancy is the chair of the audit committee. Our board of directors has determined that Mr. Clancy and Ms. Ho qualify as audit committee financial experts within the meaning of SEC regulations and the Nasdaq Listing Rules. In making this determination, our board of directors has considered the formal education and nature and scope of his or her previous experience, coupled with past and present service on various audit committees. Our audit committee assists our board of directors in its oversight of our accounting and financial reporting process and the audits of our financial statements. The audit committee met eight times during fiscal year 2018, including telephonic meetings. The audit committee's responsibilities include:

appointing, approving the compensation of, and assessing the independence of our registered public accounting firm;

overseeing the work of our registered public accounting firm, including through the receipt and consideration of reports from such firm;

reviewing and discussing with management and the registered public accounting firm our annual and quarterly financial statements and related disclosures;

recommending to our board of directors whether the audited financial statements should be included in our annual report;

monitoring our internal control over financial reporting, disclosure controls and procedures and code of business conduct and ethics;

overseeing our internal audit function;

discussing our major financial risk exposures and risks relating to data privacy and cybersecurity, and our risk management policies;

establishing policies regarding hiring employees from the registered public accounting firm and procedures for the receipt, retention and treatment of accounting related complaints and concerns;

meeting independently with our internal auditing staff, registered public accounting firm and management;

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reviewing and approving or ratifying any related person transactions; and

preparing the audit committee report required by SEC rules, which is included on page 31 of this Proxy Statement.

Compensation Committee

The members of our compensation committee are Ms. Foster, Dr. Maraganore and Mr. Clark. Ms. Foster is the chair of the compensation committee. Our compensation committee assists our board of directors in the discharge of its responsibilities relating to the compensation of our executive officers. The compensation committee met six times during fiscal year 2018. The compensation committee's responsibilities include:

reviewing and approving corporate goals and objectives relevant to CEO compensation;

making recommendations to our board of directors with respect to the compensation of our chief executive officer, and reviewing and approving the compensation of our other executive officers;

overseeing an evaluation of our senior executives;

overseeing and administering our cash and equity incentive plans;

reviewing and making recommendations to our board of directors with respect to director compensation;

reviewing and discussing with management our Compensation Discussion and Analysis, which is included on page 33 of this Proxy Statement; and

preparing the compensation committee report required by SEC rules, which is included on page 50 of this Proxy Statement.

Nominating and Corporate Governance Committee

The members of our nominating and corporate governance committee are Ms. Foster, Mr. Clark and Dr. Ho. Ms. Foster is the chair of the nominating and corporate governance committee. The nominating and corporate governance committee met five times during fiscal year 2018. The nominating and corporate governance committee's responsibilities include:

identifying individuals qualified to become board members;

recommending to our board of directors the persons to be nominated for election as directors and to each committee of our board of directors;

reviewing and making recommendations to the board of directors with respect to management succession planning;

developing and recommending corporate governance principles to the board of directors; and

overseeing periodic evaluations of the board of directors.

The processes and procedures followed by our nominating and corporate governance committee in identifying and evaluating director candidates are described above under the heading **Director Nomination Process** .

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Science and Technology Committee

The members of our science and technology committee are Dr. Scadden, Dr. Fouse and, effective February 1, 2019, Dr. Schenkein. Dr. Scadden is the chair of the science and technology committee. The science and technology committee assists our board's oversight of our research and development activities. The science and technology committee met two times during fiscal year 2018, including telephonic meetings. The committee's responsibilities include:

reviewing, evaluating, and advising our board of directors and management regarding our long-term strategic goals and objectives and the quality and direction of our research and development programs;

monitoring and evaluating trends in research and development, and recommending to our board of directors and management emerging technologies for building the company's technological strength;

recommending approaches to acquiring and maintaining technology positions (including but not limited to contracts, grants, collaborative efforts, alliances, and capital);

advising our board of directors and management on the scientific aspects of business development transactions;

regularly reviewing the company's research and development pipeline;

assisting our board of directors with its oversight responsibility for enterprise risk management in areas affecting the company's research and development; and

reviewing such other topics as delegated to the science and technology committee from time to time by our board of directors.

The Board's Role in Risk Oversight

Our board of directors has responsibility for the oversight of the company's risk management processes and, either as a whole or through its committees, regularly discusses with management our major risk exposures, the potential impact of these risks on our business and the steps we take to manage them. The risk oversight process includes receiving regular reports from board committees and members of senior management to enable our board of directors to understand the company's risk identification, risk management and risk mitigation strategies with respect to areas of potential material risk, including operations, finance, legal, regulatory, strategic and reputational risk.

The audit committee reviews information regarding liquidity and operations, and oversees our management of financial risks. Periodically, the audit committee reviews our policies with respect to risk assessment, risk management, loss prevention and regulatory compliance. Oversight by the audit committee includes direct communication with our external auditors, and discussions with management regarding significant risk exposures and

risks relating to data privacy and cybersecurity and the actions management has taken to limit, monitor or control such exposures. The compensation committee is responsible for assessing whether any of our compensation policies or programs has the potential to encourage excessive risk-taking. The nominating and corporate governance committee manages risks associated with the independence of the board, corporate disclosure practices, and potential conflicts of interest. The science and technology committee assists the board's oversight of the company's research and development activities. While each committee is responsible for evaluating certain risks and overseeing the management of such risks, the entire board of directors is regularly informed through committee reports about such risks. Matters of significant strategic risk are considered by our board of directors as a whole.

Table of Contents**Risk Considerations in our Compensation Program**

We along with our compensation committee of our board of directors have reviewed the compensation policies and practices for all of our employees and believe any risks arising from our compensation policies and programs are not reasonably likely to have a material adverse effect on our company or its operations. In reaching this conclusion, the compensation committee and we considered several factors, including the following:

the establishment of base salaries consistent with our executive officers' responsibilities and market comparable companies to ensure that our executive officers would not be motivated to take excessive risks to achieve a reasonable level of financial security;

the mix between fixed and variable, annual and long-term, and cash and equity compensation, which is intended to encourage strategies and actions that are in our company's long-term best interests;

vesting periods for equity compensation awards that reward sustained stock price appreciation;

the evaluation of company performance (which drives the amount of cash and number of shares available for grant under our contingent annual performance-based cash incentive and annual equity incentive programs, respectively) based on a variety of long- and short-term objectives in which no single objective is given substantial weight, thus diversifying the risk associated with any single indicator of performance; and

the discretion available to our compensation committee not to apply fixed formulae in assessing our company performance, thus enabling the compensation committee to, among other things, (a) eliminate the potential incentive for management to conduct activities that are in the company's annual goals, but which may not, due to new data or other inputs, ultimately prove to be in the best interest of stockholders, and (b) reward management for making decisions that are in the long-term best interest of our product development programs, even when those decisions result in the failure to meet short-term objectives.

Director Compensation

Our board of directors has adopted revisions to our non-employee director compensation policy that are designed to provide a total compensation package that enables us to attract and retain, on a long-term basis, high caliber non-employee directors. The cash and equity compensation for non-employee directors under the policy, effective January 1, 2019, is set forth in the table below.

| | Annual Cash Compensation | Number of Options/RSUs Granted |
|----------------------------|---------------------------------|---------------------------------------|
| Board of Directors: | | |
| Board Member | \$50,000 | |

\$718,000 in equity awards upon initial election (split approximately 75% in stock options and 25% in RSUs, based on value); \$360,000 in equity awards at each annual meeting thereafter (split approximately 75% in stock options and 25% in RSUs, based on value)*

| | |
|---------------------------|------------------------|
| Chair | Additional \$30,000 |
| Lead Independent Director | Additional \$25,000 |

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| | Annual Cash Compensation | Number of Options/RSUs Granted |
|---|--------------------------|--------------------------------|
| Audit Committee: | | |
| Chair | \$20,000 | |
| Member (other than Chair) | \$10,000 | |
| Compensation Committee: | | |
| Chair | \$15,000 | |
| Member (other than Chair) | \$7,500 | |
| Nominating and Corporate Governance Committee: | | |
| Chair | \$10,000 | |
| Member (other than Chair) | \$5,000 | |
| Science and Technology Committee: | | |
| Chair | \$15,000 | |
| Member (other than Chair) | \$7,500 | |

*number of units for stock option and RSU awards to be determined on the date of grant based on grant date fair value.

Under the policy, non-employee members of our board of directors also are reimbursed for travel, lodging and other reasonable expenses incurred in attending board of directors or committee meetings.

The stock options granted to our non-employee directors have an exercise price equal to the fair market value of our common stock on the date of grant and expire ten years after the date of grant. The stock options and restricted stock units, or RSUs, granted to our non-employee directors are subject to vesting based upon a director's continued service on our board of directors. The initial stock options and RSUs granted to our newly elected non-employee directors vest with respect to 25% of the shares on the first anniversary of the grant date and monthly thereafter until the fourth anniversary of the date of grant. The annual stock options and RSUs granted to our non-employee directors vest with respect to 100% of the shares on the first anniversary of the grant date. To the extent that a non-employee director has other responsibilities, such director may receive additional compensation to the extent deemed necessary by our board of directors. Directors who also are employees do not receive cash or equity compensation for service on the board of directors in addition to compensation payable for their service as employees.

For the 2018 fiscal year, our non-employee directors were compensated pursuant to our compensation guidelines that were previously recommended by our compensation committee and adopted by our board of directors in December 2017, as summarized below:

| | Annual Cash Compensation | Number of Options/RSUs Granted |
|----------------------------|--------------------------|--|
| Board of Directors: | | |
| Board Member | \$50,000 | 11,400 stock options and 2,800 RSUs upon initial election; |

5,700 stock options and 1,400 RSUs
at each annual meeting

thereafter

Chair

Additional
\$30,000

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| | Annual Cash Compensation | Number of Options/RSUs Granted |
|---|--------------------------|--------------------------------|
| Audit Committee: | | |
| Chair | \$20,000 | |
| Member (other than Chair) | \$10,000 | |
| Compensation Committee: | | |
| Chair | \$15,000 | |
| Member (other than Chair) | \$7,500 | |
| Nominating and Corporate Governance Committee: | | |
| Chair | \$10,000 | |
| Member (other than Chair) | \$5,000 | |
| Science and Technology Committee: | | |
| Chair | \$15,000 | |
| Member (other than Chair) | \$7,500 | |

The following table sets forth information concerning the compensation for our non-employee directors during the fiscal year ended December 31, 2018:

| Name | Fees Earned or Paid in Cash (\$) | Option Awards (\$)(1)(2) | Stock Awards (\$)(3)(4) | Total (\$) |
|---------------------------|----------------------------------|--------------------------|-------------------------|------------|
| Paul J. Clancy | \$ 70,000 | \$ 130,900 | \$ 353,241 | \$ 554,141 |
| Ian T. Clark | \$ 62,500 | \$ 130,900 | \$ 353,241 | \$ 546,641 |
| Kaye Foster | \$ 75,000 | \$ 130,900 | \$ 353,241 | \$ 559,141 |
| Jacquelyn A. Fouse, Ph.D. | \$ 53,750 | \$ 130,900 | \$ 353,241 | \$ 537,891 |
| Maykin Ho, Ph.D. | \$ 65,000 | \$ 130,900 | \$ 353,241 | \$ 549,141 |
| John M. Maraganore, Ph.D. | \$ 97,500 | \$ 130,900 | \$ 353,241 | \$ 581,641 |
| David Scadden, M.D. | \$ 63,125 | \$ 130,900 | \$ 353,241 | \$ 547,266 |

- (1) Represents stock options to purchase 5,700 shares of common stock granted to each of our non-employee directors during 2018 for service on our board of directors. The shares subject to these stock options vest in full on May 31, 2019. Amounts listed represent the aggregate fair value amount computed as of the grant date of the stock option awards granted during 2018 in accordance with Financial Accounting Standards Board Accounting Standards Codification Topic 718, or FASB ASC Topic 718. Assumptions used in the calculation

of these amounts are included in Note 12, Share-Based Payments, of the Notes to our Consolidated Financial Statements filed on our Annual Report on Form 10-K, filed with the SEC on February 14, 2019.

- (2) The aggregate number of shares of common stock underlying stock options outstanding as of December 31, 2018 for our non-employee directors were: Mr. Clancy: 73,100, Mr. Clark: 29,700, Ms. Foster: 42,599, Dr. Fouse: 21,700, Dr. Ho: 42,950, Dr. Maraganore: 54,825, and Dr. Scadden: 29,700.

- (3) Represents RSUs representing the contingent right to receive 1,400 shares of common stock granted to each of our non-employee directors during 2018 for service on our board of directors. The shares subject to these RSUs vest in full on May 31, 2019. Amounts listed represent the aggregate fair value amount computed as of the grant date of the RSUs granted during 2018 in accordance with FASB ASC

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Topic 718. Assumptions used in the calculation of these amounts are included in Note 12, Share-Based Payments, of the Notes to our Consolidated Financial Statements filed on our Annual Report on Form 10-K, filed with the SEC on February 14, 2019.

- (4) The aggregate number of shares of common stock underlying restricted stock units outstanding as of December 31, 2018 for our non-employee directors were: Mr. Clancy: 1,400, Mr. Clark: 1,400, Ms. Foster: 1,400, Dr. Fouse: 1,400, Dr. Ho: 1,400, Dr. Maraganore: 1,400, and Dr. Scadden: 1,400. Dr. Fouse, one of our directors who also serves as our chief executive officer, will not receive any additional compensation for her service as a director from and after February 1, 2019. Dr. Schenkein, one of our directors who also served as our president and chief executive officer until February 1, 2019, did not receive any additional compensation for his service as a director during 2018; Dr. Schenkein transitioned to the role of executive chairman of our board of directors effective February 1, 2019, and will not receive any additional compensation for his service as a director in that role.

Limitation of Liability and Indemnification

Our certificate of incorporation limits the personal liability of directors for breach of fiduciary duty to the maximum extent permitted by the Delaware General Corporation Law and provides that no director will have personal liability to us or to our stockholders for monetary damages for breach of fiduciary duty or other duty as a director. However, these provisions do not eliminate or limit the liability of any of our directors:

for any breach of the director's duty of loyalty to us or our stockholders;

for acts or omissions not in good faith or which involve intentional misconduct or a knowing violation of law;

for voting or assenting to unlawful payments of dividends, stock repurchases or other distributions; or

for any transaction from which the director derived an improper personal benefit.

Any amendment to or repeal of these provisions will not eliminate or reduce the effect of these provisions in respect of any act, omission or claim that occurred or arose prior to such amendment or repeal. If the Delaware General Corporation Law is amended to provide for further limitations on the personal liability of directors of corporations, then the personal liability of our directors will be further limited to the greatest extent permitted by the Delaware General Corporation Law.

In addition, our certificate of incorporation provides that we must indemnify our directors and officers and we must advance expenses, including attorneys' fees, to our directors and officers in connection with legal proceedings, subject to very limited exceptions.

We maintain a general liability insurance policy that covers certain liabilities of our directors and officers arising out of claims based on acts or omissions in their capacities as directors or officers. In addition, we have entered into indemnification agreements with our directors and executive officers. These indemnification agreements require us,

among other things, to indemnify each such director and executive officer for some expenses, including attorneys' fees, judgments, fines and settlement amounts incurred by him in any action or proceeding arising out of his service as one of our directors or executive officers.

Certain of our non-employee directors may, through their relationships with their employers, be insured and/or indemnified against certain liabilities incurred in their capacity as members of our board of directors.

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Insofar as indemnification for liabilities arising under the Securities Act of 1933, as amended, or the Securities Act, may be permitted to directors, executive officers or persons controlling us, in the opinion of the SEC, such indemnification is against public policy as expressed in the Securities Act and is therefore unenforceable.

Executive and Director Compensation Processes

The compensation committee generally meets at least four times annually and with greater frequency if necessary. The agenda for each meeting is usually developed by the chair of the compensation committee, in consultation with the chief executive officer or the head of human resources. The compensation committee meets regularly in executive session. However, from time to time, various members of management and other employees as well as outside advisors or consultants may be invited by the compensation committee to make presentations, to provide financial or other background information or advice or to otherwise participate in compensation committee meetings. The chief executive officer may not participate in, or be present during, any deliberations or determinations of the compensation committee regarding his or her compensation. The charter of the compensation committee grants the compensation committee full access to all of our books, records, facilities and personnel, as well as authority to obtain, at our expense, advice and assistance from internal and external legal, accounting or other advisors and consultants and other external resources that the compensation committee considers necessary or appropriate in the performance of its duties. In particular, the compensation committee has the sole authority to retain compensation consultants to assist in its evaluation of executive and director compensation, including the authority to approve the consultant's reasonable fees and other retention terms.

During the fiscal year ended December 31, 2018, the compensation committee directly engaged Radford, a compensation consultant that is affiliated with AON, to advise the compensation committee on our compensation program, which includes base salaries, annual performance-based cash incentives and equity incentive awards. Radford did not determine or make recommendations to the compensation committee regarding the specific amount or form of compensation of our executive officers or directors for fiscal year ended December 31, 2018.

Historically, the compensation committee has made (or has recommended that the independent members of the board of directors make) most of the significant adjustments to annual compensation, determined bonus and equity awards and established new performance objectives at one or more meetings held during the last quarter of the year and the first quarter of the following year. However, the compensation committee also considers matters related to individual compensation, such as compensation for new executive hires, as well as high-level strategic issues, such as the efficacy of our compensation strategy, potential modifications to that strategy and new trends, plans or approaches to compensation, at various meetings throughout the year. The compensation committee is responsible for making determinations regarding compensation of our executive officers other than our chief executive officer, making changes to pre-approved salary ranges, salary increases, equity awards, incentive payments and pre-approved equity ranges for new hires and high performers, the initiation of offerings under our 2013 employee stock purchase plan and making material changes to benefits offered to our employees. In addition, the compensation committee makes recommendations to our board of directors regarding the compensation of directors and the chief executive officer, and the determination of the size of annual evergreen increases to the number of shares reserved under our 2013 stock incentive plan and 2013 employee stock purchase plan.

Under its charter, the compensation committee may form, and delegate authority to, subcommittees, consisting of independent directors, as it deems appropriate. During fiscal year 2018, the compensation committee did not form or delegate authority to such subcommittees. During fiscal year 2018, the compensation committee delegated to the chief executive officer, or if the chief executive officer was unavailable, the chief financial officer, decision-making authority related to initial salary levels and salary adjustments, incentive payments and option grants for all non-executive officers, and non-material changes to employee benefits. Such delegated decision-making is governed

by guidelines established by the compensation committee.

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Report of the Audit Committee of the Board of Directors

The audit committee oversees the Company's financial reporting process on behalf of the board of directors. We have reviewed the Company's audited consolidated financial statements for the fiscal year ended December 31, 2018 and discussed them with Company management and PricewaterhouseCoopers LLP, the Company's independent registered public accounting firm.

We have received from, and discussed with, PricewaterhouseCoopers LLP, which is responsible for expressing an opinion on the conformity of the Company's audited consolidated financial statements with accounting principles generally accepted in the United States, its judgments as to the quality, not just the acceptability, of the Company's accounting principles and such other matters as are required to be discussed with the audit committee under generally accepted auditing standards, including the matters required to be discussed by Statement on Auditing Standards No. 1301, Communications with Audit Committees, as adopted by the Public Company Accounting Oversight Board (the PCAOB). In addition, we have received from PricewaterhouseCoopers LLP the written disclosures and the letter required by applicable requirements of the PCAOB regarding its communications with us concerning independence, have considered the compatibility of non-audit services with the auditors' independence and have discussed with PricewaterhouseCoopers LLP its independence from management and the Company.

Based on the review and discussions referred to above, we recommended to the board of directors that the audited consolidated financial statements be included in the Company's Annual Report on Form 10-K for the year ended December 31, 2018.

This report of the audit committee is not soliciting material, shall not be deemed filed with the SEC and shall not be incorporated by reference by any general statement incorporating by reference this Proxy Statement into any filing under the Securities Act of 1933, as amended, or the Securities Exchange Act of 1934, as amended, whether made before or after the date hereof and irrespective of any general incorporation language in any such filing, except to the extent that we specifically incorporate this information by reference, and shall not otherwise be deemed filed under such acts.

The foregoing report has been furnished by the audit committee.

Respectfully submitted,

The Audit Committee of the Board of Directors

Paul J. Clancy (chair)

Maykin Ho

John M. Maraganore

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The following table sets forth information regarding our executive officers as of April 8, 2019:

| Name | Age | Position(s) |
|---------------------------|------------|--------------------------|
| Jacquelyn A. Fouse, Ph.D. | 57 | Chief Executive Officer |
| Scott Biller, Ph.D. | 63 | Chief Scientific Officer |
| Chris Bowden, M.D. | 58 | Chief Medical Officer |
| Andrew Hirsch | 48 | Chief Financial Officer |

The biography of Dr. Fouse can be found under Nominees for Election to the Board of Directors.

Scott Biller, Ph.D. joined Agios in September 2010 as chief scientific officer, with more than 25 years of drug discovery and development experience. From 2003 to September 2010, Dr. Biller was vice president and head of global discovery chemistry at the Novartis Institutes for Biomedical Research, an affiliate of publicly-traded biopharmaceutical company Novartis AG. Prior to that, Dr. Biller held the positions of vice president, pharmaceutical candidate optimization at the BMS Pharmaceutical Research Institute, a division of Bristol-Myers Squibb Company, or BMS, a global biopharmaceutical company, and executive director of drug discovery chemistry for the BMS research site in Lawrenceville, New Jersey. Among his other key leadership positions at BMS, Dr. Biller was the executive director of metabolic diseases chemistry. He contributed to the drug candidate pipelines at both BMS and Novartis AG, culminating in two medicines launched worldwide (Onglyza[®] for the treatment of Type 2 diabetes and Juxtapid[®] for familial hypercholesterolemia) and three additional drugs reaching phase 3 clinical development. Dr. Biller earned a S.B. in chemistry at MIT, a Ph.D. in organic chemistry at Caltech and was an NIH Postdoctoral Fellow at Columbia University focusing on natural product synthesis.

Chris Bowden, M.D. joined Agios as chief medical officer in May 2014. He has more than 19 years of experience in clinical drug development, including the approval of several cancer medicines. Prior to joining Agios, Dr. Bowden was vice president product development oncology, franchise lead (Signaling Group) at Genentech. During Dr. Bowden's eight years at Genentech, he was responsible for the successful development of a number of novel targeted oncology medicines, including Zelboraf[®] for patients with BRAF V600E positive melanoma and Tarceva[®] for patients with EGFR activating mutation positive, non-small cell lung cancer. From 2003 to 2006, Dr. Bowden was the executive director for EMEA (Europe, Middle East, Africa) regions for BMS. In this role, he led medical affairs strategies for cancer, immunology and pain medicines. Earlier, Dr. Bowden held positions of increasing responsibility in oncology clinical development, Phases I-III at Pharmacia Corporation and Janssen Pharmaceutical, Inc., each of which is a pharmaceutical company. Prior to his industry experience, Dr. Bowden was on the oncology faculty at the University of Virginia Health Science Center where he participated in numerous industry and cooperative group trials. Dr. Bowden received his M.D. from Hahnemann University School of Medicine followed by internal medicine training at Roger Williams Medical Center and the Providence VA Medical Center, Rhode Island. He completed his medical oncology fellowship at the National Cancer Institute Medicine Branch. Dr. Bowden is board certified in internal medicine and medical oncology.

Andrew Hirsch joined Agios as chief financial officer in September 2016, and was named head of corporate development in March 2018. He has more than 20 years of experience in a range of strategic and operating roles in the biotech sector, most recently having served as president and chief executive officer of BIND Therapeutics, Inc., a biotechnology company, from March 2015 until August 2016. Prior to being named president and chief executive officer at BIND, Mr. Hirsch held several other leadership positions at the company, including chief operating officer from February 2014 to March 2015, and chief financial officer from July 2012 to March 2015. Prior to joining BIND,

Mr. Hirsch was chief financial officer at Avila Therapeutics, Inc., a biotechnology company, from June 2011 until its acquisition by Celgene Corporation in March 2012. From 2002 to 2011, Mr. Hirsch held roles of increasing responsibility at Biogen Idec, including vice president of Corporate Strategy and M&A and program executive for the Tecfidera development team. He holds an M.B.A. from the Tuck School at Dartmouth College and a B.A. in Economics from the University of Pennsylvania.

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EXECUTIVE COMPENSATION

Compensation Discussion and Analysis

Our compensation committee is responsible for reviewing and approving, or recommending for approval by the board of directors, the compensation of our named executive officers, including salary, cash and equity incentive compensation levels, severance arrangements, change in control benefits and other forms of executive compensation. This committee is also responsible for evaluating our company's performance against its goals and making related recommendations to our board of directors, assessing the performance of our named executive officers, and ensuring our compensation program is aligned with the objectives described below and competitive with those of other companies in our industry that compete with us for talent. This section discusses the principles underlying our compensation committee's policies and decisions with respect to the compensation of our named executive officers.

Our named executive officers for 2018 were as follows:

David P. Schenkein, M.D., our then president and chief executive officer;

Andrew Hirsch, our chief financial officer and head of corporate development;

Christopher Bowden, M.D., our chief medical officer;

Scott Biller, Ph.D., our chief scientific officer; and

Steve Hoerter, our then chief commercial officer.

Effective February 1, 2019, Dr. Schenkein transitioned from his role of president and chief executive officer to executive chairman, and Jacquelyn A. Fouse, Ph.D. assumed the position of chief executive officer. Effective March 15, 2019, Mr. Hoerter resigned as our chief commercial officer.

Say-on-Pay Vote Results

At our 2018 annual meeting of stockholders, we conducted a non-binding advisory vote on the compensation of our named executive officers, commonly referred to as a "say-on-pay" vote, in accordance with the Dodd-Frank Wall Street Reform and Consumer Protection Act of 2010. Over 98% of the votes cast by stockholders on this proposal, excluding broker non-votes, were cast in support of the compensation paid to our named executive officers. While this vote is a non-binding advisory vote, our compensation committee and board of directors take the voting results into account in determining the compensation of our named executive officers. Given the strong level of support evidenced by last year's say-on-pay vote, among other factors, our compensation committee decided to maintain our general approach to executive compensation and made no significant changes to our executive compensation program this year.

Our compensation committee and board of directors will continue to consider stockholder input and monitor our executive compensation program to ensure it aligns the interests of our executive officers with the interests of our stockholders and adequately addresses any stockholder concerns that may be expressed in future votes. Consistent

with the recommendation of our board of directors and the preference of our stockholders as reflected in the non-binding advisory vote on the frequency of future say-on-pay votes conducted at our 2016 annual meeting of stockholders, our stockholders will have an opportunity annually to cast an advisory vote in connection with compensation for our named executive officers.

Executive Summary

Agios made significant progress on a number of fronts in 2018, achieving development milestones across our portfolio and enhancing shareholder value.

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We continued the clinical development and commercialization of our IDH mutant inhibitor programs. We obtained approval from the U.S. Food and Drug Administration, or FDA, of TIBSOVO® (ivosidenib), our second internally discovered and developed medicine, for the treatment of adult patients with relapsed or refractory acute myeloid leukemia, or R/R AML, with an IDH1 mutation as detected by an FDA-approved test, and shipped TIBSOVO® into the distribution channel less than 24 hours after FDA approval. TIBSOVO® joins IDHIFA® (approved for R/R AML with an IDH2 mutation) as our second commercially approved medicine. We also submitted a supplemental new drug application, or sNDA, to the FDA for TIBSOVO® for the treatment of adult patients with newly diagnosed AML with an IDH1 mutation who are not eligible for standard therapy, and submitted a marketing authorization application, or MAA, to the European Medicines Agency, or EMA, for TIBSOVO® for the treatment of adult patients with R/R AML and an IDH1 mutation. On the business development front, we signed an exclusive licensing agreement with CStone Pharmaceuticals, or CStone, for the development and commercialization of ivosidenib in greater China.

In addition to our commercial and regulatory achievements, we continued to advance our clinical portfolio across our areas of focus. In the field of oncology, we accelerated our target enrollment timelines by reaching agreement with the FDA on a revised endpoint for AGILE, our phase 3 trial of ivosidenib in combination with VIDAZA® in frontline AML, and accelerated our enrollment timeline for ClarIDHy, our phase 3 trial of ivosidenib in previously-treated patients with nonresectable or metastatic cholangiocarcinoma with an IDH1 mutation. We also finalized pivotal development plans for our IDH mutant inhibitors in glioma, with plans to initiate a registration-enabling phase 3 study of vorasidenib in low-grade glioma with an IDH1 mutation by the end of 2019. We continued to drive enrollment in our other clinical studies, including our phase 1 dose escalation trial of AG-270, our MAT2A inhibitor, for which we met enrollment goals during 2018. In the field of rare genetic diseases, we initiated two pivotal trials for our pyruvate kinase-R, or PKR, activator, mitapivat, in patients with pyruvate kinase, or PK, deficiency, ACTIVATE and ACTIVATE-T, while also initiating a phase 2 proof-of-concept trial of mitapivat in thalassemia.

We advanced our research pipeline, submitting and receiving FDA clearance of an investigational new drug application, or IND, for AG-636, our dihydroorotate dehydrogenase, or DHODH, inhibitor, for the treatment of hematologic malignancies and progressing our late-stage preclinical programs. In addition, during 2018 we completed an underwritten public offering of our common stock, resulting in net proceeds to us of approximately \$516.2 million, strengthening our balance sheet and extending our cash runway. Finally, we executed a CEO-transition effective February 1, 2019, appointing Dr. Fouse as our chief executive officer effective upon Dr. Schenkein's transition to executive chairman. Based on our achievements in 2018, the compensation committee determined and the board approved that we exceeded our company goals for 2018 as a whole (125% of target).

To foster the future success of the company, we reward our executives in a manner that reinforces our pay-for-performance philosophy and culture. Consistent with our pay-for-performance philosophy, total target compensation (base salary, target annual performance-based cash incentives and equity incentive awards) for our named executive officers in 2018 continued to be primarily performance-based, as shown in the charts below.

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We have developed and periodically reassess our executive compensation program to align with current governance and best practices while ensuring our ability to achieve our stated objectives and philosophy and support our ambitious business goals:

| What We Do | What We Don't Do |
|---|---|
| Maintain an industry-specific peer group for benchmarking pay | Allow hedging or pledging of equity |
| Target pay based on market norms | × Re-price stock options |
| Deliver executive compensation primarily through performance-based pay | × Provide perquisites |
| Set challenging short- and long-term incentive award goals | × Provide supplemental executive retirement plans |
| Cap annual cash incentive program payouts at 150% of the target payout level | |
| Maintain a clawback policy for equity and incentive compensation | |
| Require minimum levels of stock ownership by executives | |
| Offer market-competitive benefits for executives that are consistent with the rest of our employees | |
| Consult with an independent advisor on compensation levels and practices | |
| <i>Compensation Objectives and Philosophy</i> | |

Our mission is to transform the lives of patients with cancer and rare genetic diseases by applying our scientific leadership in the field of cellular metabolism. Our compensation committee believes that the most effective compensation program is one that rewards sustainable value creation for stockholders, by delivering strong company performance, as well as tangible progress toward achieving our mission to improve the lives of patients. The objectives of our compensation program are to:

attract and retain superior executive officers and other employees with outstanding skills and values who contribute to our long-term success;

provide incentives that motivate and reward the achievement of performance goals that directly correlate to the enhancement of stockholder value, as well as to facilitate executive retention; and

align executives' interests with those of stockholders by rewarding the achievement of short- and long-term strategic and financial goals, which we believe serves to enhance short- and long-term value creation for our stockholders.

To achieve its objectives, our compensation committee evaluates our executive compensation program with the goal of setting total compensation at levels that align with our culture, total rewards strategy, size and life stage.

Specifically, our compensation committee targets base salaries at the 50th percentile of our peer group, as discussed below, and seeks to ensure that such salaries reflect each executive's level of experience, performance and responsibility and that such levels are competitive with those of other companies in our industry and region that compete with us for executive talent. Our compensation program links a substantial portion of our

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named executive officers' compensation to achieving scientific, business, organizational and operational goals such as regulatory submissions and approvals; progress in our clinical trials and research programs; achieving key research and development milestones; maintaining the strong financial health of the company, including implementation of appropriate financing strategies; maintaining key strategic relationships; adding to and developing internal competencies, including retention of high-performing employees; and achieving desired financial metrics. Our compensation committee targets this annual performance-based cash incentive compensation at the 50th percentile of our peer group. In addition, we provide a significant portion of our executive compensation in the form of stock options and restricted stock units, or RSUs, that vest over time, which we believe helps to retain our executives and aligns their interests with those of our stockholders by allowing them to participate in the longer term success of our company as reflected in the appreciation of our stock price. We also grant our executives performance share unit awards in limited circumstances. Our compensation committee considers a host of factors in comparison to our peers when determining equity incentive compensation for our named executive officers, including annual long-term incentive values, annual equity awards expressed as a percent of total shares outstanding, total annual and cumulative dilution, the retentive value of outstanding awards and total equity ownership. Given the dynamic biopharmaceutical market, the compensation committee does not overemphasize any one perspective. Rather, the committee takes a holistic perspective, further considering the achievement of company goals and how that impacts total shareholder return, when determining actual award levels for the named executive officers, which the committee generally targets between the 50th and the 75th percentile of our peer group. We believe that targeting overall compensation in this manner is necessary and appropriate in order to attract and retain the quality of talent we need to successfully grow our business, achieve our challenging goals, sustain strong performance, and differentiate ourselves from those companies against which we compete for talent. However, any given individual employee's compensation may vary from the targeted pay framework, based on the unique responsibilities and requirements of his or her position, his or her experience and other qualifications, internal parity relative to similar positions within the company, and individual or company performance relative to performance goals and the peer group to ensure appropriate pay-for-performance alignment. While we do not have a formal or informal policy for allocating between long-term and short-term compensation, between cash and non-cash compensation or among different forms of non-cash compensation, we generally strive to provide our named executive officers with a balance of short-term and long-term incentives to encourage consistently strong performance.

Overview of Executive Compensation Process

As a part of determining named executive officer performance and compensation, our compensation committee receives recommendations from our chief executive officer (except with respect to his or her own compensation and performance). Our chief executive officer's performance and compensation is approved by the board based upon the recommendation of our compensation committee. The evaluation of each of our named executives is based on our overall corporate performance against annual goals that are approved by the board of directors at the beginning of each year, as discussed in more detail below.

The compensation committee has the sole authority to retain, at our expense, one or more third-party compensation consultants to assist the compensation committee in performing its responsibilities. The compensation committee may terminate the services of the consultant if the compensation committee deems it appropriate. In 2018, the compensation committee utilized the services of Radford, an AON company, to assist it in fulfilling its responsibilities. Radford was retained exclusively by the compensation committee and has not been retained by management to perform any work for the company other than projects performed at the direction of the compensation committee. Radford provides analysis and recommendations regarding:

trends and emerging topics with respect to executive compensation;

peer group selection for executive compensation benchmarking;

compensation practices of our peer group, including executive severance arrangements;

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compensation philosophy and programs, including risk assessment, for executives and non-executives;

stock utilization and other metrics; and

board of directors compensation.

In addition, we subscribe to Radford's various global annual and specialized life sciences and general industry surveys on an ongoing basis. Radford advised the compensation committee on all of the principal aspects of executive compensation, including executive new hire compensation arrangements. Radford consultants attend meetings of the compensation committee, including executive sessions in which executive compensation issues are discussed, when requested to do so. Radford reports to the compensation committee and not to management, although it meets with management for purposes of gathering information for its analyses and recommendations. The compensation committee annually evaluates its engagement of compensation consultants, and selected Radford to advise with respect to compensation matters based on Radford's industry experience and reputation, which our compensation committee concluded gave Radford useful context and knowledge to advise it. The compensation committee has assessed the independence of Radford pursuant to Securities and Exchange Commission and Nasdaq rules and concluded that no conflict of interest exists that would prevent Radford from independently representing the compensation committee.

Annual base salaries for the current year, and annual performance-based cash incentives and equity incentive awards for the prior year are generally determined in the first quarter of the year based on company and individual performance of the prior year, as well as other factors, including compensation trends in the biopharmaceutical industry and among our benchmark peers. In February 2018, the compensation committee approved 2018 salaries, 2017 annual cash incentives and annual stock option and RSU awards for our named executive officers, except for the chief executive officer. With respect to our chief executive officer, the compensation committee reviewed his performance and presented to the board its recommendations for his 2018 salary and 2017 annual cash incentives and annual stock option and RSU awards for approval; these recommendations were then considered and approved by the board. In February 2019, the compensation committee approved 2019 salaries, 2018 annual cash incentives and annual stock option and RSU awards for our named executive officers, except for the chief executive officer. With respect to our chief executive officer, the compensation committee reviewed his performance and presented to the board its recommendations for his 2018 annual cash incentives for approval; these recommendations were then considered and approved by the board. Dr. Schenkein did not receive annual stock option or RSU awards in 2019 in connection with his role as chief executive officer. In August 2018, our board approved, on the recommendation of our compensation committee, 2019 base salaries, target annual performance-based cash incentive levels and initial equity awards for Dr. Fouse and Dr. Schenkein in connection with Dr. Fouse's appointment as our chief executive officer and Dr. Schenkein's transition to the role of executive chairman, each effective as of February 1, 2019, as set forth in the employee offer letters described below under *Employment, Severance and Change in Control Arrangements* *Employment Offer Letters* .

Defining and Comparing Compensation Benchmarks

The compensation committee benchmarks our executive compensation against a peer group of companies to determine competitiveness and market trends. The compensation committee reviews the companies in our peer group annually, reviews Radford's recommendations regarding which companies should be included in the peer group, and makes adjustments as necessary to ensure the peer group continues to properly reflect the market in which we compete for talented executives. The compensation committee also annually reviews the executive pay practices of other similarly-situated companies as reported by Radford through industry surveys and proxy analysis. These surveys are

specific to the biopharmaceutical and biotechnology sectors. We request customized reports of these surveys so that the compensation data reflect the practices of companies that are similar to us.

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The compensation committee considers this information when making determinations for each element of compensation.

In developing the peer group of companies to inform 2018 compensation decisions, our compensation committee, with the assistance of Radford, established a peer group of 17 publicly-traded, national and regional companies in the biopharmaceutical industry that was selected based on a balance of the following criteria:

companies whose market capitalization, number of employees, maturity of product development pipeline and area of therapeutic focus are similar to ours;

companies against which we believe we compete for executive talent; and

public companies based in the United States whose compensation and financial data are available in proxy statements or other public documents.

Based on these criteria, our peer group for 2018 was comprised of the following companies:

| | | |
|---------------------------------|-------------------------------|--------------------------------|
| ACADIA Pharmaceuticals, Inc. | Ionis Pharmaceuticals Inc. | Sarepta Therapeutics, Inc. |
| Accelaron Pharma, Inc. | Juno Therapeutics, Inc. | Seattle Genetics, Inc. |
| Alnylam Pharmaceuticals, Inc. | Neurocrine Biosciences, Inc. | Spark Therapeutics, Inc. |
| bluebird bio, Inc. | Portola Pharmaceuticals, Inc. | TESARO, Inc. |
| Clovis Oncology, Inc. | Puma Biotechnology Inc. | Ultragenyx Pharmaceutical Inc. |
| Intercept Pharmaceuticals, Inc. | Sage Therapeutics , Inc. | |

In evaluating the total compensation of our named executive officers for 2018, Radford reviewed data for the 2018 peer group, including survey data as well as data from proxy statements or other publicly available sources.

The compensation committee believes the compensation practices of our peer group provide us with appropriate compensation benchmarks for evaluating the compensation of our named executive officers. Notwithstanding the similarities of the peer group to our company, due to the nature of our business, we compete for executive talent with many companies that are larger and more established than we are or that possess greater resources than we do, as well as with prestigious academic and non-profit institutions. Other considerations, including market factors, the experience level of the executive and the executive's performance against established corporate goals and individual objectives, may require that our compensation committee vary from its historic compensation practices or deviate from its general compensation philosophy under certain circumstances.

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For the purposes of informing 2019 compensation decisions, the compensation committee, with the advice of Radford, examined the peer group list and, with reference to market capitalization, therapeutic area, stage of development, number of employees and other key business metrics, approved the following biopharmaceutical companies as our 2019 peer group:

| | | |
|---------------------------------|-------------------------------|--------------------------------|
| Alnylam Pharmaceuticals, Inc. | Ionis Pharmaceuticals Inc. | Seattle Genetics, Inc. |
| Array BioPharma Inc.* | Neurocrine Biosciences, Inc. | Spark Therapeutics, Inc. |
| bluebird bio, Inc. | Portola Pharmaceuticals, Inc. | TESARO, Inc. |
| Clovis Oncology, Inc. | Puma Biotechnology Inc. | Ultragenyx Pharmaceutical Inc. |
| Exelixis, Inc.* | Sage Therapeutics , Inc. | |
| Intercept Pharmaceuticals, Inc. | Sarepta Therapeutics, Inc. | |

*addition to 2019 peer group

The 2019 peer group represents a group of biopharmaceutical companies more similar to us in key measures than the list we used in 2018, in light of our substantial growth and the advancement of our development pipeline, focusing on companies with approximately 175 to 1,500 employees, a market capitalization between \$1.5 billion and \$14.5 billion, and a focus on orphan diseases, with a roughly equal mix of companies with a lead product candidate in the late-stage of clinical development and early commercial companies. Specifically, Juno Therapeutics, Inc. was removed from the 2018 list because it was acquired by Celgene, ACADIA Pharmaceuticals was removed from the 2018 list because it suffered a clinical development setback that affected its comparability at the time the peer group was reviewed, and Acceleron Pharma, Inc. was removed from the 2018 list because its stage of development affected its comparability at the time the peer group was reviewed. Array BioPharma Inc. and Exelixis, Inc. were added to the 2019 peer group.

Executive Compensation Elements

The primary elements of our executive compensation program are:

base salary;

annual performance-based cash incentives;

equity incentive awards;

severance and change in control benefits;

broad-based health and welfare benefits; and

broad-based 401(k) plan.

Our compensation committee uses its judgment to allocate long-term and short-term compensation for our named executive officers, in alignment with our pay-for-performance philosophy and the long-term interests of shareholders. After reviewing information provided by our compensation consultant and other relevant data,

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our compensation committee exercises its judgment to determine what it believes to be the appropriate level and mix of the various compensation components. We generally strive to provide our named executive officers with a balance of short-term and long-term incentives to encourage consistently strong performance. Ultimately, the objective in allocating between long-term and currently paid compensation is to ensure adequate base compensation to attract and retain talent, while providing incentives to maximize long-term value for our company and our stockholders.

Therefore, we provide cash compensation in the form of base salary to meet competitive salary norms and reward performance on an annual basis and in the form of incentive compensation to incent and reward performance based on specific annual goals. To further focus our executives on longer-term performance, we rely upon equity-based awards that vest over a meaningful period of time, thereby reinforcing stockholder value creation. In addition, we provide our executives with benefits that are available to all employees, including medical, vision and dental insurance; life and disability insurance; medical and dependent care flexible spending accounts; a 401(k) plan; and an opportunity to invest in our company pursuant to our employee stock purchase plan. Finally, we offer our executives severance benefits to incentivize them to continue to strive to achieve stockholder value in connection with change in control situations.

Base Salaries

Base salaries are used to recognize the experience, skills, knowledge and responsibilities required of our named executive officers. Base salaries for our named executive officers typically are established through arm's length negotiation at the time the named executive officer is hired, taking into account the position for which the named executive officer is being considered and the named executive officer's qualifications, prior experience and prior salary. None of our named executive officers is currently party to an employment agreement that provides for automatic or scheduled increases in base salary. However, on an annual basis, our compensation committee reviews and evaluates, with input from our chief executive officer (other than with respect to himself or herself), the need for adjustment of the base salaries of our named executive officers based on changes and expected changes in the scope of a named executive officer's responsibilities, including promotions, the individual contributions made by and performance of the named executive officer during the prior year, the named executive officer's performance over a period of years, overall labor market conditions, the relative ease or difficulty of replacing the executive with a well-qualified person, our overall growth and development as a company and general salary trends in our industry and among our peer group and where the named executive officer's salary falls in the salary range presented by that data. In making decisions regarding salary increases, we may also draw upon the experience of members of our board of directors with other companies. No formulaic base salary increases are provided to our named executive officers.

Based on a review of market data provided by Radford, the current compensation levels of our named executive officers and company performance and individual contributions, in February 2018, our compensation committee approved salary increases for our named executive officers, other than our chief executive officer, for 2018. With respect to our chief executive officer, the compensation committee reviewed his performance and presented to the board its recommendation for an increase in his 2018 salary, which recommendation was then considered and approved by the board. Our chief executive officer's salary increase reflected both merit and market-based adjustments; all other salary increases reflected merit-based adjustments. The 2018 base salaries of each of our named executive officers were:

| | 2017 | 2018 | % Increase |
|-----------------------------|-------------------------|-------------------------|-------------------|
| | Base Salary (\$) | Base Salary (\$) | over 2017 |
| David P. Schenkein, M.D.(1) | 585,040 | 625,000 | 6.8%(2) |
| Andrew Hirsch | 453,201 | 471,329 | 4.0%(3) |

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| | | | |
|--------------------------|---------|---------|---------|
| Christopher Bowden, M.D. | 449,174 | 467,141 | 4.0%(3) |
| Scott Biller, Ph.D. | 429,397 | 446,573 | 4.0%(3) |
| Steve Hoerter(4) | 427,451 | 444,549 | 4.0%(3) |

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- (1) Dr. Schenkein transitioned from the role of president and chief executive officer to executive chairman effective February 1, 2019.
- (2) Increase reflects merit and market-based adjustments.
- (3) Increase reflects a merit-based adjustment.
- (4) Mr. Hoerter resigned as our chief commercial officer effective March 15, 2019.

Annual Performance-based Cash Incentives

We have designed our annual performance-based cash incentive program, which is guided by specified annual corporate and individual goals and contributions, to emphasize pay-for-performance and to reward our named executive officers for our performance during the preceding year. The target pay opportunity of the annual cash incentive for the chief executive officer is determined by our board of directors, based upon the recommendation of our compensation committee, and the amount of the annual cash incentive for all other named executive officers is determined by our compensation committee. In making such determinations and recommendations, the compensation committee examined the totality of anticipated and unanticipated achievements by us and each named executive officer in the preceding year, including our performance against specific research, clinical, operational and financial company goals. In recent years, these annual company goals have primarily focused on the advancement of our lead programs.

Our compensation committee determined to formally cap annual cash incentive program payouts in any given year at 150% of the target payout level. Under our annual incentive program, cash incentive awards are determined by first establishing a cash incentive pool, which is then allocated among all eligible plan participants and adjusted based upon recommendations by our compensation committee and upon approval by our board of directors driven by performance as measured against the company's annual goals. Below is the list of the company's 2018 goals considered by our compensation committee and board of directors in their respective assessment of company performance against such 2018 goals:

Advance the clinical development and commercialization of our IDH inhibitor programs;

Prepare for potential approval of TIBSOVO® in the United States for IDH1 mutant-positive R/R AML and execute on commercial launch post-approval;

Reach agreement with FDA regarding event free survival as an acceptable endpoint for our phase 3 AGILE trial;

Continue to enroll patients in ClarIDHy, our global phase 3 trial of ivosidenib in IDH1 mutant-positive cholangiocarcinoma;

Support initiation of an intergroup sponsored phase 3 trial combination trial of ivosidenib or enasidenib in combination with standard induction and consolidation chemotherapy in frontline AML;

Advance development and regulatory discussions for ivosidenib in Europe;

Finalize pivotal development strategy for IDH inhibitors in glioma;

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Continue to drive development of our PKR activator program;

Initiate pivotal trials of mitapivat PK deficiency;

Initiate proof-of-concept trial of mitapivat in thalassemia;

Advance development of our preclinical and early clinical programs;

Continue to enroll patients in phase 1 dose escalation trial of AG-270;

File an IND for AG-636;

Continue preclinical development activities for our next wave of novel investigational medicines across the areas of cancer metabolism, rare genetic diseases and metabolic immuno-oncology;

Maintain our financial strength to ensure the execution of our long-term vision; and

Evolve and develop our organization and culture.

In December 2018, our chief executive officer recommended to our compensation committee that our company's performance against 2018 goals be assessed based on the following significant achievements in 2018:

Obtained full approval of TIBSOVO® from the FDA for the treatment of patients with R/R AML with a susceptible IDH1 mutation as detected by an FDA approved test, under priority review and ahead of the August 21, 2018 Prescription Drug User Fee Act date;

Shipped TIBSOVO® into the distribution channel within 24 hours of FDA approval, and achieved \$13.8 million of net sales for 2018;

Reached agreement with the FDA of event-free survival as an acceptable primary endpoint for the phase 3 AGILE trial of ivosidenib combination with VIDAZA® in newly diagnosed AML patients with an IDH1 mutation who are ineligible for intensive chemotherapy, and accelerated estimate for full enrollment of AGILE from 2021 to 2020;

Submitted an sNDA to the FDA for TIBSOVO® for the treatment of patients with newly diagnosed AML with an IDH1 mutation who are not eligible for standard therapy in December 2018;

Exceeded enrollment target for phase 3 ClarIDHy trial and accelerated estimate for full enrollment from 2019 to first half of 2019;

Submitted a MAA to the EMA for TIBSOVO® for the treatment of adult patients with R/R AML with an IDH1 mutation in December 2018;

Finalized pivotal development plans for vorasidenib in glioma;

Entered into an exclusive license agreement with CStone to develop and commercialize ivosidenib in greater China, resulting in a \$12 million upfront payment and the potential for \$412 million in development and commercial milestones;

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Initiated and began enrollment of ACTIVATE-T and ACTIVATE, global pivotal trials of mitapivat in 2018;

Initiated a phase 2 proof-of-concept trial of mitapivat in thalassemia in December 2018;

Met enrollment goals for phase 1 dose escalation trial of AG-270;

Submitted and received FDA clearance of an IND application for AG-636, a DHODH inhibitor, for the treatment of hematologic malignancies in November 2018;

Continued to advance our research pipeline across our three areas of focus while advancing our late-stage preclinical programs;

Completed an underwritten public offering of 8,152,986 shares of common stock, resulting in net proceeds to us of approximately \$516.2 million, and ended 2018 with a cash balance of approximately \$805 million, sufficient to meet our estimated cash needs through the at least the end of 2020; and

Continued to evolve our organization and expand internal capabilities consistent with our core values and culture, while successfully executing on our CEO transition plan.

Based on the significant advances we made against our 2018 goals, along with the overachievement on some goals, the overall company performance funding level for 2018 was set at 125% by our board of directors upon the recommendation of the compensation committee.

Our compensation committee also evaluates the individual performance of our named executive officers, with the input of our chief executive officer in the case of the evaluation of our other named executive officers, and makes recommendations to our board of directors with regard to the evaluation of our chief executive officer's individual performance. Consistent with this process, our compensation committee assessed the performance of our chief executive officer in 2018 based on our relative achievement of our corporate goals as well as his leadership in driving the execution of our strategic plans. In assessing the individual performance of our named executive officers other than our chief executive officer, our compensation committee, with the input of our chief executive officer, considered each such officer's individual contributions to the completion of our goals, and the officer's individual achievements in helping to build the company and execute on our strategy. These achievements include the following:

In 2018, Mr. Hirsch led our finance, accounting and business development functions, executing a successful follow-on public offering and executing an exclusive license agreement with CStone for the development and commercialization of ivosidenib in greater China. He also made significant progress in developing our finance, corporate development, information technology and investor relations functions.

In 2018, Dr. Bowden led our clinical and regulatory teams responsible for: obtaining FDA approval of TIBSOVO® for the treatment of adult patients with IDH1 mutant-positive R/R AML and submitting an sNDA for TIBSOVO® in newly-diagnosed IDH1 mutant-positive AML patients ineligible for standard treatment; submitting an IND for AG-636; initiating our phase 3 ACTIVATE and ACTIVATE-T trials of mitapivat; finalizing the pivotal strategy for vorasidenib in low grade glioma; and continuing enrollment across our ongoing clinical trials.

In 2018, Dr. Biller led our research and discovery science function, completing the preclinical development of AG-636, which enabled an IND submission and clearance by the FDA by

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year-end, and advancing development activities for our next wave of preclinical development programs across the areas of cancer metabolism, rare genetic diseases and metabolic immuno-oncology.

In 2018, Mr. Hoerter led our commercial function, leading our sales force in a successful launch of TIBSOVO® following approval of TIBSOVO® in 2018. He also made significant progress in continuing to strengthen our commercial operations functions.

Based on company and individual performance, our board of directors approved, upon the recommendation of the compensation committee, the 2018 cash incentive payment for our chief executive officer, and the compensation committee approved the 2018 cash incentive payments for all other named executive officers, as follows:

| | Target Award as a Percentage of Base Salary | 2018 Actual Cash Incentive Payment (\$) | 2018 Actual Cash Incentive Payment (% of Target Award) |
|-----------------------------|--|--|---|
| David P. Schenkein, M.D.(1) | 65% | 507,813 | 125% |
| Andrew Hirsch | 45% | 265,123 | 125% |
| Christopher Bowden, M.D. | 45% | 262,767 | 125% |
| Scott Biller, Ph.D. | 45% | 251,198 | 125% |
| Steve Hoerter(2) | 45% | 250,059 | 125% |

(1) Dr. Schenkein transitioned from the role of president and chief executive officer to executive chairman effective February 1, 2019.

(2) Mr. Hoerter resigned as our chief commercial officer effective March 15, 2019.

All named executive officers' 2018 annual cash incentive target award percentages increased by 5% from 2017. The 2018 cash incentive payments for all employees, including the named executive officers, were paid in 2019.

As discussed below under *2019 Executive Compensation Decisions*, the annual cash incentive target award percentages for 2019 performance for our named executive officers remain unchanged from 2018. Our board, upon the recommendation of our compensation committee, set Dr. Fouse's annual cash incentive target award percentage for 2019 performance at 65%, which is the same as Dr. Schenkein's annual cash incentive target award percentage in his role as chief executive officer for 2018.

Equity Incentive Awards

Our equity award program is the primary long-term incentive vehicle for our executives. We believe that equity grants provide our executives with a strong link to our long-term performance, create an ownership culture and help to align the interests of our executives and our stockholders. Our executives benefit from stock options and RSUs as our stock price increases through the creation of shareholder value; similarly, the performance share unit awards granted to our

executives vest only after the achievement of specified company performance milestones which benefit the company as a whole, or upon the achievement of a stock price performance threshold. Accordingly, we believe stock options, RSUs and performance share unit awards provide meaningful incentives to our executives to achieve increases in the value of our stock over time. In addition, the vesting feature of our equity grants contributes to executive retention by providing an incentive to our executives to remain employed by us during the vesting period.

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Equity compensation represents the largest at-risk component of our named executive officers' compensation arrangements. We believe that it is appropriate to align the interests of our named executive officers with those of our stockholders to achieve and sustain long-term stock price growth. Historically, we used stock options and performance share unit awards to compensate our named executive officers in the form of initial grants in connection with the commencement of employment, and generally granted stock options on an annual basis thereafter. As part of its ongoing review of our compensation strategy, our compensation committee determines the appropriate mix of the type of equity awards, based in part on recommendations from Radford. Effective in 2018, our compensation committee adjusted the mix of annual equity awards for all employees, including our named executive officers, to include RSUs with time-based vesting. Starting in 2018, the target mix for equity grants to our employees, including our named executive officers, is split approximately 75% stock options and 25% in RSUs, based on value. Our compensation committee believes that this equity mix helps to ensure that wealth creation remains tied to stock performance (through stock options), and promotes retention (via RSUs that vest over time to deliver equivalent value to stock options while using fewer authorized shares). In limited circumstances, our compensation committee, or board of directors, as applicable, grants performance share units to certain employees, including our named executive officers, in order to further promote retention and emphasize individual employees' impact on our organizational success. With the exception of the amended employment offer letter that Dr. Schenkein entered into in September 2018 in connection with his transition from president and chief executive officer to executive chairman, which provided for initial equity grants in February 2019 for this new role, none of our named executive officers are currently party to an employment agreement that provides for an automatic award of stock options, RSUs or performance share unit awards.

We grant equity awards to our named executive officers with both time-based and performance-based vesting. The stock options that we grant to our named executive officers with time-based vesting typically become exercisable as to 25% of the shares underlying the option on the first anniversary of the grant date, and as to an additional 1/48th of the shares underlying the option monthly thereafter. The RSUs we grant to our named executive officers vest in equal annual installments on each anniversary of the date of grant, until the third anniversary of such date. The performance share unit awards that we have granted to our named executive officers vest in connection with the achievement of specified company performance milestones. In addition, Dr. Fouse received a performance share unit award in connection with her appointment as chief executive officer, which award vests upon the achievement of a specified stock price performance milestone. The exercise price of all stock options equals the fair market value of shares of our common stock on the date of grant. Prior to the exercise of an option, the holder has no rights as a stockholder with respect to the shares subject to such option, including no voting rights and no right to receive dividends or dividend equivalents. Vesting for stock options ceases on termination of employment and exercise rights for stock options cease shortly after termination of employment except in the case of death or disability. Vesting of both RSUs and performance share unit awards ceases upon termination of employment.

In specified termination and change in control circumstances, equity awards held by our named executive officers are subject to accelerated vesting. See *Severance and Change in Control Benefits* below for further information.

In determining the size of the annual stock option and RSU grants to our named executive officers, our compensation committee, with the assistance from Radford, considers our company performance, individual performance, the potential for enhancing the creation of value for our stockholders, the amount of equity previously awarded to the named executive officers and the vesting terms of such prior awards, the company's broader organizational equity needs and overall dilution, as well as industry and peer group benchmark data.

Our compensation committee, or our board of directors, as applicable, made equity incentive awards in the form of stock options and RSUs to our named executive officers in the amounts set forth in the table below during 2018. In the case of each award, these grants were based on the named executive officer's existing equity incentive holdings, level

of responsibility within our company, equity ownership in relation to the peer group

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benchmark, and the compensation committee's assessment (or our board of directors' assessment, in the case of our chief executive officer) of the named executive officer's individual performance and our overall company performance in the fiscal year 2018, in each case without reference to any specific metric.

| | 2018 Equity Incentive Awards (Stock Options)(2) | 2018 Equity Incentive Awards (RSUs)(3) |
|-----------------------------|---|--|
| David P. Schenkein, M.D.(1) | 97,500 | 24,375 |
| Andrew Hirsch | 33,000 | 8,250 |
| Christopher Bowden, M.D. | 33,000 | 8,250 |
| Scott Biller, Ph.D. | 33,000 | 8,250 |
| Steve Hoerter(4) | 33,000 | 8,250 |

- (1) Dr. Schenkein transitioned from the role of president and chief executive officer to executive chairman effective February 1, 2019.
- (2) On February 16, 2018, our compensation committee, or board of directors, as applicable, approved the grant of these stock option awards at an exercise price of \$77.70 per share. The options have time-based vesting, with 25% of the shares underlying the award vesting on the first anniversary of the grant date and the remaining shares vesting monthly thereafter in equal increments over 36 months, subject to continued service.
- (3) On February 16, 2018, our compensation committee, or board of directors, as applicable, approved the grant of these RSU awards. The RSUs have time-based vesting, with one-third of the shares of common stock underlying the RSUs vesting on the first, second and third anniversaries of the grants date, subject to continued service.
- (4) Mr. Hoerter resigned as our chief commercial officer effective March 15, 2019. We evaluate our equity award program on an annual basis to ensure that it appropriately links to our long-term performance by aligning the interests of our executives and our stockholders, remains competitive with industry and peer benchmarks and is consistent with our overall equity needs and dilution levels. As discussed above, effective in 2018 our compensation committee determined to include RSUs in the mix of equity awards that we grant to our employees, including our named executive officers, which brings our equity compensation practices more in line with our peers and introduces an additional retention and compensation measure to our compensation strategy.

2019 Executive Compensation Decisions

In February 2019, our compensation committee, and our board, as applicable, approved 2019 base salaries, target annual performance-based cash incentive levels and annual stock option and RSU awards for our named executive officers, other than Dr. Schenkein, as set forth in the table below. In August 2018, our board approved, on the recommendation of our compensation committee, 2019 base salaries, target annual performance-based cash incentive levels and initial equity awards for Dr. Fouse and Dr. Schenkein in connection with Dr. Fouse's appointment as our chief executive officer and Dr. Schenkein's transition to the role of

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executive chairman, each effective as of February 1, 2019, as set forth in the employee offer letters described below under Employment, Severance and Change in Control Arrangements *Employment Offer Letters* .

| | Target Bonus Award as a Percentage of 2019 Base Salary | Change in Target Bonus Award from 2018 | 2019 Base Salary (\$) | % Base Salary Increase over 2018 | 2019 Equity Incentive Awards (Options)(4) | 2019 Equity Incentive Awards (RSUs)(5) |
|-----------------------------|---|---|------------------------------|---|--|---|
| David P. Schenkein, M.D.(1) | 65%(2) | | 175,000(2) | -72.0% | 45,719 | 8,324 |
| Andrew Hirsch | 45% | | 500,476 | 6.2%(3) | 40,000 | 10,000 |
| Christopher Bowden, M.D. | 45% | | 490,539 | 5.0%(3) | 40,000 | 10,000 |
| Scott Biller, Ph.D. | 45% | | 480,041 | 7.5%(3) | 40,000 | 10,000 |
| Steve Hoerter(6) | 45% | | 444,550 | | | |

- (1) Dr. Schenkein transitioned from the role of president and chief executive officer to executive chairman effective February 1, 2019. Dr. Schenkein's 2019 base salary, target annual performance-based cash incentive levels and initial equity awards for his role as executive chairman are listed in the table above, and were approved by our board in August 2018. Dr. Schenkein did not receive annual stock option or RSU awards in 2019 in connection with his role as chief executive officer.
- (2) In connection with his transition to executive chairman, Dr. Schenkein is entitled to receive a prorated bonus with respect to the period of time that he served as chief executive officer during 2019.
- (3) Increase reflects merit and market-based adjustments.
- (4) Our compensation committee, or board of directors, as applicable, approved the grant of these stock option awards on February 22, 2019 at an exercise price of \$58.86 per share. The options are time-based options, with 25% of the shares underlying the award vesting on the first anniversary of the grant date and the remaining shares vesting monthly thereafter in equal increments over 36 months, subject to continued service.
- (5) Our compensation committee, or board of directors, as applicable, approved the grant of these RSUs on February 22, 2019. The RSUs have time-based vesting, with one-third of the shares of common stock underlying the RSUs vesting on the first, second and third anniversaries of the grant date, subject to continued service.
- (6) Mr. Hoerter resigned as our chief commercial officer effective March 15, 2019. As a result, he did not receive a base salary increase or equity awards for 2019.

Salary increases for 2019 were made effective as of January 1, 2019. The annual cash incentive target award percentage for our named executive officers remained unchanged from 2018. Annual performance-based cash incentive program payouts for 2019 annual cash incentives will be based on our performance against specific research, clinical, operational and financial company goals and, as stated above, will be capped at 150% of the target payout level.

Severance and Change in Control Benefits

In 2016, our compensation committee adopted a Severance Plan, which applies to our named executive officers and certain other employees, which became effective as of April 22, 2016. The Severance Plan provides

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for severance benefits in the event of a termination of such named executive officer's employment by us without cause or by such employee for good reason either (i) before or more than 18 months after a change in control, or (ii) within 18 months following a change in control. The severance benefits set forth in the Severance Plan supersede the severance benefits and certain equity acceleration benefits set forth in employment offer letters with our named executive officers. Specifically, the Severance Plan eliminates single-trigger vesting on all equity grants made to our named executive officers after April 22, 2016, and provides that all unvested equity awards shall vest in full if a named executive officer's employment is terminated by an acquirer or us without cause or by such named executive officer for good reason, each within 18 months following a change in control. For equity awards that were made prior to April 22, 2016, the applicable terms, if any, of the award agreements and employment offer letter between such named executive officer and us shall continue to apply and provide that:

in the event of a termination of a named executive officer without cause or by a named executive officer for good reason not in connection with a change in control, then, subject to an effective release of claims against us, (i) Dr. Schenkein shall be entitled to receive accelerated vesting of 100% of such equity awards, and (ii) each of Drs. Biller and Bowden shall be entitled to receive accelerated vesting of 25% of the original number of shares such equity awards granted.

upon a change in control, and subject to an effective release of claims against us, each of Drs. Biller, Bowden and Schenkein will be entitled to accelerated vesting of 75% of the then unvested shares under any equity awards granted prior to April 22, 2016, with the remaining 25% of such awards continuing to vest in accordance with the vesting schedule for such awards. If a named executive officer is terminated without cause or leaves for good reason upon or within 18 months following the change in control, then his equity awards shall immediately vest and become exercisable in full.

Please refer to "Employment, Severance and Change in Control Arrangements" below for a more detailed discussion of severance and change in control benefits for our named executive officers. We also have provided estimates of the value of the severance payments made and other benefits provided to our named executive officers under specified termination circumstances under the caption "*Potential Payments Upon Termination or Change in Control*" below. We believe that providing these benefits helps us compete for executive talent. These benefits are designed to promote stability and continuity of our senior management and are intended to preserve employee morale and productivity and encourage retention in the face of the disruptive impact of an actual, threatened, or rumored change in control of the company.

Health and Welfare Benefits

Our named executive officers are eligible to participate in all of our employee benefit plans, including our medical, dental, vision, life and disability insurance plans, in each case on the same basis as other employees. We believe that these health and welfare benefits help ensure that we have a productive and focused workforce through reliable and competitive health and other benefits.

401(k) Retirement Plan

We maintain a 401(k) retirement plan that is intended to be a tax-qualified defined contribution plan under Section 401(k) of the Internal Revenue Code, or the Code. In general, all of our employees are eligible to participate, beginning on the first day of the month following commencement of their employment. The 401(k) plan includes a

salary deferral arrangement pursuant to which participants may elect to reduce their current compensation by up to the statutorily prescribed limit, equal to \$18,500 in 2018, and have the amount of the reduction contributed to the 401(k) plan. Participants who turned age 50 in 2018 were also eligible to make catch-up contributions, which in 2018 may be up to an additional \$6,000 above the statutory limit. In 2018, we

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matched employee 401(k) contributions at a rate of \$1.00 for each dollar contribution, up to 4% of eligible contributions. Matching contributions are 100% vested immediately.

Clawback Policy

Effective April 2016, we adopted a clawback policy which, in general, provides that, in the event that we are required to prepare an accounting restatement for periods ending on or after such date, we will make a reasonable attempt to recover from our current or former executive officers the pre-tax amount of certain incentive-based compensation in excess of what would have been paid to such executive officer after giving effect to the accounting restatement. For purposes of the policy, incentive-based compensation means any compensation that is granted, earned or vested based wholly or in part upon the attainment of any measures determined and presented in accordance with the accounting principles used in preparing the company's financial statements, or any measures derived wholly or in part from such financial information, stock price or total shareholder return. If the incentive-based compensation is based on our stock price or total shareholder return and the amount of excess incentive-based compensation is not calculable directly from the information in an accounting restatement, the amount recovered shall be based on a reasonable estimate of the effect of the accounting restatement on the stock price or total shareholder return upon which the incentive-based compensation was received. The policy shall be interpreted by our board of directors, or a duly established committee thereof.

Perquisites

We do not provide perquisites or personal benefits to our named executive officers.

Anti-Hedging and Pledging Policy

Our insider trading policy expressly prohibits all of our employees, including our named executive officers, as well as our directors, from engaging in speculative transactions in our stock, including short sales, puts/calls, hedging transactions and margin accounts or pledges.

No Tax Gross-ups

We do not provide for any tax gross-up payments to our named executive officers.

Accounting and Tax Considerations

We account for equity compensation paid to our employees under the rules of FASB Codification Topic 718, which rules require us to estimate and record an expense over the service period of any such award. Accounting rules also require us to record cash compensation as an expense at the time the obligation is accrued. To date, these accounting requirements have not impacted our executive compensation programs and practices.

Section 162(m) of the Internal Revenue Code generally disallows a tax deduction to public companies for compensation in excess of \$1 million paid in any taxable year to each of the company's chief executive officer, chief financial officer and three most highly compensated officers (other than the chief executive officer and chief financial officer). Historically, compensation paid to our chief financial officer and compensation that qualified under Section 162(m) as performance-based compensation was exempt from the deduction limitation. However, subject to certain transition rules, tax reform legislation signed into law on December 22, 2017,

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expanded the deduction limitation to apply to compensation in excess of \$1 million paid in any taxable year to our chief financial officer and eliminated the qualified performance-based compensation exception. As a result, for taxable years beginning after December 31, 2017, all compensation in excess of \$1 million paid to each of the executives described above (other than certain grandfathered compensation or compensation paid pursuant to certain equity awards granted during the transition period following our initial public offering) will not be deductible by us. Our board of directors and compensation committee reserve the right to use their business judgment to authorize compensation payments that may be subject to the limitations under Section 162(m) when the board or compensation committee, as applicable believe that compensation is appropriate and in the best interests of the company and our stockholders, after taking into consideration changing business conditions and performance of our employees.

Stock Ownership Guidelines

In 2016, our compensation committee established equity ownership guidelines for our directors and executive officers to further align the interests of our board of directors and named executive officers with those of stockholders. The equity ownership guidelines are as follows: our chief executive officer must own shares worth at least three times his or her base salary; our other executive officers must own shares worth at least their base salary; and our non-employee directors must own shares worth at least three times the annual cash retainer. Our chief executive officer, other executive officers and non-employee directors have five years from first being subject to these guidelines to satisfy the applicable ownership threshold.

Compensation Committee Report

The compensation committee has reviewed and discussed the Compensation Discussion and Analysis required by Item 402(b) of Regulation S-K with our management. Based on this review and discussion, the compensation committee recommended to our board of directors that the Compensation Discussion and Analysis be included in this proxy statement.

This report of the compensation committee is not soliciting material, shall not be deemed filed with the SEC and shall not be incorporated by reference by any general statement incorporating by reference this proxy statement into any filing under the Securities Act of 1933, as amended, or the Securities Exchange Act of 1934, as amended, whether made before or after the date hereof and irrespective of any general incorporation language in any such filing, except to the extent that we specifically incorporate this information by reference, and shall not otherwise be deemed filed under such acts.

The foregoing report has been furnished by the compensation committee.

Respectfully submitted,

The Compensation Committee of the Board of Directors

Kaye Foster (chair)

Ian T. Clark

John M. Maraganore

Table of Contents**Summary Compensation Table**

The following table shows information regarding the compensation of our named executive officers during the fiscal years ended December 31, 2018, 2017 and 2016.

| Name and Principal Position | Year | Salary (\$) | Bonus (\$) | Stock Awards (\$)(1) | Non-equity | | All Other Compensation (\$)(3) | Total (\$) |
|--|------|-------------|---------------|----------------------------|-----------------------------|---|--------------------------------------|------------|
| | | | | | Option Awards (\$)(1) | Incentive Plan Compensation (\$)(2) | | |
| David P. Schenkein, M.D.(4) <i>President and Chief Executive Officer</i> | 2018 | 625,000 | | 1,893,938 | 5,160,125 | 507,813 | 12,448 | 8,199,324 |
| | 2017 | 585,040 | | | 4,921,476 | 456,300 | 9,548 | 5,972,364 |
| | 2016 | 568,000 | | | 2,582,051 | 340,800 | 9,928 | 3,500,779 |
| Andrew Hirsch(5) <i>Chief Financial Officer and Head of Corporate Development</i> | 2018 | 471,329 | | 641,025 | 1,746,517 | 265,123 | 12,864 | 3,136,858 |
| | 2017 | 453,201 | | | 1,639,353 | 217,537 | 9,964 | 2,320,055 |
| | 2016 | 126,667 | | 747,450 | 4,186,746 | 176,000 | 698 | 5,237,561 |
| Christopher Bowden, M.D. <i>Chief Medical Officer</i> | 2018 | 467,141 | | 641,025 | 1,746,517 | 262,767 | 11,280 | 3,128,730 |
| | 2017 | 449,174 | | | 1,639,353 | 233,571 | 8,710 | 2,330,808 |
| | 2016 | 436,091 | | | 1,145,987 | 174,440 | 9,103 | 1,765,621 |
| Scott Biller, Ph.D. <i>Chief Scientific Officer</i> | 2018 | 446,573 | | 641,025 | 1,746,517 | 251,198 | 12,741 | 3,098,054 |
| | 2017 | 429,397 | | | 1,639,353 | 223,287 | 9,840 | 2,301,877 |
| | 2016 | 416,890 | | | 1,145,987 | 166,760 | 10,221 | 1,739,858 |
| Steve Hoerter(6) <i>Chief Commercial Officer</i> | 2018 | 444,549 | | 641,025 | 1,746,517 | 250,059 | 14,578 | 3,096,728 |
| | 2017 | 427,451 | | | 1,639,353 | 205,177 | 9,397 | 2,281,378 |
| | 2016 | 363,125 | 375,000 | 596,400 | 2,527,808 | 166,000 | 9,549 | 4,037,882 |

- (1) Amounts listed represent the aggregate fair value amount computed as of the grant date of the awards granted in accordance with FASB ASC Topic 718. Assumptions used in the calculation of these amounts are included in Note 12, Share-Based Payments, of the Notes to our Consolidated Financial Statements filed on our Annual Report on Form 10-K, filed with the SEC on February 14, 2019.
- (2) Amounts represent awards to our named executive officers under our annual performance-based cash incentive program. See *Annual Performance-based Cash Incentives* for a description of that program. Annual cash incentive compensation earned during the year is typically paid in the following year.
- (3) Amounts include a matching contribution to the company's 401(k) plan and the dollar value of company-paid life insurance and disability insurance premiums paid during the fiscal year for the named executive officer. For 2018, amounts include company matching 401(k) contributions of \$11,000 for Dr. Schenkein, Mr. Hirsch, Dr. Biller and Mr. Hoerter. In the case of Mr. Hoerter, for 2018 the amount also includes \$2,200 of qualified non-elective employer contribution to the company's 401(k) plan.

- (4) Dr. Schenkein also serves as a member of our board of directors but does not receive any additional compensation for his service as a director. Dr. Schenkein transitioned from our president and chief executive officer to executive chairman effective February 1, 2019.

- (5) Mr. Hirsch's 2016 annual base salary was \$440,000; however, as Mr. Hirsch joined the company as chief financial officer in September 2016, his base salary reported was pro-rated in 2016. In addition, Mr. Hirsch received a stock option award and performance share units upon hire.

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- (6) Mr. Hoerter's 2016 annual base salary was \$415,000; however, as Mr. Hoerter joined the company as chief commercial officer in February 2016, his base salary reported was pro-rated in 2016. In addition, Mr. Hoerter received a sign-on bonus of \$375,000 and received a stock option award and performance share units upon hire. Mr. Hoerter resigned as our chief commercial officer, effective March 15, 2019.

Pay Ratio Disclosure

As required by Section 953(b) of the Dodd-Frank Wall Street Reform and Consumer Protection Act and Item 402(u) of Regulation S-K, we are required to disclose the ratio of our median employee's annual total compensation to the annual total compensation of our principal executive officer.

The purpose of this new disclosure is to provide a measure of the equitability of pay within our company. We believe our compensation philosophy and process yield an equitable result for all of our employees. During the fiscal year ended December 31, 2018, our principal executive officer was our president and chief executive officer, David P. Schenkein, M.D. For the fiscal year ended December 31, 2018, the annual total compensation for Dr. Schenkein, as reported in the Summary Compensation Table above, was \$8,199,324. For the fiscal year ended December 31, 2018, the annual total compensation for our median employee (identified as disclosed below) was \$345,181, resulting in a pay ratio of approximately 24:1.

Consistent with the process established to identify our median employee for 2017, we identified the median employee as of October 1, 2018 by (i) aggregating for each applicable employee (A) annual base salary for salaried employees (or hourly rate multiplied by expected annual work schedule, for hourly employees), (B) target bonus for 2018, and (C) the estimated fair value of any equity awards granted during the fiscal year ended December 31, 2018 and, (ii) ranking this aggregated compensation measure for our employees from lowest to highest. This calculation was performed for all employees, excluding Dr. Schenkein.

The pay ratio reported above is a reasonable estimate calculated in a manner consistent with SEC rules based on our internal records and the methodology described above. The SEC rules for identifying the median compensated employee and calculating the pay ratio based on that employee's annual total compensation allow companies to adopt a variety of methodologies, to apply certain exclusions, and to make reasonable estimates and assumptions that reflect their employee populations and compensation practices. Therefore, the pay ratio reported by other companies may not be comparable to the pay ratio reported above, as other companies have different employee populations and compensation practices and may utilize different methodologies, exclusions, estimates and assumptions in calculating their own pay ratios.

Table of Contents**Grants of Plan-Based Awards for 2018**

The following tables sets forth information concerning each grant of an award made to a named executive officer during the fiscal year ended December 31, 2018 under any plan, contract, authorization or arrangement pursuant to which cash, securities, similar instruments or other property may be received:

| Name | Grants of Plan-Based Awards | | | | | | | |
|--------------------------------|-----------------------------|----------------|-------------|--------------|--|-------------------------------|---|--|
| | Date of Grant | Threshold (\$) | Target (\$) | Maximum (\$) | All Other Stock Awards: Number of Shares of Stock or Units (#) | Exercise Price of Awards (\$) | All Other Option Awards: Number of Securities Underlying Options (#)(2) | Grant Date Fair Value of Stock and Option Awards (\$)(2) |
| David P. Schenkein, M.D.(3)(4) | | 406,250 | 609,375 | | | | | |
| | 2/16/2018 | | | | 24,375(5) | | | 1,893,938 |
| | 2/16/2018 | | | | | 77.70 | 97,500(6) | 5,160,125 |
| Andrew Hirsch(3) | | 212,098 | 318,147 | | | | | |
| | 2/16/2018 | | | | 8,250(5) | | | 641,025 |
| | 2/16/2018 | | | | | 77.70 | 33,000(6) | 1,746,517 |
| Christopher Bowden, M.D.(3) | | 210,213 | 315,320 | | | | | |
| | 2/16/2018 | | | | 8,250(5) | | | 641,025 |
| | 2/16/2018 | | | | | 77.70 | 33,000(6) | 1,746,517 |
| Scott Biller, Ph.D.(3) | | 200,958 | 301,437 | | | | | |
| | 2/16/2018 | | | | 8,250(5) | | | 641,025 |
| | 2/16/2018 | | | | | 77.70 | 33,000(6) | 1,746,517 |
| Steve Hoerter(3)(7) | | 200,047 | 300,071 | | | | | |
| | 2/16/2018 | | | | 8,250(5) | | | 641,025 |
| | 2/16/2018 | | | | | 77.70 | 33,000(6) | 1,746,517 |

(1) Amounts shown in the threshold, target and maximum columns reflect the minimum, target and maximum amounts, respectively, payable under our annual incentive cash program as described above under Annual Performance-based Cash Incentives. Actual amounts paid are presented in the Summary Compensation Table above.

(2) Amounts listed represent the aggregate fair value amount computed as of the grant date of the awards granted in accordance with FASB ASC Topic 718. Assumptions used in the calculation of these amounts are included in Note 12, Share-Based Payments, of the Notes to our Consolidated Financial Statements filed on our Annual Report on Form 10-K, filed with the SEC on February 14, 2019.

- (3) For information on vesting acceleration upon termination of employment, see the Employment, Severance and Change in Control Arrangements section below.
- (4) Dr. Schenkein transitioned from our president and chief executive officer to executive chairman effective February 1, 2019.
- (5) On February 16, 2018, our compensation committee and the board of directors, as applicable, approved the grant of these RSUs under the 2013 Stock Incentive Plan. The RSUs are subject to time-based vesting, with one-third of the RSUs vesting each year beginning on the first anniversary of the grant date and ending on the third anniversary of the grant date, subject to continued service.
- (6) On February 16, 2018, our compensation committee, or board of directors, as applicable, approved the grant of these stock option awards under the 2013 Stock Incentive Plan at an exercise price of \$77.70 per share, the closing price of our common stock on the date of grant. The options are subject to time-based

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vesting, with 25% of the shares underlying the award vesting on the first anniversary of the grant date and the remaining shares vesting monthly thereafter in equal increments over 36 months, subject to continued service.

(7) Mr. Hoerter resigned as our chief commercial officer, effective March 15, 2019.

Outstanding Equity Awards at Fiscal Year-End

The following table sets forth information concerning outstanding equity awards for each of our named executive officers at December 31, 2018:

| Name(1) | Grant Date | Option Awards Equity Incentive Plan | | | | Option Expiration Date | Stock Awards | |
|-----------------------------|------------|---|---|---|---|------------------------|---|--|
| | | Number of Securities Underlying Unexercised Options Exercisable (#) | Number of Securities Underlying Unexercised Options Exercisable (#) | Number of Securities Underlying Unexercised Options Exercisable (#) | Number of Securities Underlying Unexercised Options Exercisable (#) | | Number of shares or units of stock that have not vested (#) | Market value of shares or units of stock that have not vested (#)(2) |
| David P. Schenkein, M.D.(3) | 04/30/2013 | 136,363 | - | - | \$ 9.05 | 04/29/2023 | | |
| | 03/05/2014 | 137,471 | - | - | \$ 31.64 | 03/04/2024 | | |
| | 03/02/2015 | 127,498 | 8,502(4) | - | \$ 107.89 | 03/01/2025 | | |
| | 02/16/2016 | 72,250 | 29,750(4) | - | \$ 39.76 | 02/15/2026 | | |
| | 02/21/2017 | 65,495 | 77,405(4) | - | \$ 50.40 | 02/20/2027 | | |
| | 02/16/2018 | - | 97,500(4) | - | \$ 77.70 | 02/15/2028 | | |
| | 02/16/2018 | - | - | - | - | - | 24,375(5) | 1,123,931 |
| Andrew Hirsch | 09/20/2016 | 70,310 | 54,690(4) | - | \$ 49.83 | 09/19/2026 | | |
| | 09/20/2016 | - | - | - | - | - | 15,000(6) | 691,650 |
| | 02/21/2017 | 21,810 | 25,790(4) | - | \$ 50.40 | 2/20/2027 | | |
| | 02/16/2018 | - | 33,000(4) | - | \$ 77.70 | 2/15/2028 | | |
| | 02/16/2018 | - | - | - | - | - | 8,250(5) | 380,408 |
| Christopher Bowden, M.D. | 05/31/2014 | 91,000 | - | - | \$ 35.16 | 05/30/2024 | | |
| | 03/02/2015 | 15,935 | 1,065(4) | - | \$ 107.89 | 03/01/2025 | | |
| | 12/21/2015 | - | - | - | - | - | 6,265(7) | 288,879 |
| | 02/16/2016 | 32,029 | 13,191(4) | - | \$ 39.76 | 02/15/2026 | | |
| | 02/21/2017 | 21,810 | 25,790(4) | - | \$ 50.40 | 2/20/2027 | | |
| | 02/16/2018 | - | 33,000(4) | - | \$ 77.70 | 02/15/2028 | | |
| | 02/16/2018 | - | - | - | - | - | 8,250(5) | 380,408 |
| Scott Biller, Ph.D. | 12/07/2010 | 1,766 | - | - | \$ 0.47 | 12/06/2020 | | |

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| | | | | | | | | |
|------------------|------------|--------|-----------|---|-----------|------------|-----------|---------|
| | 04/06/2012 | 7,218 | - | - | \$ 2.34 | 04/05/2022 | | |
| | 04/30/2013 | 4,154 | - | - | \$ 9.05 | 04/29/2023 | | |
| | 03/05/2014 | 14,862 | - | - | \$ 31.64 | 03/04/2024 | | |
| | 03/02/2015 | 63,749 | 4,251(4) | - | \$ 107.89 | 03/01/2025 | | |
| | 02/16/2016 | 32,029 | 13,191(4) | - | \$ 39.76 | 02/15/2026 | | |
| | 02/21/2017 | 21,810 | 25,790(4) | - | \$ 50.40 | 2/20/2027 | | |
| | 02/16/2018 | - | 33,000(4) | - | \$ 77.70 | 02/15/2028 | | |
| | 02/16/2018 | - | - | - | - | - | 8,250(5) | 380,408 |
| Steve Hoerter(7) | | | | | | | | |
| | 02/16/2016 | 31,476 | 29,174(4) | - | \$ 39.76 | 02/15/2026 | | |
| | 02/16/2016 | - | - | - | - | - | 15,000(9) | 691,650 |
| | 02/21/2017 | 21,810 | 25,790(4) | - | \$ 50.40 | 2/20/2027 | | |
| | 02/16/2018 | - | 33,000(4) | - | \$ 77.70 | 02/15/2028 | | |
| | 02/16/2018 | - | - | - | - | - | 8,250(5) | 380,408 |

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- (1) For information on equity acceleration benefits under specified circumstances, see Employment, Severance and Change in Control Arrangements.
- (2) Amounts listed represent the aggregate fair value amount computed as of the grant date of the awards granted in accordance with FASB ASC Topic 718. Assumptions used in the calculation of these amounts are included in Note 12, Share-Based Payments, of the Notes to our Consolidated Financial Statements filed on our Annual Report on Form 10-K, filed with the SEC on February 14, 2019.
- (3) Dr. Schenkein transitioned from the role of president and chief executive officer to executive chairman effective February 1, 2019.
- (4) The shares vest with 25% of the shares underlying the option vesting on the first anniversary of the grant date and the remaining shares vesting monthly thereafter in equal increments over 36 months, subject to continued service.
- (5) One third of the RSUs vest each year, beginning on the first anniversary of the grant date, subject to continued service. Each RSU represents a contingent right to receive one share of our common stock upon vesting.
- (6) On September 20, 2016, our compensation committee approved the grant of 15,000 performance share units to Mr. Hirsch in connection with the start of his employment. The performance-based vesting criteria were satisfied on July 20, 2018, and the performance share units vested in full on January 20, 2019. Each performance share unit represents a contingent right to receive one share of our common stock upon vesting.
- (7) On December 3, 2015, our compensation committee approved, effective December 21, 2015, the grant of 6,265 performance share units to Dr. Bowden to reflect a market adjustment to his equity position in the company. The performance-based vesting criteria were satisfied on July 20, 2018, and the performance share units vested in full on January 20, 2019. Each performance share unit represents a contingent right to receive one share of our common stock upon vesting.
- (8) Mr. Hoerter resigned as our chief commercial officer, effective March 15, 2019.
- (9) On February 15, 2016, our compensation committee approved, effective February 16, 2016, the grant of 15,000 performance share units to Mr. Hoerter in connection with the start of his employment. The performance-based vesting criteria were satisfied on July 20, 2018, and the performance share units vested in full on January 20, 2019. Each performance share unit represents a contingent right to receive one share of our common stock upon vesting.

Option Exercises and Stock Vested in 2018

The following table sets forth information concerning option exercises and stock vested for each of our named executive officers during the fiscal year ended December 31, 2018:

| | Option Awards | |
|-----------------------------|---|--|
| | Number of Shares Acquired on Exercise(#) | Value Realized on Exercise\$(1) |
| David P. Schenkein, M.D.(2) | 366,849 | 29,813,191 |
| Andrew Hirsch | | |
| Christopher Bowden, M.D. | 18,000 | 866,895 |
| Scott Biller, Ph.D. | 72,814 | 4,386,059 |
| Steve Hoerter(3) | 39,350 | 1,519,938 |

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- (1) The value realized when the stock options were exercised represents the excess of the fair market value of the shares at the time of exercise over the exercise price of the stock options.
- (2) Dr. Schenkein transitioned from the role of president and chief executive officer to executive chairman effective February 1, 2019.
- (3) Mr. Hoerter resigned as our chief commercial officer, effective March 15, 2019.

Employment, Severance and Change in Control Arrangements

Severance Benefits Plan

In 2016, our compensation committee adopted a Severance Benefits Plan, or the Severance Plan, which applies to our named executive officers and certain other officers and key employees. The Severance Plan provides for severance benefits in the event of a termination of such named executive officer's employment by us without cause or by such employee for good reason either (i) before or more than 18 months after a change in control, or (ii) within 18 months following a change in control. Except as specifically provided below, the severance benefits set forth in the Severance Plan supersede any severance benefits set forth in award agreements and/or employment offer letters with such named executive officers.

Benefits Provided Upon Termination Not in Connection with a Change in Control. Under the terms of the Severance Plan, subject to the execution and effectiveness of a release of claims against us, if a named executive officer's employment is terminated by us without cause or by such named executive officer for good reason prior to or more than 18 months following a change in control:

- (i) we are obligated (A) to pay an amount equal to his then-current monthly base salary for a period of 12 months and 100% of such named executive officer's target annual cash incentive in a lump sum, and (B) subject to certain exceptions, to contribute to the cost of COBRA coverage for health and dental insurance on the same basis as our contribution to company-provided health and dental insurance coverage in effect for active employees with the same coverage elections for a period of 12 months; and
- (ii) there will be no vesting acceleration for any equity award made to our named executive officers on or after April 22, 2016, the effective date of the Severance Plan, and for any equity awards granted prior to the effective date of the Severance Plan, the treatment of such equity awards shall be dictated by the applicable terms, if any, of the award agreements and/or employment offer letter between such named executive officer and us.

Benefits Provided Upon Termination in Connection with a Change in Control. Under the terms of the Severance Plan, subject to the execution and effectiveness of a release of claims against us, if a named executive officer's employment is terminated by us without cause or by such named executive officer for good reason within 18 months following a change in control:

- (i)

we are obligated (A) to pay an amount equal to his then-current monthly base salary for a period of 12 months and 100% of such named executive officer's target annual cash incentive in a lump sum, with the exception of Dr. Schenkein, who will be entitled to an amount equal to his then-current monthly base salary for a period of 24 months and 200% of his target annual cash incentive in a lump sum, and (B) subject to certain exceptions, to contribute to the cost of COBRA coverage for health and dental insurance on the same basis as our contribution to company-provided health and dental insurance coverage in effect for active employees with the same coverage elections for a period of 12 months, with the exception of Dr. Schenkein, who will be entitled to such insurance for a period of 24 months; and

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- (ii) any unvested equity awards shall become fully vested; provided that the treatment for such named executive officers' equity awards granted prior to the effective date of the Severance Plan shall be governed by the applicable terms, if any, of the award agreements and/or employment offer letter between such named executive officer and us.

For a discussion of the vesting acceleration for equity awards made to Drs. Schenkein, Biller and Bowden prior to April 22, 2016, the effective date of the Severance Plan, see *Employment Offer Letters*, below.

Employment Offer Letters

We have entered into employment offer letters with each of our named executive officers pursuant to which such named executive officer is employed at will, meaning each named executive officer or we may terminate the employment arrangement at any time. Such offer letters establish the named executive officer's title, initial compensation arrangements, and eligibility for benefits made available to employees generally.

In connection with his appointment as executive chairman, we entered into an amended and restated employee offer letter agreement with Dr. Schenkein, effective February 1, 2019, providing for the modified terms and conditions of his employment, including (i) an annual base salary of \$175,000; (ii) a pro-rated bonus in respect of the one-month period in 2019 that he served as our chief executive officer; (iii) a one-time grant of an option to purchase 45,719 shares of the Company's common stock, exercisable at a price per share equal to \$58.86, the closing price on the date of grant, which shall vest as to 25% of the underlying shares on the first anniversary of the date of grant and as to the remaining shares on a monthly basis thereafter; (iv) a one-time grant of 8,324 restricted stock units, each unit representing a right to receive one share of the Company's common stock, which shall vest in equal annual installments on the first, second and third anniversaries of the date of grant; (v) for any calendar year after 2019 during which he serves as executive chairman, an annual grant of equity awards with an aggregate value equal to 35% of the aggregate value of equity awards made in the ordinary course in such year to the then current chief executive officer; and (vi) severance benefits in accordance with the Severance Plan, unless his employment ends for any reason effective as of either February 1, 2020 or February 1, 2021, and except with regard to those equity awards that were made prior to April 22, 2016, which are nonetheless subject to the terms of Dr. Schenkein's original offer letter..

The terms of employment offer letters with each of Drs. Schenkein, Biller and Bowden provide for any equity awards that were made prior to April 22, 2016, the effective date of the Severance Plan, to accelerate as follows:

- (i) in the case of Dr. Schenkein (A) in the event of a change in control, 75% of his unvested shares shall vest immediately, and 25% of his unvested shares shall continue to vest, but shall be subject to immediate vesting in full if Dr. Schenkein is terminated without cause or resigns for good reason upon or within 18 months following such change in control, and (B) in the event that he is terminated without cause or resigns for good reason prior to a change in control, then all of his then unvested shares shall immediately vest in full; and
- (ii) in the case of Drs. Biller and Bowden, (A) in the event of a change in control, 75% of such officer's unvested shares shall vest immediately, and 25% of such officer's unvested shares shall continue to vest, but shall be subject to immediate vesting in full if such officer is terminated without cause or resigns for good reason upon or within 18 months following such change in control, and (B) in the event that such officer is terminated without cause or resigns for good reason prior to a change in control, then 25% of the original number of shares underlying such officer's equity awards shall immediately vest in full.

In connection with her appointment as chief executive officer, we entered into an employee offer letter agreement with Dr. Fouse, effective February 1, 2019 providing for the terms of her employment, including (i) an

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annual base salary of \$725,000; (ii) an annual target bonus equal to 65% of her base salary; (iii) a one-time grant of an option to purchase 350,346 shares of common stock at an exercise price of \$53.87 per share, which shall vest as to 25% of the underlying shares on February 1, 2020 and as to the remaining shares on a monthly basis thereafter; (iv) a one-time grant of 42,695 performance share units, each unit representing a contingent right to receive one share of common stock if, during the five-year period beginning on February 1, 2019 and ending on February 1, 2024, the closing price of our common stock equals or exceeds \$120 per share for at least 20 consecutive trading days at any time during such five-year period; and (v) severance benefits in accordance our Severance Plan, described below.

Other Agreements

We have entered into non-competition, non-solicitation, confidentiality and assignment agreements with each of our named executive officers. Under the non-competition, non-solicitation, confidentiality and assignment agreements, each named executive officer has agreed (i) not to compete with us during such officer's employment and for a period of one year after the termination of such officer's employment, (ii) not to solicit our employees or customers during his employment and for a period of one year after the termination of such officer's employment, (iii) to protect our confidential and proprietary information, and (iv) to assign to us related intellectual property that is developed during the course of such officer's employment and for a period of six months after the termination of such officer's employment, that results from tasks assigned by us or that results from the use of our property, premises, or confidential information.

Potential Payments Upon Termination or Change in Control

In 2016, our compensation committee adopted the Severance Plan, which applies to our named executive officers. The Severance Plan provides for severance benefits in the event of a termination of such named executive officer's employment by us without cause or by such employee for good reason, either (i) before or more than 18 months after a change in control, or (ii) within 18 months following a change in control. Receipt of any severance benefits under the Severance Plan require that the named executive officer comply with the provisions of any applicable non-competition, non-solicitation, and other obligations to us; and (b) execute and deliver a suitable severance agreement and release under which the named executive officer releases and discharges us and our affiliates from and on account of any and all claims between us and the named executive officer.

The following table sets forth potential payments upon termination and change in control that would be made to our named executive officers, assuming that such termination or change in control occurred on December 31, 2018, after giving effect to the Severance Plan. In addition to the amounts shown in the table below, each named executive officer would be entitled to receive payments for base salary through the date of termination and payment for any reimbursable business expenses incurred.

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| Name | Benefit | Change in Control (Without Termination of Employment) (\$) | Triggering Event | |
|--------------------------|-------------------------------------|--|---|--|
| | | | Resignation For Good Reason or Termination Without Cause Before or More Than 18 Months Following a Change In Control (\$) | Resignation For Good Reason or Termination Without Cause Upon or Within 18 Months Following a Change-in- Control (\$) |
| David P. Schenkein, M.D. | Severance Payments | | 625,000(1) | 1,250,000(7) |
| | Bonus Payment | | 406,250(2) | 812,500(8) |
| | Continuation of Benefits | | 14,229(3) | 28,458(9) |
| | Market Value of Stock Vesting(4) | 141,684(5) | 188,913(11) | 1,312,844(6) |
| | Total | 141,684 | 1,234,392 | 3,403,802 |
| Andrew Hirsch | Severance Payments | | 471,329(1) | 471,329(1) |
| | Bonus Payment | | 212,098(2) | 212,098(2) |
| | Continuation of Benefits | | 21,397(3) | 21,397(3) |
| | Market Value of Stock Vesting(4) | | | 1,072,058(6) |
| | Total | | 704,824 | 1,776,882 |
| Christopher Bowden, M.D. | Severance Payments | | 467,141(1) | 467,141(1) |
| | Bonus Payment | | 210,213(2) | 210,213(2) |
| | Continuation of Benefits | | 21,397(3) | 21,397(3) |
| | Market Value of Stock Vesting(4) | 279,482(5) | 144,007(10) | 753,050(6) |
| | Total | 279,482 | 842,758 | 1,451,801 |
| Scott Biller, Ph.D. | Severance Payments | | 446,573(1) | 446,573(1) |
| | Bonus Payment | | 200,958(2) | 200,958(2) |
| | Continuation of Benefits | | 14,229(3) | 14,229(3) |
| | Market Value of Stock Vesting(4) | 62,822(5) | 71,787(10) | 464,170(6) |
| | Total | 62,822 | 733,547 | 1,125,930 |
| Steve Hoerter | Severance Payments | | 444,549(1) | 444,549(1) |
| | Bonus Payment | | 200,047(2) | 200,047(2) |
| | Continuation of Benefits | | 21,397(3) | 21,397(3) |
| | Market Value of Stock Vesting(4) | | | 1,257,312(6) |
| | Total | | 665,993 | 1,923,305 |

- (1) Represents 12 monthly payments of each executive's monthly base salary from the time of termination.
- (2) Represents a lump sum payment equal to each executive's target annual cash incentive bonus.
- (3) Represents the cost of continued health and dental benefits. These benefits are payable until 12 months following termination.
- (4) These awards would become vested and the value of the acceleration would be equal to (i), in the case of options, the shares subject to unvested options multiplied by the excess of the then current stock price over the exercise price of the options and (ii), in the case of RSUs, the number of unvested RSUs multiplied by the then current stock price. For purposes of this table, we have calculated the value of the acceleration using the closing price of our common stock on December 31, 2018, or \$46.11 per share.
- (5) Represents the acceleration of vesting as to 75% of the unvested equity awards held by the named executive officer which were granted prior to April 22, 2016.

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- (6) Represents the acceleration of vesting as to 100% of the unvested equity awards held by the named executive officer.
- (7) Represents 24 monthly payments of executive's monthly base salary from the time of termination.
- (8) Represents a lump sum payment equal to two years of executive's target annual cash incentive bonus.
- (9) Represents the cost of continued health and dental benefits. These benefits are payable until 24 months following termination.
- (10) Represents the acceleration of vesting as to 25% of the original equity awards held by the executive which were granted prior to April 22, 2016 (or if the number of unvested shares subject to such equity award is less than 25% of the original number of shares subject to such equity award, then all remaining unvested shares subject to such equity award shall fully vest). This acceleration of vesting only occurs if the named executive officer resigns for good reason or is terminated without cause before a change in control.
- (11) Represents the acceleration of vesting as to 100% of the unvested equity awards held by the named executive officer which were granted prior to April 22, 2016. This acceleration of vesting only occurs if the named executive officer resigns for good reason or is terminated without cause before a change in control.

Securities Authorized for Issuance Under Our Equity Compensation Plans

The following table provides information about the securities authorized for issuance under our equity compensation plans as of December 31, 2018.

Equity Compensation Plan Information

| Plan category | Number of securities to be issued upon exercise of outstanding options, warrants and rights (a) | Weighted-average exercise price of outstanding options, warrants and rights (b) | Number of securities remaining available for future issuance under equity compensation plans (excluding securities reflected in column (a)) (c) |
|---------------|--|--|--|
| | | | |

| | | | |
|--|-----------|----|------------|
| Equity compensation plans approved by security holders | | | |
| 2007 Stock Incentive Plan | 383,427 | \$ | 4.45 |
| 2013 Stock Incentive Plan | 5,773,817 | \$ | 64.34(1) |
| 2013 Employee Stock Purchase Plan | | | 160,536(3) |
| Equity compensation plans not approved by security holders | | | |
| Total | 6,117,244 | \$ | 60.10 |
| | | | 2,017,017 |

- (1) The calculation does not take into account the 532,144 shares of common stock subject to outstanding restricted stock units or the 169,031 shares of common stock subject to outstanding performance-based stock units. Such shares will be issued at the time such awards vest, without any cash consideration payable for those shares.

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- (2) Our 2013 Stock Incentive Plan, or 2013 Plan, has an evergreen provision that allows for an annual increase in the number of shares available for issuance under the 2013 Plan to be added on the first day of each fiscal year, beginning with the fiscal year ending December 31, 2014 and continuing until the expiration of the 2013 Plan, equal to the least of 2,000,000 shares of our common stock, 4% of the number of shares of our common stock outstanding on the first day of the applicable fiscal year or an amount determined by our board of directors. On January 1, 2019, 2,000,000 additional shares were reserved for issuance under the 2013 Plan pursuant to this provision.
- (3) Our 2013 Employee Stock Purchase Plan, or 2013 ESPP, has an evergreen provision that allows for an annual increase in the number of shares available for issuance under the 2013 ESPP to be added on the first day of each fiscal year, beginning on January 1, 2014 and ending on December 31, 2023, in an amount equal to the least of 509,091 shares of our common stock, 1% of the total number of shares of our common stock outstanding on the first day of the applicable fiscal year or an amount determined by our board of directors. The number of shares reserved for issuance under the 2013 ESPP has not increased since the adoption of the 2013 ESPP.

Compensation Committee Interlocks and Insider Participation

For 2018, the members of our compensation committee were Ms. Foster (chair), Mr. Clark, and Dr. Maraganore, none of whom is, or ever has been, an officer or employee of our company. None of our executive officers served as a director or a member of a compensation committee (or other committee serving an equivalent function) of any other entity, one of whose executive officers served as a director or member of our compensation committee during the fiscal year ended December 31, 2018.

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CERTAIN RELATIONSHIPS AND RELATED PARTY TRANSACTIONS

Since January 1, 2018, we have engaged in the following transactions with our directors, director nominees, executive officers and holders of more than 5% of our voting securities (or their immediate family members), and affiliates of our directors, executive officers and 5% stockholders. We believe that all of the transactions described below were made on terms no less favorable to us than could have been obtained from unaffiliated third parties.

Celgene Corporation

Based solely on a Schedule 13D/A filed with the SEC on January 23, 2018, entities affiliated with Celgene Corporation, or Celgene, beneficially own more than 5% of our outstanding shares of common stock.

Public Offerings

In January 2018, we completed a public offering of 7,089,553 shares of common at a public offering price of \$67.00 per share. We received net proceeds from this offering of \$448.9 million, after deducting underwriting discounts and commissions paid by us. An affiliate of Celgene purchased 851,154 shares of our common stock in the offering for approximately \$57.0 million. In addition, we granted the underwriters the right to purchase up to an additional 1,063,433 shares of common stock, which was exercised in January 2018, resulting in additional net proceeds to us of \$67.3 million, after underwriting discounts and commissions, for total net proceeds of approximately \$516.2 million.

Termination of AG-881 Agreements

In September 2018, we and Celgene agreed to terminate our joint worldwide collaboration focused on the development and commercialization of vorasidenib products, or the AG-881 Agreements, effective as of September 4, 2018. From and after September 4, 2018, we obtained sole global rights to vorasidenib. Neither we nor Celgene will have any further financial obligation under the AG-881 Agreements, including milestones, royalties or other payments, except that (a) Celgene shall be eligible to receive royalties from us at a low single-digit percentage rate on worldwide net sales of products containing vorasidenib and (b) we and Celgene split certain agreed-upon worldwide development costs for vorasidenib until December 31, 2018 (with approximately \$1.34 million in development costs covered by Celgene from September 2018 to December 2018). In addition, for a specified period and subject to specified exceptions, Celgene and its affiliates shall be prohibited from developing, manufacturing or commercializing any product that inhibits IDH1 at specified levels of binding for any indication and we shall be prohibited from developing, manufacturing or commercializing vorasidenib in hematologic indications.

2010 Agreement

In April 2010, we entered into a collaboration agreement with Celgene. Under the terms of the 2010 Agreement, we are eligible to receive up to \$80.0 million in potential remaining milestone payments for our IDHIFA® program. The remaining potential milestone payments are comprised of: (i) up to \$55.0 million in milestone payments upon achievement of specified ex-U.S. regulatory milestone events, of which \$35.0 million relates to the first regulatory approval in any of China, Japan or a major European country, and (ii) a \$25.0 million milestone payment upon achievement of a specified ex-U.S. commercial milestone event. Pursuant to the 2010 Agreement, we may also receive royalties at tiered, low-double digit to mid-teen percentage rates on net sales of IDHIFA®. During the year ended December 31, 2018, we received a \$15.0 million milestone payment in connection with Celgene's filing of an MAA for IDHIFA® and earned \$7.2 million in royalty revenue under the 2010 Agreement.

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Effective as of August 2016, we and Celgene agreed to terminate the 2010 Agreement as to the program directed to the IDH1 target, for which ivosidenib is the lead development candidate. As a result, neither party has any further financial obligation, including royalties or milestone payments, to the other concerning ivosidenib or the IDH1 program.

Policies and Procedures for Related Party Transactions

We have adopted written policies and procedures for the review of any transaction, arrangement or relationship in which we are a participant, the amount involved exceeds \$120,000, and one of our executive officers, directors, director nominees or 5% stockholders (or their immediate family members), each of whom we refer to as a related person, has a direct or indirect material interest.

If a related person proposes to enter into such a transaction, arrangement or relationship, which we refer to as a related person transaction, the related person must report the proposed related person transaction to our principal financial officer. The policy calls for the proposed related person transaction to be reviewed and, if deemed appropriate, approved by the audit committee. Whenever practicable, the reporting, review and approval will occur prior to entry into the transaction. If advance review and approval is not practicable, the committee will review, and, in its discretion, may ratify the related person transaction. The policy also permits the chairman of the committee to review and, if deemed appropriate, approve proposed related person transactions that arise between committee meetings, subject to ratification by the committee at its next meeting. Any related person transactions that are ongoing in nature will be reviewed annually.

A related person transaction reviewed under the policy will be considered approved or ratified if it is authorized by the committee after full disclosure of the related person's interest in the transaction. As appropriate for the circumstances, the committee will review and consider:

the related person's interest in the related person transaction;

the approximate dollar value of the amount involved in the related person transaction;

the approximate dollar value of the amount of the related person's interest in the transaction without regard to the amount of any profit or loss;

whether the transaction was undertaken in the ordinary course of our business;

whether the terms of the transaction are no less favorable to us than terms that could have been reached with an unrelated third party;

the purpose of, and the potential benefits to us of, the transaction; and

any other information regarding the related person transaction or the related person in the context of the proposed transaction that would be material to investors in light of the circumstances of the particular transaction.

The committee may approve or ratify the transaction only if the committee determines that, under all of the circumstances, the transaction is in or is not inconsistent with our best interests. The committee may impose any conditions on the related person transaction that it deems appropriate.

In addition to the transactions that are excluded by the instructions to the SEC's related person transaction disclosure rule, our board of directors has determined that the following transactions do not create a

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material direct or indirect interest on behalf of related persons and, therefore, are not related person transactions for purposes of this policy:

interests arising solely from the related person's position as an executive officer of another entity (whether or not the person is also a director of such entity), that is a participant in the transaction, where (a) the related person and all other related persons own in the aggregate less than a 10% equity interest in such entity, (b) the related person and their immediate family members are not involved in the negotiation of the terms of the transaction and do not receive any special benefits as a result of the transaction, (c) the amount involved in the transaction equals less than the greater of \$1 million dollars or 2% of the annual consolidated gross revenues of the other entity that is a party to the transaction, and (d) the amount involved in the transaction equals less than 2% of our annual consolidated gross revenues; and

a transaction that is specifically contemplated by provisions of our charter or bylaws.

Our related persons transaction policy provides that transactions involving compensation of executive officers shall be reviewed and approved by the compensation committee in the manner specified in its charter.

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PROPOSAL 2 ADVISORY VOTE ON EXECUTIVE COMPENSATION

We are providing our stockholders the opportunity to vote to approve, on an advisory, non-binding basis, the compensation of our named executive officers as disclosed in this Proxy Statement in accordance with Section 14A of the Exchange Act. This proposal, which is commonly referred to as "say-on-pay," is required by the Dodd-Frank Wall Street Reform, or the Dodd-Frank Act, and Consumer Protection Act of 2010, which added Section 14A to the Exchange Act. Our executive compensation program is designed to reward value creation for stockholders and to attract, motivate, and retain our executive officers, who are critical to our success. Under this program, our named executive officers are rewarded for the achievement of our short- and long-term strategic and financial goals, which we believe serves to enhance short- and long-term value creation for our stockholders. The program contains elements of cash and equity-based compensation and is designed to align the interests of our executives with those of our stockholders and paying for performance.

The section of this Proxy Statement titled "Executive Compensation" beginning on page 33, including "Compensation Discussion and Analysis," describes in detail our executive compensation program and the decisions made by our compensation committee. As we describe in greater detail in the "Compensation Discussion and Analysis" section, our executive compensation program is designed to reward value creation for stockholders and progress towards achieving our mission and promote company performance. At the same time, we believe our program does not encourage excessive risk-taking by management. While we do not have a formal or informal policy for allocating between long-term and short-term compensation, between cash and non-cash compensation or among different forms of non-cash compensation, we generally strive to provide our named executive officers with a mix of short-term and long-term performance-based incentives to encourage strong performance, and our board of directors believes that this link between compensation and the achievement of our short- and long-term business goals has helped drive our performance over time.

Our board of directors is asking stockholders to approve a non-binding advisory vote on the following resolution:

RESOLVED, that the compensation paid to the company's named executive officers, as disclosed pursuant to the compensation disclosure rules of the SEC, including the "Compensation Discussion and Analysis," the compensation tables and any related material disclosed in this Proxy Statement, is hereby approved.

As an advisory vote, this proposal is not binding. The outcome of this advisory vote does not overrule any decision by the company or the board of directors (or any committee thereof), create or imply any change to the fiduciary duties of the company or the board of directors (or any committee thereof), or create or imply any additional fiduciary duties for the company or the board of directors (or any committee thereof). However, our compensation committee and board of directors value the opinions expressed by our stockholders in their vote on this proposal and intend to consider carefully the outcome of the vote when making future compensation decisions for named executive officers. Our board of directors has adopted a policy of providing annual advisory votes to approve the compensation of our named executive officers. The next advisory vote to approve the compensation of our named executive officers will occur at our 2020 annual meeting of stockholders.

Recommendation of the Board of Directors

OUR BOARD OF DIRECTORS UNANIMOUSLY RECOMMENDS THAT STOCKHOLDERS VOTE, ON AN ADVISORY BASIS, TO APPROVE THE COMPENSATION OF OUR NAMED EXECUTIVE OFFICERS BY VOTING FOR THIS PROPOSAL.

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PROPOSAL 3:

RATIFICATION OF APPOINTMENT OF INDEPENDENT REGISTERED PUBLIC ACCOUNTANTS

The audit committee has appointed PricewaterhouseCoopers LLP (PwC) as the company s independent registered public accounting firm for the fiscal year ending December 31, 2019 and the board of directors has directed that management submit the selection of independent registered public accountants for ratification by the stockholders at the annual meeting. PwC has served as the company s registered public accountant since May 5, 2017. Representatives of PwC are expected to be present at the annual meeting, will have an opportunity to make a statement if they so desire, and be available to respond to appropriate questions.

Stockholder ratification of the appointment of PwC as the company s independent registered public accounting firm is not required by Delaware law, our certificate of incorporation or our bylaws. However, the board of directors is submitting the audit committee s selection of PwC to the stockholders for ratification as a matter of good corporate practice. If the stockholders fail to ratify the selection, the audit committee will reconsider whether to retain that firm. Even if the selection is ratified, the audit committee in its discretion may direct the appointment of a different independent registered public accounting firm at any time during the year if the audit committee determines that such a change would be in the best interests of the company and its stockholders.

Independent Registered Public Accountants, Fees and Other Matters

At a meeting held on May 1, 2017, the audit committee approved the dismissal of Ernst & Young LLP (E&Y) as our independent registered public accounting firm, effective May 5, 2017, and the appointment of PwC as our independent registered public accounting firm, effective May 5, 2017, to perform independent audit services for the fiscal year ending December 31, 2017. The reports of E&Y on our consolidated financial statements for the fiscal years ended December 31, 2016 and 2015 did not contain an adverse opinion or disclaimer of opinion, nor were they qualified or modified as to uncertainty, audit scope, or accounting principles. During the fiscal years ended December 31, 2016 and 2015, and the subsequent interim period through May 5, 2017, there were no disagreements (as defined in Item 304(a)(1)(iv) of Regulation S-K and the related instructions) with E&Y on any matter of accounting principles or practices, financial statement disclosure, or auditing scope or procedures, which disagreement if not resolved to the satisfaction of E&Y would have caused E&Y to make reference thereto in its reports.

During the fiscal years ended December 31, 2016 and 2015, and the subsequent interim period through May 5, 2017, neither we nor anyone on our behalf consulted with PwC regarding (i) either: the application of accounting principles to a specific transaction, either completed or proposed; or the type of audit opinion that might be rendered on our financial statements, and either a written report or oral advice was provided to us that PwC concluded was an important factor considered by us in reaching a decision as to any accounting, auditing or financial reporting issue, or (ii) any matter that was the subject of a disagreement within the meaning of Item 304(a)(1)(iv) of Regulation S-K or any reportable event within the meaning of Item 304(a)(1)(v) of Regulation S-K.

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The following table represents aggregate fees billed to us for services related to the fiscal years ended December 31, 2018 and 2017, by PwC and E&Y.

| | Fiscal Year Ended December 31, 2018 | | Fiscal Year Ended December 31, 2017 | |
|-----------------------|--|----------------|--|----------------|
| | PwC | E&Y | PwC | E&Y |
| Audit Fees(1) | \$ 1,160,842 | \$ 100,000 | \$ 760,893 | \$ 183,500 |
| Audit Related Fees(2) | 10,000 | | 75,000 | |
| Tax Fees(3) | 96,031 | | 125,858 | |
| All Other Fees(4) | 6,994 | | 6,994 | |
| Total | 1,273,867 | 100,000 | 968,745 | 183,500 |

- (1) Audit fees consist of fees billed for professional services performed by PwC or E&Y, as applicable, for the audit of our annual consolidated financial statements, the review of interim consolidated financial statements, and related services that are normally provided in connection with registration statements, such as comfort letters and SEC comment letter filings.
- (2) Audit related fees consist of fees billed for assurance and related services that are reasonably related to the performance of the audit or review of our consolidated financial statements, such as fees for the adoption of new accounting standards.
- (3) Tax fees consist of fees for professional services, including tax consulting, compliance, and transfer pricing services.

- (4) All other fees consist of database subscription fees paid to PwC.

The audit committee has considered whether the provision of non-audit services is compatible with maintaining the independence of PwC, and has concluded that the provision of such services is compatible with maintaining such independence.

Pre-Approval Policies and Procedures

Our audit committee has adopted policies and procedures relating to the approval of all audit and non-audit services that are to be performed by our registered public accounting firm. These policies and procedures generally provide that we will not engage our registered public accounting firm to render audit or non-audit services unless the service is specifically approved in advance by our audit committee or the engagement is entered into pursuant to one of the pre-approval procedures described below.

From time to time, our audit committee may pre-approve specified types of services that are expected to be provided to us by our registered public accounting firm during the next 12 months. Any such pre-approval is detailed as to the particular service or type of services to be provided and is also generally subject to a maximum dollar amount.

Our audit committee may also delegate to one or more subcommittees or an individual member of our audit committee the authority to approve any audit or non-audit services to be provided to us by our registered public accounting firm. Any approval of services by a subcommittee or member of our audit committee pursuant to this delegated authority is reported on at the next meeting of our audit committee. During our 2018 and 2017 fiscal years, all of the services provided by PwC and, while it served as our registered public accounting firm, E&Y, were pre-approved by our audit committee.

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Recommendation of the Board of Directors

OUR BOARD OF DIRECTORS UNANIMOUSLY RECOMMENDS THAT THE STOCKHOLDERS VOTE TO RATIFY THE APPOINTMENT OF PRICEWATERHOUSECOOPERS LLP AS OUR INDEPENDENT REGISTERED PUBLIC ACCOUNTING FIRM FOR THE FISCAL YEAR ENDING DECEMBER 31, 2019.

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STOCKHOLDER PROPOSALS

Proposals of stockholders, including nominations for election to our board of directors, intended to be included in our proxy statement and form of proxy relating to, and presented at, our annual meeting of stockholders to be held in 2020 must be received by us no later than December 20, 2019, which is 120 days prior to the first anniversary of the mailing date of this proxy, unless the date of the 2020 annual meeting of stockholders is changed by more than 30 days from the anniversary of the Annual Meeting, in which case the deadline for such proposals will be a reasonable time before we begin to print and send our proxy materials. These proposals must comply with the requirements as to form and substance established by the SEC for such proposals in order to be included in the proxy statement.

In addition, our bylaws establish an advance notice procedure for nominations for election to our board of directors and other matters that shareholders wish to present for action at an annual meeting, but which will not be included in our proxy statement. In general, notice must be received at our principal executive offices not less than 90 calendar days before nor more than 120 calendar days before the one year anniversary of the previous year's annual meeting of stockholders. Therefore, to be presented at our 2020 annual meeting of stockholders, such a proposal must be received by us no earlier than January 30, 2020 and no later than February 29, 2020. However, if the date of the annual meeting is more than 20 days earlier or more than 60 days later than such anniversary date, notice must be received not later than the close of business 120 calendar days prior to such annual meeting and no later than the close of business on the later of 90 days prior to such annual meeting and 10 days following the day on which notice of the date of such annual meeting was mailed or public announcement of the date of such annual meeting was first made. If the stockholder fails to give notice by these dates, then the persons named as proxies in the proxies solicited by the board of directors for the 2020 annual meeting may exercise discretionary voting power regarding any such proposal. Stockholders are advised to review our bylaws which also specify requirements as to the form and content of a stockholder's notice.

Any proposals, notices or information about proposed director candidates should be sent to:

James Burns, Corporate Secretary

Agios Pharmaceuticals, Inc.

88 Sidney Street

Cambridge, MA 02139

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OTHER MATTERS

We do not know of any business that will be presented for consideration or action by the stockholders at the Annual Meeting other than that described in this Proxy Statement. If, however, any other business is properly brought before the meeting, shares represented by proxies will be voted in accordance with the best judgment of the persons named in the proxies or their substitutes.

We hope that you will attend the Annual Meeting. Whether or not you plan to attend, we urge you to vote your shares over the internet or by telephone, or to complete, date, sign and return the proxy card that may be delivered to you upon request in the accompanying postage-prepaid envelope. A prompt response will greatly facilitate arrangements for the meeting, and your cooperation will be appreciated.

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ANNUAL MEETING OF AGIOS PHARMACEUTICALS, INC.

Date: Wednesday, May 29, 2019
Time: 9:00 A.M. (Eastern Time)
Place: 88 Sidney Street, Cambridge, MA 02139
Please make your marks like this: Use dark black pencil or pen only

The Board of Directors Recommends a Vote **FOR** each of the director nominees listed in proposal 1 and **FOR** proposals 2 and 3.

1: To elect each of the three Class III director nominees set forth in the Proxy Statement for three-year terms expiring at the 2022 annual meeting of stockholders.

Nominees:

- (01) Jacquelyn A. Fouse, Ph.D.
- (02) David Scadden, M.D.
- (03) David P. Schenkein, M.D.

| | | |
|---------------------|---------------------------|-------------------|
| Vote For | Withhold Vote From | Vote For |
| All Nominees | All Nominees | All Except |

INSTRUCTIONS: To withhold authority to vote for any nominee, mark the **Vote For All Except** box and write the number(s) of the nominee(s) in the space provided to the right.

- | | |
|--|----------------------------|
| 2: To vote, on an advisory basis, to approve named executive officer compensation. | For Against Abstain |
| 3: To ratify the appointment of PricewaterhouseCoopers LLP as the independent registered public accounting firm for the fiscal year ending December 31, 2019. | For Against Abstain |

To attend the meeting and vote your shares in person, please mark this box.

Authorized Signatures - This section must be completed for your Instructions to be executed.

Please Sign Here

Please Date Above

Please Sign Here

Please Date Above

Please sign exactly as your name(s) appears on your stock certificate. If held in joint tenancy, all persons should sign. Trustees, administrators, etc., should include title and authority. Corporations should provide full name of corporation and title of authorized officer signing the proxy.

Please separate carefully at the perforation and return just this portion in the envelope provided.

Annual Meeting of Agios Pharmaceuticals, Inc.

to be held on Wednesday, May 29, 2019

for Holders as of April 8, 2019

This proxy is being solicited on behalf of the Board of Directors

VOTE BY:

INTERNET

Go To
www.proxypush.com/AGIO
Cast your vote online 24 hours a day/7 days a week.
Have your Proxy Card/Voting Instructions Form ready.

TELEPHONE

Call
866-509-2148
Use any touch-tone telephone toll-free 24 hours a day/7 days a week.
Have your Proxy Card/Voting Instruction Form ready.

OR

MAIL

Follow the simple recorded instructions.

View Meeting Documents.

OR

Mark, sign and date your Proxy Card/Voting Instruction Form.
Detach your Proxy Card/Voting Instruction Form.
Return your Proxy Card/Voting Instruction Form in the postage-paid envelope provided.

The undersigned hereby appoints Jacquelyn Fouse and Andrew Hirsch, and each or either of them, as the true and lawful attorneys of the undersigned, with full power of substitution and revocation, and authorizes them, and each of them, to vote all the shares of common stock of Agios Pharmaceuticals, Inc. which the undersigned is entitled to vote at said meeting and any adjournment thereof upon the matters specified and upon such other matters as may be properly brought before the meeting or any adjournment thereof, conferring authority upon such true and lawful attorneys to vote in their discretion on such other matters as may properly come before the meeting and revoking any proxy heretofore given.

THE SHARES REPRESENTED BY THIS PROXY WILL BE VOTED AS DIRECTED OR, IF NO DIRECTION IS GIVEN, SHARES WILL BE VOTED FOR THE ELECTION OF THE DIRECTORS LISTED IN PROPOSAL 1 AND FOR PROPOSALS 2 AND 3. THE PROXIES WILL VOTE IN THEIR DISCRETION ON ANY OTHER BUSINESS AS MAY PROPERLY COME BEFORE THE MEETING AND ANY ADJOURNMENT THEREOF.

PROXY TABULATOR FOR

AGIOS PHARMACEUTICALS, INC.

c/o MEDIANT COMMUNICATIONS

P.O. BOX 8016

CARY, NC 27512-9903

