

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 24, 2024

ORCHARD THERAPEUTICS PLC

(Exact name of Registrant as Specified in Its Charter)

England and Wales  
(State or Other Jurisdiction  
of Incorporation)

001-38722  
(Commission  
File Number)

Not Applicable  
(IRS Employer  
Identification No.)

245 Hammersmith Road  
London W6 8PW  
United Kingdom  
(Address of Principal Executive Offices; Zip Code)

Registrant's Telephone Number, Including Area Code: +44 (0) 203 808 8286

Not Applicable  
(Former Name or Former Address, if Changed Since Last Report)

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:

- 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
American Depositary Shares, each representing ten ordinary shares, nominal value £0.10 per share	ORTX	The Nasdaq Capital 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.

## Introductory Note

This Current Report on Form 8-K is being filed in connection with the completion of the previously announced Transaction (as described below) pursuant to that certain Transaction Agreement, dated October 5, 2023 (the “Transaction Agreement”), between Orchard Therapeutics plc, a public limited company incorporated under the laws of England and Wales (the “Company”), and Kyowa Kirin Co., Ltd., a Japanese joint stock company (“Kyowa Kirin”).

As previously announced, on January 22, 2024, the High Court of Justice of England and Wales (the “Court”) sanctioned the acquisition by Kyowa Kirin International plc, a wholly owned subsidiary of Kyowa Kirin, of the entire issued and to be issued share capital of the Company pursuant to a scheme of arrangement under Part 26 of the U.K. Companies Act 2006 (the “Scheme of Arrangement”, and such acquisition, the “Transaction”) at a public hearing convened by the Court. On January 24, 2024 (the “Closing Date”), the Court’s order which sanctioned the Scheme of Arrangement was delivered to the Registrar of Companies in England and Wales and, as a result, the Company and Kyowa Kirin consummated the Transaction in accordance with the Transaction Agreement and the Scheme of Arrangement.

### Item 1.02 Termination of a Material Definitive Agreement.

The information set forth in the Introductory Note of this Current Report on Form 8-K is incorporated by reference in this Item 1.02.

Concurrently with the closing of the Transaction, the Company repaid all loans and terminated all credit commitments under that certain Senior Term Facilities Agreement, dated May 24, 2019, as amended and restated on January 30, 2023, among the Company, the entities listed as original guarantors therein, MidCap Financial (Ireland) Limited, and the additional lenders party thereto from time to time.

### Item 2.01 Completion of Acquisition or Disposition of Assets.

The information set forth in the Introductory Note of this Current Report on Form 8-K is incorporated by reference into this Item 2.01.

At the Effective Time of the Scheme of Arrangement (the “Effective Time”) all voting and non-voting ordinary shares of the Company, each with a nominal value of £0.10 per share (“Company Ordinary Shares”), issued and outstanding as of the Effective Time were transferred to Kyowa Kirin International plc (a nominee of Kyowa Kirin), and the holders of Company Ordinary Shares became entitled to receive (a) \$16.00 in cash, without interest, per American Depositary Share (“ADS”), each representing 10 Company Ordinary Shares (or \$1.60 in cash, without interest, per Company Ordinary Share (the “Cash Consideration”), plus (b) 10 contractual contingent value rights (each, a “CVR”, and together with the Cash Consideration, the “Merger Consideration”) per ADS, which each represent the right to receive a contingent payment of \$0.10 in cash (or one CVR per Company Ordinary Share), without interest, if a certain milestone is achieved, pursuant to the Contingent Value Rights Agreement (the “CVR Agreement”) entered into between Kyowa Kirin and a rights agent mutually agreed to by the Company and Kyowa Kirin (the “Rights Agent”) in connection with the completion of the Transaction.

At the Effective Time, subject to all required withholding taxes:

- each (i) outstanding option to purchase Company Ordinary Shares or Company ADSs (each, a “Share Option”) granted before October 5, 2023 and each Share Option granted following October 5, 2023 that was outstanding immediately prior to the Effective Time, to the extent vested with a per share price that is less than the Cash Consideration (each, an “In-the-Money Vested Share Option”), (ii) vested and outstanding restricted share units relating to Company Ordinary Shares (each, a “Company RSU”), and (iii) vested and outstanding restricted share unit relating to the Company Ordinary Shares and subject to performance vesting (each, a “Company PSU”) was cancelled and automatically converted into the right to receive for each underlying Company Ordinary Share (in the case of Company PSUs, determined based on maximum level of achievement for all performance objectives), (y) an amount in cash from Kyowa Kirin equal to the excess of the Cash Consideration over the per share exercise price, if any, of such award and (z) one CVR; and

- each (i) unvested Company Share Option that has a per share exercise price that is less than the Cash Consideration (each, an “Unvested In-the-Money Option”), (ii) Company Share Option, whether vested or unvested, with an exercise price per share equal to or greater than the Cash Consideration, (iii) Company RSU that was outstanding and unvested immediately prior to the Effective Time and (iv) Company PSU that was outstanding and unvested as of immediately prior to the Effective Time was automatically canceled at the Effective Time without payment of any consideration being made in respect thereof.

In connection with the closing of the Transaction, Kyowa Kirin implemented a new transitional cash plan (“Transitional Cash Plan”), under which it will grant (or procure the grant of) cash awards (each, a “Transition Award”) under such Transitional Cash Plan to all holders of Unvested In-the-Money Options, and unvested Company RSUs and/or Company PSUs (the “Unvested Awards”). The value of each Transitional Award is equal to the aggregate of the value of (i) the excess, if any, of the Cash Consideration over the per share exercise price, if any, for each Company Ordinary Share underlying such Unvested Awards (in the case of Company PSUs, determined based on maximum level of achievement for all performance objectives) (the “Unvested Cash Consideration”), and (ii) one CVR for each Company Ordinary Share underlying such Unvested Award (in the case of Company PSUs, determined based on maximum level of achievement for all performance objectives) (the “Unvested CVR Consideration”), less any required deductions and withholdings for income tax and National Insurance contributions or social security contributions. The Unvested Cash Consideration and the Unvested CVR Consideration will vest and be paid pursuant to the terms of the original Unvested Award, subject to the holder’s continued service with Kyowa Kirin and its subsidiaries through each applicable vesting date; provided, however, that any portion of the Transition Award that, per the terms of the original Unvested Award, remains outstanding and unvested as of December 31, 2024, shall vest on December 31, 2024 (subject to the holder’s continued service with Kyowa Kirin and its subsidiaries through such date).

The foregoing description of the Transaction and the Transaction Agreement, and the other transactions contemplated thereby, does not purport to be complete and is qualified in its entirety by reference to the full text of the Transaction Agreement, which was filed as Exhibit 2.1 to the Current Report on Form 8-K filed by the Company with the SEC on October 5, 2023, which is incorporated herein by reference.

In connection with the closing of the Transaction, Kyowa Kirin and the Rights Agent entered into the CVR Agreement, governing the terms of the CVRs to be received by the Company’s shareholders and holders of certain Company Share Options, Company RSUs and Company PSUs. The CVRs represent the right to receive contingent payments, payable to the Rights Agent for the benefit of the holders of the CVRs, if regulatory approval is granted by the U.S. Food and Drug Administration for the commercial marketing and sale of OTL-200 in the United States of America for the treatment or prevention of metachromatic leukodystrophy (“MLD”), which includes as an intended patient population (for clarity, in addition to any other patient cohorts, if any) both (i) children (or pediatric patients) with late infantile forms of MLD, and (ii) children (or pediatric patients) with early juvenile forms of MLD, in each case that are pre-symptomatic or without clinical manifestations of the disease, on or before 11:59 p.m. Eastern Time on December 31, 2024.

The right to the contingent consideration as evidenced by the CVR Agreement is a contractual right only. The CVRs are not transferable, except in the limited circumstances specified in the CVR Agreement, are not evidenced by 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 Kyowa Kirin or the Company.

There can be no assurance that the milestone set forth in the CVR Agreement will be achieved and that the resulting payments will be required of Kyowa Kirin. The minimum payment on each CVR is zero and the maximum payment on each CVR is \$0.10 in cash.

The foregoing description of the CVR Agreement does not purport to be complete and is qualified in its entirety by reference to the full text of the CVR Agreement, a copy of which is filed as Exhibit 2.2 hereto, and is incorporated herein by reference.

**Item 3.01 Notice of Delisting or Failure to Satisfy a Continued Listing Rule or Standard; Transfer of Listing.**

The information set forth in the Introductory Note and Item 2.01 of this Current Report on Form 8-K is incorporated by reference in this Item 3.01.

In connection with the consummation of the Transaction, the Company requested that the Nasdaq Capital Market (“Nasdaq”) suspend trading of Company ADSs (Nasdaq: ORTX) effective as of the open of trading on January 24, 2024 (which suspension was granted) and file with the SEC a Notification of Removal from Listing and/or Registration on Form 25 to delist and deregister Company ADSs under Section 12(b) of the Securities Exchange Act of 1934, as amended (the “Exchange Act”). As a result, Company ADSs will no longer be listed on Nasdaq. In addition, the Company intends to file with the SEC a certification on Form 15 requesting that its reporting obligations under Sections 13(a) and 15(d) of the Exchange Act be suspended.

**Item 3.03 Material Modification to Rights of Security Holders.**

The information set forth in the Introductory Note and under Items 2.01, 3.01 and 5.03 of this Current Report on Form 8-K is incorporated by reference into this Item 3.03.

At the Effective Time, all outstanding Company Ordinary Shares were acquired by Kyowa Kirin International plc in accordance with the provisions of the Scheme of Arrangement and the laws of England and Wales, and the Company’s shareholders ceased to have any rights with respect to the Company Ordinary Shares except their rights under the Scheme of Arrangement, including the right to receive the Merger Consideration.

**Item 5.01 Changes in Control of Registrant.**

The information set forth in the Introductory Note and Item 2.01 of this Current Report on Form 8-K is incorporated by reference in this Item 5.01.

As a result of the consummation of the Transaction, a change of control of the Company occurred and the Company became an indirect wholly owned subsidiary of Kyowa Kirin.

**Item 5.02 Departure of Directors or Certain Officers; Election of Directors; Appointment of Certain Officers; Compensatory Arrangements of Certain Officers.**

The information set forth in the Introductory Note and Item 2.01 of this Current Report on Form 8-K is incorporated by reference in this Item 5.02.

In connection with the Transaction, Alicia Secor, Charles Rowland, Jim Geraghty, John Curnutte, Joanne Beck, Steven Altschuler and Marc Dunoyer each resigned from the Board of Directors of the Company (the “Board”) and from any and all committees of the Board on which they served and ceased to be directors of the Company at the Effective Time. On January 24, 2024, the Board appointed Takeyoshi Yamashita, Yasuo Fujii and Abdul Mullick as directors of the Company.

**Item 9.01 Financial Statements and Exhibits.**

(d) Exhibits.

The exhibits listed in the following Exhibit Index are filed as part of this report.

**EXHIBIT INDEX**

<b>Exhibit No.</b>	<b>Description of Exhibit</b>
2.1*	<a href="#"><u>Transaction Agreement, dated as of October 5, 2023, by and between Orchard Therapeutics plc and Kyowa Kirin Co., Ltd. (incorporated herein by reference to Exhibit 2.1 to the Company's Current Report on Form 8-K dated October 5, 2023 (SEC File No. 001-38722)).</u></a>
2.2**	<a href="#"><u>Contingent Value Rights Agreement, dated as of December 15, 2023, by and among Kyowa Kirin Co., Ltd and Wilmington Savings Fund Society, FSB.</u></a>
104	Cover Page Interactive Data File (Embedded within the Inline XBRL documents).

\* The exhibits and schedules to this Exhibit have been omitted in accordance with Item 601(a)(5) of Regulation S-K. The Registrant agrees to furnish supplementally to the SEC a copy of all omitted exhibits and schedules upon its request.

\*\* Filed herewith.

**SIGNATURES**

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.

**ORCHARD THERAPEUTICS PLC**

Date: January 24, 2024

By: /s/ Frank E. Thomas  
Frank E. Thomas  
President and Chief Operating Officer

## CONTINGENT VALUE RIGHTS AGREEMENT

THIS CONTINGENT VALUE RIGHTS AGREEMENT, dated as of December 15, 2023 (this “*Agreement*”), is entered into by and between Kyowa Kirin Co., Ltd., a Japanese joint stock company (*kabushiki kaisha*) (“*Purchaser*”), and Wilmington Savings Fund Society, FSB (the “*Rights Agent*”).

### RECITALS

WHEREAS, Purchaser and Orchard Therapeutics plc, a public limited company incorporated under the laws of England and Wales with registered number 11494381 (“*Apple*”), have entered into a Transaction Agreement, dated as of October 5, 2023 (as amended, amended and restated or otherwise modified from time to time, the “*Transaction Agreement*”), pursuant to which the entire issued share capital of Apple will be acquired by Purchaser by means of the Scheme of Arrangement (as defined in the Transaction Agreement) on the terms and subject to the conditions set out in the Transaction Agreement and the Scheme of Arrangement (the “*Transaction*”); and

WHEREAS, pursuant to the Transaction Agreement and the Scheme of Arrangement, Purchaser has agreed to provide Holders (as defined below) the right to receive one (1) contingent cash payment per Scheme Share upon the terms and subject to the conditions of this Agreement and of the Transaction Agreement and Scheme of Arrangement without interest.

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

### 1. DEFINITIONS

1.1 Definitions. Capitalized terms used but not otherwise defined herein shall have the meanings ascribed thereto in the Transaction Agreement. As used in this Agreement, the following terms shall have the following meanings:

“*Acting Holders*” means, at the time of determination, Holders of at least a majority of the outstanding CVRs as set forth on the CVR Register.

“*Apple Covered Awards*” means Apple Share Options, Apple RSUs and Apple PSUs.

“*Assignee*” has the meaning set forth in Section 7.3.

“*Commercially Reasonable Efforts*” means, with respect to OTL-200, those commercially reasonable efforts that are at least commensurate with the level of efforts that a pharmaceutical company of comparable size and resources as those of Purchaser and its Subsidiaries would devote to the development and seeking of regulatory approval for a pharmaceutical product having similar market potential as OTL-200 at a similar stage of its development or product life, taking into account its safety, tolerability and efficacy, its proprietary position and profitability (including pricing and reimbursement status), projected costs to develop such product, the competitiveness of alternative third party products, the patent and other proprietary position, including regulatory exclusivities, of such product, and the regulatory environment and other relevant technical, commercial, legal, scientific and/or medical factors. Without limiting the foregoing, Commercially Reasonable Efforts require that Purchaser: (i) shall cause Apple to devote appropriate resources and personnel with an appropriate level of education, experience and training for the relevant obligation, (ii) shall cause Apple to promptly assign responsibility for the relevant obligation to specific employees who are held accountable for progress and monitor such progress on an on-going basis, (iii) shall cause Apple to set and consistently seek to achieve specific and meaningful objectives and timelines for carrying out such obligation, and (iv) shall cause Apple to consistently make and implement decisions and allocate resources designed to advance progress with respect to relevant objectives and timelines.

“**Change of Control**” means (i) a sale or other disposition of more than 50% of the assets of either Purchaser or Apple on a consolidated basis (other than to any direct or indirect wholly owned Subsidiary of Purchaser), (ii) a merger or consolidation involving either Purchaser or Apple in which Purchaser or Apple, respectively, is not the surviving entity (other than in the case that the surviving entity is a direct or indirect wholly owned Subsidiary of Purchaser), and (iii) any other transaction involving either Purchaser or Apple in which Purchaser or Apple, respectively, is the surviving entity but in which the shareholders of Purchaser or Apple, respectively, immediately prior to such transaction own less than 50% of the surviving entity’s voting power immediately after the transaction.

“**CVR**” means a contingent value right to receive the CVR Payment upon achievement of the CVR Milestone.

“**CVR Beneficial Owners**” has the meaning set forth in [Section 2.3\(b\)](#).

“**CVR Deadline Time**” means 11:59 p.m. U.S. Eastern Time on December 31, 2024.

“**CVR Milestone**” means the granting of Regulatory Approval on or before the CVR Deadline Time.

“**CVR Milestone Non-Achievement Notice**” has the meaning set forth in [Section 2.4\(e\)](#).

“**CVR Notice**” has the meaning set forth in [Section 2.4\(a\)](#).

“**CVR Payment**” means, if the CVR Milestone is achieved prior to the CVR Deadline Time, \$0.10 in cash, per CVR, without interest.

“**CVR Register**” has the meaning set forth in [Section 2.3\(b\)](#).

“**DTC**” means The Depository Trust Company or any successor entity thereto.

“**Eligible Apple ADS Holders**” means the holders of record of Apple ADSs as of the close of business on the Business Day immediately prior to the Effective Date (as defined in the Scheme of Arrangement), which shall include Cede & Co. in respect of all Apple ADSs held through DTC as of such time.

“**Equity Award CVR Payment**” means the applicable CVR Payment that becomes payable in respect of any CVR that was issued in respect of an Apple Covered Award pursuant to the Transaction Agreement.

“**Event of Default**” has the meaning set forth in [Section 6.1](#).

“**Fee Letter**” means that certain Fee Letter, dated as of December 15, 2023, by and between the Purchaser and the Rights Agent.

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

“**Officer’s Certificate**” means a certificate signed by an executive officer or any authorized signatory of Purchaser, in his or her capacity as such an officer, and delivered to the Rights Agent.

“**Permitted Transfer**” means a transfer of CVRs (a) upon the death of a Holder by will or intestacy or 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, (b) pursuant to a court order, (c) by operation of law (including by consolidation, scheme of arrangement or merger) or without consideration in connection with the dissolution, liquidation or termination of any corporation, limited liability company, partnership or other entity, (d) in the case of CVRs held through a nominee (including CVRs held through DTC on behalf of CVR Beneficial Owners), from a nominee to a beneficial owner (and, if applicable, through an intermediary so long as such transfer ends with such beneficial owner), to the extent allowable by DTC, as applicable, (e) if Holder is a partnership or limited liability company, a distribution by the transferring partnership or limited liability company to its partners or members, as applicable or (f) as provided in [Section 2.7](#). For the avoidance of doubt, the distribution of CVRs by the Depository to the Eligible Apple ADS Holders as contemplated by this Agreement, the Scheme of Arrangement, the Deposit Agreement and the Transaction Agreement shall be a “Permitted Transfer” for all purposes hereunder.

“**Regulatory Approval**” means the granting of regulatory approval by the U.S. Food and Drug Administration (FDA) for the commercial marketing and sale of OTL-200 in the United States of America for the treatment or prevention of metachromatic leukodystrophy (MLD), which includes as an intended patient population (for clarity, in addition to any other patient cohorts, if any) both (i) children (or pediatric patients) with late infantile forms of MLD, and (ii) children (or pediatric patients) with early juvenile forms of MLD, in each case that are pre-symptomatic or without clinical manifestations of the disease. For the avoidance of doubt, “Regulatory Approval” will be determined without regard to any post-approval requirements, including without limitation any post-approval confirmatory study requirements or implementation of any risk evaluation and mitigation strategy (REMs) requirements.

“**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.

“**WSFS**” means Wilmington Savings Fund Society, FSB.

1.2 **Rules of Construction.** The following rules of interpretation shall apply to this Agreement: (i) the words “hereof”, “hereby”, “herein” and “hereunder” and words of like import used in this Agreement shall refer to this Agreement as a whole and not to any particular provision of this Agreement; (ii) the headings in this Agreement are included for convenience of reference only and shall be ignored in the construction or interpretation hereof; (iii) any singular term in this Agreement shall be deemed to include the plural, and any plural term the singular, and references to any gender shall include all genders; (iv) whenever the words “include”, “includes” or “including” are used in this Agreement, they shall be deemed to be followed by the words “without limitation”, whether or not they are in fact followed by those words or words of like import; (v) “writing”, “written” and comparable terms refer to printing, typing and other means of reproducing words (including electronic media) in a visible form; (vi) references to any Applicable Law shall be deemed to refer to such Applicable Law as amended from time to time and to any rules or regulations promulgated thereunder; (vii) references to any Person include the successors and permitted assigns of that Person; (viii) references to “dollars” and “\$” means U.S. dollars; (ix) the word “extent” in the phrase “to the extent” shall mean the degree to which a subject or other thing extends and such phrase shall not mean “if”; (x) the parties hereto have participated jointly in the negotiation and drafting of this Agreement and, in the event an ambiguity or question of intent or interpretation arises, this Agreement shall be construed as jointly drafted by the parties hereto 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, and (xi) the terms “or”, “any” and “either” are not exclusive.

## 2. CONTINGENT VALUE RIGHTS

2.1 **CVRs.** The CVRs represent the contingent rights of Holders to receive the CVR Payment upon achievement of the CVR Milestone. The initial Holders shall be determined pursuant to the terms of the Transaction Agreement and this Agreement, and a list of the initial Holders shall be furnished to the Rights Agent by or on behalf of Purchaser in accordance with Section 4.1 hereof and supplemented by Purchaser by written notice to the Rights Agent from time to time after the date hereof, including upon the issuance of additional CVRs following any exercise of the Warrants in accordance with the terms thereof.

2.2 **Non-transferable.** 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 such sale, assignment, transfer, pledge, encumbrance or disposal that is not a Permitted Transfer shall be null and void *ab initio* and of no effect.

### 2.3 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 (including issuances of CVRs after the date hereof) and transfers of CVRs as herein provided. In the case of CVRs to be received by the holders of Apple Covered Awards pursuant to the Transaction Agreement or CVRs to be received by registered holders of Scheme Shares (other than the Depositary Shares), such CVRs shall initially be registered in the name and address of the holder of such Apple Covered Awards or Scheme Shares, as applicable, as set forth in the records of Apple at the Scheme Record Time. In the case of CVRs to be received by the Depositary Custodian in respect of the Depositary Shares, (i) the CVR Register shall be updated to reflect the distribution of such CVRs to the Eligible Apple ADS Holders and such CVRs shall initially be registered in the name and address of such Eligible Apple ADS Holders as set forth in the records of the Depositary and (ii) the CVR Register will initially show one position for Cede & Co. representing all of the CVRs that are distributed in respect of Apple ADSs held through DTC on behalf of the Beneficial Owners (as defined in the Deposit Agreement) of such Apple ADSs (“**CVR Beneficial Owners**”). The Rights Agent will have no responsibility whatsoever directly to the CVR Beneficial Owners or DTC participants with respect to transfers of CVRs. The Rights Agent will have no responsibilities whatsoever with regard to the distribution of payments by DTC to such CVR Beneficial Owners.

(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 documentation reasonably requested by the Rights Agent in form reasonably satisfactory to the Rights Agent pursuant to its guidelines, duly executed by the Holder thereof, the Holder's attorney duly authorized in writing, the Holder's personal representative or the Holder's survivor, as applicable, and setting forth in reasonable detail the circumstances relating to the transfer. Upon receipt of such written notice, the Rights Agent shall, subject to its reasonable determination that the transfer instrument is in proper form, notify Purchaser that it has received such written notice. Upon receipt of such notice from the Rights Agent, Purchaser shall reasonably 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 Purchaser so reasonably determines that it does so comply, Purchaser shall reply to the Rights Agent in writing with instructions to register the transfer of such CVR in the CVR Register and notify Purchaser of the same. Upon receiving such written reply from Purchaser, the Rights Agent shall register the transfer of the CVRs in the CVR Register and notify the Purchaser of the same. No service charge shall be made for any registration of transfer of a CVR, but the Purchaser (subject at all times to the Purchaser's obligations under section 11.04 of the Transaction Agreement) and Rights Agent may require payment of a sum sufficient to cover any stamp duty or other transfer Tax or charge that is imposed in connection with any such registration of transfer. The Rights Agent shall have no duty or obligation to take any action under any section of this Agreement that requires the payment of any such Taxes or charges unless and until the Rights Agent is satisfied that all such Taxes or charges have been paid. All duly transferred CVRs registered in the CVR Register shall be the valid obligations of Purchaser 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 unless and until registered in the CVR Register.

(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 request, the Rights Agent is hereby authorized to, and shall promptly, record the change of address in the CVR Register.

## 2.4 Payment Procedures.

(a) If the CVR Milestone is achieved prior to the CVR Deadline Time, Purchaser shall, within forty (40) calendar days of the achievement of the CVR Milestone, deliver to the Rights Agent a notice (a “**CVR Notice**”) indicating the achievement of the CVR Milestone and that the Holders are entitled to receive the CVR Payment.

(b) The Rights Agent shall promptly, and in any event within ten (10) Business Days of receipt of funds pursuant to Section 4.2, (i) send each Holder at its registered address (or, in the case of Cede & Co., pursuant to the applicable procedures of DTC) a copy of the CVR Notice and (ii) pay, subject to receipt of cash from Purchaser in accordance with Section 4.2 and any letter of instruction reasonably required by the Rights Agent, the CVR Payment (excluding certain Equity Award CVR Payments to be paid via payroll systems as contemplated by the next sentence) to each of the Holders (x) by check mailed to the address of such Holder reflected in the CVR Register as of 5:00 p.m. New York City time on the date of the CVR Notice, (y) with respect to any such Holder that is due an amount in excess of \$5,000 in the aggregate who has provided the Rights Agent wiring instructions in writing as of the close of business on the date of the CVR Notice, by wire transfer of immediately available funds to the account specified on such instructions or (z) with respect to Cede & Co., by wire transfer of immediately available funds pursuant to the applicable procedures of DTC. To the extent Apple, Purchaser or any of its Subsidiaries is required under Applicable Law to deduct or withhold any employee Tax (including any employee National Insurance Contribution in the United Kingdom (or corresponding social security contribution in any other jurisdiction)) from any Equity Award CVR Payment, Purchaser shall or shall cause one of its Subsidiaries (including Apple) as an agent on its behalf to (A) promptly following the achievement of the CVR Milestone, notify the Rights Agent in writing of the Equity Award CVR Payments it intends to satisfy in this manner and (B) prior to or substantially concurrently with the payment of the CVR Payments by the Rights Agent under this Section 2.4(b), pay, through its applicable payroll system, the applicable Equity Award CVR Payments.

(c) If any funds delivered to the Rights Agent for payment to Holders as CVR Payments remain undistributed to the Holders on the date that is one year after the date of the CVR Notice, Purchaser shall be entitled to require the Rights Agent to deliver to Purchaser or its designee any funds which had been made available to the Rights Agent in connection with the CVR Payments and not disbursed to the Holders (including, all interest and other income received by the Rights Agent in respect of all funds made available to it), and, thereafter, such Holders shall be entitled to look to Purchaser (subject to abandoned property, escheat and other similar Applicable Laws) only as general creditors thereof with respect to the CVR Payment that may be payable to such Holder.

(d) Neither Purchaser, the Rights Agent nor any of their Affiliates shall be liable to any Holder for any CVR Payment delivered to a public official pursuant to any abandoned property, escheat or other similar Applicable Laws. Any amounts remaining unclaimed by such Holders at such time at which such amounts would otherwise escheat to or become property of any Governmental Authority shall become, to the extent permitted by Applicable Laws, the property of Purchaser or its designee, free and clear of all claims or interest of any Person previously entitled thereto. In addition to and not in limitation of any other indemnity obligation herein, Purchaser 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 Purchaser.

(e) If the CVR Milestone is not achieved prior to the CVR Deadline Time, Purchaser shall, within forty-five (45) calendar days of the CVR Deadline Time, deliver to the Rights Agent a notice (a “***CVR Milestone Non-Achievement Notice***”) indicating that such CVR Milestone has not been achieved. The Rights Agent shall promptly, and in any event within fifteen (15) calendar days of receipt, deliver a copy of such CVR Milestone Non-Achievement Notice to the Holders. The Rights Agent will deliver to Purchaser a certificate certifying the date of delivery of such CVR Milestone Non-Achievement Notice to the Holders. If the Rights Agent does not receive from the Acting Holders a written objection to a CVR Milestone Non-Achievement Notice within forty-five (45) calendar days after the date of delivery of such CVR Milestone Non-Achievement Notice by the Rights Agent to the Holders, the Holders will be deemed to have accepted such CVR Milestone Non-Achievement Notice, and neither Purchaser nor its Subsidiaries will have any further obligation hereunder with respect to such CVR Payment or otherwise with respect to such CVR Milestone.

**2.5 United Kingdom Tax Treatment.** The parties hereto intend that, for United Kingdom tax purposes, the CVR Payments and the Equity Award CVR Payments shall be subject to all applicable deductions and withholding (if any) as required by Applicable Law. To the extent Apple, Purchaser or any of its Subsidiaries is required under Applicable Law to deduct or withhold any employee Tax (including any employee National Insurance contributions in the United Kingdom) from any Equity Award CVR payments, then it shall do so in accordance with Section 2.4.

**2.6 No Voting, Dividends or Interest; No Equity or Ownership Interest.**

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

(b) The CVRs shall not represent any equity or ownership interest in Purchaser or in any constituent company to the Transaction or any of their respective Affiliates.

**2.7 Ability to Abandon CVR.** A Holder may at any time, at such Holder’s option, abandon all of such Holder’s remaining rights in a CVR by transferring such CVR to Purchaser or any of its Affiliates without consideration therefor. Purchaser shall notify the Rights Agent in writing of the abandonment by Holder of such CVR. Nothing in this Agreement shall prohibit Purchaser or any of its Affiliates from offering to acquire or acquiring any CVRs for consideration from the Holders, in private transactions or otherwise, in its sole discretion. Any CVRs acquired by Purchaser or any of its Affiliates shall be automatically deemed extinguished and no longer outstanding for purposes of the definition of Acting Holders and Section 5 and Section 6. Purchaser shall notify the Rights Agent in writing of any such extinguishment of a CVR.

### 3. THE RIGHTS AGENT

#### 3.1 Certain Duties and Responsibilities.

(a) Purchaser hereby appoints the Rights Agent to act as rights agent for Purchaser in accordance with the express terms and conditions set forth in this Agreement (and no implied terms and conditions), and the Rights Agent hereby accepts such appointment. The Rights Agent shall not have any liability for any actions taken, suffered or omitted to be taken in connection with this Agreement, except to the extent of its gross negligence, bad faith or willful or intentional misconduct.

(b) The Acting Holders may direct the Rights Agent to act on behalf of the Holders in enforcing any of their rights hereunder and, subject to the following sentence, the Rights Agent shall act in accordance with such direction. The Rights Agent shall be under no obligation to institute any action, suit or proceeding, or to take any other action likely to result in the incurrence of expenses by the Rights Agent, unless the Acting Holders (on behalf of the Holders) shall furnish the Rights Agent with reasonable security and indemnity for any costs and expenses that may be incurred. Any action, suit or proceeding instituted by the Rights Agent shall be brought in its name as the Rights Agent and any recovery in connection therewith shall be for the proportionate benefit of all the Holders, as their respective rights or interests may appear.

3.2 Certain Rights of the Rights Agent. 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 Purchaser 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 gross negligence, bad faith or willful or intentional misconduct on its part, incur no liability and be held harmless by Purchaser for or in respect of any action taken, suffered or omitted to be taken by it under the provisions of this Agreement in reliance upon such certificate;

(c) the Rights Agent may engage and consult with counsel of its selection and the written advice of such counsel or any opinion of counsel shall be full and complete authorization and protection and shall be held harmless by Purchaser 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 or otherwise in respect of the premises;

(f) the Rights Agent shall not be liable for or by reason of, and shall be held harmless by Purchaser 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 Purchaser only;

(g) the Rights Agent shall have no liability and shall be held harmless by Purchaser 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 Purchaser), nor shall it be responsible for any breach by Purchaser of any covenant or condition contained in this Agreement, except for any such breach resulting from the Rights Agent's gross negligence, bad faith or willful or intentional misconduct;

(h) Purchaser agrees to indemnify the Rights Agent for, and hold the Rights Agent harmless against, any loss, liability, claim, demand, suit or expense arising out of or in connection with Rights Agent's duties under this Agreement, including the reasonable out-of-pocket costs and expenses of defending Rights Agent against any claim, charge, demand, suit or loss, unless such loss has been determined by a court of competent jurisdiction to be a result of Rights Agent's gross negligence, bad faith or willful or intentional misconduct;

(i) the Rights Agent shall not be liable for consequential damages under any provision of this Agreement or for any consequential damages arising out of any act or failure to act hereunder in the absence of gross negligence, bad faith or willful or intentional misconduct on its part;

(j) Purchaser agrees (i) to pay the fees of the Rights Agent in connection with the Rights Agent's performance of its obligations hereunder, including those set out in the Fee Letter and any other fees agreed to between Purchaser and Rights Agent in writing, and (ii) to reimburse the Rights Agent promptly upon demand for all reasonable and documented out-of-pocket expenses, including all Taxes (other than income, receipt, franchise or similar Taxes) and governmental charges, incurred by the Rights Agent in the performance of its obligations under this Agreement; and

(k) 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.

(l) all funds received by 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 WSFS as agent for Purchaser and deposited in one or more bank accounts to be maintained by WSFS in its name as agent for Purchaser. Until paid pursuant to the terms of this Agreement, WSFS 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 in accordance with this paragraph, including any losses resulting from a default by any bank, financial institution or other third party.

### 3.3 Resignation and Removal; Appointment of Successor.

(a) The Rights Agent may resign at any time by giving written notice thereof to Purchaser specifying a date when such resignation shall take effect, which notice shall be sent at least sixty (60) calendar days prior to the date so specified but in no event shall such resignation become effective until a successor Rights Agent has been appointed and accepted such appointment in accordance with Section 3.4. Purchaser has the right to remove the Rights Agent at any time by specifying a date when such removal shall take effect but no such removal shall become effective until a successor Rights Agent has been appointed and accepted such appointment in accordance with Section 3.4. Notice of such removal shall be given by Purchaser to the Rights Agent, which notice shall be sent at least sixty (60) calendar 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, Purchaser shall, as soon as is reasonably practicable, appoint a qualified successor Rights Agent who shall be a stock transfer agent of national reputation in the US or the corporate trust department of an international commercial bank. Notwithstanding the foregoing, if Purchaser shall fail to make such appointment within a period of sixty (60) calendar 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 may apply to any court of competent jurisdiction for the appointment of a new Rights Agent. 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) Purchaser shall give notice of each resignation and each removal of a Rights Agent and each appointment of a successor Rights Agent through the facilities of DTC in accordance with DTC's procedures (in respect of CVRs registered in the name of Cede & Co. only) and/or by mailing written notice of such event 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 Purchaser 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 transmitted at the expense of Purchaser. Failure to give any notice provided for in this Section 3.3, however, 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) Notwithstanding anything else in this Section 3.3 to the contrary, unless consented to in writing by the Acting Holders, Purchaser shall not appoint as a successor Rights Agent any Person that is not a stock transfer agent of national reputation in the U.S. or the corporate trust department of an international commercial bank.

(e) The Rights Agent will cooperate with Purchaser and any successor Rights Agent as reasonably requested 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.

3.4 Acceptance of Appointment by Successor. Every successor Rights Agent appointed hereunder shall execute, acknowledge and deliver to Purchaser 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 Purchaser 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, trusts and duties of the retiring Rights Agent.

#### 4. COVENANTS

4.1 List of Holders. Promptly following the Effective Time (including from time to time following the issuance of additional CVRs, if any), Purchaser shall furnish or cause to be furnished to the Rights Agent, in a form reasonably satisfactory to the Rights Agent, the names and addresses of the Holders (including the Eligible Apple ADS Holders and the Holders who held an Apple Covered Award).

4.2 Milestone Payments. If the CVR Milestone is achieved prior to the CVR Deadline Time, Purchaser shall, or shall cause, promptly (but in any event no later than twenty (20) calendar days) following the delivery of the CVR Notice, deposit with the Rights Agent, for payment to the Holders in accordance with Section 2.4, the aggregate amount necessary to pay the CVR Payments to the Holders (other than holders entitled to certain Equity Award CVR Payments as set forth in Section 2.4).

4.3 Books and Records. Purchaser shall, and shall cause its Subsidiaries to, keep true, complete and accurate records in sufficient detail to enable the Holders and their consultants or professional advisors to determine the amounts payable hereunder.

4.4 Further Assurances. Purchaser agrees that it will perform, execute, acknowledge and deliver or cause to be performed, executed, acknowledged and delivered, all such further and other acts, instruments and assurances as may reasonably be required by the Rights Agent for the carrying out or performing by the Rights Agent of the provisions of this Agreement.

4.5 Commercially Reasonable Efforts. Following the Closing and until the CVR Deadline Time, Purchaser and its Subsidiaries shall use Commercially Reasonable Efforts to develop OTL-200 and achieve the CVR Milestone; it being understood, for the avoidance of doubt, that use of Commercially Reasonable Efforts does not guarantee that Purchaser will achieve the CVR Milestone by a specific date or at all. Without limiting the foregoing, neither Purchaser nor any of its controlled Affiliates shall act in bad faith for the purpose of avoiding achievement of the CVR Milestone or the payment of any CVR Payment.

## 5. AMENDMENTS

### 5.1 Amendments without Consent of Holders.

(a) Without the consent of any Holders or the Rights Agent, Purchaser 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 Purchaser such further covenants, restrictions, conditions or provisions as Purchaser shall consider to be for the protection of the Holders; *provided*, that in each case, such provisions do not 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 in the Transaction Agreement, 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 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 1933 Act, the 1934 Act or any applicable U.S. state securities or “blue sky” laws or require the publication of a prospectus under the Prospectus Regulation (EU) 2017/1129 (including as it has been retained in the United Kingdom by virtue of the European Union (Withdrawal) Act 2018, as amended);

(v) to evidence the assignment of this Agreement by Purchaser as provided in Section 7.3; or

(vi) any other amendments hereto for the purpose of adding, eliminating or changing any provisions of this Agreement, unless such addition, elimination or change is adverse to the interests of the Holders.

(b) Without the consent of any Holders, Purchaser and the Rights Agent, at any time and from time to time, may enter into one or more amendments hereto to reduce the number of CVRs, in the event any Holder agrees to renounce such Holder’s rights under this Agreement or to transfer CVRs to Purchaser pursuant to Section 2.7.

(c) Promptly after the execution by Purchaser and/or the Rights Agent of any amendment pursuant to the provisions of this Section 5.1, Purchaser shall transmit or cause the Rights Agent to transmit a notice thereof through the facilities of DTC in accordance with DTC’s procedures (in respect of CVRs registered in the name of Cede & Co. only) and/or by first class mail to the Holders at their addresses as they appear on the CVR Register, setting forth such amendment.

## 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 any Holder or the Rights Agent), with the consent of the Holders of not less than a majority of the outstanding CVRs as set forth in the CVR Register, whether evidenced in writing or taken at a meeting of the Holders, Purchaser 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 adverse to the interest of the Holders. For the avoidance of doubt, any amendment to the defined terms incorporated by reference into this Agreement, by way of an amendment to the Transaction Agreement, shall not be given effect under this Agreement if such amendment is adverse to the interest of the Holders unless the consent of the Holders of not less than a majority of the outstanding CVRs as set forth in the CVR Register consent to such amendment.

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

5.3 Execution of Amendments. Prior to executing any amendment permitted by this Section 5, the Rights Agent shall be entitled to receive, and shall be fully protected in relying upon, an opinion of counsel selected by Purchaser 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 Purchaser. The Rights Agent may, but is not obligated to, enter into any such amendment that affects the Rights Agent's own rights, powers, trusts or duties under this Agreement or otherwise.

5.4 Effect of Amendments. Upon the execution of any amendment under this Section 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.

## 6. REMEDIES OF THE HOLDERS

6.1 Event of Default. "*Event of Default*" with respect to the CVRs, means each one of the following events which shall have occurred and be continuing (whatever the reason for such Event of Default and whether it shall be voluntary or involuntary or be effected by operation of Applicable Law or pursuant to any judgment, decree or order of any court or any order, rule or regulation of any Governmental Authority):

(a) default in the payment by Purchaser pursuant to the terms of this Agreement of all or any part of the CVR Payment after a period of ten (10) Business Days after the CVR Payment shall become due and payable; or

(b) material default in the performance, or breach in any material respect, of any covenant or warranty of Purchaser hereunder (other than a default in whose performance or whose breach is elsewhere in this Section 6.1 specifically dealt with), and continuance of such default or breach for a period of ninety (90) calendar days after a written notice specifying such default or breach and requiring it to be remedied is given, which written notice states that it is a "Notice of Default" hereunder and is sent by registered or certified mail to Purchaser by the Rights Agent or to Purchaser and the Rights Agent by the Acting Holders

6.2 General Enforcement of Rights of Holders. If an Event of Default occurs and is continuing (and has not been cured or waived), then, and in each and every such case, the Acting Holders, following notice in writing to Purchaser and Rights Agent, may, at their own expense, commence a proceeding to protect the rights of the Holders, including to obtain payment for any amounts then due and payable. In the event that, at any time after the Acting Holders shall have commenced such proceeding, and before any award shall have been obtained, Purchaser shall pay or shall deposit with the Rights Agent a sum sufficient to pay all amounts which shall have become due under this Agreement and such amount as shall be sufficient to cover reasonable compensation to the Rights Agent, its agents, attorneys and counsel (if any), and all Events of Default under this Agreement shall have been cured, waived or otherwise remedied as provided herein, then and in every such case the Acting Holders, by written notice to Purchaser and to the Rights Agent, shall waive all defaults that are the subject of such proceeding, but no such waiver or rescission and annulment shall extend to or shall affect any subsequent default.

6.3 Rights upon Certain Insolvency Proceedings Involving Purchaser. In the event of an insolvency, bankruptcy or similar proceeding of Purchaser, Holders shall be entitled to assert claims in such proceeding and take related actions in pursuit of such claims to seek a payment finally determined to be due and payable pursuant to Section 2.4(b) under this Agreement and has not been paid when due with respect to any payment that may be claimed by or on behalf of Purchaser or by any creditor of Purchaser. Notwithstanding any other provision in this Agreement, the right of any Holder of any CVR to receive payment of the amounts that a CVR Notice indicates are payable in respect of such CVR on or after the applicable due date, or to commence proceedings for the enforcement of any such payment on or after such due date, shall not be impaired or affected without the consent of such Holder.

## 7. OTHER PROVISIONS OF GENERAL APPLICATION

7.1 Notices to the Rights Agent and Purchaser. Any notice or other communication required or permitted to be delivered to Purchaser or the Rights Agent under this Agreement shall be in writing sent via email and shall be given:

If to the Rights Agent, to it at:

Wilmington Savings Fund Society, FSB  
500 Delaware Avenue, 11<sup>th</sup> Floor  
Wilmington, DE 19801

Attention: Patrick Healy, Senior Vice President  
Email: [\*\*\*]

With a copy to:

McDermott Will & Emery LLP  
One Vanderbilt Avenue  
New York, NY 10017-3852

Attention: Jonathan Levine  
Email: [\*\*\*]

If to Purchaser, to it at:

Kyowa Kirin Co., Ltd.  
Otemachi Financial City Grand Cube, 1-9-2 Otemachi,  
Chiyoda-ku, Tokyo 100-0004, Japan

Attention: Corporate Strategy Department  
Email: [\*\*\*]  
[\*\*\*]  
[\*\*\*]

With a copy to:

Morrison & Foerster LLP  
Shin Marunouchi Building, 29th Fl.  
1-5-1, Marunouchi, Chiyoda-ku

Attention: Gary Smith  
Email: [\*\*\*]

or to such other email address as such party may hereafter specify for the purpose by like notice to the other party hereto. All such notices, requests and other communications shall be deemed received on the date of dispatch by the sender thereof (to the extent that no “bounce back”, “out of office” or similar message indicating non-delivery is received with respect thereto), in each case to the required recipient as set forth above, if such dispatch is made on a Business Day by 5:00 p.m. in the local time of the intended recipient or, if made on a Business Day after 5:00 p.m. in the local time of the intended recipient, such notice, request or communication shall be deemed to have been received on the next succeeding Business Day.

7.2 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 (in respect of CVRs registered in the name of Cede & Co. only) and/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.

**7.3 Purchaser Successors and Assigns.** Purchaser may assign, in its sole discretion and without the consent of any other party, any or all of its rights, interests and obligations hereunder to one or more of its controlled Affiliates (each, an “*Assignee*”) and any such Assignee may thereafter assign, in its sole discretion and without the consent of any other party, any or all of its rights, interests and obligations hereunder to one or more additional Assignees (as defined in the foregoing parenthetical); provided, however, that in connection with any assignment to an Assignee, Purchaser shall remain primarily liable for the performance of every obligation, agreement and covenant of this Agreement on the part of Purchaser to be performed or observed, and that such assignment would not be adverse to the Holders. This Agreement shall be binding upon, and shall be enforceable by and inure solely to the benefit of, the parties hereto and their respective successors and assigns. Subject to compliance with the requirements set forth in this Section 7.3 relating to assignments, this Agreement shall not restrict Purchaser’s, any Assignee’s or any of their respective successors’ ability to merge or consolidate with, or sell, issue, license or dispose of its stock or other equity interests or assets to, any other Person, or spin-off or split-off. Each of Purchaser’s successors (including following a Change of Control) and each Assignee shall, by a supplemental contingent consideration payment agreement or other acknowledgement executed and delivered to the Rights Agent, expressly assume payment of amounts on all of the CVRs and the performance of every obligation, agreement and covenant of this Agreement on the part of Purchaser to be performed or observed. The Rights Agent may not assign this Agreement without Purchaser’s written consent. Any attempted assignment of this Agreement or of any rights, interests or obligations in violation of this Section 7.3 shall be null and void *ab initio* and of no effect.

**7.4 No Third Party Beneficiaries.** Nothing in this Agreement, express or implied, shall give to any Person (other than the parties hereto, the Holders and their permitted successors and assigns hereunder, subject to the limitations set forth herein) any benefit or any legal or equitable right, remedy or claim under this Agreement or under any covenant or provision herein contained, all such covenants and provisions being for the sole benefit of the parties hereto, the Holders and their permitted successors and assigns. The Holders shall have no rights hereunder except as are expressly set forth herein. Without limitation of the rights of the Rights Agent set forth herein, the Acting Holders will have the sole right, on behalf of all Holders, to institute (or direct the Rights Agent to institute) any action or proceeding with respect to this Agreement, and no individual Holder or other group of Holders will be entitled to exercise such rights (provided that the foregoing shall not limit the ability of an individual Holder to seek a payment due from the applicable party pursuant to Section 2.4(b)) solely to the extent such payment has been finally determined to be due and payable under this Agreement and has not been paid when due). Acting Holders acting pursuant to the preceding sentence on behalf of all Holders shall have no liability to the other Holders for such actions.

**7.5 Governing Law.**

(a) This Agreement, the CVRs and all disputes, claims, actions, suits or proceedings based upon, arising out of or related to this Agreement, the CVRs or the transactions contemplated hereby, shall be governed by and construed in accordance with the laws of the State of Delaware, without regard to the conflicts of law rules or principles that would result in the application of the law of any other state or jurisdiction.

(b) Each of the parties hereto irrevocably and unconditionally agrees that any legal action or proceeding with respect to this Agreement, the CVRs, the transactions contemplated hereby or the rights and obligations arising hereunder, or for recognition and enforcement of any judgment in respect of this Agreement, the CVRs, the transactions contemplated hereby or the rights and obligations arising hereunder brought by any other party hereto or its successors or assigns, shall be brought and determined exclusively in the Delaware Court of Chancery and any state appellate court therefrom within the State of Delaware (or, solely if the Delaware Court of Chancery declines to accept jurisdiction over a particular matter, any state or federal court within the State of Delaware). Each of the parties hereto hereby irrevocably and unconditionally submits with regard to any such action or proceeding for itself and in respect of its property to the personal jurisdiction of the aforesaid courts and agrees that it will not bring any action relating to this Agreement or any of the transactions contemplated by this Agreement in any court other than the aforesaid courts. Each of the parties hereto hereby irrevocably waives, and agrees not to assert, by way of motion, as a defense, counterclaim or otherwise, in any action or proceeding with respect to this Agreement, (a) any claim that it is not personally subject to the jurisdiction of the above-named courts, (b) any claim that it or its property is exempt or immune from jurisdiction of any such court or from any legal process commenced in such courts (whether through service of notice, attachment prior to judgment, attachment in aid of execution of judgment, execution of judgment or otherwise) and (c) to the fullest extent permitted by Applicable Law, any claim that (i) the suit, action or proceeding in such court is brought in an inconvenient forum, (ii) the venue of such suit, action or proceeding is improper or (iii) this Agreement, or the subject matter hereof, may not be enforced in or by such courts. To the fullest extent permitted by Applicable Law, each of the parties hereto hereby consents to the service of process in accordance with Section 7.1 and Section 7.2 in any action relating to this Agreement, the CVRs or any of the transactions contemplated by this Agreement; provided, that nothing herein shall affect the right of any party to serve legal process in any other manner permitted by Applicable Law.

(c) EACH PARTY ACKNOWLEDGES AND AGREES THAT ANY CONTROVERSY WHICH MAY ARISE UNDER THIS AGREEMENT OR THE CVRS IS LIKELY TO INVOLVE COMPLICATED AND DIFFICULT ISSUES, AND THEREFORE EACH SUCH PARTY HEREBY IRREVOCABLY AND UNCONDITIONALLY WAIVES ANY RIGHT SUCH PARTY MAY HAVE TO A TRIAL BY JURY IN RESPECT OF ANY LITIGATION DIRECTLY OR INDIRECTLY ARISING OUT OF OR RELATING TO THIS AGREEMENT, THE CVRS, THE TRANSACTION OR THE OTHER TRANSACTIONS CONTEMPLATED HEREBY. EACH PARTY CERTIFIES AND ACKNOWLEDGES THAT (A) NO REPRESENTATIVE, AGENT OR ATTORNEY OF ANY OTHER PARTY HAS REPRESENTED, EXPRESSLY OR OTHERWISE, THAT SUCH OTHER PARTY WOULD NOT, IN THE EVENT OF LITIGATION, SEEK TO ENFORCE THE FOREGOING WAIVER, (B) EACH PARTY UNDERSTANDS AND HAS CONSIDERED THE IMPLICATIONS OF THIS WAIVER, (C) EACH PARTY MAKES THIS WAIVER VOLUNTARILY, AND (D) EACH PARTY HAS BEEN INDUCED TO ENTER INTO THIS AGREEMENT BY, AMONG OTHER THINGS, THE MUTUAL WAIVERS AND CERTIFICATIONS IN THIS SECTION 7.5(C).

7.6 Severability. In the event that any provision of this Agreement becomes or is declared by a court of competent jurisdiction to be illegal, void or unenforceable, the remainder of this Agreement shall continue in full force and effect and the application of such provision to other Persons or circumstances shall be interpreted so as reasonably to effect the intent of the parties hereto. The parties hereto further agree to replace such illegal, void or unenforceable provision of this Agreement with a valid and enforceable provision that will achieve, to the extent possible, the economic, business and other purposes of such illegal, void or unenforceable provision.

7.7 Termination. This Agreement shall be terminated and of no force or effect, the parties hereto shall have no liability hereunder (other than with respect to monies due and owing by Purchaser to Rights Agent), and no payments shall be required to be made, upon the earliest to occur of (a) the payment of the full amount of the potential CVR Payment required to be paid under the terms of this Agreement pursuant to Section 2.4, (b) the termination of the Transaction Agreement in accordance with its terms prior to the occurrence of the Effective Time and (c) sixty (60) calendar days following the CVR Deadline Time. Notwithstanding the foregoing, no such termination shall affect any rights or obligations accrued prior to the effective date of such termination (including in respect of breaches of this Agreement by Purchaser prior to such termination) or this Section 6, which shall survive the termination of this Agreement, or the resignation, replacement or removal of the Rights Agent.

7.8 Relationship of the Parties. Neither party hereto is a fiduciary or agent of the other. Under no circumstance shall either party hereto (i) be deemed to be a fiduciary or agent of the other or (ii) have the right to bind the other party hereto as its agent, in each case, by reason of this Agreement or for any other purpose.

7.9 Entire Agreement; Counterparts. This Agreement and the Transaction Agreement constitute the entire agreement and supersede all contemporaneous and prior agreements and understandings, both written and oral, among or between the parties hereto, with respect to the subject matter hereof and thereof. If and to the extent that any provision of this Agreement is inconsistent or conflicts with the Transaction Agreement, this Agreement shall govern and be controlling. This Agreement may be executed in counterparts, each of which shall be deemed an original and all of which shall constitute one and the same instrument. The exchange of a fully executed Agreement (in counterparts or otherwise) by .PDF shall be sufficient to bind the Parties to the terms and conditions of this Agreement.

[REMAINDER OF PAGE INTENTIONALLY LEFT BLANK]

IN WITNESS WHEREOF, each of the parties has caused this Agreement to be executed on its behalf by its duly authorized officers as of the day and year first above written.

**Kyowa Kirin Co., Ltd.**

By: /s/ Masashi Miyamoto  
Name: Masashi Miyamoto  
Title: Representative Director of the Board, President and  
Chief Executive Officer

**Wilmington Savings Fund Society, FSB**

By: /s/ Raye Goldsborough  
Name: Raye Goldsborough  
Title: Vice President