



88 Sidney Street, Cambridge, Massachusetts 02139

**NOTICE OF ANNUAL MEETING OF STOCKHOLDERS
TO BE HELD ON THURSDAY, MAY 20, 2021**

Dear Stockholder:

You are cordially invited to our Annual Meeting of Stockholders. To support the health and well-being of our stockholders, employees and directors in light of the ongoing coronavirus (COVID-19) pandemic, the meeting will be a virtual meeting held via the internet on Thursday, May 20, 2021, beginning at 9:00 a.m., Eastern Time. The meeting will be held for the following purposes:

1. To elect each of the three Class II director nominees set forth in the Proxy Statement, each to serve for a three-year term expiring at the 2024 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, 2021; and
4. To transact such other business as may be properly brought before the meeting or any adjournment or postponement thereof.

As noted above, due to the COVID-19 pandemic, our Annual Meeting will be a “virtual meeting” of stockholders, which will be conducted exclusively via the internet at a virtual web conference. There will not be a physical meeting location, and stockholders will not be able to attend the Annual Meeting in person. This means that you can attend the Annual Meeting online, vote your shares during the online meeting and submit questions for consideration at the online meeting. Stockholders of record as of the close of business on March 31, 2021 are entitled to vote at the meeting. In order to attend the meeting online, vote your shares electronically during the meeting and submit questions, you must register in advance at www.proxydocs.com/AGIO prior to the deadline of May 18, 2021 at 5:00 p.m., Eastern Time. Upon completing your registration, you will receive further instructions via email, including your unique links that will allow you access to the meeting and will also permit you to submit questions and examine during the Annual Meeting the list of stockholders entitled to vote at the Annual Meeting. Please be sure to follow instructions found on your Notice, proxy card and/or voting instruction form and subsequent instructions that will be delivered to you via email. In light of the ongoing public health and safety concerns related to COVID-19, we believe that hosting a “virtual meeting” will enable greater stockholder attendance and participation from any location around the world.

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, 2020 (the “2020 Annual Report”). We are mailing the Notice on or about April 8, 2021, and it contains instructions on how to access our proxy materials 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 2020 Annual Report, and a form of proxy card or voting instruction card. All stockholders who do not receive the Notice, including stockholders 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 online. Whether or not you plan to attend the Annual Meeting online, 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. Further information about how to register to attend the Annual Meeting online, attend the Annual Meeting online, vote your shares and submit questions for consideration at the meeting is included in the accompanying proxy statement.

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 8, 2021

Important Notice Regarding the Availability of Proxy Materials for the Annual Meeting of Stockholders to be Held on May 20, 2021: This Proxy Statement and our 2020 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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PROXY STATEMENT FOR THE 2021 ANNUAL MEETING OF STOCKHOLDERS

TO BE HELD ON THURSDAY, MAY 20, 2021

INFORMATION CONCERNING SOLICITATION AND VOTING

This Proxy Statement contains information about our 2021 annual meeting of stockholders, or the Annual Meeting. The Annual Meeting will be held on Thursday, May 20, 2021, beginning at 9:00 a.m. Eastern Time. To support the health and well-being of our stockholders, employees and directors in light of the ongoing coronavirus (“COVID-19”) pandemic, the meeting will be a virtual meeting held via the internet. In order to attend the Annual Meeting online, you must register in advance at www.proxydocs.com/AGIO prior to the deadline of May 18, 2021 at 5:00 p.m., Eastern Time. Upon completing your registration, you will receive further instructions via email, including your unique links that will allow you access to the meeting. Please be sure to follow instructions found on your Notice, proxy card and/or voting instruction form and subsequent instructions that will be delivered to you via email. There will not be a physical meeting location, and stockholders will not be able to attend the Annual Meeting in person.

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, 2020, or the 2020 Annual Report, available to stockholders for the first time on or about April 8, 2021.

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 20, 2021 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 March 31, 2021, you are invited to attend the Annual Meeting online 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 2020 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 8, 2021 to all stockholders entitled to vote at the Annual Meeting.

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 and time of the virtual Annual Meeting; instructions on how to attend the Annual Meeting online; 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, 2020, 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 prior to the Annual Meeting, by requesting and returning a printed proxy card, or by voting online during 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 II director nominees set forth in the Proxy Statement, each to serve for a three-year term expiring at the 2024 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, 2021.

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

Why is the 2021 Annual Meeting a virtual, online meeting?

To support the health and well-being of our stockholders, employees and directors in light of the ongoing COVID-19 pandemic, our 2021 Annual Meeting will be a virtual meeting of stockholders where stockholders will participate by accessing a website using the internet. There will not be a physical meeting location. In light of the public health and safety concerns related to the COVID-19 pandemic, we believe that hosting a virtual meeting will facilitate greater stockholder attendance and participation at our 2021 Annual Meeting by enabling stockholders to safely participate remotely from any location around the world. Our virtual meeting will be governed by our Rules of Conduct and Procedures which will be posted at proxydocs.com/AGIO in advance of the meeting. We have designed the virtual Annual Meeting to provide the same rights and opportunities to participate as stockholders have at an in-person meeting, including the right to vote and ask questions through the virtual meeting platform.

How do I virtually attend the Annual Meeting?

We will host the Annual Meeting live online via webcast. In order to attend the Annual Meeting online, you must register in advance at www.proxydocs.com/AGIO prior to the deadline of May 18, 2021 at 5:00 p.m.,

Eastern Time. Upon completing your registration, you will receive further instructions via email, including your unique links that will allow you access to the meeting. Please be sure to follow instructions found on your Notice, proxy card and/or voting instruction form and subsequent instructions that will be delivered to you via email.

Online registration for the Annual Meeting will begin on or around April 8, 2021, and you should allow ample time for the online registration.

The webcast of the Annual Meeting will start at 9:00 a.m., Eastern Time, on May 20, 2021. Instructions on how to attend and participate in the meeting online will be sent to you via email, upon completing your registration.

We will have technicians standing by and ready to assist you with any technical difficulties you may have accessing the virtual meeting at 9:00 a.m., Eastern Time on May 20, 2021. If you encounter any difficulties accessing the virtual meeting during registration or at the time of the virtual meeting, please contact technical support by following the instructions provided to you upon registration for the Annual Meeting.

Who can vote?

Only stockholders of record at the close of business on March 31, 2021, the record date for the Annual Meeting, are entitled to vote at the Annual Meeting. On this record date, there were 69,811,806 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, March 31, 2021, entitles you to one vote on each matter that is voted on.

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 prior to the Annual Meeting:** To vote over the internet prior to the Annual Meeting, 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 prior to the Annual Meeting, 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 20, 2021 to be counted.
- (2) **By Telephone prior to the Annual Meeting:** 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 20, 2021 to be counted.
- (3) **By Mail prior to the Annual Meeting:** 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 Mediant receives the proxy card no later than May 19, 2021, we will vote your shares as you direct.

- (4) **Online during the Annual Meeting:** In order to attend the Annual Meeting online and vote online during the Annual Meeting, you must register in advance at www.proxydocs.com/AGIO prior to the deadline of May 18, 2021 at 5:00 p.m., Eastern Time. You may vote your shares online while virtually attending the Annual Meeting by following instructions found on your Notice, proxy card and/or voting instruction form and subsequent instructions that will be delivered to you via email. If you vote by proxy prior to the Annual Meeting and choose to attend the Annual Meeting online, there is no need to vote again during the Annual Meeting unless you wish to change your vote.

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 prior to the Annual Meeting:** 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 prior to the Annual Meeting:** 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) **Online during the Annual Meeting:** You will receive instructions from your bank, brokerage firm, or other nominee explaining how you can register to attend the Annual Meeting online and vote your shares online during the Annual Meeting.

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 under “Over the Internet prior to the Annual Meeting” and “By Telephone prior to the Annual Meeting”. Only your latest internet or telephone vote submitted prior to the Annual Meeting is counted. You may not change your vote prior to the Annual Meeting over the internet or by telephone after 8:59 a.m., Eastern Time, on May 20, 2021.
- (2) Sign, date 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 19, 2021. Only your latest dated and timely received proxy will be counted.
- (3) Virtually attend the Annual Meeting and vote online as instructed above under “Online during the Annual Meeting”. Virtually attending the Annual Meeting alone, without voting online during the Annual Meeting, will not revoke your internet vote, telephone vote or proxy submitted by mail, as the case may be.

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 online during the Annual Meeting, which will have the effect of revoking any previously submitted voting instructions; see “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, or by mail prior to the Annual Meeting or online while virtually attending 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 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 II 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, 2021, 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.

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 virtually 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 virtually at the Annual 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, virtually or by proxy, of holders representing a majority of our outstanding common stock as of the record date, March 31, 2021, or approximately 34,905,904 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 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, 2021 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, 2021.

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 as Class II 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, 2021.

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 Class II directors, the approval, on an advisory basis, 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 do I submit a question at the Annual Meeting?

If you wish to submit a question, on the day of the Annual Meeting, beginning at 8:00 a.m. Eastern Time on Thursday, May 20, 2021, you may log into the virtual meeting platform using the unique link provided to you via email following the completion of your registration at www.proxydocs.com/AGIO, and follow the instructions there. Our virtual meeting will be governed by our Rules of Conduct and Procedures will be posted at www.proxydocs.com/AGIO in advance of the meeting. The Rules of Conduct and Procedures will address the ability of stockholders to ask questions during the meeting, including rules on permissible topics, and rules for how questions and comments will be recognized and disclosed to meeting participants. All questions received from stockholders before or during the virtual annual meeting will be posted on our website at investor.agios.com as soon as practicable following the Annual Meeting.

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.

How and when may I submit a stockholder proposal, including a stockholder nomination for director for the 2022 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 2022 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, 2020 that we filed with the SEC, we will send you one, without exhibits, free of charge. Please contact Holly Manning, Senior Director, Investor Relations. She may be contacted at 88 Sidney Street, Cambridge, Massachusetts 02139; telephone: 617-649-8600; e-mail: Holly.Manning@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 Holly Manning, Senior Director, Investor Relations. She may be contacted at 88 Sidney Street, Cambridge, Massachusetts 02139; telephone: 617-649-8600; e-mail: Holly.Manning@agios.com.

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 our annual report and proxy materials in the future, please notify your broker or contact us. If you wish to receive a separate set of our annual report and 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 or oral request to: Agios Pharmaceuticals, Inc., 88 Sidney Street, Cambridge, MA 02139, Attention: Corporate Secretary, 617-649-8600 or contact Investor Relations at 617-649-8600.

SECURITY OWNERSHIP OF CERTAIN BENEFICIAL OWNERS AND MANAGEMENT

The following table sets forth information relating to the beneficial ownership of our common stock as of March 31, 2021, 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 69,811,806 shares of our common stock outstanding as of March 31, 2021. 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, 2021 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 Wellington Management Group LLP(1)	7,877,370		—		7,877,370	11.28%
Entities affiliated with Fidelity Management & Research Company(2)	7,633,980		—		7,633,980	10.94%
Entities affiliated with Celgene Corporation(3)	7,121,658		—		7,121,658	10.20%
The Vanguard Group(4)	5,625,713		—		5,625,713	8.06%
BlackRock, Inc.(5)	4,485,000		—		4,485,000	6.42%
BB Biotech AG(6)	4,158,902		—		4,158,902	5.96%
Named Executive Officers and Directors						
Jonathan Biller	10,354		27,916		38,270	*
Christopher Bowden, M.D.	15,487		188,907		204,394	*
Bruce Car, Ph.D.	7,093		15,667		22,760	*
Jacquelyn A. Fouse, Ph.D.	58,363		259,241		317,604	*
Andrew Hirsch	—		—		—	—
Paul J. Clancy	3,278		91,801		95,079	*
Ian T. Clark	3,278		48,401		51,679	*
Kaye Foster	5,478		61,300		66,778	*
Maykin Ho, Ph.D.	3,278		61,651		64,929	*
John M. Maraganore, Ph.D.(7)	26,757		73,526		100,283	*
David Scadden, M.D.	3,849		48,401		52,250	*
David P. Schenkein, M.D.(8)	454,907		769,437		1,224,344	1.73%
All executive officers and directors as a group (12 persons)	602,793		1,713,912		2,316,705	3.24%

* Less than 1%.

- (1) Based solely on a Schedule 13G/A filed with the SEC on February 3, 2021. Wellington Management Group LLP (“Wellington”), Wellington Group Holdings LLP and Wellington Investment Advisors Holdings LLP are each deemed to be the beneficial owner of 7,877,370 shares of common stock, with respect to which each entity reported shared voting power over 7,780,029 shares and shared dispositive power over 7,877,370 shares. Wellington Management Company LLP is deemed to be the beneficial owner of 7,769,091 shares of common stock, with respect to which it reported shared voting and dispositive power over 7,769,091 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 c/o Wellington Management Company LLP 280 Congress Street, Boston, MA 02210.
- (2) Based solely on a Schedule 13G/A filed with the SEC on March 10, 2021. FMR LLC and Abigail P. Johnson are each the beneficial owners of 7,633,980 shares of common stock. FMR LLC has sole voting power over 366,858 shares of common stock and sole dispositive power over 7,633,980 shares of common stock. The address of FMR LLC is 245 Summer Street, Boston, MA 02210.
- (3) Based solely on a Schedule 13D/A filed with the SEC on November 14, 2019. 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 1,777,824 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 1,777,824 shares of common stock and shared voting and dispositive power over 5,343,834 shares of common stock. On March 31, 2021, we entered into a share repurchase agreement with Bristol-Myers Squibb Company pursuant to which we agreed to repurchase 7,121,658 shares of our common stock held by certain subsidiaries of BMS and reflected in the table above. The table above does not reflect the sale of these shares, which had not closed as of the date set forth above. 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 10, 2021. The Vanguard Group (“Vanguard”) is deemed to be the beneficial owner of 5,625,713 shares of common stock, with respect to which it reported shared voting power over 41,869 shares, sole dispositive power over 5,533,174 shares and shared dispositive power over 92,539 shares. 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 29, 2021 by BlackRock, Inc. (“BlackRock”) and certain of its subsidiaries. BlackRock is deemed to be the beneficial owner of 4,485,000 shares of common stock, with respect to which it reported sole voting power over 4,248,882 shares and sole dispositive power over 4,485,000 shares. The address of BlackRock is 55 East 52nd Street, New York, NY 10055.
- (6) Based solely on a Schedule 13G/A filed with the SEC on February 12, 2021. BB Biotech AG (“BB Biotech”) and its wholly-owned subsidiary Biotech Target N.V. (“Biotech Target”) share voting and dispositive power over 4,158,902 shares of common stock. The address of BB Biotech is Schwertstrasse 6, CH-8200 Schaffhausen, Switzerland and the address of Biotech Target is Ara Hill Top Building, Unit A-5, Pletterijweg Oost 1, Curacao.

- (7) Reflects a gift of 3,415 shares to charity on February 4, 2021.
- (8) 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.

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 II directors, Kaye Foster, Maykin Ho, Ph.D. and John M. Maraganore, Ph.D., will expire at the Annual Meeting. The nominees for Class II directors for election at the Annual Meeting are Ms. Foster and Drs. Ho and Maraganore. If any of Ms. Foster or Drs. Ho or Maraganore is elected at the Annual Meeting, such individual will be elected to serve for a three-year term that will expire at our 2024 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 Ms. Foster and Drs. Ho and Maraganore or, in the event that any of Ms. Foster and Drs. Ho and Maraganore 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.

Nominees for Election to the Board of Directors

Term Expiring at the 2024 Annual Meeting of Stockholders, if elected at the Annual Meeting (Class II)

<u>Name</u>	<u>Age</u>	<u>Present Position with Agios Pharmaceuticals, Inc.</u>
Kaye Foster	61	Director
Maykin Ho, Ph.D.	68	Director
John M. Maraganore, Ph.D.	58	Lead Independent 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 and compensation committee of Grail, Inc., or Grail, a private biotechnology company acquired by Illumina, Inc., on the board of directors and compensation and community equity committee of Resilience Inc., a private management consulting company, on the board of directors and quality and major gifts committee of Stanford Health Care, a hospital and healthcare system, on the board and human resources committee 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, including her experience in people management, compensation planning and driving and maintaining corporate culture.

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 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 and audit committee of Fibrogen, Inc., a publicly traded biotechnology company, a member of the board of directors, audit committee and chairs the nominating and corporate governance committee of Grail, a member of the board of directors and audit committee of Parexel International, a privately-held, global pharmaceutical services company, and a member of the board of directors, audit committee and science and technology committee of BioMarin Pharmaceutical Inc., a public biopharmaceutical company. Dr. Ho also serves on the board of directors, audit committee and investment committee of two non-profit research institutes: the Aaron Diamond AIDS Research Center and the Institute for Protein Innovation. In addition, she 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. Dr. Maraganore is our lead independent director. Since 2002, Dr. Maraganore has served as the chief executive officer and as a director of Alnylam Pharmaceuticals, Inc., or Alnylam, a publicly traded biopharmaceutical company. From 2002 to 2007, Dr. Maraganore also served as president of Alnylam. From 1997 to 2002, Dr. Maraganore served in a number of leadership roles including as senior vice president, strategic product development with Millennium Pharmaceuticals, Inc., a biopharmaceutical company (now Takeda Oncology), or 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., or bluebird, a publicly traded biopharmaceutical company. In addition, he was formerly a venture partner at Third Rock Ventures, L.P., and was formerly chairman of the board of directors of Regulus Therapeutics, Inc., a publicly traded biotechnology company. Dr. Maraganore serves as a strategic advisor and investor to Bria Biosciences, a private biotechnology company, and also serves in advisory roles with General Atlantic, a private equity firm, and Pictet & Cie, an investment firm. He is the former chair and current member of the Executive Committee, the Emerging Companies Section Governing Board and the Health Section Governing Board of the Biotechnology Innovation Organization (BIO). 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 35 years of experience in the biotechnology industry, bringing to our board of directors critical scientific, research and development, international and general management expertise.

Recommendation of the Board of Directors

OUR BOARD OF DIRECTORS UNANIMOUSLY RECOMMENDS THAT THE STOCKHOLDERS VOTE FOR THE ELECTION OF EACH OF MS. FOSTER AND DRs. HO AND MARAGANORE.

Members of the Board of Directors Continuing in Office

**Term Expiring at the
2022 Annual Meeting of Stockholders (Class III)**

Name	Age	Present Position with Agios Pharmaceuticals, Inc.
Jacquelyn A. Fouse, Ph.D.	59	Chief Executive Officer and Director
David Scadden, M.D.	68	Director
David P. Schenkein, M.D.	63	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, or Celgene, 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, from 2006 to 2007, and as its senior vice president and chief financial officer from 2002 to 2006. Prior to her time with Alcon she held a variety of senior leadership roles with international companies. Dr. Fouse is also a director and

member of the audit committee and nominating and corporate governance committee of Incyte Corporation, or Incyte, a publicly traded biopharmaceutical company, and was a director of Perrigo Company, a public pharmaceutical manufacturer, Celgene and Dick's Sporting Goods, Inc. a publicly traded sporting goods retailer. 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. She also earned a Masters in Environmental Management from the Yale University School of Forestry and Environmental Studies. 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 international and 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 directors and nominating and governance and science and technology committees of Editas Medicine, Inc., a publicly traded biotechnology company, and on the boards of directors of the private biotechnology companies LifeVault Bio and Clear Creek Bio, Inc. Dr. Scadden is also a scientific founder of Fate Therapeutics, a publicly traded biotechnology company, and a scientific founder and member of the board of directors and scientific advisory board of Magenta Therapeutics, a publicly traded biotechnology company. He is the recipient of numerous honors including the E. Donnell Thomas and the Dameshek awards from the American Society of Hematology and awards from the Doris Duke Charitable Trust, the Ellison Medical Foundation, the Burroughs Wellcome 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 field of hematology.

David P. Schenkein, M.D. has served as a member of our board of directors since August 2009, as our executive chairman from February 2019 to February 2020, and as the chairman of our board of directors since February 2020. 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 30 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. (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, 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 Prime Medicine, Inc., a private biotechnology company, Leyden Labs, Inc., a private biotechnology company, and on the board of directors, nominating and corporate governance committee and science and technology committee of Denali Therapeutics Inc., a publicly traded biotechnology company. Dr. Schenkein was formerly on the board of directors of Foundation Medicine, Inc., a publicly traded biotechnology company and on the board of directors, compensation committee and nominating and corporate governance committee of bluebird. 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 of hematology and his extensive background in the biotechnology industry, including his roles at Genentech and Millennium, provide a critical contribution to our board of directors.

**Term Expiring at the
2023 Annual Meeting of Stockholders (Class I)**

<u>Name</u>	<u>Age</u>	<u>Present Position with Agios Pharmaceuticals, Inc.</u>
Paul J. Clancy	59	Director
Ian T. Clark	60	Director

Paul J. Clancy has served as a member of our board of directors since September 2013. Mr. Clancy has more than 35 years of experience in financial management and strategic planning. Mr. Clancy served as a senior advisor from November 2019 through July 2020 and as the Executive Vice President and Chief Financial Officer from July 2017 through October 2019 of Alexion Pharmaceuticals, Inc., a publicly traded biopharmaceutical company. Mr. Clancy served as the Executive Vice President and Chief Financial Officer at Biogen Inc. (formerly known as Biogen Idec), a biopharmaceutical company, or Biogen from August 2007 through June 2017. 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. Mr. Clancy is on the board of directors, audit committee and compensation committee of Incyte and on the board of directors and audit and finance committee of Exact Sciences, a publicly traded molecular diagnostics company. Mr. Clancy is also on the board of directors, audit committee and compensation committee of Xilio Therapeutics, a private biotechnology company. Mr. Clancy is currently a Senior Visiting Lecturer of Finance at Cornell University. 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 multinational companies.

Ian T. Clark has served as a member of our board of directors since December 2016. Mr. Clark has more than 35 years of experience in the biotechnology industry. Mr. Clark currently serves as an operating partner at Blackstone Life Sciences, a private investment firm. Prior to that, he was chief executive officer and a member of the board of directors of Genentech, a position he held from 2010 to 2016. Mr. Clark served for 12 years on the Genentech Executive Committee, initially as EVP of Commercial Operations. Before that, he served as SVP and General Manager of BioOncology. Prior to Genentech, Mr. Clark spent 23 years in positions of increasing responsibility at Novartis, Sanofi-Synthelabo SA (Aventis SA), Ivax Pharmaceuticals, Inc. and G.D. Searle, LLC, a subsidiary of Monsanto Corporation, working in the UK, Canada, Eastern Europe and France. Mr. Clark is a member of the boards of directors of the following publicly traded biotechnology companies: Takeda Pharmaceutical Company Limited (also on the compensation committee); Guardant Health, Inc. (lead independent director, chair of the compensation committee and a member of the audit committee); Corvus Pharmaceuticals, Inc. (also on the compensation committee and nominating and corporate governance committees); Olema Pharmaceuticals, Inc.; and AvroBio, Inc. (also chair of the compensation committee). He was formerly on the boards of directors of Forty Seven, Inc. (prior to its acquisition by Gilead Sciences, Inc. or Gilead), Shire plc (prior to its acquisition by Takeda), Kite Pharma, Inc. (prior to its acquisition by Gilead), Vernalis plc, and TerraVia Holdings, Inc. Mr. Clark served on the Board of the Biotechnology Industry Association, and as a member of the economic advisory council of the 12th District of the Federal Reserve. He also served on the BioFulcrum Board of the Gladstone Institute, as Chair of the External Advisory Board to Southampton University’s Institute for Life Sciences and as an advisor to Blackstone Life Sciences. 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 international and leadership experience.

CORPORATE GOVERNANCE

General

We believe that good corporate governance is important to ensure that Agios 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 corporate 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 Holly Manning, Senior Director, Investor Relations, at 88 Sidney Street, Cambridge, Massachusetts 02139; telephone: 617-649-8600; e-mail: Holly.Manning@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.

Environmental Sustainability and Social Responsibility

At Agios, we are committed to building a sustainable business that provides long-term value for all our stakeholders. We support environmental, social and governance (ESG) initiatives that are aligned with our culture and values and that may positively impact the patients we serve, our employees, our communities and our world. Our vision to make the world a better place is foundational to Agios. We are driven to improve the lives of those fighting life-threatening and life-altering diseases, including genetically defined diseases that have often been overlooked or neglected.

More information about our ESG initiatives can be found in our 2021 Environmental, Social and Governance Report, available on the "Investors" tab of our website www.agios.com.

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 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.

In March 2021, 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 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, Mr. Clark, Ms. Foster and Dr. Maraganore, who comprise our compensation committee, and Mr. Clark, Ms. Foster and Dr. Ho, who comprise 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

Our corporate governance guidelines provide that the nominating and corporate governance committee shall periodically assess our board of directors' leadership structure, including whether the offices of chief executive officer and chairman of the board of directors should be separate and why the board of directors' leadership structure is appropriate given the specific characteristics or circumstances of our company. These guidelines provide the board of directors with flexibility to determine whether the two roles should be combined or separated based upon our needs and the board of directors' assessment of its leadership from time to time. We currently separate the roles of chief executive officer and chairman of the board of directors. Separating the duties of the chairman of the board from the duties of the chief executive officer allows our chief executive

officer to focus on our day-to-day business, while allowing the chairman of the board to lead the board of directors in its fundamental role of providing advice to and independent oversight of management. Specifically, the chairman of our board of directors presides over meetings of the board of directors, facilitates communications between management and the board of directors and assists with other corporate governance matters.

Because Dr. Schenkein, the chairman of our board of directors, is not independent within the meaning of the Nasdaq Listing Rules, 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 chairman and our chief executive officer;
- Working with our 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 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;
- 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 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. Our lead independent director or the chair of our nominating and corporate 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 thirteen times during our fiscal year 2020, including telephonic and virtual 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.

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. All of our then-serving directors attended our 2020 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 nine times during fiscal year 2020, 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;
- 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, Mr. Clark and Dr. Maraganore. 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 2020. 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 34 of this Proxy Statement; and
- preparing the compensation committee report required by SEC rules, which is included on page 52 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 four times during fiscal year 2020. 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 and human capital matters;
- 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”.

Science and Technology Committee

The members of our science and technology committee are Dr. Scadden, Dr. Fouse and 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 four times during fiscal year 2020, including telephonic meetings. The science and technology 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, reputational and human capital 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.

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

	<u>Annual Cash Compensation</u>	<u>Number of Options/RSUs Granted</u>
Board of Directors:		
Board Member	\$50,000	\$718,000 in equity awards upon initial election (split approximately 75% in stock options and 25% in restricted stock units, or 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)*
Chairman	Additional \$30,000	—
Lead Independent Director	Additional \$25,000	—
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 shares 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 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 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 initial RSUs granted to our newly elected non-employee directors vest as to one-third of the underlying shares each year following the grant date. 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.

The following table sets forth information concerning the compensation for our non-employee directors during the fiscal year ended December 31, 2020:

Name	Fees Earned or Paid in Cash (\$)	Option Awards (\$)(1)(2)	Stock Awards (\$)(3)(4)	All Other Compensation (\$)	Total (\$)
Paul J. Clancy	70,000	270,043	89,948	—	429,991
Ian T. Clark	62,500	270,043	89,948	—	422,491
Kaye Foster	75,000	270,043	89,948	—	434,991
Maykin Ho, Ph.D.	65,000	270,043	89,948	—	424,991
John M. Maraganore, Ph.D.	92,500	270,043	89,948	—	452,491
David Scadden, M.D.	65,000	270,043	89,948	—	424,991
David P. Schenkein, M.D.	80,208	270,043	89,948	14,781(5)	454,981

- (1) Represents stock options to purchase 8,096 shares of common stock granted to each of our non-employee directors during 2020 for service on our board of directors. The shares subject to these stock options vest in full on May 28, 2021. Amounts listed represent the aggregate fair value amount computed as of the grant date of the stock option awards granted during 2020 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 15, Share-Based Payments, of the Notes to our Consolidated Financial Statements in our Annual Report on Form 10-K filed with the SEC on February 25, 2021.
- (2) The aggregate number of shares of common stock underlying stock options outstanding as of December 31, 2020 for our non-employee directors were: Mr. Clancy: 90,115, Mr. Clark: 46,715, Ms. Foster: 59,614, Dr. Ho: 59,965, Dr. Maraganore: 71,840, Dr. Scadden: 46,715, and Dr. Schenkein: 806,049.
- (3) Represents RSUs representing the contingent right to receive 1,686 shares of common stock granted to each of our non-employee directors during 2020 for service on our board of directors. The shares subject to these RSUs vest in full on May 28, 2021. Amounts listed represent the aggregate fair value amount

computed as of the grant date of the RSUs granted during 2020 in accordance with FASB ASC Topic 718. Assumptions used in the calculation of these amounts are included in Note 15, Share-Based Payments, of the Notes to our Consolidated Financial Statements in our Annual Report on Form 10-K filed with the SEC on February 25, 2021.

- (4) The aggregate number of shares of common stock underlying RSUs outstanding as of December 31, 2020 was 15,360 for Dr. Schenkein and 1,686 shares for each of our other non-employee directors.
- (5) Reflects compensation paid to Dr. Schenkein for his service as executive chairman during January 2020 including one month of his \$175,000 annual salary and company-paid life insurance premium.

Dr. Fouse, one of our directors who also serves as our chief executive officer, did not receive any additional compensation for her service as a director from and after February 1, 2019, when she assumed the role of chief executive officer. The compensation that we pay to Dr. Fouse as our chief executive officer is disclosed under “Executive Compensation—Summary Compensation Table.” Dr. Schenkein, one of our directors who also served as our president and chief executive officer until February 1, 2019, and as the executive chairman of our board of directors from February 1, 2019 until February 1, 2020, did not receive any additional compensation for his service as a director while in those roles. 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 as described below under “Executive Compensation—Employment, Severance and Change in Control Arrangements—*Employment Offer Letters*”. Effective February 1, 2020, Dr. Schenkein transitioned from executive chairman to chairman and is compensated pursuant to the non-employee director compensation policy described above.

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.

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. From time to time, various members of management and other employees as well as outside advisors or consultants are invited by the compensation committee to make presentations, provide financial or other background information or advice or 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, 2020, the compensation committee directly engaged Radford, a compensation consultant which is part of the Rewards Solutions practice at Aon plc, to advise the compensation committee on our compensation program for executive officers and directors, 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, 2020.

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 2020, the compensation committee did not form or delegate authority to such subcommittees. During fiscal year 2020, 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.

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, 2020 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 by the applicable requirements of the SEC and 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, 2020.

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

EXECUTIVE OFFICERS

The following table sets forth information regarding our executive officers as of April 1, 2021:

<u>Name</u>	<u>Age</u>	<u>Position(s)</u>
Jacquelyn A. Fouse, Ph.D.	59	Chief Executive Officer
Jonathan Biller	57	Chief Financial Officer, Head of Legal and Corporate Affairs
Chris Bowden, M.D.	60	Chief Medical Officer
Bruce Car, Ph.D.	59	Chief Scientific Officer
Darrin Miles	47	Chief Commercial Officer

The biography of Dr. Fouse can be found under “Members of the Board of Directors Continuing in Office.”

Jonathan Biller joined Agios as chief legal officer in December 2019 and was appointed to the role of chief financial officer, head of legal and corporate affairs in September 2020. He has more than 30 years of legal, tax and treasury experience. Prior to joining Agios, Mr. Biller held leadership roles at Celgene, leading Celgene’s global legal function as executive vice president and general counsel from July 2018 to November 2019, and serving as senior vice president, tax and treasury from 2011 to June 2018, in which role he was responsible for the company’s global tax and treasury functions including its capital allocation strategy and tax policy. Prior to Celgene, Mr. Biller was general counsel, chief tax officer and secretary for Bunge. Prior to Bunge, Mr. Biller held roles of increasing responsibility at Alcon, Inc., during which time it was a publicly traded company. Mr. Biller began his career at law firm Hopkins & Sutter rising to the level of partner and was also partner at Foley & Lardner LLP after the firms merged. He holds a B.A. from Brown University and a J.D. from Yale Law School.

Chris Bowden, M.D. joined Agios as chief medical officer in May 2014. He has more than 20 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.

Bruce Car, Ph.D. joined Agios as chief scientific officer in January 2020. Prior to joining Agios, Dr. Car was at Bristol-Myers Squibb Company, or BMS, a global biopharmaceutical company, and its predecessor companies, since 2005, serving most recently as the interim head, BMS drug discovery. While at BMS, Dr. Car helped bring more than 200 drug candidates to clinical trials and 13 marketed drugs, including ELIQUIS®, OPDIVO® and FARXIGA®, to patients, holding roles of increasing responsibility in drug discovery, covering all therapeutic areas, drug platforms and stages of discovery. Dr. Car also led the BMS research and development campus in Bangalore, India, for six years across all aspects of discovery and later established a translational medicine function at that site. In 2017, he became the first head of the BMS translational medicine

group, where he successfully built a cohesive function of over 300 scientists and specialists covering biomarkers through pharmaco-diagnostics. Among his responsibilities in translational medicine, Dr. Car led the research and development computational biology and machine learning capabilities and pharmaco-diagnostic technology platforms. Dr. Car received veterinary medicine and Masters in pathology degrees from The University of Melbourne, Victoria, Australia, and his Ph.D. from Cornell University, where he is currently an adjunct professor. Dr. Car undertook his postdoctoral studies in immunology and inflammation at the Theodor Kocher Institute, University of Berne and ETH/University of Zurich.

Darrin Miles was appointed chief commercial officer in January 2021. Prior to that, he served as our senior vice president, U.S. commercial and global marketing from April 2019 to January 2021, as vice president, oncology program leadership from October 2016 to April 2019, and as vice president, oncology marketing and market research from July 2015 to October 2016, in which roles he was instrumental in setting the strategy for our IDH portfolio and leading the program team through the development and approval of TIBSOVO®. Prior to joining Agios, Mr. Miles spent 14 years at Genentech, where he held roles of increasing responsibility including marketing, sales management, reimbursement and lifecycle leadership supporting a broad portfolio of brands including Rituxan®, Herceptin®, Tarceva® and Lucentis®. Mr. Miles also led the U.S. launches of next generation anti-HER2 treatments Perjeta® and Kadcyra®. Prior to Genentech, he worked for five years at ALZA Corporation and SmithKline Beecham (now GSK) in numerous roles including marketing, market analytics and organization development. Mr. Miles holds a B.S. in applied economics with a minor in biological sciences from Cornell University.

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, or NEOs, 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 NEOs, 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 NEOs.

Our NEOs for 2020 were as follows:

- Jacquelyn A. Fouse, Ph.D., our chief executive officer;
- Jonathan Biller, our chief legal officer and, effective September 11, 2020, our chief financial officer, head of legal and corporate affairs;
- Christopher Bowden, M.D., our chief medical officer;
- Bruce Car, Ph.D., our chief scientific officer; and
- Andrew Hirsch, our former chief financial officer and head of corporate development until September 11, 2020.

Say-on-Pay Vote Support and Stockholder Engagement

Our compensation committee strives to ensure that our executive compensation program aligns with the interest of our stockholders and adheres to our pay-for-performance philosophy. Our executive compensation program has historically received very strong stockholder support, including, for example, the support of over 97% of votes cast on the non-binding advisory vote on the compensation of our NEOs, commonly referred to as a "say-on-pay" vote, at our annual meetings of stockholders in 2019 and 2018. At our 2020 annual meeting of stockholders, approximately 65% of votes cast supported our executive compensation program. We were disappointed by this lower level of support. While these votes are non-binding advisory votes, our compensation committee and board of directors take the voting results into account in determining the compensation of our NEOs. To understand our stockholders' concerns, we reached out in September 2020 to stockholders representing approximately 80% of our outstanding shares. We ultimately held discussions with stockholders representing approximately 45% of our outstanding shares. Kaye Foster, a member of the board of directors and also chair of our compensation committee, participated in some of these discussions. Other participants in these discussions included our investor relations professionals, our chief people officer and our chief financial officer, head of legal and corporate affairs.

Over the course of several months, senior management, our compensation committee and Radford, our compensation committee's independent consultant, analyzed and discussed what was learned in this stockholder outreach process. In general, we learned that the decline in support for our "say-on-pay" proposal last year was primarily related to (i) the one-time equity awards granted to Dr. Fouse, our chief executive officer, in connection with the start of her employment in February 2019, (ii) the preference for increased use of performance based long-term equity awards, and (iii) the desire for more fulsome disclosure of the performance metrics underlying such awards. Below are selected highlights from this process:

What We Heard

- Standard options are not sufficiently performance-driven, as stock prices may be influenced by trends within a company’s given industry
- The amount of pay for our chief executive officer was outsized in 2019
- Desire for more disclosure around performance share unit, or PSU, performance metrics

What We’ve Done

- In 2021, all of our NEOs received PSUs as part of their annual long-term incentive compensation package; in 2021, our chief executive officer was granted options and PSUs, but no time-based restricted stock units, or RSUs
- Equity grants to our chief executive officer in 2020 and 2021 are reflective of market and peer annual practices, and are significantly lower than her 2019 new hire grant
- We are providing additional disclosure on PSU metrics in this proxy statement – see “*Executive Compensation Elements—Performance Share Units (PSUs)*” below

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 continue to have an opportunity annually to cast an advisory vote in connection with compensation for our NEOs.

Executive Summary

2020 was a transformational year for Agios. Notwithstanding the difficulties faced by our company, our industry and the world as a whole due to the COVID-19 pandemic, we successfully navigated a number of operational challenges and disruptions and continued to make notable progress and achieve significant corporate milestones on the clinical, commercial and business development fronts.

We continued to advance the development of our lead genetically defined disease, or GDD, program, mitapivat, across three distinct therapeutic areas. We completed enrollment in both ACTIVATE and ACTIVATE-T, our phase 3 clinical trials evaluating mitapivat in adults with pyruvate kinase (PK) deficiency who are not regularly transfused and those who are regularly transfused, respectively. In December 2020, we reported topline data for ACTIVATE demonstrating that the trial’s primary endpoint was met and statistically significant improvements over placebo were observed across key secondary endpoints. In June 2020 we also reported clinical data for mitapivat in two other indications which further demonstrate the potential of this molecule in hemolytic anemias: we reported phase 1 data demonstrating clinical proof of concept for mitapivat of in sickle cell disease, or SCD, and we reported updated phase 2 data showing sustained hemoglobin responses in non-transfusion dependent α - and β - thalassemia patients. Based on the strength of these data, we were able to engage in a productive dialogue with regulators that informed the design of our pivotal clinical trials for mitapivat, which we expect to initiate in both indications by the end of 2021. Finally, we successfully initiated a phase 1 trial of AG-946, or next-generation PKR activator, in healthy volunteers.

We continued to advance development of our oncology programs in both hematologic malignancies and solid tumors. We finished fiscal year 2020 with \$121.1 million in net product revenue from sales of TIBSOVO® (ivosidenib tablets), exceeding our guidance for the year. In addition, in September 2020 we announced final overall survival data from our phase 3 ClarIDHy trial of TIBSOVO® in previously treated patients with an isocitrate dehydrogenase-1, or IDH1, mutant cholangiocarcinoma, demonstrating a consistent trend in improved overall survival in patients treated with TIBSOVO® compared to those in the placebo arm of the trial. These data supported a supplemental new drug application, or sNDA, that we submitted to the FDA in the first quarter of

2021 for TIBSOVO® as a potential treatment for patients with previously treated IDH1 mutant cholangiocarcinoma. Despite facing logistical challenges in site initiations and patient enrollment as a result of the COVID-19 pandemic, we also continued to drive enrollment in the ongoing clinical trials of our IDH inhibitors, including the phase 3 INDIGO trial of vorasidenib in patients with grade 2 non-enhancing glioma with an IDH mutation, our phase 3 AGILE trial combining TIBSOVO® and VIDAZA® (azacitidine) in newly diagnosed acute myeloid leukemia, or AML, patients with an IDH1 mutation who are ineligible for intensive chemotherapy, and the myelodysplastic syndrome, or MDS, arm of our phase 1 trial of TIBSOVO® in patients with advanced hematologic malignancies with an IDH1 mutation. In October 2020, we announced the withdrawal of our European Marketing Authorization Application for TIBSOVO® for the treatment of adult patients with relapsed or refractory AML with an IDH1 mutation, based on feedback from the European Medicine Agency's Committee for Medicinal Products for Human Use that the available data from our single arm, uncontrolled phase 1 trial did not sufficiently support a positive benefit-risk balance for the proposed indication. Although we were disappointed by this development, we continued to drive enrollment in our two ongoing phase 3 trials of TIBSOVO® in patients with newly diagnosed AML. We also continued to enroll patients in our phase 1 trial of AG-270 in multiple tumor types carrying an methylthioadenosine phosphorylase (MTAP) deletion. In the first half of 2020 we completed a strategic prioritization exercise and made the decision to cease internal development of AG-636 as a result of limited clinical trial enrollment in our phase 1 trial for lymphoma.

Under the leadership of Bruce Car, Ph.D., who joined us as chief scientific officer on January 6, 2020, we advanced our pre-clinical research pipeline and progressed multiple programs to validated program or development candidate status.

We also made significant progress on the business development front, announcing two transformational transactions that reshaped the Agios of the future. First, in June 2020, we announced the sale of our tiered, sales-based royalty rights on worldwide net sales of Bristol Myers Squibb's IDHIFA® (enasidenib), as well as our rights to receive up to \$55.0 million in outstanding regulatory milestone payments from Bristol Myers Squibb, to Royalty Pharma for \$255.0 million. We concluded the year with the announcement of the sale of our oncology business to Servier Pharmaceuticals, LLC, or Servier, an independent global pharmaceutical company, for cash consideration of up to \$2.0 billion and royalties on U.S. net sales of TIBSOVO® and vorasidenib. Following the closing of the transaction on March 31, 2021, we believe we are positioned to move forward with a singular focus on accelerating and expanding our GDD portfolio, including our mitapivat and AG-946 clinical programs and robust pipeline of therapeutic candidates. The proceeds from the transaction allow us to focus on rapidly advancing our GDD portfolio for patients in need, strengthen our capital structure while returning up to \$1.2 billion to our stockholders post-closing, and achieve capital markets independence while still participating in the future success of sales of TIBSOVO® and vorasidenib.

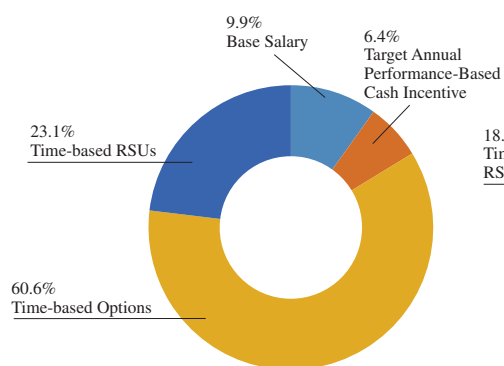
In addition to the above achievements, we managed our cash spending in line with our board-approved budget, attracted, retained and fostered the development of our human capital and effectively managed the impact of the COVID-19 pandemic on our business, switching to a remote-based work environment and supporting our employee base during this difficult time. During 2020, the onset of the COVID-19 pandemic provided an opportunity and an imperative to demonstrate our commitment to the health and wellbeing of our employees, our patients and our community. We immediately implemented an action plan that took into consideration local and national public health guidelines, input from our employees and our desire to reduce the risk of transmission to our employees and communities. Throughout the pandemic, our ability to embrace flexibility has helped us maintain productivity to deliver for patients.

Based upon an evaluation of our performance against each of our goals for 2020 and the weighting assigned to each, the board determined, upon recommendation from the compensation committee, that we achieved an overall result of 105% for the year.

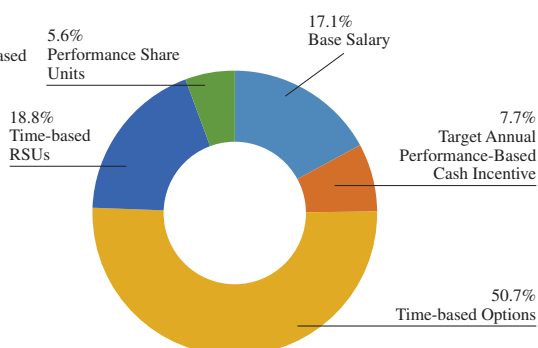
Executive Compensation Overview

To foster the future success of the company, we reward our executives in a manner that reinforces our pay-for-performance philosophy and culture. Our compensation committee is committed to ensuring that a substantial portion of executive compensation is “at-risk” and variable. As such, on average, 90% of Dr. Fouse’s total direct compensation for 2020 and, on weighted average, 83% of our other NEOs’ (excluding Mr. Hirsch, who left the Company in September 2020, and Mr. Biller, who did not received equity awards during 2020) total direct compensation for 2020 is variable and directly affected by both the company’s and each NEO’s performance. 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 NEOs with a balance of short-term and long-term incentives to encourage consistently strong performance.

CEO 2020 Target Compensation Mix



Other Named Executive Officers 2020 Target Compensation Mix



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

- ✓ Maintain an industry-specific peer group for benchmarking pay
- ✓ Target pay based on market norms
- ✓ Deliver executive compensation primarily through performance-based pay
- ✓ Set challenging short- and long-term incentive award goals
- ✓ Cap annual performance-based 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

What We Don't Do

- × Allow hedging or pledging of equity
- × Re-price stock options
- × Offer perquisites or personal benefits beyond limited perquisites for new hires
- × Provide supplemental executive retirement plans

Compensation Objectives and Philosophy

Our mission is to transform the lives of patients with GDDs by applying our scientific leadership in the field of cellular metabolism and adjacent areas of biology. 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, life stage and industry peers. 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 that compete with us for executive talent. Our compensation program also provides annual performance-based compensation with targets at the 50th percentile of our peer group and based on performance against objectives including, among other things, 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.

In addition, we provide a significant portion of our executive compensation in the form of equity compensation through the grants of stock options, RSUs that vest over time, and PSUs that vest upon the achievement of specified corporate or stock price milestones. We believe our long-term incentive awards facilitate retention and aligns the interests of our NEOs 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. Our compensation committee considers a number of factors when comparing to our peers in determining equity incentive compensation for our NEOs, including annual long-term incentive values, annual equity awards expressed as a percentage 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 view, further considering the achievement of company goals and how that impacts total stockholder return, as well as market data of our peer group, when determining actual award levels for the NEOs. 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.

Overview of Executive Compensation Process

As a part of determining NEO performance and compensation, our compensation committee receives recommendations from our chief executive officer (except with respect to his or her own performance and compensation). Our chief executive officer's performance and compensation is approved by our board of directors 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 2020, the compensation committee utilized the services of Radford, which is part of the Rewards Solutions practice at Aon plc, 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;
- 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. 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 2020, the compensation committee approved 2020 base salaries, 2019 performance-based cash incentive awards and annual stock option and RSU awards for our continuing NEOs at that time, other than Mr. Biller and Dr. Car. Mr. Biller, who joined the company as chief legal officer effective December 3, 2019, and Dr. Car, who joined the company as chief

scientific officer effective January 6, 2020, received stock option, RSU and PSU awards in connection with the start of their employment but did not receive a 2019 annual performance-based cash incentive award or annual equity awards in 2020.

In February 2021, the compensation committee approved 2021 base salaries, 2020 performance-based cash incentive awards and annual stock option, RSU and PSU awards for our NEOs, other than Mr. Hirsch, who resigned effective September 11, 2020.

The terms of the arrangements with Dr. Car and Mr. Hirsch are described in more detail below under “Employment, Severance and Change in Control Arrangements—*Employment Offer Letters*” and “Employment, Severance and Change in Control Arrangements —*Severance Benefits Plan*”, respectively.

Use of Comparator Peer Group

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

In developing the peer group of companies to inform 2020 compensation decisions, our compensation committee, with the assistance of Radford, established a peer group of 14 publicly traded, national and regional companies in the biopharmaceutical industry based on a balance of the following criteria:

- companies with a market capitalization of between \$900 million and \$10.5 billion, approximately 200 to 1,700 employees, and an early commercial-stage portfolio with a focus on orphan diseases;
- 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 2020 was comprised of the following companies:

ACADIA Pharmaceuticals Inc.	Exelixis, Inc.	Portola Pharmaceuticals, Inc.
Alnylam Pharmaceuticals, Inc.	FibroGen, Inc.	Sage Therapeutics, Inc.
Amicus Therapeutics, Inc.	Intercept Pharmaceuticals, Inc.	Sarepta Therapeutics, Inc.
bluebird bio, Inc.	Nektar Therapeutics	Ultragenyx Pharmaceutical Inc.
Blueprint Medicines Corporation	Neurocrine Biosciences, Inc.	

The compensation committee believes the compensation practices of our peer group provide us with appropriate compensation benchmarks for evaluating the compensation of our NEOs. 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.

For the purposes of informing 2021 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 2021 peer group:

ACADIA Pharmaceuticals Inc.	Blueprint Medicines Corporation	Intercept Pharmaceuticals, Inc.
Accelaron Pharma, Inc.*	Exelixis, Inc.	Nektar Therapeutics
Amicus Therapeutics, Inc.	FibroGen, Inc.	Sage Therapeutics, Inc.
bluebird bio, Inc.	Global Blood Therapeutics, Inc.*	Ultragenyx Pharmaceutical Inc.

*addition to 2021 peer group

The 2021 peer group represents a group of biopharmaceutical companies more similar to us in key measures than the list we used in 2020, in light of the current state of our development pipeline, focusing on companies with approximately 225 to 2,000 employees, a market capitalization between \$1.0 billion and \$14 billion, and an early commercial-stage portfolio with a focus on orphan diseases. Specifically, Portola Pharmaceuticals, Inc. was removed from the 2021 peer group because it was acquired, and Alnylam Pharmaceuticals, Inc., Neurocrine Biosciences, Inc. and Sarepta Therapeutics, Inc. were removed from the 2021 peer group because their market capitalization moved outside of our targeted range. Accelaron Pharma, Inc. and Global Blood Therapeutics, Inc. were added to the 2021 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 NEOs, in alignment with our pay-for-performance philosophy and the long-term interests of stockholders. After reviewing information provided by our compensation consultant and other relevant data, 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 NEOs 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 incentivize 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 or upon the achievement of significant corporate milestones, 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 NEOs. Base salaries for our NEOs typically are established through arm's length negotiation at the time the NEO is hired, taking into account the position for which the NEO is being considered and the NEO's qualifications, prior experience and salary expectations. None of our NEOs is currently party to an employment agreement that provides for automatic or scheduled increases in base salary and we do not provide formulaic base salary increases to our NEOs. However, on an annual basis, our compensation committee reviews and evaluates, with input from our chief executive officer (other than with respect to herself), the need for adjustment of the base salaries of our NEOs based on changes and expected changes in the scope of a NEO's responsibilities, including promotions, the individual contributions made by and performance of the NEO during the prior year, the NEO'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 NEO'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.

Based on a review of market data provided by Radford, the current compensation levels of our NEOs and company performance and individual contributions, in February 2020, our compensation committee approved 2020 salary increases for Dr. Fouse, Dr. Bowden and Mr. Hirsch. Mr. Biller and Dr. Car assumed their roles on December 3, 2019 and January 6, 2020, respectively, and did not receive an annual base salary increase for 2020. The 2019 and 2020 annual base salaries of each of our NEOs were:

	2019	2020	
	<u>Base Salary (\$)</u>	<u>Base Salary (\$)</u>	<u>% Change</u>
Jacquelyn A. Fouse, Ph.D.	725,000	746,750	3.0%(1)
Jonathan Biller(2)	500,000	500,000	0.0%
Christopher Bowden, M.D.	490,539	515,007	5.0% (3)
Bruce Car, Ph.D.(4)	—	500,000	N/A
Andrew Hirsch(5)	500,476	515,490	3.0%(1)

(1) Increase reflects merit-based adjustment.

(2) Mr. Biller was appointed chief legal officer effective December 3, 2019, and was appointed chief

financial officer, head of legal and corporate affairs effective September 11, 2020.

- (3) Increase reflects merit- and market-based adjustments.
- (4) Dr. Car was appointed chief scientific officer effective January 6, 2020.
- (5) Mr. Hirsch resigned as chief financial officer effective September 11, 2020.

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 NEOs 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 NEOs 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 NEO 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 2020 goals and relative weighting assigned to each goal, as considered by our compensation committee and board of directors in their respective assessment of company performance in 2020.

- Maximize patient impact and value creation of IDH inhibitors globally: *relative weighting 35%*
 - Achieve target for annual net sales of TIBSOVO®;
 - Achieve enrollment target for select clinical trials by year-end; and
 - Complete certain activities related to plans for evaluating regulatory strategy for TIBSOVO® in R/R AML by the end of the first quarter and for cholangiocarcinoma by the end of the year.
- Drive PKR program development, building global leadership in PKD while advancing thalassemia and SCD: *relative weighting 35%*
 - Deliver topline pivotal clinical trial data to support regulatory strategy for mitapivat in PKD by year-end;
 - Achieve proof of concept for mitapivat in SCD by the end of the second quarter;
 - Complete certain activities related to plans for evaluating clinical and regulatory strategy for mitapivat in the second and third quarters;
 - Achieve enrollment target for phase 1 clinical trial of AG-946 by year-end; and
 - Present phase 2 clinical data for mitapivat in thalassemia by the end of the second quarter.
- Drive towards clinical proof-of-concept for early clinical programs (MAT2A, DHODH): *relative*

weighting 15%

- Achieve specified clinical trial milestone for phase 1 dose escalation and expansion trial by year-end; and
- Complete certain activities related to evaluation of clinical development plans for AG-636 by year-end.
- Advance our discovery pipeline in our three focus areas through our own efforts and with external partners: *relative weighting 15%*
- Achieve specified number of development candidates and validated programs by year-end.

Certain of these corporate goals include highly sensitive and competitive data, including pre-clinical, clinical and financial targets. We do not disclose the specific portions of these goals because we believe that such disclosure would result in competitive harm to us. We purposely set these goals at challenging levels. Revealing certain elements of these goals could potentially reveal insights about our pre-clinical, clinical, regulatory and strategic plans or objectives that our competitors or potential collaborators could use against us.

In addition to these goals, our compensation committee and board of directors evaluated the company's performance with respect to maintaining financial strength and developing our strategy, organization and culture to ensure execution of our long-term vision. These factors included managing our financials in line with board-approved budget; cultivating our human capital by attracting, developing and retaining talented employees while managing employee attrition; and maintaining and improving organizational health and resiliency. In evaluating these factors, our compensation committee and board of directors apply a discretionary modifier of 80% to 120% to the company's weighted goal achievement in order to arrive at our overall company performance score for the year.

In December 2020, our chief executive officer recommended to our compensation committee that our company's performance against 2020 goals be assessed based on achievements against these goals during the year, which achievements are discussed above under "Compensation Discussion and Analysis—*Executive Summary*". Based on the company's achievement against our 2020 goals, the overall company performance score for 2020 was determined to be 105% by our board of directors upon the recommendation of the compensation committee. Below is our relative weighted performance against our 2020 goals, as determined by our board of directors.

2020 Corporate Goals	Weighting	Assessment (out of 100%)	Weighted Performance
<i>Maximize patient impact and value creation of IDH inhibitors globally</i>	35%	90%	31%
<i>Drive PKR development, building global leadership in PKD while advancing thalassemia and SCD</i>	35%	128%	45%
<i>Drive towards clinical proof-of-concept for early clinical programs</i>	15%	50%	8%
<i>Advance discovery pipeline in our three focus areas through our own efforts and with external partners</i>	<u>15%</u>	<u>110%</u>	<u>16%</u>
Total	100%		100%
<i>Maintain financial strength and develop strategy, organization and culture to ensure execution of company's long-term vision (80%-120% multiplier)</i>		105%	105%
Adjusted Company Performance Score = Total Weighted Performance X Multiplier			105%

Our compensation committee also evaluates the individual performance of our NEOs, with the input of our chief executive officer in the case of the evaluation of our other NEOs 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 Dr. Fouse in 2020 based on our relative achievement of our corporate goals as well as her leadership in driving the execution of our strategic plans.

In assessing the individual performance of our NEOs other than our chief executive officer and Mr. Hirsch (who resigned effective September 11, 2020), 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 2020, Mr. Biller led our legal function and, effective September 11, 2020, our finance, accounting and investor relations functions, facilitating the sale of our IDHIFA[®] royalty rights to RPI, managing the successful negotiation of a definitive agreement to sell our oncology business to Servier, and making significant progress in developing our legal, finance and investor relations functions.
- In 2020, Dr. Car led our research and discovery science function, advancing multiple research efforts that yielded development candidates and validated programs and elucidating the potential of our PKR activator program across multiple hemolytic anemias.
- In 2020, Dr. Bowden led our clinical and regulatory teams responsible for: completing database lock and a positive topline data readout for our phase 3 trial ACTIVATE; completing the final overall survival analysis for our ClarIDHy trial; achieving clinical proof-of-concept for mitapivat in SCD based on phase 1 trial results; and continuing enrollment across our ongoing clinical trials, including our phase 3 trials INDIGO and AGILE.

Based on company and individual performance, our board of directors approved, upon the recommendation of the compensation committee, the 2020 cash incentive payment for our chief executive officer, and the compensation committee approved the 2020 cash incentive payments for our other NEOs, as follows:

	Target Award as a Percentage of Base Salary	2020 Actual Cash Incentive Payment (\$)	2020 Actual Cash Incentive Payment (% of Target Award)
Jacquelyn A. Fouse, Ph.D.	65%	509,657	105%
Jonathan Biller	45%	258,750	115%
Christopher Bowden, M.D.	45%	243,341	105%
Bruce Car, Ph.D.	45%	236,250	105%
Andrew Hirsch(1)	45%	231,971	100%

- (1) Mr. Hirsch resigned as our chief financial officer effective September 11, 2020 and received his 2020 target annual cash incentive payment in connection with his departure pursuant to the terms of the Severance Benefits Plan described below under “—*Employment, Severance and Change in Control Arrangements—Benefits Provided Upon Termination Not in Connection with a Change in Control.*”

The 2020 cash incentive payments for all employees, including the NEOs, were paid in 2021.

As discussed below under “—2021 Executive Compensation Decisions”, the annual cash incentive target award percentages for 2021 performance for our continuing NEOs remain unchanged from 2020.

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 stockholder value; similarly, the PSU 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 option, RSU and PSU awards provide meaningful incentives to our executives that increase 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 with us during the vesting period.

Equity compensation represents the largest at-risk component of our NEOs’ compensation arrangements. We believe that it is appropriate to align the interests of our NEOs with those of our stockholders to achieve and sustain long-term stock price appreciation. We typically use stock option, RSU and PSU awards to compensate our NEOs in the form of initial grants in connection with the commencement of employment, and generally grant stock options and RSUs on an annual basis thereafter. In addition, from time to time we grant certain employees, including our NEOs, supplemental RSU and/or PSU awards in order to further promote retention and emphasize individual employees’ impact on our organizational success.

We grant equity awards to our NEOs with both time-based and performance-based vesting. The stock options that we grant to our NEOs 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 NEOs vest in equal annual installments on each anniversary of the date of grant, until the third anniversary of such date. The PSU awards that we have granted to our NEOs vest in connection with the achievement of specified company performance or stock price milestones. 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 upon 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 RSU and PSU awards ceases upon termination of employment.

In specified termination and change in control circumstances, equity awards held by our NEOs are subject to accelerated vesting. See “—Severance and Change in Control Benefits” below for further information.

Annual Equity Grants

Our compensation committee determined that the mix for the 2020 annual equity grants to our NEOs (other than our chief executive officer) be split approximately 75% in stock options and 25% in RSUs, based on value. The compensation committee believes that this equity mix helps to ensure that compensation remains tied to stock performance and achievement of corporate milestones (through stock options) and promotes retention (via RSUs that vest over time to deliver equivalent value to stock options while using fewer authorized shares). None of our NEOs are currently party to an employment agreement that provides for an automatic award of equity grants, other than initial equity awards made in connection with the start of employment.

In determining the size of the annual stock option and RSU grants to our NEOs, 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 NEOs 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. 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.

Our compensation committee, or our board of directors, as applicable, made annual equity incentive awards in the form of stock options and RSUs to our NEOs in the amounts set forth in the table below during 2020. In the case of each award, these grants were based on the NEO’s existing equity incentive holdings, level of responsibility within our company, equity ownership in relation to the peer group benchmark, and the compensation committee’s assessment (or our board of directors’ assessment, in the case of our chief executive officer) of the NEO’s individual performance and our overall company performance, in each case without reference to any specific metric.

	<u>2020 Annual Stock Option Awards(1)</u>	<u>2020 Annual RSU Awards(2)</u>
Jacquelyn A. Fouse, Ph.D.	137,000	34,000
Jonathan Biller(3)	—	—
Christopher Bowden, M.D.	45,000	12,000
Bruce Car, Ph.D.(4)	—	—
Andrew Hirsch(5)	45,000	12,000

- (1) Effective February 14, 2020, our compensation committee, or board of directors, as applicable, approved the grant of these stock option awards to our NEOs, other than Mr. Biller and Dr. Car. The options have an exercise price of \$51.51 per share, the closing price on the date of grant, and are subject to 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.
- (2) Effective February 14, 2020, our compensation committee, or board of directors, as applicable, approved the grant of these RSU awards to our NEOs, other than Mr. Biller and Dr. Car. The RSUs are subject to 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.
- (3) Mr. Biller was appointed as our chief legal officer effective December 3, 2019 and did not receive an annual equity incentive award in 2020. For information about his new hire equity grants, see “—*New Hire Equity Grants*” below.
- (4) Dr. Car was appointed as our chief scientific officer effective January 6, 2020 and did not receive an annual equity incentive award in 2020. For information about his new hire equity grants, see “—*New Hire Equity Grants*” below.
- (5) Mr. Hirsch resigned as chief financial officer effective September 11, 2020.

New Hire Equity Grants

Mr. Biller joined the company as chief legal officer effective December 3, 2019 and Dr. Car joined the company as chief scientific officer effective January 6, 2020. In connection with the start of Mr. Biller’s and Dr. Car’s employment, the compensation committee granted them equity awards in the form of stock options, RSUs and PSUs, as reflected in the table below.

	New-Hire Stock Option Awards(1)	New-Hire RSU Awards(2)	New Hire PSU Awards(3)
Jonathan Biller	78,824	17,259	25,568
Bruce Car, Ph.D.	47,007	10,311	20,622

- (1) Reflects options granted to Mr. Biller and Dr. Car on December 3, 2019 and January 6, 2020, respectively. The options have an exercise price of \$39.11 per share and \$48.49 per share, respectively, the closing price on the date of grant. The options are subject to 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.
- (2) Reflects RSUs granted to Mr. Biller and Dr. Car on December 3, 2019 and January 6, 2020, respectively. The RSUs are subject to 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.
- (3) Reflects PSUs granted to Mr. Biller and Dr. Car on December 3, 2019 and January 6, 2020, respectively. The PSUs vest as to one-third of the underlying shares of common stock upon the achievement of each of three specified research, regulatory and commercial milestones, subject to continued service.

Performance Share Units (PSUs)

In addition to annual stock option and RSU grants, we occasionally grant PSUs to our executive officers when we want to further align executive incentives with critical value drivers for the business. We have multiple PSU programs for our executives under which PSUs will vest only after the achievement of specified performance or stock price milestones, as described below.

2019 PSU Program

The PSUs granted to Dr. Fouse at the start of her employment vest if, during the five-year period beginning on February 1, 2019 and ending on February 1, 2024, the closing price of the issuer's common stock equals or exceeds \$120 per share for at least 20 consecutive trading days at any time during such five-year period.

The other PSUs that we granted to our NEOs during 2019, and to Dr. Car upon the start of his employment in January 2020, vest as to one-third of the underlying shares upon our compensation committee's determination of the achievement of each of the following milestones, the performance period for which expires on December 31, 2022:

- Meeting our internal forecast for sales of TIBSOVO® in the untreated AML population over the 12 months following FDA approval of a company-sponsored sNDA submission for TIBSOVO® as a monotherapy for the treatment of patients with newly diagnosed AML (*milestone deemed achieved by the compensation committee in April 2020*);
- Receipt of written notice by the FDA of its approval of a new drug application for a company-sponsored submission for an investigational drug in the company's PKR program (*milestone not yet achieved*); and
- Receipt of written notice by the FDA of its acceptance of a company-sponsored investigational new drug application for a development candidate with a new mechanism of action for the company (*milestone not yet achieved*).

2021 PSU Program

The PSUs that we granted to our NEOs in February 2021 vest as to one-half of the underlying shares upon our compensation committee's determination of the achievement of each of the following milestones, the performance period for which expires on December 31, 2024:

- Exceeding our internal target for the number of PKD patients treated with mitapivat in the 12 months from start of U.S. commercial launch (*milestone not yet achieved*); and
- Meeting the primary endpoint in a pivotal trial of mitapivat in thalassemia (*milestone not yet achieved*).

We believe that PSUs further increase the performance orientation of our executive compensation program and that the milestones under our 2021 PSU program and the remaining milestones under our 2019 PSU program align with the strategic evolution of our company following the sale of our oncology business to Servier.

2021 Executive Compensation Decisions

In February 2021, our compensation committee, and our board, as applicable, approved 2021 base salaries, target annual performance-based cash incentive levels and annual stock option, RSU and PSU awards for our NEOs, other than Mr. Hirsch who resigned in September 2020, as set forth in the table below.

	Target Bonus Award as a Percentage of 2021 Base Salary	Change in Target Bonus Award from 2020	2021 Base Salary (\$)	% Base Salary Increase over 2020	2021 Equity Incentive Awards (Options)(1)	2021 Equity Incentive Awards (RSUs)(2)	2021 Equity Incentive Awards (PSUs)(3)
Jacquelyn A. Fouse, Ph.D.	65%	—	760,000	1.8%(4)	163,000	—	35,000
Jonathan Biller	45%	—	530,140	6.0%(5)	44,000	11,000	10,000
Christopher Bowden, M.D.	45%	—	530,547	3.0%(4)	44,000	11,000	10,000
Bruce Car, Ph.D.	45%	—	515,000	3.0%(4)	44,000	11,000	10,000

- (1) Effective February 10, 2021, our compensation committee, or board of directors, as applicable, approved the grant of these stock option awards at an exercise price of \$56.68 per share, the closing price on the date of grant. The options are subject to 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.
- (2) Effective February 10, 2021, our compensation committee, or board of directors, as applicable, approved the grant of these RSUs. The RSUs are subject to 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.
- (3) Effective February 10, 2021, our compensation committee, or board of directors, as applicable, approved the grant of these PSUs. The PSUs vest as to one-half of the underlying shares of common stock upon the achievement of each of two specified clinical and commercial milestones, subject to continued service.

- (4) Increase reflects merit-based adjustment.
- (5) Increase reflects merit-based adjustment and recognition of the additional responsibilities of Mr. Biller upon his appointment to chief financial officer, head of legal and corporate affairs effective September 11, 2020.

Salary increases for 2021 were made effective as of January 1, 2021. The annual cash incentive target award percentage for our continuing NEOs remained unchanged from 2020. Annual performance-based cash incentive program payouts for 2021 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 NEOs and certain other employees, and which became effective as of April 22, 2016. The Severance Plan provides for severance benefits in the event of a termination of such NEO'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 NEOs. Specifically, the Severance Plan eliminates single-trigger vesting on all equity grants made to our NEOs after April 22, 2016 and provides that all unvested equity awards shall vest in full if a NEO's employment is terminated by an acquirer or us without cause or by such NEO for good reason, each within 18 months following a change in control.

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 NEOs. We also have provided estimates of the value of the severance payments made and other benefits provided to our NEOs 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 NEOs 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 \$19,500 in 2020, and have the amount of the reduction contributed to the 401(k) plan. Participants who turned age 50 in 2020 were also eligible to make “catch-up” contributions, which in 2020 may be up to an additional \$6,500 above the statutory limit. In 2020, we 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 stockholder return. If the incentive-based compensation is based on our stock price or total stockholder 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 stockholder 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

Consistent with our pay-for-performance philosophy, we provide only limited perquisites to our executives in connection with new hires. We do not provide personal perquisites such as automobile leases, driver services or provide aircraft for personal use.

Anti-Hedging and Pledging Policy

Our insider trading policy expressly prohibits all of our employees, including our NEOs, 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 NEOs.

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, 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) 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 NEOs 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

Summary Compensation Table

The following table shows information regarding the compensation of our NEOs during the fiscal years ended December 31, 2020, 2019 and 2018.

Name and Principal Position	Year	Salary (\$)	Bonus (\$)	Stock Awards (\$)(1)	Option Awards (\$)(1)	Non-equity Incentive Plan Compensation (\$)(2)	All Other Compensation (\$)(3)	Total (\$)
Jacqualyn A. Fouse, Ph.D. <i>Chief Executive Officer</i>	2020	746,750	—	1,751,340	4,593,651	509,657	10,577	7,611,975
	2019	664,583	—	2,538,474	12,875,608	424,125	105,204	16,607,994
	2018	—	—	—	—	—	—	—
Jonathan Biller <i>Chief Financial Officer, Head of Legal and Corporate Affairs</i>	2020	500,000	—	—(4)	—(4)	258,750	10,578	769,328
	2019	39,583(5)	110,000(5)	1,008,334	2,025,036	—	936	3,183,889
	2017	—	—	—	—	—	—	—
Christopher Bowden, M.D. <i>Chief Medical Officer</i>	2020	515,007	—	618,120	1,508,875	243,341	9,203	2,894,546
	2019	490,539	—	915,590	1,609,989	198,668	12,422	3,227,208
	2018	467,141	—	641,025	1,746,517	262,767	11,280	3,128,730
Bruce Car, Ph.D.(6) <i>Chief Scientific Officer</i>	2020	494,318	—	833,301	1,499,943	236,250	164,231	3,228,043
	2019	—	—	—	—	—	—	—
	2018	—	—	—	—	—	—	—
Andrew Hirsch <i>Former Chief Financial Officer</i>	2020	361,234	—	618,120	1,508,875	—	397,893(7)	2,886,122
	2019	500,476	—	1,923,145	1,609,989	202,693	12,528	4,248,831
	2018	471,329	—	641,025	1,746,517	265,123	12,864	3,136,858

- 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 15, Share-Based Payments, of the Notes to our Consolidated Financial Statements in our Annual Report on Form 10-K, filed with the SEC on February 25, 2021. For Dr. Car, the 2020 amount in the “Stock Awards” column also reflects the grant date fair value of PSUs granted in connection with the start of his employment, based upon the probable outcome at the time of grant. The value of Dr. Car’s 2020 PSU award at the grant date assuming that the highest level of performance conditions was achieved would be \$999,961. For PSUs granted in 2019, the amounts in the “Stock Awards” column includes the grant date fair value of such awards based upon the probable outcome at the time of grant. The value of the 2019 PSU awards for Dr. Fouse, Mr. Hirsch and Dr. Bowden at the grant date assuming that the highest level of performance conditions were achieved would be \$2,299,956, \$1,000,913 and \$981,033, respectively. The value of the 2019 PSU award for Mr. Biller at the grant date assuming that the highest level of performance conditions was achieved would be \$999,964.
- Amounts represent awards to our NEOs 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.
- For 2020, amounts include a matching contribution under the company’s 401(k) plan for Dr. Fouse, Mr. Biller, Dr. Bowden, Dr. Car and Mr. Hirsch of \$9,957, \$9,958, \$8,583, \$4,167 and \$11,200 respectively. Amounts also include the dollar value of company-paid life insurance and disability insurance premiums paid during the fiscal year for the NEO.
- Mr. Biller joined the company as chief legal officer effective December 3, 2019, and as a result did not receive an annual equity award in 2020.

- (5) Mr. Biller's 2019 annual base salary was \$500,000, which was pro-rated in 2019 for the period in which he served. In addition, Mr. Biller received a sign-on bonus of \$110,000 in 2019.
- (6) Dr. Car's 2020 annual base salary was \$500,000; however, as Dr. Car joined the company as chief scientific officer effective January 6, 2020, his base salary reported was pro-rated in 2020. In addition, Dr. Car received stock option, RSU and PSU awards upon hire. In addition to the items set forth in footnote (3) above, the amount in the "All Other Compensation" column for Dr. Car includes \$159,454 in relocation assistance provided by the Company in connection with the start of his employment.
- (7) Mr. Hirsch resigned as our chief financial officer effective September 11, 2020. In connection with his resignation, he received severance benefits under the Severance Benefits Plan, as described below under "*—Employment, Severance and Change in Control Arrangements—Benefits Provided Upon Termination Not in Connection with a Change in Control.*" In addition to the items set forth in footnote (3) above, the amount in the "All Other Compensation" column for 2020 includes the following 2020 compensation paid to Mr. Hirsch subsequent to his resignation, pursuant to the terms of the Severance Benefits Plan: \$154,256 in salary and \$231,971 in bonus.

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 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, 2020, our principal executive officer was our chief executive officer Dr. Jacquelyn Fouse. For the fiscal year ended December 31, 2020, the total compensation for Dr. Fouse, as reported in the Summary Compensation Table above, was **\$7,611,975**. For the fiscal year ended December 31, 2020, the total compensation for our median employee (identified as disclosed below) was **\$253,211**, resulting in a pay ratio of approximately **30:1**.

Consistent with Instruction 2 to Item 402(u) of Regulation S-K, the applicable SEC rule, we may identify our median employee for purposes of providing such pay ratio disclosure once every three years and calculate and disclose total compensation for that employee each year; provided that, during the last completed fiscal year, there has been no change in the employee population or employee compensation arrangements that we reasonably believe would result in a significant change to the pay ratio disclosure. We identified the median employee for 2019 as of October 1, 2019 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 2019, and (C) the estimated fair value of any equity awards granted during the fiscal year ended December 31, 2019 and, (ii) ranking this aggregated compensation measure for our employees from lowest to highest. Amounts paid in currencies other than U.S. Dollars were converted based on the average annual exchange rate as of October 1, 2019. This calculation was performed for all employees, excluding Dr. Fouse. We have reviewed the changes in our employee population and employee compensatory arrangements during 2020 and, based on that review, determined that there has been no change in our employee population or employee compensatory arrangements that would significantly impact the pay ratio disclosure and require us to identify a new median employee. The median employee whose compensation was disclosed in our prior year proxy statement received a promotion during 2020 that resulted in that employee's compensation being anomalous. We have substituted that employee with a different employee whose compensation is substantially similar to the original median employee based on the compensation measure used to select the original median employee described above.

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.

Grants of Plan-Based Awards for 2020

The following tables sets forth information concerning each grant of an award made to a NEO during the fiscal year ended December 31, 2020 under any plan, contract, authorization or arrangement pursuant to which cash, securities, similar instruments or other property may be received. The cash awards were made under our annual cash incentive program and the equity awards were made under our 2013 Stock Incentive Plan. Each grant was authorized by our compensation committee, or board of directors, as applicable. For more information on equity acceleration benefits under specified circumstances, see “—Employment, Severance and Change in Control Arrangements.”

Name	Date of Grant	Estimated Future Payouts Under Non-Equity Incentive Plan Awards(1)			Estimated Future Payouts Under Equity Incentive Plan Awards			All Other Stock Awards: Number of Shares of Stock or Units(#)	All Other Option Awards: Number of Securities Underlying Options (#)(2)	Exercise or Base Price of Option Awards (\$)(3)	Grant Date Fair Value of Stock and Option Awards (\$)(4)
		Threshold (\$)	Target (\$)	Maximum (\$)	Threshold (#)	Target (#)	Maximum (#)				
Jacquelyn A. Fouse, Ph.D.	2/14/2020	—	485,388	728,081				34,000(5)			1,751,340
	2/14/2020								137,000	51.51	4,593,651
Jonathan Biller		—	225,000	337,500							
Christopher Bowden, M.D.	2/14/2020	—	231,753	347,630				12,000(5)			618,120
	2/14/2020								45,000	51.51	1,508,875
Bruce Car, Ph.D.(6)	1/06/2020	—	225,000	337,500				10,311(5)			499,980
	1/06/2020				6,874(7)	20,622(7)	— (7)				333,320
	1/06/2020								47,007	48.49	1,499,943
Andrew Hirsch(8)	2/22/2019	—	231,971	347,956				12,000(5)			618,120
	2/14/2020								45,000	51.51	1,508,875

- (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) Options subject to time-based vesting criteria established by the compensation committee and described in the footnotes to the Outstanding Equity Awards at Fiscal Year End table below.
- (3) The exercise price per share of these stock options is equal to the closing price of our common stock on the grant date.
- (4) 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 15, Share-Based Payments, of the Notes to our Consolidated Financial Statements in our Annual Report on Form 10-K, filed with the SEC on February 25, 2021. For PSUs, these amounts reflect the grant date fair value of such awards based upon the probable outcome at the time of grant.

- (5) RSUs subject to time-based vesting criteria established by the compensation committee and described in the footnotes to the Outstanding Equity Awards at Fiscal Year End table below.
- (6) Dr. Car joined the company as chief scientific officer effective January 6, 2020, and received stock option, RSU and PSU awards upon hire.
- (7) PSUs vest upon the achievement of specified corporate milestones established by the compensation committee and described in the footnotes to the Outstanding Equity Awards at Fiscal Year End table below. Threshold payout assumes one-third of the specified performance vesting criteria are achieved. Target payout assumes 100% of the specified performance vesting criteria are achieved.
- (8) Mr. Hirsch resigned as our chief financial officer effective September 11, 2020. Following his resignation, Mr. Hirsch served as a strategic advisor to the company for approximately two months and, consequently, the exercise period for Mr. Hirsch's vested stock option awards was extended through January 29, 2021. Mr. Hirsch's unvested RSU and PSU awards were forfeited at the time of his resignation.

Outstanding Equity Awards at Fiscal Year-End

The following table sets forth information concerning outstanding equity awards for each of our NEOs at December 31, 2020. For more information on equity acceleration benefits under specified circumstances, see “-Employment, Severance and Change in Control Arrangements.”

Name	Grant Date	Option Awards					Stock Awards				
		Number of Securities Underlying Unexercised Options Exercisable (#)	Number of Securities Underlying Unexercised Options (#)(1)	Equity Incentive Plan Awards: Number of Securities Underlying Unexercised Options (#)	Option Exercise Price (\$)	Option Expiration Date	Number of Shares or Units of Stock That Have Not Vested (#)(2)	Market Value of Shares or Units of Stock That Have Not Vested (#)(3)	Equity Incentive Plan Awards: Number of Unearned Shares, Units or Other Rights That Have Not Vested (#)(4)	Equity Incentive Plan Awards: Market or Payout Value of Unearned Shares, Units or Other Rights That Have Not Vested (#)(3)	
Jacquelyn A. Fouse, Ph.D.	12/03/2017	11,992	4,008	-	\$ 61.17	12/03/2027					
	05/31/2018	5,700	-	-	\$ 93.50	05/31/2028					
	02/01/2019	160,572	189,774	-	\$ 53.87	02/01/2029					
	02/01/2019	-	-	-	-	-	-	-	42,695(5)	1,849,974	
	04/11/2019	-	-	-	-	-	-	-	12,379(6)	536,382	
	02/14/2020	-	137,000	-	\$ 51.51	02/14/2030	-	-	-	-	
	02/14/2020	-	-	-	-	-	34,000	1,473,220			
Jonathan Biller	12/03/2019	19,706	59,118	-	\$ 39.11	12/03/2029					
	12/03/2019	-	-	-	-	-	11,506	498,555			
	12/03/2019	-	-	-	-	-			8,523(6)	369,302	
Christopher Bowden, M.D.	05/31/2014	15,723	-	-	\$ 35.16	05/30/2024					
	03/02/2015	17,000	-	-	\$107.89	03/01/2025					
	02/16/2016	45,220	-	-	\$ 39.76	02/15/2026					
	02/21/2017	45,616	1,984	-	\$ 50.40	02/20/2027					
	02/16/2018	23,368	9,632	-	\$ 77.70	02/16/2028					
	02/16/2018	-	-	-	-	-	2,750	119,158			
	02/22/2019	18,330	21,670	-	\$ 58.86	02/22/2029					
	02/22/2019	-	-	-	-	-	6,667	288,881			
	04/11/2019	-	-	-	-	-			5,280(6)	228,782	
	02/14/2020	-	45,000	-	\$ 51.51	02/14/2030					
	02/14/2020	-	-	-	-	-	12,000	519,960			
Bruce Car, Ph.D.	01/06/2020	-	47,007	-	\$ 48.49	01/06/2030					
	01/06/2020	-	-	-	-	-	10,311	446,776			
	01/06/2020	-	-	-	-	-			6,874(6)	297,850	
Andrew Hirsch(7)	09/20/2016	125,000	-	-	\$ 49.83	01/29/2021					
	02/21/2017	43,632	-	-	\$ 50.40	01/29/2021					
	02/16/2018	21,992	-	-	\$ 77.70	01/29/2021					
	02/22/2019	16,664	-	-	\$ 58.86	01/29/2021					

- (1) 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.
- (2) Represents RSUs, each unit representing a contingent right to receive one share of common stock. One-third of the shares underlying the units vest on the first, second and third anniversary of the grant date, subject to continued service.
- (3) Amounts shown are based on a price of \$43.33 per share, which was the closing price of our common stock as reported on the Nasdaq Global Select Market on December 31, 2020, the last trading day of the year.

- (4) These amounts represent the number of PSUs granted assuming threshold performance conditions are met.
- (5) Represents PSUs, 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.
- (6) Represents PSUs, each unit representing a contingent right to receive one share of common stock. One-third of the shares underlying the units vest upon the achievement of each of three specified research, regulatory and commercial milestones, as determined by our compensation committee. The first performance milestone was deemed to be achieved in April 2020. The performance period ends on December 31, 2022.
- (7) Following his resignation as chief financial officer in September 2020, Mr. Hirsch served as a strategic advisor to the company for approximately two months and, consequently, the exercise period for Mr. Hirsch's vested stock option awards was extended through January 29, 2021.

Option Exercises and Stock Vested in 2020

The following table sets forth information concerning option exercises and stock vested for each of our NEOs during the fiscal year ended December 31, 2020:

	Option Awards		Stock Awards	
	Number of Shares Acquired on Exercise(#)	Value Realized on Exercise\$(1)	Number of Shares Acquired on Vesting(#)	Value realized on Vesting\$(2)
Jacquelyn A. Fouse, Ph.D.	—	—	12,379	492,437
Jonathan Biller	—	—	14,276	606,157
Christopher Bowden, M.D.	55,277	825,410	11,363	528,707
Bruce Car, Ph.D.	—	—	6,874	273,448
Andrew Hirsch(3)	—	—	17,363	800,505

- (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) The value realized when the stock awards vested represents the number of shares underlying the units vested multiplied by market value of the shares on the vesting date.
- (3) Mr. Hirsch resigned as chief financial officer effective September 11, 2020.

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 NEOs and certain other officers and key employees. The Severance Plan provides for severance benefits in the event of a termination of such NEO'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 NEOs.

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 NEO's employment is terminated by us without cause or by such NEO 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 or her then-current monthly base salary for a period of 12 months and 100% of such NEO'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 NEOs 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 NEO and us. All equity awards held by NEOs granted prior to the effective date of the Severance Plan are fully vested.

Prior to September 11, 2020, Mr. Hirsch was eligible for the same benefits as described above. Following his resignation effective September 11, 2020, Mr. Hirsch was compensated under the Severance Plan in accordance with the terms set forth under "Benefits Provided Upon Termination Not in Connection with a Change in Control" above. The amounts Mr. Hirsch received under the Severance Plan are set forth in the Summary Compensation Table above and the notes thereto.

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 NEO's employment is terminated by us without cause or by such NEO for good reason within 18 months following a change in control:

- (i) we are obligated (A) to pay an amount equal to his or her then-current monthly base salary for a period of 12 months and 100% of such NEO's target annual cash incentive in a lump sum, with the exception of Dr. Fouse, who will be entitled to an amount equal to her then-current monthly base salary for a period of 24 months and 200% of her 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. Fouse, who will be entitled to such insurance for a period of 24 months; and
- (ii) any unvested equity awards shall become fully vested; provided that the treatment for such NEOs' 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 NEO and us. All equity awards held by NEOs granted prior the effective date of the Severance Plan are fully vested.

Prior to his resignation effective September 11, 2020, Mr. Hirsch was eligible for the same benefits as described above.

Employment Offer Letters

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

In connection with his appointment as chief scientific officer, we entered into an employee offer letter agreement with Dr. Car, effective January 6, 2020 providing for the terms of his employment, including (i) an annual base salary of \$500,000; (ii) an annual target bonus equal to 45% of his base salary; (iii) a one-time grant of an option to purchase 47,007 shares of common stock at an exercise price of \$48.49 per share, the closing price on the date of grant, which shall vest as to 25% of the underlying shares on January 6, 2021 and as to the remaining shares on a monthly basis thereafter in equal increments over 36 months; (iv) a one-time grant of 10,311 RSUs, each unit representing a contingent right to receive one share of common stock, which shall vest in equal annual installments on the first, second and third anniversaries of the date of grant; (v) a one-time grant of 20,622 PSUs, each unit representing a contingent right to receive one share of common stock, which shall vest as to one-third of the underlying shares of common stock upon the achievement of specified research, regulatory and commercial milestones; (vi) relocation assistance covering reasonable relocation costs; and (vii) 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 NEOs. Under the non-competition, non-solicitation, confidentiality and assignment agreements, each NEO 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 NEOs. The Severance Plan provides for severance benefits in the event of a termination of such NEO'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. See "*—Severance Benefits Plan*" for a further description of the terms of the Severance Plan. Receipt of any severance benefits under the Severance Plan require that the NEO 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 NEO releases and discharges us and our affiliates from and on account of any and all claims between us and the NEO.

The following table sets forth potential payments upon termination and change in control that would be made to our NEOs, other than Mr. Hirsch, assuming that such termination or change in control occurred on December 31, 2020, after giving effect to the Severance Plan. In addition to the amounts shown in the table below, each NEO would be entitled to receive payments for base salary through the date of termination and payment for any reimbursable business expenses incurred.

Name	Benefit	Triggering Event		
		Change in Control (Without Termination of Employment) (\$)	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 (\$)
Jacqualyn A. Fouse, Ph.D.	Severance Payments	—	746,750(1)	1,493,500(2)
	Bonus Payment	—	485,388(3)	970,775(4)
	Continuation of Benefits	—	13,119(5)	26,238(6)
	Market Value of Stock Vesting	—	—	4,396,002(7)
	Total	—	1,245,257	6,886,515
Jonathan Biller	Severance Payments	—	500,000(1)	500,000(1)
	Bonus Payment	—	225,000(3)	225,000(3)
	Continuation of Benefits	—	11,776(5)	11,776(5)
	Market Value of Stock Vesting	—	—	1,486,593(7)
	Total	—	736,776	2,223,369
Christopher Bowden, M.D.	Severance Payments	—	515,007(1)	515,007(1)
	Bonus Payment	—	231,753(3)	231,753(3)
	Continuation of Benefits	—	23,552(5)	23,552(5)
	Market Value of Stock Vesting	—	—	1,385,607(7)
	Total	—	770,311	2,155,918
Bruce Car, Ph.D.	Severance Payments	—	500,000(1)	500,000(1)
	Bonus Payment	—	225,000(3)	225,000(3)
	Continuation of Benefits	—	13,119(5)	13,119(5)
	Market Value of Stock Vesting	—	—	1,042,476(7)
	Total	—	738,119	1,780,595
Andrew Hirsch(8)	Severance Payments	—	515,490(1)	—
	Bonus Payment	—	231,971(3)	—
	Continuation of Benefits	—	17,664(5)	—
	Market Value of Stock Vesting	—	—	—
	Total	—	765,124	—

- (1) Represents 12 monthly payments of each executive's monthly base salary from the time of termination.
- (2) Represents 24 monthly payments of executive's monthly base salary from the time of termination.
- (3) Represents a lump sum payment equal to each executive's target annual cash incentive bonus.
- (4) Represents a lump sum payment equal to two years of executive's target annual cash incentive bonus.
- (5) Represents the cost of continued health and dental benefits. These benefits are payable until 12 months following termination.
- (6) Represents the cost of continued health and dental benefits. These benefits are payable until 24 months following termination.
- (7) Represents the acceleration of vesting as to 100% of the unvested equity awards held by the NEO. 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 and PSUs, the number of unvested RSUs and/or PSUs 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, 2020, or \$43.33 per share.

- (8) Mr. Hirsch resigned as chief financial officer effective September 11, 2020. Values in the table above show actual compensation to be received by Mr. Hirsch following his resignation pursuant to the Severance Plan.

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, 2020.

Equity Compensation Plan Information

Plan category	Number of securities to be issued upon exercise of outstanding options, warrants and rights	Weighted-average exercise price of outstanding options, warrants and rights	Number of securities remaining available for future issuance under equity compensation plans (excluding securities reflected in column (a))
	(a)	(b)	(c)
Equity compensation plans approved by security holders			
2007 Stock Incentive Plan	157,235	\$ 8.36	—
2013 Stock Incentive Plan	7,455,113	\$ 59.78(1)	2,971,884(2)
2013 Employee Stock Purchase Plan	—	—	471,353(3)
Equity compensation plans not approved by security holders	—	—	—
Total	7,612,348	\$ 58.46(1)	3,443,237

- (1) The calculation does not take into account the 1,284,378 shares of common stock subject to outstanding RSUs or the 184,924 shares of common stock subject to outstanding PSUs. Such shares will be issued at the time such awards vest, without any cash consideration payable for those shares.
- (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, 2021, 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. On January 1, 2021, 509,091 additional shares were reserved for issuance under the 2013 ESPP pursuant to this provision.

Compensation Committee Interlocks and Insider Participation

For 2020, 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, 2020.

CERTAIN RELATIONSHIPS AND RELATED PARTY TRANSACTIONS

Since January 1, 2020, 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 November 14, 2019, entities affiliated with Celgene Corporation, or Celgene, beneficially own more than 5% of our outstanding shares of common stock.

In April 2010, we entered into a collaboration agreement, or the 2010 Agreement, with Celgene. Pursuant to the 2010 Agreement, we were eligible to receive royalties at tiered, low-double digit to mid-teen percentage rates on net sales of IDHIFA®. On June 11, 2020, we sold such royalty rights to Royalty Pharma, Inc., or RPI. During the year ended December 31, 2020, we earned \$10.2 million in royalty revenue under the 2010 Agreement. Following the sale of our royalty rights to RPI, we remained eligible to receive a \$25.0 million milestone payment upon achievement of a specified ex-U.S. commercial milestone event.

Effective as of March 31, 2021, we transferred to Servier our rights and obligations under the 2010 Agreement, including the right to receive the remaining \$25.0 million milestone payment described above.

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 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.

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 NEOs 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 NEOs 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 34, 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 NEOs 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 NEOs, 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 NEOs. Our board of directors has adopted a policy of providing annual advisory votes to approve the compensation of our NEOs. The next advisory vote to approve the compensation of our NEOs will occur at our 2022 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 NEOs BY VOTING ‘FOR’ THIS PROPOSAL.

**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, 2021 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 participate in 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

The following table represents aggregate fees billed to us for services related to the fiscal years ended December 31, 2020 and 2019 by PwC.

	Fiscal Year Ended December 31,	
	2020	2019
Audit Fees(1)	\$1,360,000	\$1,287,000
Audit Related Fees(2)	—	—
Tax Fees(3)	26,155	111,963
All Other Fees(4)	6,356	6,356
Total	\$1,392,511	\$1,405,319

- (1) Audit fees consist of fees billed for professional services performed 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.

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 2020 and 2019 fiscal years, all of the services provided by PwC were pre-approved by our audit committee.

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, 2021.

STOCKHOLDER PROPOSALS

Proposals of stockholders 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 2022 must be received by us no later than December 9, 2021, which is 120 days prior to the first anniversary of the mailing date of this proxy, unless the date of the 2022 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 stockholders 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 2022 annual meeting of stockholders, such a proposal must be received by us no earlier than January 20, 2022 and no later than February 19, 2022. 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 earlier 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 2022 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:

William Cook, Corporate Secretary
Agius Pharmaceuticals, Inc.
88 Sidney Street
Cambridge, MA 02139

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 virtually attend the Annual Meeting. Whether or not you plan to attend the Annual Meeting virtually, 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.