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**UNITED STATES  
SECURITIES AND EXCHANGE COMMISSION**  
Washington, D.C. 20549

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**FORM 8-K**

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**CURRENT REPORT**  
Pursuant to Section 13 or 15(d)  
of the Securities Exchange Act of 1934

**August 4, 2014**  
Date of Report (Date of earliest event reported)

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**JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY**

(Exact name of registrant as specified in its charter)

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**Ireland**  
(State or other jurisdiction  
of incorporation)

**001-33500**  
(Commission  
File No.)

**98-1032470**  
(IRS Employer  
Identification No.)

**Fourth Floor, Connaught House,  
1 Burlington Road, Dublin 4, Ireland**  
(Address of principal executive offices, including zip code)

**011-353-1-634-7800**  
(Registrant's telephone number, including area code)

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

As previously announced, on July 1, 2014, Jazz Pharmaceuticals Public Limited Company (the “Company”), Jazz Pharmaceuticals International II Limited (“Buyer”), a wholly-owned subsidiary of the Company, and Gentium S.p.A., a majority-owned subsidiary of the Company (“Gentium” and together with Buyer and the Company, “Jazz Pharmaceuticals”), entered into an Assignment Agreement (the “Assignment Agreement”) with Sigma-Tau Pharmaceuticals, Inc. (“Sigma-Tau”).

Pursuant to the terms of the Assignment Agreement, Sigma-Tau agreed to sell, transfer and assign to Buyer all of Sigma-Tau’s right, title and interest in and to the License and Supply Agreement, dated December 7, 2001, as amended, between Gentium and Sigma-Tau (the “License Agreement”), pursuant to which Sigma-Tau held certain rights to market defibrotide in North America, Central America and South America, and Buyer agreed to assume certain liabilities of Sigma-Tau under the License Agreement. Sigma-Tau also agreed to sell, transfer and assign to Buyer all of Sigma-Tau’s and its affiliates’ right, title and interest in and to certain know-how and certain other assets that, in each case, relate to defibrotide.

On August 4, 2014, the parties closed the transaction contemplated by the Assignment Agreement (the “Closing”). At the Closing, Buyer made an initial upfront payment of \$75 million to Sigma-Tau. Buyer is also obligated to make milestone payments of up to \$175 million to Sigma-Tau (the “Milestone Payments”) comprised of: (i) \$25 million upon the acceptance for filing by the U.S. Food and Drug Administration (“FDA”) of the first new drug application (“NDA”) for defibrotide for veno-occlusive disease (“VOD”); and (ii) up to an additional \$150 million based on the timing of potential FDA approval of defibrotide for VOD. The Company has guaranteed Buyer’s payment obligations under the Assignment Agreement, including to make the Milestone Payments and to cover any indemnification claims that are Buyer’s responsibility. The terms of the Assignment Agreement, including the consideration, were negotiated between Jazz Pharmaceuticals and Sigma-Tau at arms’ length.

The foregoing description of the Assignment Agreement and the transaction contemplated thereby does not purport to be complete and is subject to, and qualified in its entirety by reference to, the full text of the Assignment Agreement, which is filed as Exhibit 2.1 hereto and incorporated herein by reference. The Company has sought confidential treatment for certain portions of the Assignment Agreement pursuant to a confidential treatment request submitted to the Securities and Exchange Commission.

The Assignment Agreement has been included solely to provide investors and security holders with information regarding its terms. It is not intended to be a source of financial, business or operational information, or to provide any other factual information, about the rights acquired pursuant to, or any of the parties to, the Assignment Agreement. The representations, warranties and covenants contained in the Assignment Agreement are made only for purposes of the Assignment Agreement and are made as of specific dates; are solely for the benefit of the parties (except as specifically set forth therein); may be subject to qualifications and limitations agreed upon by the parties in connection with negotiating the terms of the Assignment Agreement, including being qualified by confidential disclosures made for the purpose of allocating contractual risk between the parties, instead of establishing matters as facts; and may be subject to standards of materiality and knowledge applicable to the contracting parties that differ from those applicable to investors or security holders. Investors and security holders should not rely on the representations, warranties and covenants or any description thereof as characterizations of the actual state of facts or condition of the rights acquired pursuant to, or any of the parties to, the Assignment Agreement. Moreover, information concerning the subject matter of the representations, warranties and covenants may change after the date of the Assignment Agreement, as applicable, which subsequent information may or may not be fully reflected in public disclosures.

### *Relationships*

The Company became the indirect majority shareholder of Gentium in January 2014 pursuant to a tender offer and ultimately acquired more than 99% of the outstanding voting securities of Gentium (the “Gentium Acquisition”). Sigma-Tau is an affiliate of Sigma-Tau Finanziaria S.p.A., which together with its affiliated entities beneficially owned, based on public filings, approximately 17% of the outstanding voting securities of Gentium immediately prior to the Gentium Acquisition and was also represented on Gentium’s board of directors.

**Item 9.01. Financial Statements and Exhibits.****(d) Exhibits**

<u>Exhibit Number</u>	<u>Description</u>
2.1	Assignment Agreement, dated July 1, 2014, by and among Jazz Pharmaceuticals International II Limited, Sigma-Tau Pharmaceuticals, Inc., Jazz Pharmaceuticals plc and Gentium S.p.A.*

\* Schedules have been omitted pursuant to Item 601(b)(2) of Regulation S-K. The Company undertakes to furnish supplemental copies of any of the omitted schedules upon request by the Securities and Exchange Commission. In addition, confidential treatment has been requested as to certain portions of this exhibit, which portions have been omitted and filed separately with the Securities and Exchange Commission.

**Forward-Looking Statements**

*This Current Report on Form 8-K contains forward-looking statements, including, but not limited to, statements related to the potential acceptance for filing of and potential approval of an NDA for defibrotide, and other statements that are not historical facts. These forward-looking statements are based on the Company's current expectations and inherently involve significant risks and uncertainties. Actual results and the timing of events could differ materially from those anticipated in such forward-looking statements as a result of these risks and uncertainties, which include, without limitation, risks and uncertainties associated with the Company's ability to successfully manage the risks associated with integrating defibrotide into the Company's product portfolio; the possibility that the Company may fail to realize the anticipated benefits (commercial or otherwise) from the acquisition of rights to defibrotide in the Americas from Sigma-Tau; the inherent uncertainty associated with the regulatory approval process, including the risks that the Company may be required to conduct additional time-consuming and costly clinical trials in order to obtain any regulatory approval of defibrotide in the United States and that the Company may otherwise be unable to obtain or maintain any regulatory approvals for defibrotide in the United States; the difficulty and uncertainty of pharmaceutical product development, including the timing and cost thereof; and those other risks detailed from time-to-time under the caption "Risk Factors" and elsewhere in the Company's Securities and Exchange Commission filings and reports (Commission File No. 001-33500), including the Quarterly Report on Form 10-Q for the quarter ended March 31, 2014 and future filings and reports by the Company, including the Quarterly Report on Form 10-Q for the quarter ended June 30, 2014. The Company undertakes no duty or obligation to update any forward-looking statements contained in this Current Report on Form 8-K as a result of new information, future events or changes in its expectations.*

**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.

JAZZ PHARMACEUTICALS PUBLIC LIMITED  
COMPANY

By: /s/ Suzanne Sawochka Hooper  
Name: Suzanne Sawochka Hooper  
Title: Executive Vice President and General Counsel

Date: August 5, 2014

## EXHIBIT INDEX

**Exhibit  
Number**

**Description**

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\* Schedules have been omitted pursuant to Item 601(b)(2) of Regulation S-K. The Company undertakes to furnish supplemental copies of any of the omitted schedules upon request by the Securities and Exchange Commission. In addition, confidential treatment has been requested as to certain portions of this exhibit, which portions have been omitted and filed separately with the Securities and Exchange Commission.

[\*] = Certain confidential information contained in this document, marked by brackets, has been omitted and filed separately with the Securities and Exchange Commission pursuant to Rule 24b-2 of the Securities Exchange Act of 1934, as amended.

*Execution Version*

ASSIGNMENT AGREEMENT

by and among

JAZZ PHARMACEUTICALS INTERNATIONAL II LIMITED,

SIGMA-TAU PHARMACEUTICALS, INC.,

JAZZ PHARMACEUTICALS PLC

and

GENTIUM S.P.A.

Dated July 1, 2014

## ASSIGNMENT AGREEMENT

This Assignment Agreement (this “Agreement”) is entered into on July 1, 2014 (the “Execution Date”), by and among (i) Jazz Pharmaceuticals International II Limited, a Bermuda company limited by shares (“Buyer”), (ii) Sigma-Tau Pharmaceuticals, Inc., a Nevada corporation (the “Company”), (iii) solely for the purposes of Sections 2.7(b), 6.10, 7.3, 8.1, 8.4, 8.7, 8.8, 8.10, 8.11 and 8.12, Jazz Pharmaceuticals Public Limited Company, an Irish company (“Parent”), and (iv) solely for the purposes of Sections 6.10, 7.3, 8.2, 8.4, 8.8, 8.10, 8.11 and 8.12, Gentium S.p.A. (“Gentium”), an Italian corporation. Buyer and the Company are referred to collectively herein as the “Parties” and individually as a “Party.”

## PRELIMINARY STATEMENTS

Whereas, the Company has certain rights to defibrotide under and in connection with the License Agreement (as defined below); and

Whereas, subject to the terms and conditions set forth herein, the Company desires to sell, convey, transfer, assign and deliver to Buyer, and Buyer desires to purchase and acquire from the Company, the Company’s right, title and interest in and to the License Agreement, to assume the Company’s future obligations and liabilities under the License Agreement, and to purchase and acquire the Company’s title and interest to the Compound-Related Assets (as defined below), all as per the terms and conditions set forth herein.

Now, therefore, in consideration of the premises and the mutual promises herein made, and in consideration of the representations, warranties, and covenants herein contained, the Parties agree as follows.

ARTICLE 1  
DEFINITIONS

Unless the context otherwise requires, the terms defined in this Article 1 shall have the meanings herein specified for all purposes of this Agreement, applicable to both the singular and plural forms of any of the terms defined herein. When a reference is made in this Agreement to Sections, such reference shall be to a Section of this Agreement unless otherwise indicated. Whenever the words “include,” “includes” or “including” are used in this Agreement, they shall be deemed to be followed by the words “without limitation.”

“Affiliate” means, with respect to any Person, (a) any other Person that controls, is controlled by, or is under common control with such Person (including through multiple levels of subsidiaries), or (b) any officer or director of such Person. For purposes of this definition, the term “control” of a Person shall mean the possession, directly or indirectly, of the power to direct or cause the direction of the management or policies, whether through the ownership of voting securities, by contract or otherwise. For the mere sake of clarity, Sigma Tau Industrie

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Farmaceutiche Riunite S.p.A. is, and shall be considered, an Affiliate of the Company and Parent is, and shall be considered, an Affiliate of Buyer.

“Agreement” has the meaning set forth in the preamble hereto.

“Allocation” has the meaning set forth in Section 5.5(a).

“Approval” means any approval, registration, license or authorization from the FDA or any other Governmental Body required for the research, development, manufacture, storage, transport, distribution, marketing, use, offer for sale, or sale of the Compound or Product.

“Assumed Liabilities” has the meaning set forth in Section 2.3.

“Business Day” means any day that is not a Saturday, Sunday or any other day on which banks are required or authorized by Law to be closed in the State of New York.

“Buyer” has the meaning set forth in the preamble hereto.

“Buyer Indemnitees” has the meaning set forth in Section 6.1.

“Claim Notice” has the meaning set forth in Section 6.6.

“Closing” has the meaning set forth in Section 2.6(a).

“Closing Date” has the meaning set forth in Section 2.6(a).

“Code” means the Internal Revenue Code of 1986, as amended, and any applicable rules and regulations thereunder, and any successor to such statute, rules or regulations.

“Company” has the meaning set forth in the preamble hereto.

“Company Indemnitees” has the meaning set forth in Section 6.2.

“Compound” means the compound whose International Non-proprietary Name (INN) is defibrotide, including [ \* ].

“Compound-Related Assets” has the meaning set forth in Section 2.2.

“Confidential Information” means (a) the ST Know-How, and (b) all other information or material possessed or developed by or on behalf of, or disclosed to, Gentium or the Company or their respective Affiliates, pursuant to the License Agreement or in relation to the Compound or the Product or the development, promotion, marketing, distributing, or selling thereof, including, without limitation, information or materials on substances, formulations, techniques, technology, equipment, data, reports, Know-How, sources for supply, patent positions and business plans; provided that the “Confidential Information” shall not include information or material that is (i) already in the public domain as of the Execution Date by reason of prior publication or

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otherwise; or (ii) part of public domain after the Execution Date through no act, omission or fault of the Company or its Affiliates.

“Consent” means, with respect to any Person, any consent, approval, authorization, permission or waiver of, or registration, declaration or other action or filing with or exemption by such Person.

“Contract” means any contract, obligation, commitment, lease, license, purchase order, bid or other agreement with outstanding rights or obligations on the part of any party thereto, together with all amendments thereto.

“Cost Sharing Agreement” means the amendment, dated October 12, 2007, to the License and Supply Agreement between Gentium and the Company, as assignee of Sigma-Tau Industrie Farmaceutiche Riunite S.p.A., dated December 7, 2001, as amended.

“Disputing Party” has the meaning set forth in Section 8.12(b).

“DOJ” has the meaning set forth in Section 7.1.

“Effective Date” has the meaning set forth in Section 2.6(a).

“Execution Date” has the meaning set forth in the preamble hereto.

“FDA” means the United States Food and Drug Administration or any successor agency thereto.

“FTC” has the meaning set forth in Section 7.1.

“Fundamental Representations” has the meaning set forth in Section 6.8.

“Gentium” has the meaning set forth in the preamble hereto.

“Gentium Consent” means the consent of Gentium for the assignment of the License Agreement and the Compound-Related Assets to Buyer hereunder, in the form attached hereto as Exhibit A.

“Governmental Body” means any foreign or domestic federal, state, municipal or local government or quasi-governmental body (including any self-policing, self-regulatory or self-reporting industry groups and including any social security authority) or any department, agency, subdivision, committee, court or other tribunal of any of the foregoing.

“HSR Act” means the Hart-Scott-Rodino Antitrust Improvements Act of 1976, as amended, and the rules and regulations promulgated thereunder.

“HSR Clearance” means all applicable waiting periods under the HSR Act with respect to the transactions contemplated under this Agreement have expired or have been terminated.

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“HSR Clearance Date” means the earliest date on which the Parties have actual knowledge that all applicable waiting periods under the HSR Act with respect to the transactions contemplated under this Agreement have expired or have been terminated.

“HSR Filing” means filings by the Parties with the United States Federal Trade Commission and the Antitrust Division of the United States Department of Justice of a Notification and Report Form for Certain Mergers and Acquisitions (as that term is defined in the HSR Act) with respect to the matters set forth in this Agreement, together with all required documentary attachments thereto.

“Indemnified Party” has the meaning set forth in Section 6.5(a).

“Indemnifying Party” has the meaning set forth in Section 6.5(a).

“Indication” has the meaning set forth in Section 2.7(a).

“JAMS Rules” has the meaning set forth in Section 8.12(c).

“Know-How” means any and all information, know-how, data, results, knowledge, techniques, discoveries, inventions, specifications, designs, regulatory filings, trade secrets, technology, methods, processes, discoveries, inventions, invention disclosures, developments, specifications, protocols, formulations, formulae, software, algorithms, marketing reports, expertise, technology, test data (including pharmacological, biological and chemical, biochemical, preclinical test data, clinical test data and data resulting from non-clinical studies), manufacturing processes, information, records, stability data and other study data and procedures, business or financial information or information of any type whatsoever, in any tangible or intangible form.

“Law” means any foreign or domestic federal, state, municipal or local law, statute, code, ordinance, regulation, Order, rule, consent agreement, constitution, treaty or other requirement of any Governmental Body.

“Liability” means any liability, obligation, debt, charge, cost, expense, loss of any nature, penalty, or commitment of any kind or nature, whether known or unknown, asserted or unasserted, absolute or contingent, accrued or unaccrued, disclosed or undisclosed, liquidated or unliquidated, or due or to become due.

“License Agreement” means the License and Supply Agreement between Gentium and the Company, as assignee of Sigma-Tau Industrie Farmaceutiche Riunite S.p.A., dated December 7, 2001, as amended, including the Cost Sharing Agreement and all amendments thereto.

“Loss” means any and all losses, Liabilities, injury, fines, costs (including cost of investigation), claim, demand, settlement, judgment, award, damages, diminution in value, Tax, fee, penalties, charge or expense (including related Taxes) of any nature (including reasonable

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attorneys' fees and expenses and litigation, settlement and judgment and interest costs and any reasonable legal or other expenses incurred in connection with investigating or defending any claims or actions).

“Milestone Event” has the meaning set forth in Section 2.7(a).

“Milestone Payment” has the meaning set forth in Section 2.7(a).

“NDA” means a New Drug Application, as defined in the United States Federal Food, Drug, and Cosmetic Act, as amended, and applicable regulations promulgated thereunder by the FDA.

“Order” means any order, award, decision, injunction, judgment, ruling, decree, charge, writ, subpoena or verdict entered, issued, made or rendered by any Governmental Body or arbitrator.

“Parent” has the meaning set forth in the preamble hereto.

“Party” has the meaning set forth in the preamble hereto.

“Permit” means any approval, license, franchise, Consent, exemption, permit, certificate, certificate of occupancy or Order issued by any Person.

“Person” means any individual, partnership, corporation, limited liability company, association, joint stock company, trust, joint venture, unincorporated organization, other business entity, or Governmental Body.

“Proceeding” means any claim, demand, dispute, action, audit, lawsuit, litigation, investigation or arbitration (in each case, whether civil, criminal or administrative) pending by or before any Governmental Body or arbitrator.

“Product” means any pharmaceutical product in intravenous formulation containing the Compound as an active ingredient.

“Representative” means, with respect to a particular Person, any employee, manager, officer, director, agent, consultant, advisor or other representative of such Person, including legal counsel, accountants and financial advisors.

“Retained Liabilities” has the meaning set forth in Section 2.4.

“Special Representations” has the meaning set forth in Section 6.9.

“ST Fin” has the meaning set forth in Section 8.7.

“ST Know-How” means any and all Know-How [ \* ] and any additional Know-How [ \* ], including, if any, information concerning: [ \* ].

[\*] = Certain confidential information contained in this document, marked by brackets, has been omitted and filed separately with the Securities and Exchange Commission pursuant to Rule 24b-2 of the Securities Exchange Act of 1934, as amended.

“Tax” or “Taxes” means any federal, state, local, or foreign income, gross receipts, license, payroll, employment, excise, severance, stamp, occupation, premium, windfall profits, environmental (including taxes under Code §59A), customs duties, capital stock, franchise, profits, withholding, social security (or similar), unemployment, disability, real property, personal property, sales, use, transfer, registration, value added, escheat, alternative or add-on minimum, estimated, or other tax of any kind whatsoever, including any interest, penalty, or addition thereto, whether disputed or not.

“Tax Deduction” means a deduction or withholding for or on account of Tax from a payment under or in connection with this Agreement.

“Tax Return” means any return, declaration, report, claim for refund, or information return or statement relating to Taxes, including any schedule or attachment thereto, and including any amendment thereof.

“Third Party” means any Person other than the Company, Buyer, any Affiliate of the Company and any Affiliate of Buyer.

“Third-Party Claim” has the meaning set forth in Section 6.5(a).

“Transfer Taxes” has the meaning set forth in Section 8.3.

“Unpaid Pre-Effective Date Costs” has the meaning set forth in the Gentium Consent.

“Up-Front Amount” has the meaning set forth in Section 2.6(c).

## ARTICLE 2 ASSIGNMENT; CLOSING

2.1 License Agreement. Effective upon the Effective Date, the Company sells, transfers and assigns to Buyer, which acquires, purchases and succeeds in, all of the Company’s right, title and interest in and to the License Agreement as well as those Liabilities of Company and its Affiliates under the License Agreement as and to the extent specified in Section 2.3.

2.2 Compound-Related Assets. Effective upon the Effective Date, the Company sells, conveys, transfers, assigns and delivers to Buyer, which acquires, purchases and succeeds in, all of the Company’s and its Affiliates’ right, title and interest in and to (a) [ \* ], (b) all [ \* ], if any, and (c) all [ \* ], if any ((a)-(c) collectively, the “Compound-Related Assets”).

2.3 Assumed Liabilities. Subject to the terms and conditions hereof, as of the Effective Date, Buyer assumes and shall satisfy, perform, pay, discharge and be liable for the following Liabilities (collectively, the “Assumed Liabilities”):

(a) all Liabilities to Third Parties and Affiliates of Buyer to the extent arising out of or relating to the prosecution, ownership, operation, maintenance, sale, lease or use of the Compound-Related Assets on or after the Effective Date; and

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(b) all Liabilities of the Company or its Affiliates under or in connection with the License Agreement to the extent arising on or after the Effective Date and to the extent that such Liabilities are not Retained Liabilities.

For clarity, Buyer is not assuming any Liability of the Company or its Affiliates other than as specifically identified in this Agreement.

2.4 Retained Liabilities. Without prejudice to any other provision of this Agreement, Assumed Liabilities shall not include, Buyer shall not be required to assume and shall have no obligation to perform or discharge, and the Company shall retain the following Liabilities (the “Retained Liabilities”):

(a) all Liabilities to Third Parties and Affiliates of the Company to the extent arising out of or relating to the prosecution, ownership, operation, maintenance, sale, lease or use of the Compound-Related Assets before the Effective Date; and

(b) the Liabilities specifically retained by Company pursuant to Sections 4(b) and 4(c) of the Gentium Consent.

The Parties acknowledge that, due to Buyer’s assumption of Liability pursuant to Section 2.3(a) and the releases set forth in Section 2.5(a) herein and Section 2 and Section 3 of the Gentium Consent, the Company and its Affiliates do not have any Liability and/or obligation under or in connection with the License Agreement or the Compound-Related Assets other than the Retained Liabilities and the Liabilities and obligations expressly assumed by the Company under this Agreement, including pursuant to Article 4 and Section 5.4.

#### 2.5 Release.

(a) Effective upon the Effective Date, Buyer (also on behalf of any of its Affiliates) hereby fully and unconditionally releases and forever discharges the Company, ST Fin and/or any of their respective Affiliates from any and all claims, causes of action, charges, complaints, demands, actions, and Liabilities, known or unknown, suspected or unsuspected, legal or equitable, under or in connection with the Compound-Related Assets and/or the License Agreement (including in connection with its validity and enforceability, other than due to actions or inactions of the Company or its Affiliates, and/or fulfillment of the obligations thereunder by Gentium and/or Gentium’s assignees) with the sole exception of (i) the Retained Liabilities and (ii) the Liabilities and obligations expressly assumed by the Company under this Agreement, including pursuant to Article 4 and Section 5.4.

(b) Effective upon the Effective Date, the Company (also on behalf of any of its Affiliates) hereby fully and unconditionally releases and forever discharges Buyer, Parent and/or any of their respective Affiliates from any and all claims, causes of action, charges, complaints, demands, actions, and Liabilities, known or unknown, suspected or unsuspected, legal or equitable, under or in connection with the Compound-Related Assets and/or the License Agreement (including in connection with its validity and enforceability (other than due to actions

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or inactions of Buyer or its Affiliates)) with the sole exception of (i) the Assumed Liabilities, (ii) the Liabilities and obligations specifically retained by Gentium under the Gentium Consent, and (iii) the Liabilities and the obligations expressly assumed by Buyer, Gentium, the Parent and their Affiliates under this Agreement, including pursuant to Article 3 and Section 5.4.

## 2.6 Closing.

(a) Closing. The closing of the transactions contemplated hereby (the “Closing”) will take place at the offices of Cooley LLP, 3175 Hanover Street, Palo Alto, CA 94304, on the third (3<sup>rd</sup>) Business Day following the satisfaction or waiver of all conditions to the obligations of the Parties to effect the Closing or on such other date as Buyer and the Company may mutually agree in writing (the “Closing Date”); provided that a Party has not earlier terminated this Agreement pursuant to Section 7.4 prior to such date. The Closing shall take place through exchange of the deliverables as provided under Section 2.6(b) and (c) below. The Closing Date on which both the actions under Section 2.6(b) and (c) are fulfilled is referred to as the “Effective Date.”

(b) Closing Deliverables by the Company. On the Closing Date, Company shall deliver or cause to be delivered to Buyer all tangible embodiments of the Compound-Related Assets, available to the Company or its Affiliates.

(c) Closing Deliverables by Buyer. On the Closing, Buyer shall deliver or cause to be delivered to Company an amount equal to seventy-five million U.S. Dollars (\$75,000,000) (the “Up-Front Amount”) less the amount of the Unpaid Pre-Effective Date Costs by wire transfer of immediately available funds to an account (or accounts) designated in writing by the Company.

(d) Conditions to Closing Obligations of the Company. The obligation of the Company to effect the Closing is subject to the satisfaction, or written waiver by the Company, of each of the following conditions:

(i) Each of the representations and warranties of Buyer contained in this Agreement shall be true and correct in all material respects, both individually and when considered collectively with other representations and warranties of Buyer contained in this Agreement, as of the Effective Date as if made on the Effective Date, except in each case where the failure of such representations and warranties to be accurate, individually or in the aggregate, does not and would not be reasonably expected to have a material adverse effect on the ability of Buyer or Company to consummate the transactions contemplated by this Agreement;

(ii) Each of the representations and warranties of Buyer set forth in Sections 3.1 and 3.2 of this Agreement shall be true and correct in all respects;

[\*] = Certain confidential information contained in this document, marked by brackets, has been omitted and filed separately with the Securities and Exchange Commission pursuant to Rule 24b-2 of the Securities Exchange Act of 1934, as amended.

(iii) A certificate executed by a director of Buyer no earlier than the third (3<sup>rd</sup>) Business Day following the HSR Clearance Date, confirming satisfaction of the conditions set forth in Section 2.6(d)(i) and (ii); and

(iv) HSR Clearance.

(e) Conditions to Closing Obligations of Buyer. The obligation of Buyer to effect the Closing is subject to the satisfaction, or written waiver by Buyer, of each of the following conditions:

(i) Each of the representations and warranties of the Company contained in this Agreement shall be true and correct in all material respects, both individually and when considered collectively with other representations and warranties of the Company contained in this Agreement, as of the Effective Date as if made on the Effective Date, except in each case where the failure of such representations and warranties to be accurate, individually or in the aggregate, does not and would not be reasonably expected to materially adversely affect the value to Buyer or Company of the transactions contemplated by this Agreement;

(ii) Each of the representations and warranties of the Company set forth in Sections 4.1 and 4.2 of this Agreement shall be true and correct in all respects;

(iii) A certificate executed by a director of the Company no earlier than the third (3<sup>rd</sup>) Business Day following the HSR Clearance Date, confirming satisfaction of the conditions set forth in Section 2.6(e)(i) and (ii); and

(iv) HSR Clearance.

**2.7 Contingent Payments.**

(a) Milestone Payments. Within [ \* ] days after the first occurrence of each of the milestone events set forth in the chart below (each, a “Milestone Event”, and collectively, the “Milestone Events”), Buyer shall pay to the Company the milestone payment set forth opposite such Milestone Event in the chart below (each, a “Milestone Payment”, and collectively, the “Milestone Payments”):

<u>Milestone Event</u>	<u>Milestone Payment</u>
1. First acceptance for filing by the FDA of the first NDA for the Product for [ * ] hematopoietic stem cell transplant patients with veno-occlusive disease, [ * ] ([ * ] the “ <u>Indication</u> ”), wherein such first acceptance occurs [ * ]	Twenty-five million U.S. Dollars (\$25,000,000)
2. First approval by the FDA of the first NDA for the	

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Milestone Event	Milestone Payment
Product for any Indication, wherein such first approval is received:	
A. [ * ]	[ * ] U.S. Dollars (\$[ * ])
B. [ * ]	[ * ] U.S. Dollars (\$[ * ])
C. [ * ]	[ * ] U.S. Dollars (\$[ * ])
D. [ * ]	[ * ] U.S. Dollars (\$[ * ])
E. [ * ]	[ * ] U.S. Dollars (\$[ * ])
F. [ * ]	[ * ] U.S. Dollars (\$[ * ])
G. [ * ]	[ * ] U.S. Dollars (\$[ * ])
H. [ * ]	[ * ] U.S. Dollars (\$[ * ])
I. [ * ]	[ * ] U.S. Dollars (\$[ * ])

Each Milestone Payment is payable one time only, regardless of the number of times the corresponding Milestone Event is achieved. No Milestone Payment shall be owed for Milestone Event 1 if the date of first acceptance for filing by the FDA of the first NDA for the Product for the Indication is [ \* ]. Only one Milestone Payment shall be owed for achievement of Milestone Event 2, and such Milestone Payment shall be the amount that corresponds with the date of receipt of the first approval by the FDA of the first NDA for the Product for any Indication, and no Milestone Payment shall be owed for Milestone Event 2 if the date of receipt of the first approval by the FDA of the first NDA for the Product for any Indication is [ \* ]. Under no circumstances shall Buyer be obligated to pay the Company as Milestone Payments more than one hundred and seventy-five million U.S. Dollars (\$175,000,000) pursuant to this Section 2.7(a).

(b) Withholding Taxes.

(i) The Buyer and the Parent shall make all payments to be made by it under this Agreement without any Tax Deduction, unless a Tax Deduction is required by Law. The Buyer shall promptly upon becoming aware that same must make a Tax Deduction (or that there is a change in the rate or the basis of a Tax Deduction) notify the Company accordingly. If Buyer or the Parent is required to make a Tax Deduction, Buyer or the Parent, as the case may be, shall make that Tax Deduction and any payment required in connection with that Tax Deduction within the time allowed and in the

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minimum amount required by Law. If a Tax Deduction is required by Law to be made by Buyer or the Parent and such Tax Deduction results from (1) any action taken by Buyer or Parent, including a transfer or assignment of Buyer's or Parent's payment obligations under this Agreement, or a change in the jurisdiction of Tax residence of Buyer or the Parent or (2) Tax laws of Buyer or Parent's jurisdiction of Tax residence (including any change in Tax Law), then the amount of the payment due from Buyer or the Parent to the Company shall be increased to an amount which (after making any Tax Deduction, including any and all Tax Deductions imposed on additional payments) leaves an amount equal to the payment which would have been due to the Company if no such Tax Deductions had been required. Within [ \* ]days of making either a Tax Deduction or any payment required in connection with that Tax Deduction, Buyer or the Parent, as the case may be, shall deliver to the Company evidence reasonably satisfactory to the Company that the Tax Deduction has been made or (as applicable) any appropriate payment paid to the relevant taxing authority. Only in cases other than those set forth under (1) and (2) above, the amount withheld by Buyer or Parent and remitted to the appropriate Governmental Body as a Tax Deduction pursuant to the above provisions shall be treated for all purposes of this License Agreement as having been delivered and paid to the Company.

(ii) The Company will provide Buyer with an IRS Form W-9 on or prior to the Effective Date, and, after the Effective Date will, or will cause its successors and assignees to, provide any Tax forms that can be legally provided and that establish an exemption from, or reduction to, otherwise applicable withholding Taxes, if any. The Company represents that, as of the Execution Date, it is the beneficial owner for United States federal income tax purposes of the Up-Front Amount referenced under Section 2.6(c) and any Milestone Payments. The Buyer and the Parent represent that, as of the Execution Date, no Tax Deduction is required to be made in connection with the payment of the Up-Front Amount referenced under Section 2.6(c) or in connection with any Milestone Payment.

### ARTICLE 3 REPRESENTATIONS AND WARRANTIES OF BUYER

Buyer represents and warrants to the Company that the following statements are correct and true as of the Execution Date and the Effective Date:

3.1 Organization of Buyer. Buyer is a Bermuda company limited by shares duly formed, validly existing, and in good standing under the Laws of its jurisdiction of incorporation.

3.2 Authorization of Transaction. Buyer has full power and authority to execute and deliver this Agreement and to perform its obligations hereunder. The execution and delivery by Buyer of this Agreement and the performance by Buyer of the transactions contemplated hereby have been duly approved by all requisite corporate or other applicable action of Buyer. Assuming the due authorization, execution and delivery of this Agreement by the Company, this Agreement constitutes the valid and legally binding obligation of Buyer, enforceable against it in

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accordance with the terms of this Agreement, except as such enforceability may be limited by bankruptcy, insolvency, reorganization, moratorium, mandatory provisions of any applicable Law and other laws affecting creditors generally and by the availability of equitable remedies.

3.3 Non-contravention. Assuming the due authorization, execution and delivery of this Agreement by the Company, neither the execution or the delivery of this Agreement, nor the consummation of the transactions contemplated hereby and thereby, will (a) violate or conflict with any Law or Order to which Buyer is subject or (b) violate or conflict with any provision of the organizational documents of Buyer. Except for the HSR Filings and the HSR Clearance, Buyer is not required to give any notice to, make any filing with, or obtain any Consent or Permit of any Governmental Body or other Person in order to consummate the transactions contemplated by this Agreement.

3.4 Brokers' Fees. Buyer is not party to any Contract as a result of which a broker, finder or agent could hold any payment obligation against the Company with respect to the transactions contemplated by this Agreement.

3.5 Litigation. The Buyer is not party to any Proceedings pending or, to Buyer's knowledge, threatened in writing by a Third Party challenging, or that would have the effect of preventing, delaying, making illegal or otherwise interfering with, the transactions contemplated by this Agreement.

3.6 License Agreement.

(a) Assuming the validity of the Gentium Consent and the acknowledgment and agreement of Gentium under Section 8.2, [ \* ] Buyer has the right to be assigned the Company's right, title, interest and obligations in the License Agreement pursuant to Section 2.1 of this Agreement; provided, however, for the avoidance of doubt, Buyer is [ \* ].

(b) Neither Buyer nor its Affiliates have any actual knowledge on or prior to the Effective Date that the License Agreement is not assignable by Company as a matter of interpretation of applicable Law. Neither Buyer nor its Affiliates have any actual knowledge on or prior to the Effective Date that Buyer does not have the right to be assigned the Company's right, title, interest and obligations in the ST Know-How pursuant to Section 2.2 of this Agreement as a matter of interpretation of applicable Law.

ARTICLE 4  
REPRESENTATIONS AND WARRANTIES OF THE COMPANY

The Company represents and warrants to Buyer that the following statements are correct and true as of the Execution Date and the Effective Date:

4.1 Organization. The Company is duly organized, validly existing, and in good standing under the Laws of the State of Nevada.

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4.2 Authority; Binding Nature of Agreements. The Company has full power and authority to execute and deliver this Agreement and to perform its obligations hereunder. The execution and delivery by the Company of this Agreement and the performance by the Company of the transactions contemplated hereby have been duly approved by all requisite corporate or other applicable action of the Company. Assuming the due authorization, execution and delivery of this Agreement by Buyer and the validity and effectiveness of the Gentium Consent, this Agreement constitutes the valid and legally binding obligation of the Company, enforceable against it in accordance with the terms of this Agreement, except as such enforceability may be limited by bankruptcy, insolvency, reorganization, moratorium, mandatory provisions of any applicable Law and other laws affecting creditors generally and by the availability of equitable remedies.

4.3 Non-contravention. Assuming the due authorization, execution and delivery of this Agreement by Buyer and the validity of the Gentium Consent, neither the execution or delivery of this Agreement, nor the consummation of the transactions contemplated hereby or thereby, will (a) violate or conflict with any Law or Order to which the Company is subject or (b) violate or conflict with any provision of the organizational documents of the Company. Except for the HSR Filings, the HSR Clearance, and the Gentium Consent, the Company is not required to give any notice to, make any filing with, or obtain any Consent or Permit of any Governmental Body or other Person in order to consummate the transactions contemplated by this Agreement.

4.4 Brokers' Fees. The Company is not party to any Contract as a result of which a broker, finder or agent could hold any payment obligation against Buyer with respect to the transactions contemplated by this Agreement.

4.5 Compound-Related Assets. The Company is not aware of any [ \* ] related to the Compound or Product [ \* ] which have been issued, granted, applied for, and/or made in favor of, or by or on behalf of, the Company or its Affiliates other than those that will be transferred to Buyer at Closing pursuant to Section 5.2(a). Assuming the validity of the Gentium Consent, neither the execution, delivery or performance of this Agreement nor the consummation of the transactions contemplated hereby will (a) cause the grant, assignment or transfer to any Person (other than Buyer) of any license or other right or interest under, to or in any of the Compound-Related Assets, if any, under any Contract to which the Company or its Affiliates is a party, or (b) conflict with, result in a breach of, constitute a default under, or otherwise result in any material change under any Contract to which the Company or its Affiliates is a party, which would have an adverse effect on the Compound-Related Assets or the License Agreement or the Parties' rights thereto or thereunder. Except for this Agreement and the License Agreement, the Company is not party to any Contract granting any Person any right to the Compound-Related Assets, if any.

#### 4.6 License Agreement.

(a) The Company is party to the License Agreement and, other than the assignment contemplated by Section 2.1 of this Agreement, has not assigned, delegated,

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transferred, licensed, sublicensed, granted any options to, or pledged to any Person, any of its right, title or interest or obligations under the License Agreement.

(b) Assuming the validity of the Gentium Consent and the acknowledgment and agreement of Gentium under Section 8.2, [ \* ] the Company has the right to assign its right, title, interest and obligations in the License Agreement to Buyer pursuant to Section 2.1 of this Agreement; provided, however, for the avoidance of doubt, Company is [ \* ].

(c) Neither Company nor its Affiliates have any actual knowledge on or prior to the Effective Date that the License Agreement is not assignable by Company as a matter of interpretation of applicable Law.

4.7 ST Know-How. The Company has not created (or failed to take actions to avoid the creation of) any encumbrance, pledge or security interest on the ST Know-How; the Company is not subject to any Order relating to the ST Know-How nor is the Company a party to any contractual obligations to Third Parties with respect to ST Know-How. Assuming the validity of the Gentium Consent, to the knowledge of the Company, the Company has the right to assign its and its Affiliates' right, title and interest in the ST Know-How to Buyer pursuant to Section 2.2 of this Agreement; provided, that for the avoidance of doubt, [ \* ]. Neither Company nor its Affiliates have any actual knowledge on or prior to the Effective Date that the Company does not have the right to assign its right, title, interest and obligations in the ST Know-How to Buyer pursuant to Section 2.2 of this Agreement as a matter of interpretation of applicable Law. Except for the license granted to Gentium pursuant to Section 2.5 of the License Agreement and the assignment contemplated by Section 2.2 of this Agreement, the Company has not granted to any Person any license of, option to obtain a license of, or other right with respect to, any ST Know-How. There are no pending or, to the Company's knowledge, Proceedings or Orders threatened in writing to the Company in any jurisdiction or territory between the Company and any Third Party relating to the ST Know-How. To the Company's knowledge, neither the Company nor any of its Affiliates owns or holds any invention, patent and/or patent application relating to the Compound or Product (any such invention, patent or patent application, a "Compound-Related Patent").

4.8 Contracts. Except for the License Agreement, this Agreement and the Contracts listed in Schedule 4.8, the Company and its Affiliates are not parties to any Contract related to the Compound or Product (including the research, development, manufacture, storage, transport, distribution, marketing, use, offer for sale, or sale thereof and further including any license, option to obtain license, or right to use, any patent, Know-How, trademark, copyright or application therefor of any Person that is related to the Compound or Product).

4.9 Litigation. The Company is not and has not been during the preceding [ \* ] party to any Proceedings or Orders pending or, to the Company's knowledge, threatened in writing by a Third Party (a) relating to or affecting the License Agreement or (b) challenging, or that would have the effect of preventing, delaying, making illegal or otherwise interfering with, the transactions contemplated hereby. To the actual knowledge of the Company, no claim or dispute has been brought in writing against the Company or its Affiliates and neither the Company nor

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its Affiliates has received any written notification from a Third Party relating to a potential claim or dispute that might reasonably give rise to the commencement of any Proceeding or Order relating to the License Agreement or the transactions contemplated hereby.

ARTICLE 5  
LIMITATION OF REPRESENTATIONS AND LIABILITIES; ADDITIONAL COVENANTS;

5.1 Limitations on Representation and Liabilities. Each Party acknowledges and agrees that (a) neither the other Party nor any of its Affiliates nor any of their shareholders, members, partners, trustees, beneficiaries, directors, officers, managers, employees, attorneys, accountants, contractors, consultants, agents or representatives, nor any Person purporting to represent any of the foregoing, makes or has made any other express or any implied representation or warranty with respect to the License Agreement, the Compound-Related Assets and any other matter contemplated herein other than those expressly set forth in this Agreement and (b) all guarantees, representations and warranties which would arise under applicable Law are herein excluded and derogated, to the fullest extent legally possible, except solely for those representations and warranties expressly set forth herein.

5.2 Compound-Related Assets, ST-Know How and Gentium Consent.

(a) Notwithstanding the first sentence of Section 4.5, if any Compound-Related Assets under (b) or (c) of the definition exist as of the Effective Date, such Compound-Related Assets shall be deemed assigned and transferred as of the Effective Date, where legally permitted, from the Company to Buyer pursuant to the provisions herein and the Parties shall take all reasonable actions, at the sole cost and expense of the Company, to effect and formalize such assignment and transfer. Additionally, if any additional tangible embodiments of the Compound-Related Assets that existed as of the Effective Date that were not transferred to Buyer pursuant to Section 2.6(b) become available to the Company or its Affiliates, then Company shall promptly transfer such tangible embodiments of the Compound-Related Assets that existed as of the Effective Date to Buyer.

(b) Notwithstanding the last sentence of Section 4.7, if any Compound-Related Patent exists as of the Effective Date, such Compound-Related Patent shall be deemed a Compound-Related Asset and assigned and transferred as of the Effective Date, where legally permitted, from the Company to Buyer pursuant to the provisions herein and the Parties shall take all reasonable actions, at the sole cost and expense of the Company, to effect and formalize such assignment and transfer.

(c) The Parties acknowledge that most of the ST Know-How has been based, developed and/or elaborated on the basis of data, information, material, know-how, reports provided or supplied by Gentium to the Company under the License Agreement or in any case on the basis of data and information shared between Gentium and the Company thereunder.

(d) Company (also on behalf of any of its Affiliates) and Buyer (also on behalf of any of its Affiliates) each covenant not to challenge or contest the validity or

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enforceability of the Gentium Consent or the assignability of the License Agreement as a matter of interpretation of applicable Law.

5.3 Further Actions. The Parties hereby agree to take all further reasonable actions, at the sole cost and expense of the requesting Party, that are necessary to give effect to the transfer of the Compound-Related Assets existing as of the Effective Date, carry out the purposes of this Agreement or consummate the transactions contemplated hereby; provided, that nothing in this Section 5.3 limits the obligations of the Parties to take those actions otherwise expressly agreed to in this Agreement at their own cost and expense unless otherwise expressly noted.

5.4 Confidentiality. Upon Buyer's request after the Effective Date, the Company shall destroy or transfer to Buyer all electronic or tangible copies of the Confidential Information in the possession or control of the Company or any of its Affiliates (except for one copy which may be kept in the Company's restricted file for record-keeping purposes only and except as otherwise required by applicable Law or Order). The Company agrees not to, and to cause its Affiliates not to, disclose or use any Confidential Information except as may be required in order to perform their respective obligations under this Agreement or, after compliance with the obligations set forth in this Section 5.4, as may be required by applicable Law or Order. In the event that the Company is requested or required pursuant to written or oral question or request for information or documents in any legal proceeding, interrogatory, subpoena, civil investigation demand, or similar process to disclose any Confidential Information, the Company will, where legally permitted, notify Buyer promptly of the request or requirement so that Buyer may seek an appropriate protective order or waive compliance with the provisions of this Section 5.4. If, in the absence of a protective order or the receipt of a waiver hereunder, the Company is, on the advice of counsel, compelled to disclose any Confidential Information, the Company may disclose such Confidential Information; provided, however, that the Company shall use its reasonable efforts to obtain, at the request of Buyer, an order or other assurance that confidential treatment will be accorded to such portion of the Confidential Information required to be disclosed as Buyer shall designate. The foregoing provisions shall not apply to any Confidential Information that is generally available to the public immediately prior to the time of the Company's disclosure unless such Confidential Information is so available due to the unauthorized actions of the Company.

#### 5.5 Taxes.

(a) Allocation. Promptly after the Effective Date, the Parties shall mutually agree upon an allocation among the License Agreement and each of the several Compound-Related Assets (the "Allocation") for the payments received by the Company pursuant to this Agreement. Buyer and the Company shall report the transactions hereunder consistently with the Allocation for all federal, state, local and applicable foreign Tax purposes, and shall take no position inconsistent with the Allocation on any Tax Return or before any Tax authority unless required to do so pursuant to a final determination within the meaning of Section 1313 of the Code or any similar provision of applicable Law.

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(b) **Tax Liabilities.** Notwithstanding any provision of this Agreement or the Gentium Consent to the contrary, Buyer is not assuming, and Company will indemnify Buyer for, any Tax liabilities (i) of the Company or (ii) relating to the License Agreement or Compound-Related Assets through the Effective Date, and the Company is not assuming, and Buyer will indemnify the Company for, any Tax liabilities (1) of Buyer or (2) relating to the License Agreement or Compound-Related Assets after the Effective Date.

5.6 **Cooperation.** For a period of [ \* ] following the Effective Date the Company agrees to make its personnel set forth on Exhibit B available to Buyer for in-person meetings (which shall not be more than [ \* ]) or telephone conferences, on reasonable notice and during normal business hours, and to make available to Buyer all ST Know-How retained in the memory of such personnel that has not previously been transferred to Buyer pursuant to this Agreement; provided, however, that Buyer shall promptly reimburse the Company for any reasonable out-of-pocket expenses incurred by it (including travel expenses) in connection with providing assistance to Buyer pursuant to this Section 5.6.

## ARTICLE 6 INDEMNIFICATION

6.1 **Indemnification by the Company.** Subject to the terms and conditions of this Article 6, the Company shall indemnify and hold harmless Buyer, Parent and their Affiliates and their respective Representatives, successors and assigns (the "Buyer Indemnitees") from and against, and shall compensate and reimburse the Buyer Indemnitees for, all Losses that any Buyer Indemnitee may actually suffer, incur, or otherwise become subject to, directly or indirectly, resulting from, arising out of, in connection with, or caused by (a) any breach or inaccuracy of any representation or warranty made by the Company in this Agreement or in the certificate referred to under Section 2.6(e)(ii), (b) any breach of, or failure to perform, any covenant, agreement, commitment or obligation, on the part of the Company (on behalf of itself and its Affiliates) in this Agreement or under the Gentium Consent, (c) the Retained Liabilities, or (d) any Proceeding or any claim by any Person (including any Affiliate of the Company) relating to or arising from any breach or alleged breach, Liability or matter of the type referred to in clause (a), (b) or (c) of this sentence (including any Proceeding commenced for the purpose of enforcing any of its rights under this Article 6).

6.2 **Indemnification by Buyer.** Subject to the terms and conditions of this Article 6, Buyer shall indemnify and hold harmless the Company, ST Fin and their Affiliates and their respective Representatives, successors and assigns (the "Company Indemnitees") from and against, and shall compensate and reimburse the Company Indemnitees for, all Losses that any Company Indemnitee may actually suffer, incur, or otherwise become subject to, directly or indirectly, resulting from, arising out of, in connection with, or caused by (a) any breach or inaccuracy of any representation or warranty made by Buyer in this Agreement or in the certificate referred to under Section 2.6(d)(ii), (b) any breach of, or failure to perform, any covenant, agreement, commitment or obligation, on the part of Buyer, Parent or Gentium (on behalf of themselves and their respective Affiliates) in this Agreement or under the Gentium Consent, (c) the Assumed Liabilities as well as the Liabilities specifically

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retained by Gentium pursuant to Section 4(a) of the Gentium Consent; or (d) any Proceeding or any claim by any Person (including any Affiliate of Buyer) relating to or arising from any breach or alleged breach, Liability or matter of the type referred to in clause (a), (b), or (c) of this sentence (including any Proceeding commenced for the purpose of enforcing any of its rights under this Article 6).

6.3 Setoff. Following the Effective Date, Buyer shall have the right to withhold and deduct any sum [ \* ] payable by the Company to Buyer pursuant to the terms of this Agreement from any amount payable by Buyer to the Company under this Agreement. The withholding and deduction of any such sum shall operate for all purposes as a complete discharge (to the extent of such sum) of the obligation to pay the amount from which such sum was withheld and deducted.

6.4 Exclusivity of Remedies. After the Effective Date, except in the case of [ \* ] by the Indemnifying Party or its Affiliates, the [ \* ] shall be exclusive and, without limitation to the generality of the foregoing, after the Effective Date, [ \* ]. Prior to the Effective Date, this Agreement may be terminated only in accordance with Section 7.4. Notwithstanding the foregoing, nothing in this Article 6 shall be deemed to prohibit or limit any Party's right at any time after the Effective Date to seek injunctive or equitable relief for the failure of any Party to perform any covenant or agreement contained in this Agreement. Notwithstanding any other provision set forth herein, under no circumstance whatsoever shall either Party be entitled to rescind or terminate this Agreement after the Effective Date.

#### 6.5 Defense of Third Party Claims.

(a) If any Person (other than a Buyer Indemnitee or Company Indemnitee) initiates a Proceeding against any Buyer Indemnitee or Company Indemnitee, as applicable (the "Indemnified Party"), with respect to any matter that the Indemnified Party shall be entitled to make a claim for indemnification against the other Party (such Proceeding with respect to such matter, a "Third-Party Claim", and such Party, the "Indemnifying Party") under Section 6.1 or 6.2, as the case may be, then the Indemnified Party must promptly notify the Indemnifying Party in writing of the existence of such Third-Party Claim and must deliver copies of any documents served on the Indemnified Party with respect to the Third-Party Claim; provided, however, that any failure on the part of an Indemnified Party to so notify an Indemnifying Party shall not limit any of the obligations of the Indemnifying Party under this Article 6, except to the extent such failure materially prejudices the defense of such Proceeding.

(b) Upon receipt of the notice described in Section 6.5(a), and subject to Section 6.5(c), the Indemnifying Party will have the right, at its own costs and expense, to defend the Indemnified Party against the Third-Party Claim, provided that (i) the Indemnifying Party's counsel is reasonably satisfactory to the Indemnified Party, (ii) the Third-Party Claim involves only money damages and does not involve the seeking of an injunction or other equitable relief, (iii) the Indemnifying Party conducts the defense of the Third-Party Claim actively and diligently, (iv) the Indemnifying Party keeps the Indemnified Party apprised of all material developments, including settlement offers, with respect to the Third-Party Claim and permits the Indemnified Party to participate in the defense of the Third-Party Claim and (v) any settlement of

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the Third Party Claim shall be subject to the provisions set forth under Section 6.5(e). So long as the Indemnifying Party is conducting the defense of the Third-Party Claim in accordance with this Section 6.5(b), then the Indemnifying Party will not be responsible for any attorneys' fees or other expenses incurred by the Indemnified Party regarding the Third-Party Claim.

(c) Notwithstanding anything to the contrary under Section 6.5(b) or Section 6.5(d), Buyer shall have the right to control, [ \* ], the defense of any Third-Party Claim [ \* ], provided that (A) the conditions under (i), (iii) and (iv) of Section 6.5(b) shall apply *mutatis mutandis* as if Buyer were the Indemnifying Party; and (B) any settlement of the Third-Party Claim by Buyer shall be subject to the provisions of Section 6.5(e) or Section 6.5(f), as the case may be, depending on whether Buyer is the Indemnifying Party or the Indemnified Party. Notwithstanding the foregoing sentence, if Buyer and the applicable Company Indemnitee(s) are defendants in such Third-Party Claim and [ \* ], after consultation with outside counsel, that there is [ \* ] or if there can reasonably be expected to be a [ \* ], then the provisions set forth under [ \* ] and (x) the Company Indemnitee shall [ \* ]; (y) the conditions under [ \* ]; and (z) [ \* ].

(d) In the event that any of the conditions to the Indemnifying Party's right to assume the defense of a Third-Party Claim under Section 6.5(b) is or becomes unsatisfied, and subject to Section 6.5(c), (i) the Indemnified Party may decide to defend against the Third-Party Claim, provided that in such case the defense shall be [ \* ]; (ii) the Indemnifying Party will reimburse the Indemnified Party promptly and periodically for the costs of defending against the Third-Party Claim (including reasonable attorneys' fees and expenses for one counsel); (iii) the Indemnified Party will keep the Indemnifying Party apprised of all material developments, including settlement offers, with respect to the Third-Party Claim and permit the Indemnifying Party to participate in the defense of the Third-Party Claim, at the sole costs and expense of the Indemnifying Party; and (iv) the Indemnifying Party will remain responsible for any Losses the Indemnified Party may suffer resulting from, arising out of or caused by the Third-Party Claim to the extent provided in this Article 6.

(e) In case the Indemnifying Party has the control of the Third Party Claim pursuant to Section 6.5(b) or 6.5(c) and [ \* ], the Indemnifying Party shall not have the right to consent to the entry of any judgment or enter into any settlement or compromise of such Third-Party Claim without the consent of the Indemnified Party (which consent shall not be unreasonably withheld or delayed) except under the conditions specified in this Section 6.5(e). If the Indemnifying Party elects to enter into a settlement of the Third Party Claim without the consent of the Indemnified Party, then (i) such settlement (1) must [ \* ] and (2) must [ \* ] and [ \* ] and (ii) the Indemnifying Party must pay all monetary obligations specified in such settlement in full, even if such amounts exceed the amount of the maximum liability of the Indemnifying Party under this Agreement. In addition, if the Indemnified Party does not consent to a proposed settlement which the Indemnifying Party would be willing to accept and which satisfies the criteria set forth in the preceding sentence, then the Indemnifying Party shall [ \* ].

(f) In case the Indemnified Party has the control of the Third Party Claim pursuant to Section 6.5(d) or 6.5(c) and [ \* ], the Indemnified Party shall not have the right to consent to the entry of any judgment or enter into any settlement or compromise of such Third-

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Party Claim without the consent of the Indemnifying Party (which consent shall not be unreasonably withheld or delayed) except under the conditions specified in this Section 6.5(f). If the Indemnified Party elects to enter into a settlement of the Third Party Claim without the consent of the Indemnifying Party, (i) the Indemnifying Party will not be bound by the determination of such Third-Party Claim so compromised without the Indemnifying Party's consent and [ \* ], in light of the [ \* ] and (ii) the Indemnifying Party will not be subject to or responsible for any non-monetary obligations under such settlement.

6.6 Procedure for Direct Claims. In the event that any Person believes that it is entitled or may become entitled, to claim indemnification, compensation or reimbursement under this Article 6, such Person shall notify the applicable Indemnifying Party in writing (the "Claim Notice"). The Claim Notice shall include a non-binding estimate of the amount of Losses suffered, incurred or paid or to be suffered, incurred or paid by such Person and contain a description in reasonable detail of the facts and circumstances supporting such Person's claim for indemnification. The Parties hereby agree to resolve any disputes concerning matters set forth in a Claim Notice in accordance with the procedures set forth in Section 8.12.

6.7 Limitation of Liability. EXCEPT FOR AMOUNTS PAYABLE TO ANY PERSON (OTHER THAN A PARTY) BY AN INDEMNIFIED PARTY FOR WHICH IT SEEKS REIMBURSEMENT OR INDEMNIFICATION PROTECTION FROM THE OTHER PARTY PURSUANT TO SECTION 6.1 or SECTION 6.2, AND EXCEPT FOR BREACH OF SECTION 5.4 HEREOF, IN NO EVENT SHALL EITHER PARTY, ITS DIRECTORS, OFFICERS, EMPLOYEES, AGENTS OR AFFILIATES BE LIABLE TO THE OTHER PARTY FOR ANY INDIRECT, INCIDENTAL, SPECIAL, PUNITIVE, EXEMPLARY OR CONSEQUENTIAL DAMAGES, WHETHER BASED UPON A CLAIM OR ACTION OF CONTRACT, WARRANTY, NEGLIGENCE, STRICT LIABILITY OR OTHER TORT, OR OTHERWISE, ARISING OUT OF THIS AGREEMENT, UNLESS SUCH DAMAGES ARE DUE TO THE WILLFUL MISCONDUCT OR FRAUD OF THE LIABLE PARTY. For clarification, the foregoing sentence shall not be interpreted to limit or to expand the express rights specifically granted in the sections of this Agreement.

6.8 Limitations on Indemnification Obligations. Except with respect to willful misconduct or fraud, in no event shall the aggregate liability of the Company for breach of its representations and warranties and/or covenants under this Agreement exceed: (a) with respect to [ \* ] other than with respect to [ \* ], an aggregate amount equal to [ \* ] plus [ \* ] that [ \* ]; and (b) with respect to [ \* ], an aggregate amount equal to [ \* ] plus [ \* ] that [ \* ]. "Fundamental Representations" mean the representations and warranties of the Company under [ \* ]. Notwithstanding anything contained herein to the contrary, except with respect to [ \* ], it is understood and agreed that the maximum liability of the Company for [ \* ] cannot exceed an aggregate amount equal to [ \* ] plus [ \* ] that [ \* ].

6.9 Time Limitation for Claim for Breach of Representations and Warranties. Notice of claims for breach of representations and warranties under Article 3 and Article 4, other than with respect to the Fundamental Representations and the Special Representations, shall be notified, under penalty of forfeiture, respectively by the Company and Buyer, not later than the

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date falling on [ \* ]. Notice of claims for breach of the Fundamental Representations and the Special Representations shall be notified, under penalty of forfeiture, by Buyer, not later than the date falling on the earlier of (a) [ \* ] and (b) [ \* ]. “Special Representations” mean the representations and warranties of the Company under [ \* ].

6.10 No Duplication. Notwithstanding anything contained herein to the contrary, the Buyer Indemnitees and the Company Indemnitees, as applicable, shall not be entitled to recover more than once under this Article 6 or the indemnification obligations set forth in the Gentium Consent for any single indemnifiable Loss; provided, however, that the foregoing shall not be interpreted as limiting the ability of the Buyer Indemnitees or the Company Indemnitees to recover the aggregate amount of all Losses for which they are entitled to indemnification, including the aggregate Losses of different Buyer Indemnitees or Company Indemnitees arising from a single indemnification claim.

#### ARTICLE 7 HSR MATTERS AND EFFECTIVENESS OF AGREEMENT

7.1 HSR Filing. Each Party will, as soon as practicable after, and in any event within [ \* ] Business Days after, the Execution Date (or such later time as may be agreed to in writing by the Parties), file with the United States Federal Trade Commission (“FTC”) and the Antitrust Division of the United States Department of Justice (“DOJ”), any HSR Filing required with respect to the transactions contemplated hereby. The Parties will cooperate with one another to the extent necessary in the preparation of any such HSR Filing. Each Party shall request early termination of the waiting period under the HSR Act. Each Party will be responsible for its own costs and expenses (other than filing fees, which Buyer will pay) associated with any HSR Filing. Each Party shall use diligent efforts to ensure that its representations and warranties set forth in this Agreement shall remain true and correct as of the Effective Date and shall use diligent efforts to ensure the satisfaction of the conditions to Closing for which it is responsible.

7.2 HSR Clearance. In furtherance of obtaining HSR Clearance for an HSR Filing filed under Section 7.1, each Party will use commercially reasonable efforts to resolve as promptly as practicable any objections that may be asserted and to respond as promptly as practicable to requests for supplemental information that may be issued from the FTC, the DOJ or other Governmental Body with respect to this Agreement or the transactions contemplated by this Agreement under any antitrust, competition or trade regulatory Law. Each Party’s commercially reasonable efforts shall include, but will not be limited to, such Party’s counsel’s undertaking to: (a) cooperate and keep the other Party’s counsel appropriately informed of any communications from the FTC, the DOJ or other Governmental Body with respect to this Agreement or the transactions contemplated by this Agreement under any antitrust, competition or trade regulatory Law, and subject to applicable Law, permit the other parties to review in advance any proposed written communication to any such Governmental Body, (b) not participate in any substantive meeting with any such Governmental Body in respect of any filing, investigation or inquiry concerning this Agreement or the transactions contemplated hereby unless it consults with the other Party in advance and, to the extent permitted by such Governmental Body, gives the other Party the opportunity to attend, and (c) furnish the other

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Party with copies of all material correspondence, and written communications between such Party and its Affiliates and their respective Representatives on the one hand, and any such Governmental Body or its staff, on the other hand, with respect to this Agreement or the transactions contemplated hereby, provided however that a Party may elect to restrict, to the other Party's outside counsel, materials that it believes to be valuation related or competitively sensitive. In connection with obtaining such HSR Clearance from the FTC, the DOJ or any other Governmental Body, Buyer and its Affiliates will not be required to (a) sell, hold separate, divest (including through a license or a reversion of licensed or assigned rights), transfer, dispose of or agree to any limitation of restrictions on, any assets (including any assets or rights obtained under this Agreement), operations, rights, product lines, businesses or interest therein of Buyer or any of its Affiliates (or consent to any of the foregoing actions) or (b) litigate or otherwise formally oppose any determination (whether judicial or administrative in nature) by a governmental authority seeking to impose any of the restrictions referenced in clause (a) of this Section 7.2.

7.3 Term and Effectiveness of Certain Provisions. Except for the provisions set forth under Section 2.6, Article 3 (Representations and Warranties of Buyer), Article 4 (Representation and Warranties of Company), Section 5.1 (Limitation on Representations and Liabilities), Section 5.2 (Compound-Related Assets, ST Know-How and Gentium Consent), Section 5.4 (Confidentiality), Article 6 (Indemnification) except for Section 6.3 (Set-Off), Article 7 (HSR Matters and Effectiveness of Agreement) and Article 8 (Miscellaneous), and the definitions used herein, which shall each become effective on the Execution Date, the provisions of this Agreement shall become effective only upon the Effective Date.

7.4 Termination. Before the Effective Date, this Agreement can be terminated only according to the following:

(a) immediately upon written notice by a Party to the other Party, if the HSR Clearance Date has not occurred (i) on or prior to [ \* ] days after the effective date of the latest HSR Filing made by the Parties pursuant to Section 7.1 and (ii) on or prior to [ \* ] days from the Execution Date; provided, that no Party may terminate this Agreement pursuant to this Section 7.4(a)(ii) if such Party is in breach of its obligations to make the HSR Filings under this Agreement;

(b) immediately upon written notice by Buyer to the Company, if the Company has materially breached its covenants or obligations herein or in case of breach of its representations and warranties, which breach would render the condition in Section 2.6(e)(i) incapable of being satisfied; and

(c) immediately upon written notice by the Company, to Buyer if Buyer has materially breached its covenants or obligations herein or in case of breach of its representations and warranties, which breach would render the condition in Section 2.6(d)(i) incapable of being satisfied.

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7.5 [\*] to be Transferred. At least [\*] prior to Closing, the Company will provide Buyer with a copy of all [\*], if any, and all [\*] related to the Compound or Product, if any, in each case that will be transferred to Buyer at Closing.

ARTICLE 8  
MISCELLANEOUS

8.1 Guarantee by Parent. Parent hereby guarantees to the Company the full, prompt, and complete fulfillment of any and all the payment obligations of Buyer (or its assignee hereunder) towards the Company which arise under this Agreement, including those under Sections 2.6(c) and 2.7 and Article 6 of this Agreement subject only to (a) Parent receiving written notification of any payment default by Buyer under this Agreement (which notice shall specify the nature and amount of such payment default and shall be sent to Parent at: 3180 Porter Drive, Palo Alto, California 94304, Attention: General Counsel, or at such other address as specified by Parent in writing); and (b) such amount remaining unpaid [\*] Business Days after Parent's receipt of such notice. Parent hereby represents and warrants to the Company that Parent has full power and authority to execute this Agreement solely for the purposes of this Section 8.1 and the other provisions of this Agreement expressly applicable to the Parent, to deliver this guarantee and to perform its obligations with respect to this guarantee, and this guarantee under this Section 8.1 represents a valid and legal obligation of the Parent, enforceable according to its terms herein, except as such enforceability may be limited by bankruptcy, insolvency, reorganization, moratorium, mandatory provisions of any applicable Law and other laws affecting creditors generally and by the availability of equitable remedies. The Parent has the financial capability to meet the guarantee obligations provided herein. The execution by Parent of this Agreement solely for the purposes of this Section 8.1 and the other provisions of this Agreement expressly applicable to the Parent, delivery by Parent of this guarantee and the performance by Parent of its obligations with respect to this guarantee have been duly approved by all requisite corporate or other applicable action of Parent. Parent hereby consents, and agrees to be bound by, the provisions set forth in this Section 8.1 and the other provisions of this Agreement expressly applicable to the Parent.

8.2 Acknowledgment of Gentium. Gentium hereby acknowledges and agrees that (a) [\*] and (b) to its knowledge, [\*].

8.3 Transfer Taxes. Notwithstanding any provision of this Agreement or the Gentium Consent to the contrary, the Company shall be responsible for payment of United States state or local sales or use tax, if any, applicable to the amounts payable in connection with this Agreement. All transfer, documentary, stamp, registration, value added and other such taxes and fees, including any penalties and interest, (other than United States state or local sales or use tax) imposed in connection with this Agreement ("Transfer Taxes") shall be borne [\*]. The party responsible under applicable Law for the filing of any Tax Return or other documents with respect to any Transfer Tax shall prepare and file such Tax Return at its sole expense (and the other party shall promptly remit payment for [\*] of such Transfer Taxes).

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8.4 Press Releases and Public Announcements. Except as provided in this Section 8.4, each Party, Gentium and Parent shall treat and hold as confidential all of the terms and conditions of this Agreement and the transactions contemplated thereby; provided, however, that each Party, Gentium and Parent may disclose such information to its legal counsel, accountants, or other advisors on an as-needed basis so long as any such Person is bound by a confidentiality obligation with respect thereto similar to the obligation set forth in this Section 8.4. Either Party may issue a press release, or other similar written public announcement, regarding this Agreement or the transactions contemplated hereby, and the Parties (a) shall give each other a reasonable opportunity to review and comment upon any such press release or other similar written public announcement and (b) shall not issue any such press release or similar written public announcement prior to such consultation, except in each case as otherwise required by applicable Law (in which case such Party will use commercially reasonable efforts to advise the other Party prior to making the disclosure). Buyer and the Company shall be permitted to make oral and written public statements and disclosures generally consistent with the content of prior disclosures in any press release or other similar written public announcement permitted under this Section 8.4. For the avoidance of doubt, after the Effective Date, Buyer shall not be restricted in its right to publish information, articles or studies regarding the Compound or Product or press releases relating thereto. The Company acknowledges that Buyer is a subsidiary of a public company and has certain disclosure obligations under applicable Law (including applicable regulations promulgated by applicable security exchanges) to make certain public disclosures with respect to this Agreement and to file a copy of this Agreement with the United States Securities and Exchange Commission. Nothing contained herein shall prohibit Buyer, the Company and their Affiliates from making any public disclosures or filings required under applicable Laws, provided that in such case where the disclosure or filing relates to the transactions contemplated under this Agreement (except to the extent the disclosure is only replicating one or more prior disclosures made in accordance with this Section 8.4), the disclosing Party or its Affiliate shall use commercially reasonable efforts (where legally possible) to advise, and consult with, the other Party prior to making the disclosure.

8.5 No Third-Party Beneficiaries. Other than the Buyer Indemnitees and the Company Indemnitees as set forth herein, this Agreement shall not confer any rights or remedies upon any Person other than the Parties and their respective successors and permitted assigns.

8.6 Entire Agreement. This Agreement constitutes the entire agreement between the Parties and supersedes any prior understandings, agreements, or representations by or among the Parties, written or oral, to the extent they relate in any way to the subject matter hereof; provided, however, that the Confidential Disclosure Agreement, dated August 26, 2013, between Parent and Sigma-Tau Finanziaria S.p.A., an Affiliate of the Company, is not superseded by this Agreement and remains in full force and effect.

8.7 Assignment. This Agreement shall be binding upon and inure to the benefit of the Parties named herein and their respective successors and permitted assigns. Neither this Agreement nor any right or obligation hereunder may be assigned or delegated prior to the Effective Date, in whole or part, by either Party without the prior express written consent of the other; provided, that after the Effective Date (a) the Company may assign or delegate any right

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or obligations hereunder (including the right to receive payment from the Parent under Section 8.1), in whole or in part, to Sigma Tau Finanziaria S.p.A. (“ST Fin”) and/or any Affiliate of ST Fin and (b) Buyer may assign or delegate any right or obligation hereunder, in whole or in part, to any of its Affiliates; in each case under (a) and (b), a prior written notice shall be given to the other Party. Any permitted assignee shall assume all obligations of its assignor under this Agreement, and any permitted assignment shall be binding on the successors of the assigning Party. Parent may assign its obligations hereunder to [ \* ], provided that [ \* ]. Parent may [ \* ]. Any purported assignment in violation of this Section 8.7 shall be void.

8.8 Counterparts. This Agreement may be executed in one or more counterparts (including by means of facsimile or other electronic transmission), each of which shall be deemed an original but all of which together will constitute one and the same instrument.

8.9 Headings. The section headings contained in this Agreement are inserted for convenience only and shall not affect in any way the meaning or interpretation of this Agreement.

8.10 Notices. All notices, requests, demands, claims, and other communications hereunder will be in writing. Any notice, request, demand, claim, or other communication hereunder shall be deemed duly given (a) when delivered personally to the recipient, (b) when sent by electronic mail or facsimile, on the date of transmission to such recipient, (c) one Business Day after being sent to the recipient by reputable overnight courier service (charges prepaid), or (d) four Business Days after being mailed to the recipient by certified or registered mail, return receipt requested and postage prepaid, and, in each case, addressed to the intended recipient as set forth below:

If to the Company:	Sigma-Tau Pharmaceuticals, Inc. 9841 Washingtonian Blvd. Suite 500 Gaithersburg, MD 20878 Attention: CEO - General Counsel Fax: 1 (301) 948-1862
Copies (which shall not constitute notice) to:	Gianni Origoni Grippo Cappelli Partners Via delle Quattro Fontane, 20 00184 Rome Attn: Francesco Gianni - Alfredo D’Aniello Fax: +39-064871101
	Thompson Hine LLP 335 Madison Avenue, 12th Floor New York, New York 10017 Attn: Frank Chaiken; Corby Baumann Fax: (212) 344-6101

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If to Buyer: Jazz Pharmaceuticals International II Limited  
Clarendon House  
2 Church Street  
Hamilton HM 11  
Bermuda  
Attn: David J Doyle, Director  
Fax: +1 (441) 298 7855

Copies (which shall not constitute notice) to: Jazz Pharmaceuticals International II Limited  
c/o Jazz Pharmaceuticals Public Limited Company  
Fourth Floor, Connaught House  
One Burlington Road  
Dublin 4 Ireland  
Attn: Company Secretary  
Fax: +353 (1) 634-7850

Jazz Pharmaceuticals  
3180 Porter Drive  
Palo Alto, CA 94304  
Attn: Suzanne Sawochka Hooper, Executive Vice President & General Counsel  
Fax: (650) 496-3781

Cooley LLP  
3175 Hanover Street  
Palo Alto, CA 94304-1130  
Attn: Marya A. Postner  
Fax: (650) 849-7400

Either Party may change the address to which notices, requests, demands, claims, and other communications hereunder are to be delivered by giving the other Party notice in the manner herein set forth.

8.11 Governing Law. To the fullest extent permitted by applicable Law, this Agreement shall be governed by and construed in accordance with the domestic Laws of the State of New York without giving effect to any choice or conflict of Law provision or rule (whether of the State of New York or any other jurisdiction) that would cause the application of the Laws of any other jurisdiction other than the State of New York, unless the application of such other Law is mandatory according to the Law governing the License Agreement.

8.12 Dispute Resolution.

(a) The Parties recognize that disputes may arise from time to time which relate to either Party's rights and/or obligations hereunder. It is the objective of the Parties to establish procedures to facilitate the resolution of disputes arising under this Agreement in an

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expedient manner by mutual cooperation and without resort to litigation. To accomplish this objective, the Parties, Parent and Gentium agree to follow the procedures set forth in this Section 8.12 to resolve any controversy or claim arising out of, relating to or in connection with any provision of this Agreement, if and when a dispute arises under this Agreement.

(b) With respect to all disputes arising among the Parties, Parent and Gentium (or any subset thereof) (each a “Disputing Party”) under this Agreement, including any alleged breach under this Agreement or any issue relating to the interpretation, application, enforcement, termination or validity of this Agreement, if the Disputing Parties are unable to resolve such dispute within [ \* ] days after such dispute is first identified by either Disputing Party in writing to the other, the Disputing Parties shall refer such dispute to the Chief Operating Officer or Chief Executive Officer of the Disputing Parties (or any senior executive reporting directly to either Disputing Party’s Chief Executive Officer) for attempted resolution by good faith negotiations within [ \* ] days after such notice is received.

(c) If the Chief Operating Officer(s) and/or or Chief Executive Officer(s) (or any senior executive reporting directly to either Disputing Party’s Chief Executive Officer) of the Disputing Parties are not able to resolve such disputed matter within [ \* ] days and either Disputing Party wishes to pursue the matter, each such dispute, controversy or claim shall be finally resolved by binding arbitration administered by JAMS pursuant to JAMS’ Comprehensive Arbitration Rules and Procedures then in effect (the “JAMS Rules”), except as otherwise provided herein. The arbitration shall be governed by the United States Federal Arbitration Act, 9 U.S.C. §§ 1-16, to the exclusion of any inconsistent state Laws. The Disputing Parties adopt and agree to implement the JAMS Optional Arbitration Appeal Procedure (as it exists on the Execution Date of this Agreement) with respect to any final award in an arbitration arising out of or related to this Agreement. Judgment on the arbitration award may be entered in any court having jurisdiction thereof. Any situation not expressly covered by this Agreement shall be decided in accordance with the JAMS Rules. The Disputing Parties agree that:

(i) The arbitration shall be conducted by a single neutral, independent and impartial arbitrator selected in accordance with JAMS Rules, and any appeal under the JAMS Optional Arbitration Appeal Procedure shall be conducted by a panel of three neutral, independent and impartial arbitrators selected in accordance with JAMS Rules. The place of arbitration and appeal shall be New York, NY and all proceedings and communications shall be in English.

(ii) Either Disputing Party may apply to the arbitrator for interim injunctive relief until the arbitration award is rendered or the controversy is otherwise resolved. Either Disputing Party also may, without waiving any remedy under this Agreement, seek from any state or federal court in New York, NY any injunctive or provisional relief necessary to protect the rights or property of that Disputing Party pending the arbitration award. The arbitrator shall have no authority to award punitive or any other type of damages not measured by a Disputing Party’s compensatory damage. Each Disputing Party shall bear its own costs and expenses and attorneys’ fees and an

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equal share of the arbitrator's fees and any administrative fees of arbitration regardless of the outcome of such arbitration.

(iii) Except to the extent necessary to confirm an award or as may be required by Law, neither a Disputing Party nor an arbitrator may disclose the existence, content, or results of an arbitration without the prior written consent of both Disputing Parties. In no event shall an arbitration be initiated after the date when commencement of a legal or equitable proceeding based on the dispute, controversy or claim would be barred by the applicable New York statute of limitations.

8.13 Amendments and Waivers. No amendment of any provision of this Agreement shall be valid unless the same shall be in writing and signed by Buyer and the Company. No waiver by any Party of any provision of this Agreement or any default, misrepresentation, or breach of warranty or covenant hereunder, whether intentional or not, shall be valid unless the same shall be in writing and signed by the Party making such waiver nor shall such waiver be deemed to extend to any prior or subsequent default, misrepresentation, or breach of warranty or covenant hereunder or affect in any way any rights arising by virtue of any prior or subsequent such occurrence.

8.14 Severability. Any term or provision of this Agreement that is invalid or unenforceable in any situation in any jurisdiction shall not affect the validity or enforceability of the remaining terms and provisions hereof or the validity or enforceability of the offending term or provision in any other situation or in any other jurisdiction; in such case the Parties shall negotiate in good faith in order to agree the terms of mutually satisfactory provisions, achieving as closely as possible the same commercial effect, to be substituted for the provisions so found to be void or unenforceable.

8.15 Expenses. Except as otherwise provided herein, each Party will bear its own costs and expenses (including legal fees and expenses) incurred in connection with this Agreement and the transactions contemplated hereby.

8.16 Construction. The Parties have participated jointly in the negotiation and drafting of this Agreement. In the event an ambiguity or 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 of the provisions of this Agreement. Any reference to any Law shall be deemed also to refer to all rules and regulations promulgated thereunder, unless the context requires otherwise.

\* \* \*

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IN WITNESS WHEREOF, each of Buyer, the Company, Parent and Gentium have executed this Agreement as of the date first above written.

**BUYER:**

Jazz Pharmaceuticals International II Limited

By: /s/ Kevin Insley

Name: Kevin Insley

Title: President

**COMPANY:**

Sigma-Tau Pharmaceuticals, Inc.

By: /s/ Dave Lemus & /s/ Marco Maria Brughera

Name: Dave Lemus & Marco Maria Brughera

Title: CEO & President STPI

**Solely for the Purposes of Sections 2.7(b),  
6.10, 7.3, 8.1, 8.4, 8.7, 8.8, 8.10, 8.11 and 8.12:**

Jazz Pharmaceuticals PLC

By: /s/ Fintan Keegan

Name: Fintan Keegan

Title: EVP Technical Operations

**Solely for the Purposes of Sections 6.10, 7.3,  
8.2, 8.4, 8.8, 8.10, 8.11 and 8.12:**

Gentium S.p.A.

By: /s/ Salvatore Calabrese

Name: Salvatore Calabrese

Title: Senior VP CFO/COO

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Exhibit List

Exhibit A – Form of Gentium Consent

Exhibit B – Company Personnel

Schedule 4.8

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