UNITED STATES
SECURITIES AND EXCHANGE COMMISSION
WASHINGTON, D.C. 20549

FORM 8-K

CURRENT REPORT
PURSUANT TO SECTION 13 OR 15(D) OF
THE SECURITIES EXCHANGE ACT OF 1934

Date of report (Date of earliest event reported): January 28, 2020

ALEXION PHARMACEUTICALS, INC.

(Exact name of registrant as specified in its charter)

Delaware	**000-27756**	**13-3648318**
(State or other jurisdiction of incorporation or organization)	**(Commission File Number)**	**(I.R.S. Employer Identification No.)**

121 Seaport Boulevard, Boston, Massachusetts 02210

(Address of Principal Executive Offices) (Zip Code)

Registrant's telephone number, including area code: (475) 230-2596

Check the appropriate box below if the Form 8-K filing is intended to simultaneously satisfy the filing obligation of the registrant under any of the following provisions (*see* General Instruction A.2. below):

☐ Written communications pursuant to Rule 425 under the Securities Act (17 CFR 230.425)

☐ Soliciting material pursuant to Rule 14a-12 under the Exchange Act (17 CFR 240.14a-12)

☐ Pre-commencement communications pursuant to Rule 14d-2(b) under the Exchange Act (17 CFR 240.14d-2(b))

☐ Pre-commencement communications pursuant to Rule 13e-4(c) under the Exchange Act (17 CFR 240.13e-4(c))

Securities registered pursuant to Section 12(b) of the Act:

Title of each class	Trading Symbol(s)	Name of each exchange on which registered
Common Stock, par value $0.0001 per share	ALXN	Nasdaq Global Select Market

Indicate by check mark whether the registrant is an emerging growth company as defined in Rule 405 of the Securities Act of 1933 (§230.405 of this chapter) or Rule 12b-2 of the Securities Exchange Act of 1934 (§240.12b-2 of this chapter).

☐ Emerging Growth Company

If an emerging growth company, indicate by check mark if the registrant has elected not to use the extended transition period for complying with any new or revised financial accounting standards provided pursuant to Section 13(a) of the Exchange Act. ☐

Item 1.01 Entry into a Material Definitive Agreement.

As previously disclosed, on October 15, 2019, Alexion Pharmaceuticals, Inc. ("Alexion") entered into an Agreement and Plan of Merger with Beagle Merger Sub, Inc., a Delaware corporation and a wholly owned subsidiary of Alexion ("Merger Subsidiary"), and Achillion Pharmaceuticals, Inc. ("Achillion"), pursuant to which, among other things, upon the terms and subject to the conditions thereof, Merger Subsidiary merged with and into Achillion, with Achillion surviving as a wholly owned subsidiary of Alexion (the "Merger"). The Merger became effective on January 28, 2020. At the effective time of the Merger, each share of Achillion common stock (other than certain excluded shares as described in the Merger Agreement) automatically converted into the right to receive (1) $6.30 in cash, without interest, and (2) one CVR pursuant to the CVR Agreement (each as defined below).

In connection with its acquisition of Achillion, Alexion entered into a Contingent Value Rights Agreement (the "CVR Agreement") with Computershare Inc. as rights agent. Each contingent value right ("CVR") entitles its holder to receive a payment in cash of (1) $1.00 upon the achievement of a Clinical Trial Milestone (as defined in the CVR Agreement) relating to the development of Achillion's product candidate ACH-5228 prior to January 28, 2024 and (2) $1.00 upon Alexion's first receipt of approval by the FDA of a new drug application or other regulatory approval application which approval grants Alexion the right to market and sell Achillion's product candidate ACH-4471 in the United States prior to July 28, 2024.

The CVRs are not transferable except under certain limited circumstances described in the CVR Agreement, are not evidenced by a certificate or other instrument and are not registered or listed for trading. The CVRs do not have any voting or dividend rights and do not represent any equity or ownership interest in Alexion, Achillion or any of their affiliates. The maximum aggregate amount potentially payable by Alexion pursuant to the CVRs is approximately $306 million.

References to, and descriptions of, the CVR Agreement as set forth herein are not intended to be complete and are qualified in their entirety by the full text of the agreement, which is attached to this report as Exhibit 10.1, and is incorporated by reference in this Item 1.01.

Item 7.01 Regulation FD Disclosure.

On January 28, 2020, Alexion issued a press release announcing the consummation of the Merger. A copy of the press release is attached as Exhibit 99.1 hereto.

The information furnished under this Item 7.01, including Exhibit 99.1, shall not be deemed "filed" for purposes of Section 18 of the Securities Exchange Act of 1934, as amended (the "Exchange Act"), or otherwise subject to the liabilities under that section and shall not be deemed to be incorporated by reference into any filing under the Securities Act of 1933, as amended or the Exchange Act, except as expressly set forth by specific reference in such filing. In addition, Exhibit 99.1 contains statements intended as "forward-looking statements" that are subject to the cautionary statements about forward-looking statements set forth in such exhibit.

Item 9.01 Financial Statements and Exhibits.

(d) Exhibits.

Exhibit Number	Description
10.1	Contingent Value Rights Agreement dated as of January 28, 2020 among Alexion Pharmaceuticals, Inc. and Computershare Inc.
99.1	Press Release issued by Alexion Pharmaceuticals, Inc. dated January 28, 2020
104	Cover Page Interactive Data File (embedded within the Inline XBRL document)

Signature

Pursuant to the requirements of the Securities Exchange Act of 1934, the registrant has duly caused this report to be signed on its behalf by the undersigned hereunto duly authorized.

Date: January 28, 2020

ALEXION PHARMACEUTICALS, INC.

By: /s/ Doug Barry
Name: Doug Barry
Title: Vice President, Corporate Law

CONTINGENT VALUE RIGHTS AGREEMENT

Dated as of

January 28, 2020

among

Alexion Pharmaceuticals, Inc.

and

Computershare Inc.

as Rights Agent

B5092239.1

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CONTINGENT VALUE RIGHTS AGREEMENT

CONTINGENT VALUE RIGHTS AGREEMENT, dated as of January 28, 2020 (this "**Agreement**"), by and between Alexion Pharmaceuticals, Inc., a Delaware corporation ("**Parent**"), and Computershare Inc., as rights agent (the "**Rights Agent**").

WHEREAS, this Agreement is entered into pursuant to the Agreement and Plan of Merger, dated October 15, 2019 (the "**Merger Agreement**"), by and among Achillion Pharmaceuticals, Inc., a Delaware corporation (the "**Company**"), Parent and Beagle Merger Sub, Inc., a Delaware corporation wholly owned by Parent ("**Merger Subsidiary**"), pursuant to which Merger Subsidiary will merge with and into the Company (the "**Merger**") with the Company surviving (the "**Surviving Corporation**"), on the terms and subject to the conditions set forth therein;

WHEREAS, pursuant to and subject to the terms and conditions of the Merger Agreement, Parent has agreed to provide to the Holders of shares of common stock, par value $0.001 per share, of the Company (the "**Shares**") and Holders of In the Money Options (as defined in the Merger Agreement) (the "**Option Holders**") the right to receive the Regulatory Approval Milestone Payment and the Clinical Trial Milestone Payment (each, as defined below) during the Regulatory Approval Milestone Period and the Clinical Trial Milestone Period, respectively (each as defined below);

WHEREAS, pursuant to this Agreement, the potential amount payable per CVR with respect to the Regulatory Approval Milestone Payment is $1.00 in cash, without interest; and

WHEREAS, pursuant to this Agreement, the potential amount payable per CVR with respect to the Clinical Trial Milestone Payment is $1.00 in cash, without interest.

NOW, THEREFORE, in consideration of the foregoing and the consummation of the transactions referred to above, Parent and the Rights Agent agree, for the equal and proportionate benefit of all Holders (as hereinafter defined), as follows:

Article 1
DEFINITIONS

Section 1.1 *Definitions*. Capitalized terms used in this Agreement and not otherwise defined shall have the meanings assigned to them in the Merger Agreement. For purposes of this Agreement, the following terms shall have the following meanings:

"**Adaptive Trial**" means a Clinical Trial that is designed to allow for a prospectively planned modification from a Phase II Clinical Trial to a Phase III Clinical Trial based on accumulating data from subjects in such Clinical Trial (as permitted by the FDA).

"**Assignment Transaction**" means any transaction (including a sale of assets, spin-off, split-off or licensing transaction), other than a Change in Control, pursuant to which (a) any rights of Parent or any of its Affiliates (including Intellectual Property rights) (i) necessary for the development or commercialization of any Product or (ii) useful for the development or commercialization of any Product (other than, in the case of any useful but not necessary rights, to the extent that the applicable transaction would not reasonably be expected to result in a material delay in achievement of any of the Milestones) or (b) all or substantially all of the assets used or held for use in connection with any Product, in each case (in respect of the foregoing (a) and (b)) are, directly or indirectly, disposed of, sold, licensed, assigned, conveyed, or transferred to or acquired by any Person other than by Parent or any of Parent's direct or indirect wholly-owned subsidiaries (such Person, the "**Acquiror**"). An "Assignment Transaction" shall not apply to (a) sales of a Product made by Parent or its Affiliates to distributors in the ordinary course, or ordinary course licensing arrangements between Parent and its Affiliates, on the one hand, and third party distributors, contract research organizations or contract manufacturers on the other hand, entered into in the ordinary course of business for purposes of developing, manufacturing, distributing and selling a Product, in each case, on Parent's or its Affiliates' behalf and (b) licenses granted by Parent or its Affiliates so long as Parent and its Affiliates retain any and all necessary rights and useful rights (other than, in the case of any useful but not necessary rights, to the extent that the applicable license would not reasonably be expected to result in a material delay in achievement of any of the Milestones) to develop and obtain approval by the FDA and any other applicable Regulatory Authority to market and sell the Products. For purposes of this definition, any delay that reasonably would be expected to result in failure to achieve any of the Milestones hereunder shall be deemed to be a "material delay."

"**Board of Directors**" means the board of directors of Parent or any other body performing similar functions, or any duly authorized committee of that board.

"**Board Resolution**" means a copy of a resolution of the Board of Directors that has been certified in writing by the chairman of the Board of Directors, the chief executive officer, chief financial officer, executive vice president, company secretary or a deputy company secretary of Parent to have been duly adopted by the Board of Directors and to be in full force and effect on the date of such certification, which has been delivered to the Rights Agent.

"**Business Day**" means any day other than a Saturday or Sunday or a day on which commercial banks are authorized or required by Law or executive order to be closed in New York City.

"**Change in Control**" means (a) a merger or consolidation involving Parent in which Parent is not the surviving entity, (b) any transaction involving Parent in which Parent is the surviving entity but in which the stockholders of Parent immediately prior to such transaction own less than fifty percent (50%) of Parent's voting power immediately after the transaction or (c) any sale of all or substantially all of Parent's assets.

"**Clinical Trial**" means a clinical study of a pharmaceutical product conducted on human subjects.

"**Clinical Trial Milestone**" means, and will be deemed to occur upon, the earlier of (a) first dosing of the first patient with the Clinical Trial Product in the first Phase III Clinical Trial, (b) the Conversion Date for the first Converted Trial of the Clinical Trial Product (and, for clarity, the first dosing of the first patient in an Adaptive Trial prior to the Conversion Date shall not constitute dosing of a first patient in a Phase III Clinical Trial), and (c) the first submission of a new drug application to market and sell the Clinical Trial Product in the United States.

"Clinical Trial Milestone Payment" means $1.00 per CVR.

"**Clinical Trial Milestone Period**" means the period commencing as of the date of this Agreement and ending 11:59 p.m., Eastern Time, on that date that is four (4) years after the date of this Agreement.

"**Clinical Trial Product**" means any pharmaceutical product, including all forms, presentations, doses and formulations, containing the Company's compound, ACH-5228 or any salt, free-base, hydrate, solvate, polymorph, isomer, enantiomer, metabolite, prodrug or other derivative thereof, whether as the sole active ingredient or in combination with other active ingredients.

"**Commercially Reasonable Efforts**" means, with respect to each Product, using such efforts and resources typically used by Parent for the development and commercialization of similar products at similar development stages taking into account, as applicable, such Product's advantages and disadvantages, product profile, efficacy, safety, toxicity, tolerability, regulatory authority-approved labeling and pricing, the competitiveness of alternative products in the marketplace or under development, the current or future status as an orphan product, the patent coverage and proprietary position of such Product, the likelihood of development success or regulatory approval, the regulatory structure involved, the anticipated profitability of such Product, and other relevant scientific, technical and commercial factors typically considered in good faith by Parent in connection with such similar products. For clarity, Commercially Reasonable Efforts does not mean that either of Parent or any of its Affiliates guarantee either of the Milestones will be achieved or that either of the Milestones will be achieved by a specific date, and the fact that a Milestone is not actually achieved is not, in and of itself, dispositive evidence that Parent or any of its Affiliates did not in fact utilize its Commercially Reasonable Efforts in attempting to achieve such Milestone. For clarity, the application of Commercially Reasonable Efforts will not necessarily require Parent to disadvantage any particular currently available competing products or products currently under development by Parent or any of its Affiliates or which may in the future enter development by Parent or any of its Affiliates, the success of which may reduce the prospects of achieving the relevant Milestone. Any payments payable under this Agreement, including Milestone Payments, may not be taken into account in determining Commercially Reasonable Efforts.

"**Conversion Date**" means, with respect to a Converted Trial, the date when the first action specified in the protocol for the corresponding Adaptive Trial is taken following the decision to modify such Adaptive Trial to proceed as a Phase III Clinical Trial.

"**Converted Trial**" means an Adaptive Trial that is proceeding as a Phase III Clinical Trial following a prospectively planned modification from a Phase II Clinical Trial based on accumulating data from subjects in such Adaptive Trial.

"**CVRs**" means the contingent value rights of Holders to receive cash payments in the amounts and subject to the terms and conditions set forth in this Agreement.

"**Holder**" means a Person in whose name a CVR is registered in the CVR Register at the applicable time.

"**Majority Holders**" means, at the time of determination, Holders of at least the majority of the outstanding CVRs as set forth in the CVR Register.

"**Milestone(s)**" means the Clinical Trial Milestone and/or the Regulatory Approval Milestone, as applicable.

"**Milestone Payment(s)**" means the Clinical Trial Milestone Payment and/or the Regulatory Approval Milestone Payment, as applicable.

"**Milestone Payment Date**" means the date that is selected by Parent not more than fifteen (15) Business Days following the date of the achievement of the applicable Milestone.

"**Milestone Period**" means the Clinical Trial Milestone Period and/or the Regulatory Approval Milestone Period, as applicable.

"**Officer's Certificate**" means a certificate signed by the chief executive officer, chief financial officer, or an executive vice president, in each case of Parent, in his or her capacity as such an officer, and delivered to the Rights Agent or any other person authorized to act on behalf of Parent.

"**Opinion of Counsel**" means a written opinion of counsel, who may be counsel for Parent or its direct or indirect wholly-owned subsidiaries.

"**Party**" shall mean each of the Rights Agent and Parent.

"**Permitted Transfer**" means a transfer of CVRs (a) upon death of a Holder by will or intestacy; (b) by instrument to an *inter vivos* or testamentary trust in which the CVRs are to be passed to beneficiaries upon the death of the trustee, (c) pursuant to a court order; (d) by operation of Law (including by consolidation or merger) or without consideration in connection with the dissolution, liquidation or termination of any corporation, limited liability company, partnership or other entity; or (e) in the case of CVRs payable to a nominee, from a nominee to a beneficial owner (and, if applicable, through an intermediary) or from such nominee to another nominee for the same beneficial owner, in each case to the extent allowable by The Depository Trust Company ("**DTC**").

"**Phase II Clinical Trial**" means a Clinical Trial of the Clinical Trial Product within the scope of 21 C.F.R. § 312.21(b), as amended from time to time.

"**Phase III Clinical Trial**" means a Clinical Trial of the Clinical Trial Product that is intended as a pivotal study to gather effectiveness data to support the submission of a marketing application within the scope of 21 C.F.R. § 312.21(c), as amended from time to time.

"**Product(s)**" means the Clinical Trial Product and/or the Regulatory Approval Product, as applicable.

"**Program Transaction**" means any Assignment Transaction pursuant to which a Third Party is (a) assigned, or exclusively licensed for any and all uses, all Intellectual Property necessary or useful for the development or commercialization of Regulatory Approval Products or Clinical Trial Products or (b) assigned all or substantially all of the assets used or held for use in connection with Regulatory Approval Products or Clinical Trial Products.

"**Regulatory Approval Milestone**" means, and will be deemed to occur upon, Parent's or its Affiliates' (or their respective successors or assigns) first receipt of approval by the FDA of a new drug application or other regulatory approval application which approval grants Parent or its Affiliates (or their respective successors or assigns) the right to market and sell the Regulatory Approval Product in the United States.

"**Regulatory Approval Milestone Payment**" means $1.00 per CVR.

"**Regulatory Approval Milestone Period**" means the period commencing as of the date of this Agreement and ending 11:59 p.m., Eastern Time, on that date that is fifty-four (54) months after the date of this Agreement.

"**Regulatory Approval Product**" means any pharmaceutical product, including all forms, presentations, doses and formulations, containing the Company's compound, ACH-4471 or any salt, free-base, hydrate, solvate, polymorph, isomer, enantiomer, metabolite, prodrug or other derivative thereof, whether as the sole active ingredient or in combination with other active ingredients.

"**Rights Agent**" means the Rights Agent named in the first paragraph of this Agreement, until a successor Rights Agent becomes such pursuant to the applicable provisions of this Agreement, and thereafter "Rights Agent" shall mean such successor Rights Agent.

Section 1.2 *Additional Definitions*. For purposes of this Agreement, each of the following terms shall have the meaning specified in the Section set forth opposite to such term:

Term	**Section**
Acquiror	1.1
Aggregate Clinical Trial Milestone Payment	2.4(a)
Aggregate Regulatory Approval Milestone Payment	2.4(b)
Aggregate Milestone Payments	2.4(b)
Agreement	Preamble
Assignee	6.10
Company	Recitals
CVR Register	2.3(b)
DTC	1.1

Equity Awards Schedule	2.3(b)
Merger	Recitals
Merger Agreement	Recitals
Merger Subsidiary	Recitals
Milestone Achievement Certificate	2.4(a)
Option Holders	Recitals
Parent	Preamble
Rights Agent	Preamble
Shares	Recitals
Surviving Corporation	Recitals

Section 1.3 *Other Definitional Provisions*. Unless the context expressly otherwise requires:

(a) the words "hereof," "hereto," "herein," and "hereunder," and words of similar import, when used in this Agreement, shall refer to this Agreement as a whole and not to any particular provision of this Agreement;

(b) the terms defined in the singular have a comparable meaning when used in the plural, and vice versa;

(c) the terms "Dollars" and "$" mean United States Dollars;

(d) references herein to a specific Article, Section, or Annex shall refer, respectively, to Articles and Sections of, and Annexes to, this Agreement;

(e) wherever the word "include," "includes," or "including" is used in this Agreement, it shall be deemed to be followed by the words "without limitation";

(f) the term "or" will not be deemed to be exclusive;

(g) references herein to any gender include the other gender; and

(h) any Law defined or referred to herein will refer to such Law as amended and the rules and regulations promulgated thereunder.

ARTICLE 2
CONTINGENT VALUE RIGHTS

Section 2.1 *CVRs*. The CVRs represent the rights of Holders to receive contingent cash payments pursuant to this Agreement. The initial Holders shall be the (i) holders of Shares other than Dissenting Shares immediately prior to the Effective Time and (ii) holders of Company Stock Options immediately prior to the Effective Time whose Company Stock Options are converted into the right to receive the Cash Merger Consideration pursuant to Section 2.06 of the Merger Agreement. *Nontransferable*. The CVRs may not be sold, assigned, transferred, pledged, encumbered or in any other manner transferred or disposed of, in whole or in part, other than through a Permitted Transfer. Any attempted sale, assignment, transfer, pledge, encumbrance or disposition of CVRs in whole or in part, that is not a Permitted Transfer, will be void ab initio and of no effect.

Section 2.2 *No Certificate; Registration; Registration of Transfer; Change of Address*.

(a) The CVRs shall not be evidenced by a certificate or other instrument.

(b) The Rights Agent shall keep a register (the "**CVR Register**") for the purpose of registering CVRs and Permitted Transfers. The CVR Register will initially show one position for Cede & Co. representing all Shares held by DTC on behalf of street name holders held by such holders as of immediately prior to the Effective Time. The Rights Agent will have no responsibility whatsoever directly to the street name holders with respect to transfers of CVRs. With respect to any payments to be made under Section 2.4 below, the Rights Agent will accomplish the payment to any former street name holders of Shares by sending one lump sum payment to DTC. The Rights Agent will have no responsibilities whatsoever with regard to the distribution of payments by DTC to such street name holders. In the case of Option Holders, the CVRs shall be registered in the names and addresses of such Option Holder and in a denomination equal to the number of Shares subject to the outstanding number of Shares underlying the outstanding In the Money Options held by such Option Holder immediately prior to the Effective Time, and, in each case, as set forth in a schedule delivered by the Company to Parent (the "**Equity Awards Schedule**"). The Parent shall deliver the Equity Awards Schedule to the Rights Agent pursuant to Section 4.1 for registration by the Rights Agent of the Option Holders to

the CVR Register. Except as otherwise provided herein, once registered on the CVR Register, such Option Holders shall be deemed "Holders" pursuant to this Agreement and entitled to all rights, and privileges and subject to all obligations under this Agreement.

(c) Subject to the restrictions on transferability set forth in Section 2.2, every request made to transfer a CVR must be in writing and accompanied by a written instrument of transfer and other requested documentation in form reasonably satisfactory to Parent and the Rights Agent pursuant to its customary policies and guidelines, duly executed by the Holder thereof, the Holder's attorney duly authorized in writing, the Holder's personal representative duly authorized in writing, or the Holder's survivor (with written documentation evidencing such person's status as the Holder's survivor), and setting forth in reasonable detail the circumstances relating to the transfer. Upon receipt of such written notice by the Rights Agent, the Rights Agent shall, subject to its reasonable determination that the transfer instrument is in proper form, notify Parent that it has received such written notice. Upon receipt of such notice from the Rights Agent, Parent shall determine whether the transfer otherwise complies with the other terms and conditions of this Agreement (including the provisions of Section 2.2), and if the Parent determines that it does so comply, Parent shall instruct the Rights Agent in writing to register the transfer of the CVRs in the CVR Register. Each of Parent and the Rights Agent may require payment, from any such Holder and any such transferee, of a sum sufficient to cover any stamp or other Tax or other charge of any nature whatsoever that is imposed by a Governmental Authority or Taxing Authority in connection with any such registration of transfer. Neither Parent nor the Rights Agent shall have any duty or obligation to take any action under any section of this Agreement that requires the payment by a Holder or a transferee of a CVR of applicable Taxes or charges unless and until each of Parent and the Rights Agent is satisfied that all such Taxes or charges have been paid by such Holder or such transferee. All duly transferred CVRs registered in the CVR Register shall be the valid obligations of Parent and shall entitle the transferee to the same benefits and rights under this Agreement as those held immediately prior to the transfer by the transferor. No transfer of a CVR shall be valid until registered in the CVR Register in accordance with this Agreement.

(d) A Holder may make a written request to the Rights Agent to change such Holder's address of record in the CVR Register. The written request must be duly executed by the Holder. Upon receipt of such written notice, the Rights Agent is hereby authorized to promptly record the change of address in the CVR Register.

Section 2.3 *Payment Procedures.*

(a) If the Clinical Trial Milestone occurs at any time prior to the expiration of the Clinical Trial Milestone Period, then, on or prior to the applicable Milestone Payment Date, Parent will deliver or cause to be delivered to the Rights Agent (i) a certificate (a "**Milestone Achievement Certificate**") certifying the date of the satisfaction of the Clinical Trial Milestone, that the Holders are entitled to receive the Clinical Trial Milestone Payment and whether Parent is electing to pay the aggregate amount of Clinical Trial Milestone Payment payable to all Option Holders directly to the Surviving Company, and (ii) a wire transfer of Dollars in immediately available funds to an account designated by the Rights Agent, in the aggregate amount equal to the number of CVRs (as reflected in the CVR Register) then outstanding multiplied by the amount of the Clinical Trial Milestone Payment (the "**Aggregate Clinical Trial Milestone Payment**"), provided that Parent may instead, at its discretion, pay directly to the Surviving Corporation the aggregate amount of Clinical Trial Milestone Payment payable to all Option Holders. After receipt of the Milestone Achievement Certificate and wire transfer described in the foregoing sentence, the Rights Agent will promptly (and in any event, within five (5) Business Days) pay (x) by one lump sum wire payment to DTC for any Holder who is a former street name holder of Shares, (y) by one lump sum wire payment to the Surviving Corporation for all Option Holders (unless Parent has elected to pay such amount itself to the Surviving Corporation as indicated in the Milestone Achievement Certificate), and (z) for all other Holders, by check mailed, first-class postage prepaid, to the address of each Holder set forth in the CVR Register or by other method of delivery as specified by the applicable Holder in writing and reasonably acceptable to the Rights Agent (such amount in (x), (y) and (z) together, an amount in Dollars equal to Aggregate Clinical Trial Milestone Payment). The Rights Agent shall hold the Aggregate Clinical Trial Milestone Payment pursuant to Section 2.4(g) until such payment is made in accordance with the foregoing sentence. Notwithstanding the foregoing, in no event shall Parent be required to pay the Clinical Trial Milestone Payment more than once, and Parent shall not be required to pay the Clinical Trial Milestone Payment if the Clinical Trial Milestone occurs after the expiration of the Clinical Trial Milestone Period.

(b) If the Regulatory Approval Milestone occurs at any time prior to the expiration of the Regulatory Approval Milestone Period, then, on or prior to the applicable Milestone Payment Date, Parent will deliver or cause to be delivered to the Rights Agent (i) a Milestone Achievement Certificate certifying the date of the satisfaction of the Regulatory Approval Milestone, that the Holders are entitled to receive the Regulatory Approval Milestone Payment, and whether Parent is electing to pay the aggregate amount of Regulatory Approval Milestone Payment payable to all Option Holders directly to the Surviving Company, and (ii) a wire transfer of Dollars in immediately available funds to an account designated by the Rights Agent, in the aggregate amount equal to the number of CVRs (as reflected in the CVR Register) then outstanding multiplied by the amount of the Regulatory Approval Milestone Payment (the "**Aggregate Regulatory Approval Milestone Payment**", together with the Aggregate Clinical Trial Milestone Payment, the "**Aggregate Milestone Payments**"), provided that Parent may instead, at its discretion, pay directly to the Surviving Corporation the aggregate amount of Regulatory Approval Milestone Payment payable to all Option Holders. After receipt of the Milestone Achievement Certificate and wire transfer described in the foregoing sentence, the Rights Agent will promptly (and in any event, within five (5) Business Days) pay (x) by one lump sum wire payment to DTC for any Holder who is a former street name holder of Shares, (y) by one lump sum wire payment to the Surviving Corporation for

all Option Holders (unless Parent has paid such amount itself to the Surviving Corporation as indicated in the Milestone Achievement Certificate) and (z) for all other Holders, by check mailed, first-class postage prepaid, to the address of each Holder set forth in the CVR Register or by other method of delivery as specified by the applicable Holder in writing and reasonably acceptable to the Rights Agent (such amount in (x), (y) and (z) together, an amount in Dollars equal to Aggregate Regulatory Approval Milestone Payment). The Rights Agent shall hold the Aggregate Regulatory Approval Milestone Payment pursuant to Section 2.4(g) until such payment is made in accordance with the foregoing sentence. Notwithstanding the foregoing, in no event shall Parent be required to pay the Regulatory Approval Milestone Payment more than once and Parent shall not be required to pay the Regulatory Approval Milestone Payment if the Regulatory Approval Milestone occurs after the expiration of the Regulatory Approval Milestone Period.

(c) Except in respect of the CVRs corresponding to In the Money Options (which CVRs shall be subject to the Tax withholding provisions set forth in Section 2.08 of the Merger Agreement and which withholdings shall be made by Parent or its Affiliate), each of Parent and the Rights Agent shall be entitled to deduct and withhold from either of the Milestone Payments, if payable, such amounts as may be required to be deducted and withheld with respect to the applicable Milestone Payment or CVR under the Code, and the rules and regulations thereunder, or any other applicable provision of state, local or foreign Law relating to Taxes, as may be reasonably determined by Parent and communicated to the Rights Agent in writing. Prior to making any such deduction or withholding, other than ordinary course payroll withholding and reporting, if applicable, Parent shall instruct the Rights Agent to solicit IRS Form W-9 or IRS Form W-8, or any other appropriate forms or information from each Holder in order to avoid or reduce such deduction and withholding, and the Milestone Payment may be reasonably delayed in order to gather such forms or information. Provided that the Parent has timely delivered all information and documents reasonably requested by the Rights Agent to effect any tax withholdings requested by the Parent from the Milestone Payments, the Rights Agent shall, not more than ten (10) Business Days following its receipt of the Milestone Payment, deliver any amounts withheld by it in respect of Taxes to the Parent for the purposes of remitting such amounts over to the relevant Governmental Authorities. To the extent such amounts are so deducted and withheld and timely remitted to the relevant Governmental Authorities, such amounts shall be treated for all purposes under this Agreement as having been paid to the person to whom such amounts would otherwise have been paid. As required by applicable Law, Parent or the Rights Agent (at the direction and expense of Parent), as applicable, shall deliver to the Person to whom such amounts would otherwise have been paid an original IRS Form 1099 or other reasonably acceptable evidence of such withholding.

(d) Any portion of any Milestone Payment that remains undistributed to the Holders twelve (12) months after the date of the applicable Milestone Achievement Certificate shall be delivered by the Rights Agent to Parent, upon demand, and any Holder shall thereafter look only to Parent for payment of such Milestone Payment, without interest, but such Holder shall have no greater rights against Parent than those accorded to general unsecured creditors of Parent under applicable Law.

(e) Neither Parent nor the Rights Agent shall be liable to any person in respect of any Milestone Payment delivered to a public official in compliance with any applicable state, federal or other abandoned property, escheat or similar Law. In addition to and not in limitation of any other indemnity obligation herein, Parent agrees to indemnify and hold harmless the Rights Agent with respect to any liability, penalty, cost or expense the Rights Agent may incur or be subject to in connection with transferring such property to Parent. The indemnification provided by this Section 2.4(e) shall survive the resignation, replacement or removal of the Rights Agent and the termination of this Agreement.

(f) Except to the extent any portion of the Milestone Payment is required to be treated as imputed interest and except as otherwise required pursuant to applicable Law, the Parties hereto intend to treat the Milestone Payments payable with respect to Shares for all Tax purposes as consideration for the Shares, pursuant to the Merger Agreement. Parent shall, and shall cause the Surviving Corporation to, report imputed interest on the CVRs as required by applicable Law.

(g) All funds received by the Rights Agent under this Agreement that are to be distributed or applied by the Rights Agent in the performance of services hereunder (the "Funds") shall be held by the Rights Agent as agent for Parent and deposited in one or more bank accounts to be maintained by the Rights Agent in its name as agent for Parent. Until paid pursuant to the terms of this Agreement, the Rights Agent will hold the Funds through such accounts in: deposit accounts of commercial banks with Tier 1 capital exceeding $1 billion or with an average rating above investment grade by S&P (LT Local Issuer Credit Rating), Moody's (Long Term Rating) and Fitch Ratings, Inc. (LT Issuer Default Rating) (each as reported by Bloomberg Finance L.P.). The Rights Agent shall have no responsibility or liability for any diminution of the Funds that may result from any deposit made by the Rights Agent in accordance with this paragraph, including any losses resulting from a default by any bank, financial institution or other third party. The Rights Agent may from time to time receive interest, dividends or other earnings in connection with such deposits. The Rights Agent shall not be obligated to pay such interest, dividends or earnings to the Parent, any Holder or any other Person.

Section 2.4 *No Voting, Dividends or Interest; No Equity or Ownership Interest in Parent.*

(a) The CVRs will not have any voting or dividend rights, and interest will not accrue on any amounts payable on the CVRs to any Holder.

(b) The CVRs will not represent any equity or ownership interest in Parent or in any constituent company to the Merger or any of their respective Affiliates.

Section 2.5 *Enforcement of Rights of Holders*. Any actions seeking the enforcement of the rights of Holders hereunder may only be brought by the Majority Holders.

ARTICLE 3
THE RIGHTS AGENT

Section 3.1 *Certain Duties and Responsibilities*. The Rights Agent shall not have any liability for or in respect of any actions taken, suffered or omitted to be taken in connection with its acceptance and administration of this Agreement and the exercise and performance of its duties hereunder, except to the extent of its gross negligence, bad faith or willful or intentional misconduct (each as determined by a final non-appealable judgment of a court of competent jurisdiction).

Section 3.2 *Certain Rights of the Rights Agent*. The Parent hereby appoints the Rights Agent to act as rights agent for the Parent in accordance with the express terms and conditions hereof and the Rights Agent undertakes to perform such duties and only such duties as are specifically set forth in this Agreement, and no implied covenants or obligations shall be read into this Agreement against the Rights Agent. In addition:

(a) the Rights Agent may rely and shall be protected and held harmless by Parent in acting or refraining from acting upon any resolution, certificate, statement, instrument, opinion, report, notice, request, direction, consent, order or other paper or document believed by it in good faith to be genuine and to have been signed or presented by the proper party or parties;

(b) whenever the Rights Agent shall deem it desirable that a matter be proved or established prior to taking, suffering or omitting any action hereunder, the Rights Agent may rely upon an Officer's Certificate, which certificate shall be full authorization and protection to the Rights Agent, and the Rights Agent shall, in the absence of bad faith on its part, incur no liability and be held harmless by Parent for or in respect of any action taken, suffered or omitted to be taken by it under the provisions of this Agreement in good faith reliance upon such certificate;

(c) the Rights Agent may engage and consult with counsel of its selection (who may be an employee of the Rights Agent) and the written advice of such counsel or any Opinion of Counsel shall be full and complete authorization and protection to the Rights Agent, and the Rights Agent shall be held harmless by Parent in respect of any action taken, suffered or omitted by it hereunder in good faith and in reliance thereon;

(d) the permissive rights of the Rights Agent to do things enumerated in this Agreement shall not be construed as a duty;

(e) the Rights Agent shall not be required to give any note or surety in respect of the execution of such powers;

(f) the Rights Agent shall not be liable for or by reason of, and shall be held harmless by Parent with respect to any of the statements of fact or recitals contained in this Agreement or be required to verify the same, but all such statements and recitals are and shall be deemed to have been made by Parent only;

(g) the Rights Agent shall have no liability and shall be held harmless by Parent in respect of the validity of this Agreement or the execution and delivery hereof (except the due execution and delivery hereof by the Rights Agent and the enforceability of this Agreement against the Rights Agent assuming the due execution and delivery hereof by Parent), nor shall it be responsible for any breach by Parent of any covenant or condition contained in this Agreement;

(h) Parent agrees to indemnify the Rights Agent for, and hold the Rights Agent harmless against, any loss, liability, damage, judgment, fine, penalty, claim, demand, suit or expense (individually and collectively, "**losses**") for any action taken, suffered or omitted to be taken by the Rights Agent or arising out of or in connection with the Rights Agent's duties under this Agreement, including the reasonable and documented out-of-pocket costs and expenses of defending the Rights Agent against any such losses, unless such losses has been determined by a final non-appealable judgment of a court of competent jurisdiction to be a result of Rights Agent's gross negligence, bad faith or willful or intentional misconduct;

(i) anything to the contrary notwithstanding, the aggregate liability of the Rights Agent arising in connection with this Agreement, whether in contract, or in tort, or otherwise, is limited to, and shall not exceed, the amounts paid hereunder by the Parent to the Rights Agent, as fees, but not including reimbursable expenses, during the twelve (12) months immediately preceding the event for which recovery from the Rights Agent is being sought;

(j) the Rights Agent shall not be liable for special, punitive, indirect, consequential or incidental losses or damages of any kind whatsoever (including but not limited to lost profits) under any provision of this Agreement even if the Rights Agent has been advised of the possibility or likelihood of such damages;

(k) Parent agrees (i) to pay the fees and expenses of the Rights Agent in connection with this Agreement as set forth on Schedule 1 attached hereto, and (ii) to reimburse the Rights Agent for all Taxes and governmental charges, reasonable and documented out-of-pocket expenses and other charges of any kind and nature incurred by the Rights Agent in the execution of this Agreement (other than Taxes imposed on or measured by the Rights Agent's net income and franchise or similar Taxes imposed on it (in lieu of net income Taxes)). The Rights Agent shall also be entitled to reimbursement from Parent for all reasonable and documented out-of-pocket expenses paid or incurred by it in connection with the administration by the Rights Agent of its duties hereunder;

(l) no provision of this Agreement shall require the Rights Agent to expend or risk its own funds or otherwise incur any financial liability in the performance of any of its duties hereunder or in the exercise of its rights if there shall be reasonable grounds for believing that repayment of such funds or adequate indemnification against such risk or liability is not reasonably assured to it;

(m) the Rights Agent shall not be deemed to have knowledge of any event of which it was supposed to receive notice thereof hereunder but did not, and the Rights Agent shall be fully protected and shall incur no liability for failing to take action in connection therewith, unless and until it has received such notice in writing;

(n) unless otherwise specifically prohibited by the terms of this Agreement, the Rights Agent and any shareholder, affiliate, director, officer or employee of the Rights Agent may buy, sell or deal in any securities of Parent or the Company or become pecuniarily interested in any transaction in which Parent or the Company may be interested, or contract with or lend money to Parent or the Company or otherwise act as fully and freely as though it were not the Rights Agent under this Agreement. Nothing herein shall preclude the Rights Agent from acting in any other capacity for Parent, the Company or for any other Person;

(o) the Rights Agent may execute and exercise any of the rights or powers hereby vested in it or perform any duty hereunder either itself (through its directors, officers and employees) or by or through its attorneys or agents, and the Rights Agent shall not be liable, answerable or accountable for any act, default, neglect or misconduct of any such attorneys or agents, absent gross negligence, bad faith or willful or intentional misconduct (each as determined by a final non-appealable judgment of a court of competent jurisdiction) in the selection and continued employment thereof;

(p) the Rights Agent shall neither be responsible for, nor chargeable with, knowledge of, nor have any requirements to comply with, the terms and conditions of any other agreement, instrument or document, including, the Merger Agreement, nor shall the Rights Agent be required to determine if any person or entity has complied with any such agreements, instruments or documents, nor shall any additional obligations of the Rights Agent be inferred from the terms of such agreements, instruments or documents even though reference thereto may be made in this Agreement; and

(q) the Rights Agent shall be under no responsibility for the validity or sufficiency of this Agreement or the execution and delivery hereof (except the due execution hereof by the Rights Agent) or in respect of the validity or execution of the CVRs, nor shall it be responsible for any breach by Parent or any other Person of any covenant or condition contained in this Agreement or any CVR. The Rights Agent shall not have any duty or responsibility in the case of the receipt of any written demand from any Holder with respect to any action or default by the Parent or any other Person including, without limiting the generality of the foregoing, any duty or responsibility to initiate or attempt to initiate any proceedings at law or otherwise or to make any demand upon the Parent or any other Person.

The provisions of this Section 3.2 shall survive the termination of this Agreement, the resignation, replacement or removal of the Rights Agent, and the payment, termination and the expiration of the CVRs.

Section 3.3 *Resignation and Removal; Appointment of Successor*.

(a) The Rights Agent may resign at any time by giving written notice thereof to Parent specifying a date when such resignation shall take effect, which notice shall be sent at least thirty (30) days prior to the date so specified, but in no event shall such resignation become effective until a successor Rights Agent has been appointed. Parent has the right to remove the Rights Agent at any time by a Board Resolution specifying a date when such removal shall take effect, but no such removal shall become effective until a successor Rights Agent has been appointed. Notice of such removal shall be given by Parent to the Rights Agent, which notice shall be sent at least thirty (30) days prior to the date so specified.

(b) If the Rights Agent provides notice of its intent to resign, is removed or becomes incapable of acting, Parent, by a Board Resolution, shall promptly, appoint a qualified successor Rights Agent. If Parent shall fail to make such appointment within thirty (30) days after giving notice of such removal or after it has been notified in writing of such resignation or incapacity by the resigning or incapacitated Rights Agent then the incumbent Rights Agent or any Holder may apply to any court of competent jurisdiction for the appointment of a new Rights Agent. Any successor Rights Agent, unless otherwise consented to in writing by the Majority Holders, shall be a stock transfer agent of national reputation or the corporate trust department of a

commercial bank. The successor Rights Agent so appointed shall, forthwith upon its acceptance of such appointment in accordance with Section 3.4, become the successor Rights Agent.

(c) Parent shall give notice of each resignation and each removal of a Rights Agent and each appointment of a successor Rights Agent by delivering a notice of such event through the facilities of DTC in accordance with DTC's procedures or mailing it by first-class mail to the Holders as their names and addresses appear in the CVR Register. Each notice shall include the name and address of the successor Rights Agent. If Parent fails to send such notice within ten (10) Business Days after acceptance of appointment by a successor Rights Agent, the successor Rights Agent shall cause the notice to be mailed at the expense of Parent. Failure to give any notice provided for in this Section 3.3, however, and any defect therein, shall not affect the legality or validity of the resignation or removal of the Rights Agent or the appointment of the successor Rights Agent, as the case may be.

(d) In connection with any resignation or removal, the Rights Agent will cooperate with Parent and any successor Rights Agent in connection with the transition of the duties and responsibilities of the Rights Agent to the successor Rights Agent, including transferring the CVR Register to the successor Rights Agent.

Section 3.4 *Acceptance of Appointment by Successor*. Every successor Rights Agent appointed hereunder shall execute, acknowledge and deliver to Parent and to the retiring Rights Agent an instrument accepting such appointment and a counterpart of this Agreement, and thereupon such successor Rights Agent, without any further act, deed or conveyance, shall become vested with all the rights, powers, trusts and duties of the retiring Rights Agent. On request of Parent or the successor Rights Agent, the retiring Rights Agent shall execute and deliver an instrument transferring to the successor Rights Agent all the rights, powers and trusts of the retiring Rights Agent.

ARTICLE 4
COVENANTS

Section 4.1 *List of Holders*. Parent shall furnish or cause to be furnished to the Rights Agent, promptly after the Effective Time and in no event later than ten (10) Business Days following the Effective Time, in such form as Parent receives from the Company's transfer agent (or other agent performing similar services for the Company), the names and addresses of the Holders and, with respect to Option Holders, in such form as set forth in the Equity Awards Schedule. Until such list of Holders and Option Holders are furnished to the Rights Agent, the Rights Agent shall have no duties, responsibilities or obligations with respect to such Holders or Option Holders.

Section 4.2 *Payment of Milestone Payment*. If a Milestone has been achieved on or prior to the expiration of the applicable Milestone Period, Parent will duly deposit or cause to be deposited with the Rights Agent, on or prior to the Milestone Payment Date, the applicable Milestone Payment to be made to the Holders in accordance with the terms of this Agreement. Such amounts shall be considered paid on the Milestone Payment Date if on such date the Rights Agent has received (prior to 11 am Eastern Standard Time) in accordance with this Agreement Dollars in immediately available funds sufficient to pay all such amounts then due.

Section 4.3 *Assignment Transactions*.

(a) Parent shall not, and shall cause its Affiliates, including the Surviving Corporation, not to, consummate any Assignment Transaction unless: such Transaction is (A) a Program Transaction; (B) the Acquiror is a pharmaceutical or biotechnology company with (1) substantial experience in conducting clinical development of, and filing for and obtaining approval in accordance with all applicable Laws to place on the market and sell in the United States, pharmaceutical products for human use and (2) a development, regulatory and scientific infrastructure, that is at least reasonably comparable to that of Parent and its Affiliates; (C) the Acquiror expressly assumes in writing all of Parent's and its Affiliates' obligations under this Agreement with respect to the applicable Products by an assumption agreement, executed and delivered to the Rights Agent, in form attached as Annex A; and (D) Parent has delivered to the Rights Agent an Officer's Certificate stating that such transaction complies with this Section 4.3(a) and all conditions precedent herein related to such transaction have been complied with.

(b) Notwithstanding Section 4.3(a), Parent may, in its sole discretion and without the consent of any other party (including any Holder), consummate any Change in Control; provided that Parent or the Surviving Corporation, as applicable, will cause the Person acquiring Parent to expressly assume in writing Parent's and the Surviving Corporation's (as applicable) obligations, duties and covenants under this Agreement. No later than five (5) Business Days prior to the consummation of any Change in Control, Parent will deliver to the Rights Agent an Officer's Certificate, stating that such Change in Control complies with this Section 4.3(b) and that all conditions precedent herein relating to such transaction have been satisfied.

Section 4.4 *Commercially Reasonable Efforts*. During each Milestone Period, Parent (and its successors and assigns) shall, and shall cause its (and their) Affiliates to, use Commercially Reasonable Efforts to achieve the Milestones. Notwithstanding the foregoing, Parent shall have no obligation to develop the Clinical Trial Product in any indication other than paroxysmal nocturnal hemoglobinuria.

Section 4.5 *Tax Reporting*. The Rights Agent shall comply with all applicable Laws, including as such Laws relate to Tax reporting and with withholding with respect to any Milestone Payments made pursuant to this Agreement, other than any payroll reporting requirements with respect to any Milestone Payments made under Section 2.4 of this Agreement.

Section 4.6 *No Conflict*. Parent will not enter into any agreement with any Third Party that is, or otherwise take any actions or inactions, in conflict with this Agreement in any material respect or adversely affect the performance of its obligations under this Agreement.

Section 4.7 *Compliance with Applicable Laws*. Parent agrees that its development and regulatory activities in connection with the Clinical Trial Product and Regulatory Approval Product will be carried out in compliance with all applicable Laws in all material respects.

ARTICLE 5
AMENDMENTS

Section 5.1 *Amendments without Consent of Holders.*Without the consent of any Holders, Parent, when authorized by a Board Resolution, and the Rights Agent at any time and from time to time, may enter into one or more amendments hereto, for any of the following purposes:

(i) to evidence the succession of another Person as a successor Rights Agent and the assumption by any such successor of the covenants and obligations of the Rights Agent herein;

(ii) to add to the covenants of Parent such further covenants, restrictions, conditions or provisions as Parent shall consider to be reasonably necessary or desirable for the protection of the Holders; provided that, in each case, such provisions do not materially adversely affect the interests of the Holders;

(iii) to cure any ambiguity, to correct or supplement any provision herein that may be defective or inconsistent with any other provision herein, or to make any other provisions with respect to matters or questions arising under this Agreement; provided that, in each case, such provisions do not materially adversely affect the interests of the Holders;

(iv) as may be necessary or appropriate to ensure that the CVRs are not subject to registration under the Securities Act, the Exchange Act or any applicable state securities or "blue sky" Laws; provided that, such amendments do not materially adversely affect the interests of the Holders;

(v) to reduce the number of CVRs, in the event any Holder agrees to renounce such Holder's rights under this Agreement in accordance with Section 6.11;

(vi) subject to Section 4.3, to evidence the succession of another Person to Parent and the assumption by any such successor of the covenants of Parent contained herein;

(vii) to evidence the assignment of this Agreement by Parent as provided in Section 4.3; or

(viii) any other amendment to this Agreement that would provide any additional rights or benefits to the Holders or that does not materially adversely affect the legal rights under this Agreement of any such Holder.

(b) Promptly after the execution by Parent and the Rights Agent of any amendment pursuant to the provisions of this Section 5.1, Parent shall deliver to the Rights Agent pursuant to Section 6.2 and shall deliver (or cause the Rights Agent to deliver) a notice thereof through the facilities of DTC in accordance with DTC's procedures or by first class mail to the Holders at their addresses as they appear on the CVR Register, setting forth such amendment.

Section 5.2 *Amendments with Consent of Holders*.

(a) Subject to Section 5.1 (which amendments pursuant to Section 5.1 may be made without the consent of the Holders), with the prior consent of the Majority Holders, whether evidenced in writing or taken at a meeting of the Holders, Parent, when authorized by a Board Resolution, and the Rights Agent may enter into one or more amendments hereto for the purpose of adding, eliminating or changing any provisions of this Agreement, even if such addition, elimination or change is materially adverse to the interest of the Holders.

(b) Promptly after the execution by Parent and the Rights Agent of any amendment pursuant to the provisions of this Section 5.2, Parent shall deliver (or cause the Rights Agent to deliver) a notice thereof through the facilities of DTC in accordance with DTC's procedures or by first class mail to the Holders at their addresses as they appear on the CVR Register, setting forth such amendment.

Section 5.3 *Execution of Amendments*. In executing any amendment permitted by this Article 5, the Rights Agent shall be entitled to receive, and shall be fully protected in relying upon, an Opinion of Counsel stating that the execution of such amendment is authorized or permitted by this Agreement. Each amendment to this Agreement shall be evidenced by a writing signed by the Rights Agent and Parent. The Rights Agent may, but is not obligated to, enter into any such amendment that affects the Rights Agent's own rights, powers, privileges, covenants or duties under this Agreement or otherwise, and the Rights Agent shall not be bound by amendments not executed by it.

Section 5.4 *Effect of Amendments*. Upon the execution of any amendment under this Article 5, this Agreement shall be modified in accordance therewith, such amendment shall form a part of this Agreement for all purposes and every Holder shall be bound thereby.

ARTICLE 6
MISCELLANEOUS AND GENERAL

Section 6.1 *Termination*. Subject to the survival provisions contained in Section 2.4(e) and Section 3.2, this Agreement will be terminated and of no force or effect, the Parties will have no liability hereunder (other than with respect to monies due and owing by Parent to the Rights Agent) and no payments will be required to be made, upon the earlier to occur of (a) the payment by the Rights Agent to each Holder of both Milestone Payments required to be paid under the terms of this Agreement in accordance with Section 2.4(a) and 2.4(b), (b) the delivery of a written notice of termination duly executed by Parent and the Majority Holders and (c) the expiration of both Milestone Periods. For the avoidance of doubt, the termination of this Agreement will not affect or limit the right to receive the Milestone Payments under Section 2.4 to the extent earned prior to termination of this Agreement and the provisions applicable thereto will survive the expiration or termination of this Agreement.

Section 6.2 *Notices to the Rights Agent and Parent*. Any notices or other communications required or permitted under, or otherwise given in connection with, this Agreement shall be in writing and shall be deemed to have been duly given (a) when delivered or sent if delivered in person or sent by facsimile transmission (provided confirmation of facsimile transmission is obtained), (b) on the next Business Day if transmitted by national overnight courier, (c) on the date delivered if sent by email (provided confirmation of email receipt is obtained), or (d) two (2) Business Days after being sent by registered or certified mail, in each case as follows:

If to Parent:

Alexion Pharmaceuticals, Inc.
121 Seaport Boulevard
Boston, Massachusetts 02210
Attention: General Counsel
Email: ellen.chiniara@alexion.com

With a copy to:

Foley Hoag LLP
Seaport Trade Center West
155 Seaport Boulevard
Boston, Massachusetts 02210
Attention: Mark A. Haddad
Email: mhaddad@foleyhoag.com

If to Rights Agent:

Computershare Inc.
150 Royall Street
Canton, MA 02021
Attention: Corporate Actions

With a copy to:

Computershare Inc.
150 Royall Street
Canton, MA 02021
Attention: General Counsel

or to such other persons or addresses as may be designated in writing by the Party to receive such notice as provided above.

Section 6.3 *Notice to Holders*. Where this Agreement provides for notice to Holders, such notice shall be sufficiently given (unless otherwise herein expressly provided) if in writing and transmitted through the facilities of DTC in accordance with DTC's procedures or mailed, first-class postage prepaid, to each Holder affected by such event, at the Holder's address as it appears in the CVR Register, not later than the latest date, and not earlier than the earliest date, if any, prescribed for the giving of such notice. In any case where notice to Holders is given by mail, neither the failure to mail such notice, nor any defect in any notice so mailed, to any particular Holder shall affect the sufficiency of such notice with respect to other Holders.

Section 6.4 *Counterparts*. This Agreement may be signed in any number of counterparts, each of which shall be an original, with the same effect as if the signatures thereto and hereto were upon the same instrument. This Agreement shall become effective when each Party hereto shall have received a counterpart hereof signed by the other Party hereto. Until and unless each Party has received a counterpart hereof signed by each other Party hereto, this Agreement shall have no effect and no Party shall have any right or obligation hereunder (whether by virtue of any other oral or written agreement or other communication). Signatures to this Agreement transmitted by facsimile transmission, by electronic mail in PDF form, or by any other electronic means designed to preserve the original graphic and pictorial appearance of a document, will be deemed to have the same effect as physical delivery of the paper document bearing the original signatures.

Section 6.5 *Governing Law; Jurisdiction; WAIVER OF JURY TRIAL*. This Agreement shall be governed by and construed in accordance with the laws of the State of Delaware, without regard to the conflicts of law rules of such State that would cause the application of the laws of any other jurisdiction. The Parties hereto agree that any Proceeding seeking to enforce any provision of, or based on any matter arising out of or in connection with, this Agreement or the transactions contemplated hereby shall be brought in the Court of Chancery of the State of Delaware in and for New Castle County, Delaware and any state appellate court therefrom or, if (but only if) such court lacks subject matter jurisdiction, the United States District Court sitting in New Castle County, Delaware and any appellate court therefrom. Each Party hereto hereby irrevocably submits to the exclusive jurisdiction of such court in respect of any legal action, suit or proceeding arising out of or relating to this Agreement or the transactions contemplated hereby, and hereby waives, and agrees not to assert, as a defense in any such action, suit or proceeding, any claim that it is not subject personally to the jurisdiction of such court, that the action, suit or proceeding is brought in an inconvenient forum, that the venue of the action, suit or proceeding is improper or that this Agreement or the transactions contemplated hereby may not be enforced in or by such courts. Each Party hereto agrees that notice or the service of process in any action, suit or proceeding arising out of or relating to this Agreement or the transactions contemplated hereby shall be properly served or delivered if delivered in the manner contemplated by Section 6.2 or in any other manner permitted by law. EACH OF THE PARTIES HERETO HEREBY IRREVOCABLY WAIVES ANY AND ALL RIGHT TO TRIAL BY JURY IN ANY LEGAL PROCEEDING ARISING OUT OF OR RELATED TO THIS AGREEMENT OR THE TRANSACTIONS CONTEMPLATED HEREBY.

Section 6.6 *Other Remedies*. Except as otherwise provided herein, any and all remedies herein expressly conferred upon a Party will be deemed cumulative with and not exclusive of any other remedy conferred hereby, or by Law or equity upon such Party, and the exercise by a Party of any one remedy will not preclude the exercise of any other remedy.

Section 6.7 *Entire Agreement*. As between the Parent and the Rights Agent, this Agreement alone contains the entire understanding of the Parties with respect to the subject matter hereof and supersede all prior agreements and understandings, both written and oral, among the Parties with respect to the subject matter hereof. If and to the extent that any provision of this Agreement is inconsistent or conflicts with the Merger Agreement with respect to the Rights Agent or its rights, duties, obligations, protections and liabilities hereunder, this Agreement shall govern and be controlling.

Section 6.8 *Third-Party Beneficiaries; Action by Majority Holders*. Parent and the Rights Agent hereby agree that the respective covenants and agreements set forth herein are intended to be for the benefit of, and shall be enforceable by, the Majority Holders, who are intended third-party beneficiaries hereof. Parent and the Rights Agent further agree that this Agreement and their respective covenants and agreements set forth herein are solely for the benefit of Parent, the Rights Agent, the Holders and their respective permitted successors and assigns hereunder in accordance with and subject to the terms of this Agreement, and nothing in this Agreement, express or implied, will confer upon any Person other than Parent, the Rights Agent, the Holders and their permitted successors and assigns hereunder any benefit or any legal or equitable right, remedy or claim hereunder. Except for the rights of the Rights Agent set forth herein, the Majority Holders will have the sole right, on behalf of all Holders, by virtue of or under any provision of this Agreement, to institute any action or proceeding at Law or in equity or in bankruptcy or otherwise upon or under or with respect to this Agreement, and no individual Holder or other group of Holders will be entitled to exercise such rights. The Parties hereby agree that irreparable damage would occur in the event that any provision of this Agreement were not performed in accordance with its specific terms or were otherwise breached, and that money damages or other legal remedies will not be an adequate remedy for any such damages. Accordingly, the Parties acknowledge and hereby agree that in the event of any breach by Parent or Assignee (as such term is defined below), on the one hand, or the Rights Agent, on the other hand, of any of their respective covenants or obligations set forth in this Agreement, Parent or Assignee, on the one hand, and the Rights Agent, on the other hand, shall be entitled to (and the other Party will not oppose) an injunction or injunctions to prevent or restrain breaches of this Agreement, by the other(s) (as applicable), and to specific enforcement of the terms and provisions of this Agreement.

Section 6.9 *Severability*. If any term, provision, covenant or restriction of this Agreement is held by a court of competent jurisdiction or other Governmental Authority to be invalid, void or unenforceable, the remainder of the terms, provisions, covenants and restrictions of this Agreement shall remain in full force and effect and shall in no way be affected, impaired or invalidated so long as the economic or legal substance of the transactions contemplated hereby is not affected in any manner materially adverse to any Party; provided, that if any such excluded provision, or the application thereof, shall materially and adversely affect the rights, immunities, liabilities, duties, responsibilities or obligations of the Rights Agent, the Rights Agent shall be entitled to resign immediately. Upon such a determination, the Parties agree to modify this Agreement so as to effect the original intent of the Parties as closely as possible in an acceptable manner, in order that the transactions contemplated hereby be consummated as originally contemplated to the fullest extent possible.

Section 6.10 *Assignment*. This Agreement shall not be assignable provided, however, that (a) Parent may assign this Agreement to a Person (each such Person, an "**Assignee**") (i) which is a direct or indirect wholly-owned subsidiary of Parent; provided that Parent remains jointly and severally liable, (ii) with the prior consent of the Majority Holders, whether evidenced in writing or taken at a meeting of the Holders, or (iii) in connection with a transaction involving an Assignment Transaction conducted in compliance with Section 4.3 and (b) the Rights Agent may assign this Agreement to a successor Rights Agent appointed in accordance with Section 3.3.

Section 6.11 *Benefits of Agreement*. Notwithstanding anything to the contrary contained herein, any Holder may at any time agree to renounce, in whole or in part, whether or not for consideration, such Holder's rights under this Agreement by written notice to the Rights Agent and Parent, which notice, if given, shall be irrevocable. Parent may, in its sole discretion, at any time, offer consideration to Holders in exchange for their agreement to irrevocably renounce their rights hereunder.

Section 6.12 *Legal Holidays*. In the event that any Milestone Payment Date shall not be a Business Day, then (notwithstanding any provision of this Agreement to the contrary) payment need not be made on such date, but may be made, without the accrual of any additional interest thereon on account of such Milestone Payment Date not being a Business Day, on the next succeeding Business Day with the same force and effect as if made on such Milestone Payment Date.

Section 6.13 *Interpretation; Construction*.

(a) The table of contents and headings herein are for convenience of reference only, do not constitute part of this Agreement and shall not be deemed to limit or otherwise affect any of the provisions hereof.

(b) The Parties have participated jointly in negotiating and drafting this Agreement. In the event that an ambiguity or a question of intent or interpretation arises, this Agreement shall be construed as if drafted jointly by the Parties, and no presumption or burden of proof shall arise favoring or disfavoring any Party by virtue of the authorship of any provision of this Agreement.

[Signature Pages Follow.]

IN WITNESS WHEREOF, this Agreement has been duly executed and delivered by the duly authorized officers of the Parties hereto as of the date first written above.

ALEXION PHARMACEUTICALS, INC.

By: /s/ Aradhana Sarin
Name: Aradhana Sarin
Title: Executive Vice President, Chief Financial Officer

COMPUTERSHARE INC.

By: /s/ Neda Sheridan
Name: Neda Sheridan
Title: Regional Manager

[*Signature Page to Contingent Value Rights Agreement*]

B5092239.1

Annex A

Form of Assignment and Assumption Agreement

ASSIGNMENT AND ASSUMPTION AGREEMENT, made as of [ ] (this "**Agreement**"), between [***Parent***], a Delaware corporation ("**Assignor**"), and [●], a [ ] ("**Assignee**"). Unless otherwise defined herein, capitalized terms used in this Agreement shall have the meanings given to them in the CVR Agreement referred to below.

W I T N E S S E T H:

WHEREAS, Assignor and [●], as rights agent (the "**Rights Agent**") are parties to a Contingent Value Rights Agreement dated as of [●], 2020 (the "**CVR Agreement**"); and

WHEREAS, Assignor and Assignee desire to execute and deliver this Agreement evidencing the transfer to and assumption by Assignee of both (a) the due and punctual payment of any Milestone Payment and (b) the performance or observance of every covenant and agreement of the CVR Agreement to be performed or observed on the part of Assignor to be performed and observed and the assumption thereof of Assignee;

NOW, THEREFORE, in consideration of the premises and for other good and valuable consideration, the receipt and sufficiency of which is hereby acknowledged, Assignor and Assignee hereby agree as follows:

1. Assignment and Assumption. Effective as of [●] (the "**Assignment Date**"), Assignor hereby assigns to Assignee, and Assignee hereby accepts the assignment of, assumes and becomes responsible for, (a) the due and punctual payment of any Milestone Payment and (b) the performance or observance of every covenant and agreement of the CVR Agreement to be performed or observed on the part of Assignor.

2. Successors and Assigns. This Agreement shall be binding upon and shall inure to the benefit of the respective parties hereto and their respective successors and assigns.

3. Governing Law. This Agreement shall be governed by, construed and enforced in accordance with the Laws of Delaware, without giving effect to the principles of conflicts of Laws thereof.

4. Counterparts. This Agreement may be executed in one or more counterparts, each of which shall be deemed an original but all of which together will constitute one and the same instrument.

[Signature Page Follows]

IN WITNESS WHEREOF, this Agreement has been duly executed and delivered by the duly authorized officers of the parties hereto as of the date first written above.

[ASSIGNOR]

By:
Name:
Title:

[ASSIGNEE]

By:
Name:
Title:

Alexion Completes Acquisition of Achillion

– Acquisition adds two clinical-stage Factor D inhibitors to Alexion's pipeline and provides promising development platform for additional complement-mediated diseases –

BOSTON – January 28, 2020 - Alexion Pharmaceuticals, Inc. (NASDAQ:ALXN) today announced it has completed its acquisition of Achillion Pharmaceuticals, Inc. The acquisition adds two clinical-stage oral small molecule Factor D inhibitors to Alexion's pipeline and provides the foundation and expertise for a broader oral Factor D inhibition development platform with the potential to treat numerous additional complement-mediated diseases.

"The acquisition of Achillion adds two clinical-stage Factor D inhibitors to our growing pipeline, representing important continued momentum in expanding and diversifying our portfolio and advancing our mission of transforming the lives of people with rare diseases," said Ludwig Hantson, Ph.D., Chief Executive Officer of Alexion. "We believe oral Factor D inhibition holds great promise in treating people with multiple rare, complement-mediated diseases, providing the opportunity to significantly expand our portfolio into new therapeutic areas and to help many more patients. We look forward to the expertise the Achillion team brings to Alexion, which, combined with our own complement biology and rare disease development and commercialization experience, will enable us to collectively accelerate progress of the development programs. We are committed to maintaining continuity in the programs currently underway and will be moving quickly to advance Achillion's efforts."

Alexion will continue development of Achillion's oral Factor D inhibitor portfolio, which includes two clinical-stage medicines-in-development – danicopan (ACH-4471) and ACH-5228 – as well as multiple compounds in preclinical development. Phase 3 development is being initiated for danicopan as an add-on therapy for PNH patients with extravascular hemolysis (EVH). Danicopan is also in Phase 2 development for C3G, and ACH-5228 is in Phase 2 development for PNH.

About Factor D

Factor D is an essential serine protease and critical control point in the alternative pathway (AP) of the complement system, a part of the innate immune system. Achillion's complement platform is focused on advancing oral small molecules that inhibit the AP and can potentially be used in the treatment of immune-related diseases in which complement AP plays a critical role. Potential indications currently being evaluated for these compounds include paroxysmal nocturnal hemoglobinuria (PNH) and C3 glomerulopathy (C3G).

About Paroxysmal Nocturnal Hemoglobinuria (PNH)

Paroxysmal nocturnal hemoglobinuria (PNH) is a chronic, progressive, debilitating and life-threatening ultra-rare blood disorder characterized by hemolysis (destruction of red blood cells) that is mediated by uncontrolled activation of the complement system, a component of the body's immune system. Patients with PNH may experience a wide range of signs and symptoms, such as fatigue, difficulty swallowing, shortness of breath, abdominal pain, erectile dysfunction, dark-colored urine and anemia. The most devastating consequence of chronic hemolysis is thrombosis, which can occur in blood vessels throughout the body, damaging vital organs and causing premature death. PNH is primarily a disease of intravascular hemolysis (IVH), where the red blood cell destruction occurs within the blood vessels. C5 inhibition addresses the complications of IVH and the increases in LDH that cause thrombosis and even death in patients with PNH. However, a small portion of patients – less than 10 percent – receiving a C5 inhibitor continue to experience clinical extravascular hemolysis (EVH), where the red blood cell destruction occurs outside the blood vessels. As a result, these patients are transfusion dependent despite treatment but do not have bone marrow failure or aplastic anemia. Inhibiting Factor D in the alternative pathway (AP) of the complement system offers the possibility of selectively blocking AP activity and protecting against the destruction of red blood cells, while leaving the rest of the complement system intact to fight infection.

About C3 Glomerulopathy (C3G)

C3G is an ultra-rare kidney disease for which there is no approved treatment. The disease is characterized by the deposition of C3 protein fragments in the filtering units (glomeruli) of the kidney, caused by overactivation of the complement alternative pathway (AP). Over time, the chronic deposition of C3 fragments results in permanent kidney damage and kidney failure. Today, C3G patients are treated with steroids and broad-acting immunosuppressants to slow the progression of kidney damage. Oral Factor D inhibitors have demonstrated proof-of-mechanism to interrupt the overactivation of the AP and reduce C3 fragment deposition, providing a potential treatment approach for targeting the underlying cause of C3G.

About Alexion

Alexion is a global biopharmaceutical company focused on serving patients and families affected by rare diseases through the discovery, development and commercialization of life-changing medicines. As the global leader in complement biology and inhibition for more than 20 years, Alexion has developed and commercializes two approved complement inhibitors to treat patients with paroxysmal nocturnal hemoglobinuria (PNH) and atypical hemolytic uremic syndrome (aHUS), as well as the first and only approved complement inhibitor to treat anti-acetylcholine receptor (AChR) antibody-positive generalized myasthenia gravis (gMG) and neuromyelitis optica spectrum disorder (NMOSD). Alexion also has two highly innovative enzyme replacement therapies for patients with life-threatening and ultra-rare metabolic disorders, hypophosphatasia (HPP) and lysosomal acid lipase deficiency (LAL-D). In addition, the company is developing several mid-to-late-stage therapies, including a copper-binding agent for Wilson disease, an anti-neonatal Fc receptor (FcRn) antibody for rare Immunoglobulin G (IgG)-mediated diseases and an oral Factor D inhibitor as well as several early-stage therapies, including one for light chain (AL) amyloidosis, a second anti-FcRn therapy, a second oral Factor D inhibitor and a third complement inhibitor. Alexion focuses its research efforts on novel molecules and targets in the complement cascade and its development efforts on the core therapeutic areas of hematology, nephrology, neurology, metabolic disorders and cardiology. Headquartered in Boston, Massachusetts, Alexion has offices around the globe and serves patients in more than 50 countries. This press release and further information about Alexion can be found at: www.alexion.com.

[ALXN-G]

Forward-Looking Statement

This press release includes forward-looking statements related to Alexion's acquisition of Achillion, including: the acquisition provides the foundation for a broader oral Factor D inhibition development platform with the potential to treat numerous additional complement-mediated diseases; the acquisition of Achillion represents important continued momentum in expanding and diversifying our portfolio; oral Factor D inhibition holds great promise in treating multiple rare, complement-mediated diseases, providing the opportunity to significantly expand our portfolio into new therapeutic areas; we are committed to maintaining continuity in the programs currently underway and will be moving quickly to accelerate Achillion's efforts; and Alexion will continue development of Achillion's oral Factor D inhibitor portfolio. A number of important factors could cause actual results to differ materially from those indicated by such forward-looking statements, including: the anticipated benefits of the Achillion platform and therapies not being realized; future clinical trials of Achillion products not proving that the therapies are safe and effective to the level required by regulators; decisions of regulatory authorities regarding the adequacy of the research and clinical tests, marketing approval or material limitations on the marketing of Achillion products; delays or failure of product candidates to obtain regulatory approval; delays or the inability to launch product candidates due to regulatory restrictions; unanticipated expenses; interruptions or failures in the manufacture and supply of products and product candidates; failure to satisfactorily address matters raised by the FDA and other regulatory agencies; the possibility that results of clinical trials are not predictive of safety and efficacy results of products in broader patient populations; the possibility that clinical trials of product candidates could be delayed or terminated prior to completion for a number of reasons; the adequacy of pharmacovigilance and drug safety reporting processes; and a variety of other risks set forth from time to time in Alexion's or Achillion's filings with the SEC, including but not limited to the risks discussed in Alexion's Quarterly Report on Form 10-Q for the period ended September 30, 2019 and in its other filings with the SEC and the risks discussed in Achillion's Quarterly Report on Form 10-Q for the period ended September 30, 2019 and in its other filings with the SEC. Alexion disclaims any obligation to update any of these forward-looking statements to reflect events or circumstances after the date hereof, except when a duty arises under law.

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