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

FORM 8-K/A
(Amendment No. 1)

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

Date of Report (Date of earliest event reported):

December 19, 2013

**JAZZ PHARMACEUTICALS PUBLIC LIMITED
COMPANY**

(Exact name of registrant as specified in its charter)

Ireland
(State or other jurisdiction of
incorporation)

001-33500
(Commission
File Number)

98-1032470
(IRS Employee
Identification No.)

**Fourth Floor, Connaught House,
One Burlington Road, Dublin 4, Ireland**
011-353-1-634-7800
(Address of principal executive offices, including zip code)

Not Applicable
(Former name or former address, if changed since last report.)

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

- Written communications pursuant to Rule 425 under the Securities Act (17 CFR 230.425)
 - Soliciting material pursuant to Rule 14a-12 under the Exchange Act (17 CFR 240.14a-12)
 - Pre-commencement communications pursuant to Rule 14d-2(b) under the Exchange Act (17 CFR 240.14d-2(b))
 - Pre-commencement communications pursuant to Rule 13e-4(c) under the Exchange Act (17 CFR 240.13e-4(c))
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Item 1.01. Entry into a Material Definitive Agreement.

Tender Offer Agreement

As previously reported on a Current Report on Form 8-K filed with the Securities and Exchange Commission on December 19, 2013 (the “Initial Form 8-K”), on December 19, 2013, Jazz Pharmaceuticals Public Limited Company (“Jazz Pharmaceuticals”), Jazz Pharmaceuticals Italy S.r.l., an Italian *società a responsabilità limitata* (“Purchaser”) and a wholly-owned subsidiary of Jazz Pharmaceuticals, and Gentium S.p.A., a *società per azioni* incorporated in Italy (“Gentium”), entered into a Tender Offer Agreement (the “Tender Offer Agreement”), pursuant to which Jazz Pharmaceuticals will commence a tender offer (the “Offer”) to purchase all of the outstanding ordinary shares of Gentium, no par value per share (“Ordinary Shares”), and all outstanding American Depositary Shares, each representing one Ordinary Share and evidenced by an American Depositary Receipt issued by the Bank of New York, as depositary (the “ADSs” and, together with the Ordinary Shares, the “Company Shares”), of Gentium at a purchase price of \$57.00 per Company Share, net to the seller in cash, without interest thereon, less any required withholding taxes.

Support Agreements

In connection with the Tender Offer Agreement, certain shareholders of Gentium, including certain members of its board of directors and its management team, entered into support agreements with Jazz Pharmaceuticals (collectively, the “Support Agreements”) pursuant to which they each agreed, among other things, to: (i) promptly tender all of their Company Shares into the Offer; (ii) not transfer their Company Shares, subject to certain limited exceptions; (iii) vote their Company Shares against, among other things, any alternative acquisition transaction with respect to Gentium; and (iv) not solicit any employees of Gentium for a period of either one year or eighteen months following the closing of the Offer. Each of the Support Agreements provides that, if the Tender Offer Agreement is terminated under certain circumstances involving, among other things, an alternative acquisition proposal, then the restrictions on transfer of Company Shares and the voting covenants contained therein will continue in effect for a period of six months following the termination of the Tender Offer Agreement. Approximately 15% of the total outstanding Company Shares (and approximately 22% of the number of fully diluted Company Shares) as of the date of the Tender Offer Agreement are subject to the Support Agreements.

The Support Agreements with certain directors and the members of senior management of Gentium also include a covenant not to compete with Gentium in the United States, Canada, Europe or Japan for a period of three years following the closing of the Offer.

Item 9.01 of the Initial Form 8-K is being amended by this Amendment No. 1 to Form 8-K (this “Report”) to include the Tender Offer Agreement as Exhibit 2.1 and the form of Support Agreement as Exhibit 99.3. Such information should be read in conjunction with the Initial Form 8-K. The summary of the Tender Offer Agreement and the transactions contemplated thereby set forth above and in the Initial 8-K does not purport to be complete and is subject to, and qualified in its entirety by, the full text of the Tender Offer Agreement attached as Exhibit 2.1, which is incorporated herein by reference. The summary of the Support Agreements set forth above does not purport to be complete and is qualified in its entirety by reference to the form of Support Agreement filed as Exhibit 99.3 hereto, and is incorporated into this Report by reference. The Tender Offer Agreement has been attached to provide investors with information regarding its terms. It is not intended to provide any other factual information about Jazz Pharmaceuticals, Purchaser or Gentium, their respective businesses, or the actual conduct of their respective businesses during the period prior to the consummation of the Offer. The Tender Offer Agreement contains representations and warranties that are the product of negotiations among the parties thereto, and solely for the benefit of, each other as of specified dates. In particular, the assertions embodied in the representations and warranties contained in the Tender Offer Agreement are qualified by information in confidential disclosure letter provided by the parties thereto in connection with the signing of the Tender Offer Agreement. This disclosure letter contains information that modifies, qualifies and creates exceptions to the representations and warranties set forth in the Tender Offer Agreement. Moreover, certain representations and warranties in the Tender Offer Agreement were used for the purpose of allocating risk between Jazz Pharmaceuticals, Purchaser and Gentium, rather than establishing matters of fact and may be subject to standards of materiality applicable to the contracting parties that differ from those applicable to investors. Accordingly, you should not rely on the representations and warranties in the Tender Offer Agreement as characterizations of the actual state of facts about Jazz Pharmaceuticals, Purchaser or Gentium or any of their respective businesses.

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

<u>Exhibit Number</u>	<u>Description</u>
2.1	Tender Offer Agreement, dated December 19, 2013, by and among Jazz Pharmaceuticals Public Limited Company, Jazz Pharmaceuticals Italy S.r.l. and Gentium S.p.A.*
99.1	Joint Press Release of Jazz Pharmaceuticals and Gentium issued on December 19, 2013**
99.2	Jazz Pharmaceuticals investor presentation first made available on December 19, 2013**
99.3	Form of Support Agreement

* Schedules and certain exhibits to the Tender Offer Agreement have been omitted pursuant to Item 601(b)(2) of Regulation S-K. Jazz Pharmaceuticals hereby undertakes to furnish supplementally copies of any omitted schedules and exhibits upon request by the SEC.

** Previously filed.

“Safe Harbor” Statement under the Private Securities Litigation Reform Act of 1995

This Amendment No. 1 to Form 8-K contains forward-looking statements, including, but not limited to, statements related to the anticipated consummation of the Offer and the timing and benefits thereof, the expected financing for the Offer, and other statements that are not historical facts. These forward-looking statements are based on Jazz Pharmaceuticals’ 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 related to Jazz Pharmaceuticals’ ability to complete the Offer on the proposed terms and schedule, including risks and uncertainties related to the satisfaction of closing conditions. There can be no assurance that Jazz Pharmaceuticals will be able to complete the transactions contemplated by the Tender Offer Agreement on the anticipated terms, or at all. Additional risks and uncertainties relating to Jazz Pharmaceuticals and its business can be found under the caption “Risk Factors” and elsewhere in Jazz Pharmaceuticals plc’s SEC filings and reports (Commission File No. 001-33500), including in the Quarterly Report on Form 10-Q for the quarter ended September 30, 2013 filed with the SEC. Jazz Pharmaceuticals undertakes no duty or obligation to update any forward-looking statements contained in this Amendment No. 1 to Form 8-K as a result of new information, future events or changes in its expectations.

Additional Information and Where to Find It

The tender offer for the outstanding shares of Gentium (including those shares represented by American Depositary Shares) referenced in this Current Report on Form 8-K has not yet commenced. This announcement is for informational purposes only and is neither an offer to purchase nor a solicitation of an offer to sell shares of Gentium, nor is it a substitute for the tender offer materials that Jazz Pharmaceuticals and its acquisition subsidiary will file with the U.S. Securities and Exchange Commission (the “SEC”) upon commencement of the tender offer. At the time the tender offer is commenced, Jazz Pharmaceuticals and its acquisition subsidiary will file tender offer materials on Schedule TO, and Gentium will file a Solicitation/Recommendation Statement on Schedule 14D-9 with the SEC with respect to the tender offer. The tender offer materials (including an Offer to Purchase, a related Letter of Transmittal and certain other tender offer documents) and the Solicitation/Recommendation Statement will contain important information. Holders of shares of Gentium are urged to read these documents when they become available because they will contain important information that holders of Gentium securities should consider before making any decision regarding tendering their securities. The Offer to Purchase, the related Letter of Transmittal and certain other tender offer documents, as well as the Solicitation/Recommendation Statement, will be made available to all holders of shares of Gentium at no expense to them. Investors and security holders may obtain free copies of these documents (when they are available) and other related documents filed with the SEC at the SEC’s web site at <http://www.sec.gov> or by (i) directing a request to Jazz Pharmaceuticals plc, c/o Jazz Pharmaceuticals,

Inc., 3180 Porter Drive, Palo Alto, California 94304, U.S.A., Attention: Investor Relations, (ii) calling +353 1 634 7892 (Ireland) or + 1 650 496 2800 (U.S.) or (iii) sending an email to investorinfo@jazzpharma.com. Investors and security holders may obtain free copies of the documents filed with the SEC on Jazz Pharmaceuticals' website at www.jazzpharmaceuticals.com under the heading "Investors" and then under the heading "SEC Filings".

In addition to the Offer to Purchase, the related Letter of Transmittal and certain other tender offer documents, as well as the Solicitation/Recommendation Statement, Jazz Pharmaceuticals and Gentium file annual, quarterly (except in the case of Gentium) and special reports and other information with the SEC. You may read and copy any reports or other information filed by Jazz Pharmaceuticals or Gentium at the SEC public reference room at 100 F Street, N.E., Washington, D.C. 20549. Please call the Commission at 1-800-SEC-0330 for further information on the public reference room. Jazz Pharmaceuticals' and Gentium's filings with the SEC are also available to the public from commercial document-retrieval services and at the website maintained by the SEC at <http://www.sec.gov>.

SIGNATURE

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

JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY

Date: December 20, 2013

By: /s/ Suzanne Sawochka Hooper

Name: Suzanne Sawochka Hooper

Title: Executive Vice President and General Counsel

EXHIBIT INDEX

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** Previously filed.

TENDER OFFER AGREEMENT

by and among

JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY

JAZZ PHARMACEUTICALS ITALY S.R.L.

and

GENTIUM S.P.A.

Dated as of December 19, 2013

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TENDER OFFER AGREEMENT

This TENDER OFFER AGREEMENT (this “**Agreement**”) is made and entered into as of December 19, 2013 (the “**Agreement Date**”) by and among Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland (“**Parent**”), Jazz Pharmaceuticals Italy S.r.l., an Italian *società a responsabilità limitata* and a wholly owned subsidiary of Parent (“**Purchaser**”) and Gentium S.p.A., a *società per azioni* incorporated in Italy (the “**Company**”). All capitalized terms used in this Agreement shall have the respective meanings ascribed thereto in Article I hereof.

WITNESSETH:

WHEREAS, it is proposed that Purchaser shall, as promptly as practicable, commence a tender offer (the “**Offer**”) to acquire all of the outstanding shares of Company Common Stock and Company ADSs (together, the “**Company Shares**”) (other than any Company Shares owned by Parent, Purchaser or any of their Subsidiaries), in each case (and without duplication for shares of Company Common Stock underlying Company ADSs), at a price of \$57.00 per Company Share, net to the holder thereof in cash (such amount, or any different amount per Company Share that may be paid pursuant to the Offer, being hereinafter referred to as the “**Offer Price**”), without interest and subject to any required withholding or other Taxes, all upon the terms and subject to the conditions set forth herein.

WHEREAS, the Board of Directors of each of Parent and Purchaser, as well as the Company Board, has, by the unanimous vote of all directors of the Company, other than one director that has deemed himself conflicted, (a) determined that this Agreement is advisable, (b) determined that the Offer is at a price and on terms that are fair to and in the best interests of their respective shareholders and, with respect to the Company Board, the other Company stakeholders and (c) approved this Agreement and the transactions contemplated hereby, all upon the terms and subject to the conditions set forth herein.

WHEREAS, in order to induce Parent and Purchaser to enter into this Agreement and to consummate the Offer, concurrently with the execution and delivery of this Agreement: (a) certain holders of Company Shares are executing and delivering Support Agreements in favor of Parent and Purchaser (the “**Support Agreements**”); (b) certain directors of the Company are executing and delivering to Purchaser resignation letters that would become effective at or around the Acceptance Time; (c) certain key employees are entering into certain agreements, including consulting or retention agreements and/or confidentiality and intellectual property assignment agreements, with the Company and its Subsidiaries at the direction of Parent and Purchaser; and (d) certain key employees of the Company are entering into noncompetition and non-solicitation agreements in favor of Parent.

NOW, THEREFORE, in consideration of the foregoing premises and the representations, warranties, covenants and agreements set forth herein, as well as other good and valuable consideration, the receipt and sufficiency of which are hereby acknowledged and accepted, and intending to be legally bound hereby, Parent, Purchaser and the Company hereby agree as follows:

ARTICLE I
DEFINITIONS & INTERPRETATIONS

Section 1.1 Certain Definitions. For all purposes of and under this Agreement, the following capitalized terms shall have the following respective meanings:

(a) “**Acceptance Time**” shall mean the first time as of which Purchaser accepts any Company Shares for payment pursuant to the Offer; provided that any rights of Purchaser or Parent that arise under this Agreement as a result of the Acceptance Time having occurred shall be subject to payment by Purchaser for such Company Shares.

(b) “**Acquisition Proposal**” shall mean any inquiry of or communication to the Company or any offer or proposal, or indication of interest in making an offer or proposal (other than an offer, proposal or indication of interest in making an offer or proposal by Parent or Purchaser), in each case, relating to any Acquisition Transaction.

(c) “**Acquisition Transaction**” shall mean any transaction or series of related transactions (other than the transactions contemplated by this Agreement) involving: (i) any acquisition or purchase from the Company or any of its Subsidiaries by any Person or “group” (as defined in or under Section 13(d) of the Exchange Act), directly or indirectly, of more than a fifteen percent (15%) interest in the total outstanding voting securities of the Company or any of its Subsidiaries (it being understood that Company ADSs constitute voting securities for all purposes under this Agreement), (ii) any tender offer (including self-tender) or exchange offer that if consummated would result in any Person or “group” (as defined in or under Section 13(d) of the Exchange Act) beneficially owning fifteen percent (15%) or more of the total outstanding voting securities of the Company or any of its Subsidiaries (an “**Acquisition Transaction Tender Offer**”); (iii) any merger, consolidation, business combination, share exchange, issuance of securities, acquisition of securities, reorganization, recapitalization or other similar transaction involving the Company or any of its Subsidiaries, the business(es) of which, individually or in the aggregate, constitute more than fifteen percent (15%) of the assets of the Company and its Subsidiaries, taken as a whole, pursuant to which the shareholders of the Company or such Subsidiary immediately preceding such transaction hold less than eighty-five percent (85%) of the voting equity interests in the surviving or resulting entity of such transaction or any parent entity thereof; (iv) any sale, lease, exchange, transfer, license or disposition (in each case, other than in the ordinary course of business) of more than fifteen percent (15%) of the assets of the Company and its Subsidiaries, taken as a whole (measured by the lesser of book or fair market value thereof); or (v) any combination of the foregoing.

(d) “**Action**” shall mean any action, claim, suit, litigation, proceeding (public or private) or criminal prosecution by or before any Governmental Entity.

(e) “**Affiliate**” shall mean, with respect to any Person, any other Person that directly or indirectly controls, is controlled by or is under common control with such Person. For purposes of the immediately preceding sentence, the term “control” (including, with correlative meanings, the terms “controlling,” “controlled by” and “under common control with”), as used with respect to any Person, means the possession, directly or indirectly, of the power to direct or cause the direction of the management and policies of such Person, whether through ownership of voting securities, by contract or otherwise.

(f) "**API**" shall mean any active pharmaceutical ingredient that is supplied by or on behalf of the Company or any of its Subsidiaries to any Person in any country in the world as of the date of this Agreement, including Urokinase and Sulglicotide.

(g) "**beneficially own**" or "**beneficial ownership**" with respect to any securities shall mean having "beneficial ownership" of such securities (as determined pursuant to Rule 13d-3 under the Exchange Act), including pursuant to any agreement, arrangement or understanding, whether or not in writing.

(h) "**Business Day**" shall have the meaning ascribed thereto in Rule 14d-1(g)(3).

(i) "**Code**" shall mean the Internal Revenue Code of 1986, as amended, and the rules and regulations promulgated thereunder, or any successor statute, rules and regulations thereto.

(j) "**Company 20-F**" shall mean the Company's Annual Report on Form 20-F for the fiscal year ended December 31, 2012, filed with the SEC on April 1, 2013, and amended by the Company on Form 20-F/A (Amendment No. 1), filed with the SEC on April 30, 2013.

(k) "**Company ADSs**" means shall mean the American Depositary Shares, each representing one (1) share of Company Common Stock and which are currently listed on NASDAQ.

(l) "**Company Board**" shall mean the Board of Directors of the Company.

(m) "**Company Common Stock**" shall mean the ordinary shares, no par value per share, of the Company.

(n) "**Company Contract**" shall mean any Contract: (a) to which the Company or any Subsidiary of the Company is a party; (b) to the Knowledge of the Company, by which the Company or any Subsidiary of the Company is bound or under which the Company or any Subsidiary of the Company has any obligation; or (c) to the Knowledge of the Company, under which the Company or any Subsidiary of the Company has any legally enforceable right or interest.

(o) "**Company Equity Plans**" shall mean (i) the Company's 2004 Equity Incentive Plan (including the Company's 2004 Italy Stock Award Sub-Plan), as amended and restated, (ii) the Company's 2007 Stock Option Plan, as amended and (iii) any other compensatory equity award plans or Contracts of the Company, including option plans or Contracts assumed by the Company pursuant to a merger or acquisition.

(p) "**Company Intellectual Property Rights**" shall mean (i) all Intellectual Property Rights solely owned by the Company or any of its Subsidiaries or co-owned by the Company or any of its Subsidiaries, on the one hand, and one or more third parties on the other, and (ii) all Intellectual Property Rights which are licensed to the Company or any of its Subsidiaries (other than implied (and not express) licenses and other than licenses accompanying computer software that have not been modified or customized for the Company or any of its Subsidiaries).

(q) "**Company IP Contracts**" shall mean all Contracts in effect as of the date of this Agreement under which any third party has licensed, granted, assigned or conveyed to the Company

or any of its Subsidiaries any right, title or interest in or to any Material Company IP Rights (other than “shrink wrap,” “click through” or similar license agreements accompanying widely available computer software that have not been modified or customized for the Company or any of its Subsidiaries).

(r) “**Company Material Adverse Effect**” shall mean any fact, circumstance, occurrence, event, change or effect (an “Effect”) that, individually or when taken together with all other Effects, has had or is reasonably likely to have a material adverse effect on the business, operations, assets (including intangible assets), financial condition or results of operations of the Company taken as a whole with its Subsidiaries; provided, however, that, none of the following Effects, by itself or when aggregated with any one or more other Effects, shall be deemed to be or constitute a Company Material Adverse Effect and none of the following Effects, by itself or when aggregated with any one or more other Effects, shall be taken into account when determining whether a Company Material Adverse Effect has occurred or is reasonably likely to occur: Effects arising out of or resulting from (A) (1) general market, economic or political conditions in Italy or worldwide or (2) conditions (or any changes therein) in the industries in which the Company or any of its Subsidiaries conduct business, in each case, including any acts of terrorism or war, weather conditions or other force majeure events, in the case of each of clauses (1) and (2), to the extent that such Effects do not have and are not reasonably likely to have a disproportionate impact on the Company and its Subsidiaries, taken as a whole, relative to other companies operating in the same industries in which the Company or any of its Subsidiaries conduct business; (B) the announcement of the execution of this Agreement or the pendency of the Offer, including any loss of, or adverse change in, the relationship of the Company or any of its Subsidiaries with its employees, licensors, licensees, customers, distributors, partners or suppliers resulting or arising therefrom but excluding any Action made or brought by any Company Shareholder in its capacity as a shareholder which, together with its Affiliates, beneficially owns 10% or more of the Company Shares; (C) changes in GAAP or other accounting standards (or the interpretation thereof by a third party); (D) changes in applicable Law or regulatory conditions (or the interpretation thereof by a third party), to the extent that such changes do not have a disproportionate impact on the Company and its Subsidiaries, taken as a whole, relative to other companies operating in the same industries in which the Company or any of its Subsidiaries conduct business; (E) changes in the trading price or trading volume of the Company ADSs, in and of themselves (it being understood that any underlying cause of any such change may, subject to the other terms of this definition, be taken into consideration when determining whether a Material Adverse Effect has occurred or is reasonably likely to occur); or (F) any failure by the Company or any of its Subsidiaries to meet any public estimates or expectations of the Company’s bookings, revenue, earnings or other financial performance or results of operations for any period, or any failure by the Company or any of its Subsidiaries to meet any internal budgets, plans or forecasts of its bookings, revenues, earnings, earnings per share or other financial performance or results of operations (it being understood that any underlying cause of any such failure may, subject to the other terms of this definition, be taken into consideration when determining whether a Material Adverse Effect has occurred or is reasonably likely to occur).

(s) “**Company Options**” shall mean any options to purchase Company Shares outstanding under any of the Company Equity Plans.

(t) “**Confidentiality Agreement**” shall mean the letter agreement regarding non-disclosure and other obligations, by and between Parent and the Company dated September 30, 2013, which will continue in full force and effect in accordance with its terms.

(u) “**Consultant Agreement**” shall mean each independent contractor or consultant agreement or other consultant-related Contract between the Company or any of its Subsidiaries and any independent contractors or consultants.

(v) “**Continuing Employees**” shall mean all employees of the Company (i) who are offered and timely accept employment by Parent or any Subsidiary of Parent, (ii) who continue their employment with the Company at the request of Parent, or (iii) who remain or become employees of the Company or Parent or any Subsidiary of Parent, as required by applicable Law.

(w) “**Contract**” shall mean any written or oral legally binding bond, debenture, note, mortgage, indenture, guarantee, license, lease, purchase or sale order or other contract, commitment, agreement, instrument or obligation, including all amendments thereto, and including any of the foregoing that has been terminated or has expired and has ongoing obligations or under which any liabilities of any kind may exist.

(x) “**Defibrotide**” shall mean the product currently under development and sold by the Company and/or its Subsidiaries under the trademark Defibrotide.

(y) “**Deposit Agreement**” shall mean that certain Deposit Agreement by and among the Company, The Bank of New York, as depository, and the owners and beneficial owners of American Depositary Receipts issued thereunder, dated as of June 15, 2005, as amended.

(z) “**DOJ**” shall mean the United States Department of Justice, or any successor thereto.

(aa) “**EMA**” shall mean the European Medicines Agency, or any successor thereto.

(bb) “**Employee Benefit Plan**” shall mean each plan, fund, program, agreement, arrangement or scheme sponsored, maintained or contributed to (or required to be contributed to) by the Company or any of its Subsidiaries, and any communications establishing or creating obligations related thereto, with respect to which the Company or any of its Subsidiaries has any liability, contingent or otherwise, for any current or former employees, directors, independent contractors, consultants or other persons engaged by the Company or any of its Subsidiaries providing for bonus or other incentive compensation, stock purchase, stock option and other equity compensation, severance or other termination benefits, salary continuation, unemployment compensation, retention or change of control benefits, deferred compensation, vacation, profit sharing, retirement benefits, health benefits, insurance coverage, death or disability benefits, fringe benefits, or any other employee benefits or other similar benefits.

(cc) “**Employment Agreement**” shall mean each management, employment, relocation, repatriation, expatriation, visa, work permit or other employment-related Contract between the Company or any of its Subsidiaries and any employees.

(dd) “**Environmental Law**” shall mean all applicable Laws issued, promulgated, approved or entered relating to the protection of the environment (including ambient air, surface water, groundwater, land, or plant or animal life or other natural resource) or human health or safety, or otherwise relating to the production, use, emission, storage, treatment, transportation, recycling,

disposal, discharge, release or other handling of any Hazardous Materials or any products or wastes containing any Hazardous Materials including any Laws related to product take-back, packaging, or content requirements, or the investigation, clean-up or other remediation or analysis of or liability related to Hazardous Materials.

(ee) “**Exchange Act**” shall mean the Securities Exchange Act of 1934, as amended, and the rules and regulations promulgated thereunder, or any successor statute, rules and regulations thereto.

(ff) “**FASB ASC**” shall mean the Accounting Standards Codification of the Financial Accounting Standards Board.

(gg) “**FDA**” shall mean the United States Food and Drug Administration, or any successor thereto.

(hh) “**Financing Sources**” shall mean the entities that have committed to provide or otherwise entered into agreements in connection with the Purchaser’s financing of the Offer (the “**Debt Financing**”), including the lenders under the Purchaser Credit Facility and the parties to the Financing Commitment, and any joinder agreements, credit agreements, indentures or other definitive documentation relating thereto, together with their Affiliates and their and their Affiliates’ officers, directors, employees, agents and representatives and their respective successors and assigns.

(ii) “**FTC**” shall mean the United States Federal Trade Commission, or any successor thereto.

(jj) “**GAAP**” shall mean, as the context may require, Italian GAAP with reference to the Company’s financial statements, Swiss GAAP in relation to Gentium GmbH’s financial statements and U.S. GAAP with reference to the consolidated financial statements filed with the SEC by the Company.

(kk) “**Governmental Entity**” shall mean any EU, EU Member State, United States, federal, state, provincial, local, municipal, district, foreign or other jurisdiction of any nature government, any government or subdivision thereof or any other governmental, administrative, judicial, arbitral, legislative, executive, regulatory or self-regulatory authority, instrumentality, agency, commission or body.

(ll) “**Hazardous Material**” shall mean any hazardous waste, substance, material, emission or chemical pollutant or contaminant (including petroleum and petroleum products, PCBs, urea-formaldehyde, radon, mold, CFCs and all other ozone-depleting substances, lead or lead based paints or materials, friable asbestos or asbestos containing materials, and any other material regulated under, or that can result in liability under, any Environmental Law).

(mm) “**Intellectual Property Rights**” shall mean any and all of the following and all worldwide common law and statutory rights in or arising out of: (i) all issued patents, pending patent applications and abandoned patents and applications provided that they can be revived (which for purposes of this Agreement shall include utility models, design patents, industrial designs, certificates of invention and applications for certificates of invention and priority rights) in any country or region, including all reissues, re-examinations, divisionals, renewals, extensions, provisionals, continuations and

continuations-in-part thereof (“**Patents**”); (ii) copyrights, copyright registrations and applications therefor and all copyrightable works (including databases and other compilations of information, mask works and semiconductor chip rights), and all other rights corresponding thereto throughout the world however denominated, including all rights of authorship, use, publication, reproduction, distribution, performance, transformation, moral rights and rights of ownership of copyrightable works and all registrations and rights to register and obtain renewals and extensions of registrations, together with all other interests accruing by reason of international copyright (“**Copyrights**”); (iii) industrial designs and any registrations and applications therefor; (iv) rights in trademarks, registered trademarks, applications for registration of trademarks, service marks, registered service marks, applications for registration of service marks, trade names, registered trade names and applications for registration of trade names, and Internet domain name registrations; and including all intent to use any of the foregoing if not registered or subject to a pending application (“**Trademarks**”); (v) rights in trade secrets (including in any information that derives economic value (actual or potential) from not being generally known to other persons who can obtain economic value from its disclosure and any other “trade secrets” as defined in the Uniform Trade Secrets Act and under corresponding foreign statutory and common law) and rights in other proprietary or confidential information including rights to limit the use or disclosure thereof by any Person (“**Trade Secrets**”); (vi) rights with respect to databases and data collections; (vii) rights in internet domain names; (viii) rights in computer software (excluding unmodified, commercially-available software); (ix) moral rights; (x) any other proprietary, intellectual or industrial property rights of any kind or nature; and (xi) the rights to sue for and remedies against past, present and future infringements of any or all of the foregoing, and rights of priority and protection of interests therein under the Laws of any jurisdiction worldwide.

(nn) “**IRS**” shall mean the United States Internal Revenue Service, or any successor thereto.

(oo) “**Italian GAAP**”: shall mean the rules of Italian Law applicable to the drafting of financial statements, as integrated by, interpreted and applied in accordance with the accounting principles (*principi contabili*) issued by (x) the *Consiglio Nazionale dei Dottori Commercialisti and the Consiglio Nazionale dei Ragionieri*, as amended by the OIC (*Organismo Italiano di Contabilità*), and (y) directly by the OIC (*Organismo Italiano di Contabilità*), in each case in effect as of the reference date of the financial statements.

(pp) “**Knowledge**” of the Company, with respect to any matter in question, shall mean the actual knowledge of such matter by any of the executive officers or directors of the Company.

(qq) “**Law**” shall mean any EU, EU Member State, United States, federal, state, local or foreign law, statute, ordinance, rule, code, regulation, order, industry codes of conduct, judgment, injunction or decree.

(rr) “**Leased Real Property**” shall mean all real property leased for the benefit of the Company or any of its Subsidiaries pursuant to a Contract providing for annual aggregate rent in excess of \$100,000.

(ss) “**Licensed Company IP**” shall mean all Company Intellectual Property Rights that are licensed, exclusively or non-exclusively, to the Company or any of its Subsidiaries.

(tt) "**Lien**" shall mean any pledge, claim, lien, charge, mortgage, hypothecation, option, right of first refusal, encumbrance or security interest of any kind or nature whatsoever (including any limitation on voting, sale, transfer or other disposition or exercise of any other attribute of ownership) other than (i) Liens disclosed on the consolidated balance sheet of such Person included in the most recent annual report filed by such Person with the SEC prior to the Agreement Date, (ii) Liens for Taxes and other similar governmental charges and assessments which are not yet due and payable or liens for Taxes being contested in good faith by any appropriate Action for which adequate reserves have been established to the extent required by GAAP; (iii) liens of landlords other than in relation to where the relevant obligations of the tenant have not been breached or violated, and liens of carriers, warehousemen, mechanics and materialmen and other like liens arising in the ordinary course of business; (iv) security given in the ordinary course of business to any public utility, Governmental Entity or other statutory or public authority, (v) defects, imperfections or irregularities in title, covenants, easements and rights-of-way (unrecorded and of record) and other similar Liens (or other encumbrances of any type) on zoning, building and other similar codes or restrictions relating to real property, in each case that do not adversely affect in any material respect the current use of the applicable real property owned, leased or held for use by the Company or any of its Subsidiaries; (vii) pledges or deposits to secure obligations under workers' compensation laws or similar legislation; (viii) Liens imposed by applicable Law (other than Tax law); (ix) Liens imposed on the underlying fee interest in Leased Real Property that are not caused by the Company or any of its Subsidiaries; and (x) non-exclusive licenses of Intellectual Property Rights granted by the Company or its Subsidiaries in the ordinary course of business.

(uu) "**Material**" shall, solely for purposes of Sections Section 3.8(h), Section 3.11(a)-(c) and Section 3.12(c), mean any amount in excess of \$20,000 per event, incident or occasion, or in potential value or liability.

(vv) "**Material Company IP Rights**" shall mean (i) all Patents within the Company Intellectual Property Rights covering or claiming Defibrotide, or its use or manufacture and (ii) all material Company Intellectual Property Rights (other than Patents) relating to or used in Defibrotide, or its use or manufacture.

(ww) "**NASDAQ**" shall mean the NASDAQ Global Market, or any successor inter-dealer quotation system operated by The NASDAQ Stock Market, Inc., or any successor thereto.

(xx) "**Order**" shall mean any judgment, decision, decree, injunction, ruling, writ, assessment or order of any Governmental Entity that is binding on any Person or its property under applicable Law.

(yy) "**Owned Company IP**" shall mean those Company Intellectual Property Rights that are owned by the Company or any of its Subsidiaries.

(zz) "**Owned Real Property**" shall mean all real property owned by the Company or any of its Subsidiaries.

(aaa) "**Person**" shall mean any individual, corporation (including any non-profit corporation), general partnership, limited partnership, limited liability partnership, joint venture, estate, trust, company (including any limited liability company or joint stock company), firm or other enterprise, association, organization, entity or Governmental Entity.

(bbb) “**Product**” shall mean (a) any pharmaceutical product that has received regulatory approval or that is offered for commercial sale by or on behalf of the Company or any of its Subsidiaries in any jurisdiction in the world as of the date of this Agreement; or (b) any product candidate, including any reformulation of a product described under clause (a), that the Company or any of its Subsidiaries is developing or seeking regulatory approval for as of the date of this Agreement. For clarity, Defibrotide is a Product.

(ccc) “**Purchaser Credit Facility**” shall mean that certain Credit Facility, dated June 12, 2012, as amended, modified or otherwise supplemented from time to time, by and among Purchaser, certain subsidiaries of the Purchaser, Barclays Bank PLC, as administrative agent, and the lenders from time to time party thereto.

(ddd) “**Purchaser Material Adverse Effect**” shall mean any Effect that, individually or when taken together with all other Effects, does, or would be reasonably likely to, prevent or materially impair or delay the performance by Purchaser or Parent of any of their material obligations under this Agreement or to accept for payment and to pay for the Company Shares validly tendered (and not withdrawn) pursuant to the Offer on the terms set forth in this Agreement prior to the Termination Date, or if extended pursuant to Section 7.1(b), the Extended Termination Date.

(eee) “**Regulatory Authority**” shall mean any EU, EU Member State, United States federal, national, or multinational governmental health regulatory agency or authority within a regulatory jurisdiction, with the authority to grant approvals, licenses, registrations or authorizations necessary for the development, manufacture, use and sale of a pharmaceutical product, including the FDA, the EMA, European Commission and the national competent authority of the Republic of Italy.

(fff) “**Sarbanes-Oxley Act**” shall mean the Sarbanes-Oxley Act of 2002, as amended.

(ggg) “**SEC**” shall mean the United States Securities and Exchange Commission, or any successor thereto.

(hhh) “**Securities Act**” shall mean the Securities Act of 1933, as amended, and the rules and regulations promulgated thereunder, or any successor statute, rules or regulations thereto.

(iii) “**Specified Representations**” shall mean the representations and warranties of the Company contained in Section 3.2(a) and Section 3.2(b) (*Capital Stock*), Section 3.4 (*Authority*), Section 3.19 (*Takeover Statutes*), Section 3.22 (*No Vote Required*), Section 3.23 (*Opinion of Financial Advisor*) and Section 3.25 (*Rule 14d-10*) of this Agreement.

(jjj) “**Subsidiary**” of any Person shall mean (i) a corporation more than fifty percent (50%) of the combined voting power of the outstanding voting securities of which is owned, directly or indirectly, by such Person or by one or more other Subsidiaries of such Person or by such Person and one or more other Subsidiaries thereof, (ii) a partnership of which such Person, or one or more other Subsidiaries of such Person or such Person and one or more other Subsidiaries thereof, directly or indirectly, is the general partner and has the power to direct the policies, management and

affairs of such partnership, (iii) a limited liability company of which such Person or one or more other Subsidiaries of such Person or such Person and one or more other Subsidiaries thereof, directly or indirectly, is the managing member and has the power to direct the policies, management and affairs of such company, (iv) any other Person (other than a corporation, partnership or limited liability company) in which such Person, or one or more other Subsidiaries of such Person or such Person and one or more other Subsidiaries thereof, directly or indirectly, has at least a majority ownership and power to direct the policies, management and affairs thereof, or (iv) any representative office, sales office or branch in the United States.

(kkk) “**Sulglicotide**” shall mean the active pharmaceutical ingredient currently sold by the Company or its Subsidiaries under the name sulglicotide.

(lll) “**Superior Proposal**” shall mean any bona fide written Acquisition Proposal involving an Acquisition Transaction that, if consummated, would result in a Person or “group” (as defined in or under Section 13(d) of the Exchange Act) owning more than fifty percent (50%) of the outstanding voting securities of the Company or more than fifty percent (50%) of the assets of the Company and its Subsidiaries, taken as a whole, which the Company Board shall have determined in good faith (after consultation with its financial advisor and its outside legal counsel) is more favorable to the holders of Company Shares (in their capacity as such) and other stakeholders of the Company than the transactions contemplated by this Agreement.

(mmm) “**Swiss GAAP**” shall mean generally accepted accounting principles, as applied in Switzerland and the rules of Swiss law applicable to the drafting of financial statements.

(nnn) “**Tax**” shall mean (i) any and all taxes, including net income, gross income, gross receipts, capital gains, alternative, minimum, sales, consumption, use, social services, goods and services, value added, harmonized sales, ad valorem, transfer, franchise, profits, registration, license, lease, service, service use, withholding, payroll, wage, employment, unemployment, pension, health insurance, excise, severance, stamp, occupation, premium, property, windfall profits, environmental, customs, duties or other taxes, fees, assessments, social security contributions or charges of any kind whatsoever, together with any interest and any penalties, additions to tax or additional amounts with respect thereto; (ii) any liability for payment of amounts described in clause (i) whether as a result of transferee liability, of being a member of any group of entities for any period or otherwise through operation of law; and (iii) any liability for the payment of amounts described in clauses (i) or (ii) as a result of any tax sharing, tax indemnity or tax allocation agreement or any other express or implied agreement to indemnify any other Person.

(ooo) “**Tax Returns**” shall mean any return (including any information return), report, statement, declaration, estimate, schedule, notice, notification, form, election, certificate or other document, and any amendment or supplement to any of the foregoing, with respect to income Taxes.

(ppp) “**U.S. GAAP**” shall mean generally accepted accounting principles, as applied in the United States.

(qqq) “**Urokinase**” shall mean the active pharmaceutical ingredient currently sold by the Company or its Subsidiaries under the name urokinase.

Section 1.2 Additional Definitions. The following capitalized terms shall have the respective meanings ascribed thereto in the respective sections of this Agreement set forth opposite each of the capitalized terms below:

<u>Term</u>	<u>Section Reference</u>
401(k) Plan	Section 6.8(f)
Acceptance Time Board Appointments	Section 6.11
Acquisition Agreement	Section 6.2(a)
Adjusted Outstanding Share Number	Section 2.1(a)
Agreement	Preamble
Agreement Date	Preamble
Antitrust Authority	Section 6.3(c)
Antitrust Laws	Section 6.3(c)
Business	Section 4.14
Company	Preamble
Company Board Recommendation	Section 3.4(b)
Company Board Recommendation Change	Section 6.2(a)
Company Bylaws	Section 3.1(b)
Company Charter	Section 3.1(b)
Company Disclosure Letter	Article III
Company Registered IP	Section 3.18(b)
Company SEC Documents	Section 3.6(a)
Company Shareholders	Section 2.1(a)
Company Senior Management	Section 4.9
Company Shares	Recitals
Delaware Federal Court	Section 8.10
Designated Courts	Section 8.10
D&O Insurance	Section 6.9(b)
Effects	Section 1.1(r)
Fairness Opinion	Section 3.22
Financing	Section 4.5(b)
Financing Commitment	Section 4.5(b)
Indebtedness	Section 5.2(h)
Indemnified Parties	Section 6.9(a)
Initial Director Resignations	Section 6.11
Jefferies	Section 3.21
Material Contract	Section 3.15(a)
Maximum Annual Premium	Section 6.9(b)
Minimum Condition	Section 2.1(a)
Offer	Recitals
Offer Closing	Section 2.1(d)
Offer Documents	Section 2.1(f)
Offer Price	Recitals
Offer to Purchase	Section 2.1(f)

Parent	Preamble
Pre-Closing Period	Section 6.4
Products	Section 3.10(b)
Purchaser	Preamble
Purchaser Disclosure Letter	Article IV
Permits	Section 3.10(a)
Representatives	Section 6.1(a)
Required Company Shareholder Vote	Section 3.22
Schedule 14D-9	Section 2.2(a)
Schedule TO	Section 2.1(f)
Specified Acquisition Transaction	Section 7.3(b)(ii)
Specified Definitive Acquisition Agreement	Section 7.1(d)(ii)
Strategic Investors	Section 3.2
Takeover Laws	Section 3.19
Termination Date	Section 7.1
Termination Fee	Section 7.3(b)(iii)

Section 1.3 Certain Interpretations.

(a) Unless otherwise indicated, all references herein to Sections, Articles, Annexes, Exhibits or Schedules, shall be deemed to refer to Sections, Articles, Annexes, Exhibits or Schedules of or to this Agreement, as applicable.

(b) Unless otherwise indicated, the words “include,” “includes” and “including,” when used herein, shall be deemed in each case to be followed by the words “without limitation.” The words “hereof,” “herein” and “hereunder” and words of like import used in this Agreement, unless otherwise stated, shall refer to this Agreement as a whole and not to any particular provision of this Agreement.

(c) The table of contents and headings set forth in this Agreement are for convenience of reference purposes only and shall not affect or be deemed to affect in any way the meaning or interpretation of this Agreement or any term or provision hereof.

(d) When reference is made herein to a Person, such reference shall be deemed to include all direct and indirect Subsidiaries of such Person unless otherwise indicated or the context otherwise requires.

(e) Unless otherwise indicated, all references herein to the Subsidiaries of a Person shall be deemed to include all direct and indirect Subsidiaries of such Person unless otherwise indicated or the context otherwise requires.

(f) The parties hereto agree that they have been represented by counsel during the negotiation and execution of this Agreement and, therefore, waive the application of any Law, regulation, holding or rule of construction providing that ambiguities in an agreement or other document will be construed against the party drafting such agreement or document.

(g) For purposes of this Agreement, whenever the context requires: the singular number shall include the plural, and vice versa; the masculine gender shall include the feminine and neuter genders; the feminine gender shall include the masculine and neuter genders; and the neuter gender shall include masculine and feminine genders.

(h) References to “\$” or “dollars” refer to United States dollars unless otherwise noted.

(i) References to “Made Available” shall mean that such documents or information referenced: (i) were delivered to Parent or its Representatives prior to the execution and delivery of this Agreement; (ii) were contained in the Company’s electronic data room maintained by Merrill Corporation prior to the execution and delivery of this Agreement; or (iii) were publicly available on the EDGAR website all times during the forty-eight (48) hours prior to the Agreement Date.

(j) References to “independent contractors” or “consultants” shall not include Persons which provide material services to two or more entities and are in the business of regularly providing such services to multiple Persons at the same time.

ARTICLE II

THE OFFER

Section 2.1 The Offer.

(a) Terms of Offer; Conditions to Offer. Provided that this Agreement shall not have been terminated pursuant to Article VII hereof, as promptly as reasonably practicable and, in any event, within fifteen (15) Business Days after the Agreement Date, Purchaser shall commence (within the meaning of Rule 14d-2 under the Exchange Act) the Offer to purchase all of the Company Shares at a price per Company Share, subject to the terms of Section 2.1(b) hereof, equal to the Offer Price. The Offer will be a single offer made to all holders of Company Shares. The obligation of Purchaser to accept for payment and to pay for any Company Shares tendered shall be subject only to (i) the condition (the “**Minimum Condition**”) that, prior to the then scheduled expiration date of the Offer (as it may be extended from time to time pursuant to Section 2.1(c) hereof), there be validly tendered in accordance with the terms of the Offer (not including as tendered Company Shares tendered pursuant to guaranteed delivery procedures and not actually delivered in settlement or satisfaction of such guarantee prior to the expiration of the Offer) and not withdrawn a number of Company Shares that, together with the Company Shares then owned by Parent or Purchaser or any of their respective Subsidiaries (if any), represents at least sixty-six and two thirds percent (66 2/3%) of the Adjusted Outstanding Share Number, and (ii) the other conditions set forth in Annex A hereto. For purposes of this Agreement, the “**Adjusted Outstanding Share Number**” shall be the sum of: (A) the aggregate number of outstanding Company Shares immediately prior to the Acceptance Time; plus (B) the aggregate number of Company Shares issuable upon the conversion, exchange, settlement or exercise, as applicable, of all options and other rights to acquire, or securities convertible into or exchangeable for, Company Shares that are outstanding immediately prior to the Acceptance Time in the case of each of clauses (A) and (B), without duplication for shares of Company Common Stock underlying Company ADSs. Parent and Purchaser each expressly reserves the right to increase the Offer Price or to make any other changes in the terms and conditions of the Offer; provided, however, that unless otherwise provided by this Agreement or previously approved by the

Company in writing, neither Parent nor Purchaser shall make any change to the terms and conditions of the Offer that (A) decreases the Offer Price, (B) changes the form of consideration to be paid in the Offer, (C) reduces the number of Company Shares sought to be purchased in the Offer, (D) imposes conditions to the Offer in addition to the conditions to the Offer set forth in Annex A hereto, (E) amends, modifies or supplements the conditions to the Offer set forth in Annex A hereto in any manner adverse to the holders of Company Shares, (F) extends the Offer in any manner other than pursuant to and in accordance with the terms of Section 2.1(c) or Section 2.1(e) hereof, (G) amends or waives the Minimum Condition (other than a Permitted Minimum Condition Modification), or (H) amends any of the terms of the Offer in any manner adverse to the holders of Company Shares (collectively, the “**Company Shareholders**”). The conditions to the Offer set forth in Annex A hereto are for the sole benefit of Parent and Purchaser and may be waived by Parent or Purchaser, in whole or in part, at any time and from time to time, in its sole discretion, other than the Minimum Condition, which may be waived by Parent or Purchaser only with the prior written consent of the Company. The failure by Parent or Purchaser at any time to exercise any of the foregoing rights shall not be deemed a waiver of any such right, and each such right shall be deemed an ongoing right that may be asserted at any time and from time to time. For purposes of this Agreement, a “**Permitted Minimum Condition Modification**” shall mean a modification to the definition of Minimum Condition, which may be effected at Parent’s election and in Parent’s sole discretion by delivering written notice of such modification to the Company prior to the Acceptance Time, to replace the reference to “sixty-six and two thirds percent (66 2/3%) of the Adjusted Outstanding Share Number” in such definition with “a majority of the aggregate number of outstanding Company Shares immediately prior to the Acceptance Time (without duplication for shares of Company Common Stock underlying Company ADSs)” (for the avoidance of doubt, effecting a Permitted Minimum Condition Modification shall not constitute a waiver of the Minimum Condition for purposes of this Agreement).

(b) Adjustments to Offer Price. The Offer Price shall be adjusted appropriately to reflect the effect of any stock split, reverse stock split, stock dividend (including any dividend or distribution of securities convertible into Company Shares), reorganization, recapitalization, reclassification, combination, exchange of shares or other like change with respect to Company Shares occurring on or after the Agreement Date and prior to Purchaser’s acceptance for payment of, and payment for, Company Shares pursuant to the Offer.

(c) Extension and Expiration of Offer. Subject to the terms and conditions of this Agreement and the Offer, the Offer shall initially be scheduled to expire at midnight, New York Time, on the date that is twenty (20) Business Days (for this purpose calculated in accordance with Section 14d-1(g)(3) under the Exchange Act) after the date the Offer is commenced (within the meaning of Rule 14d-2 under the Exchange Act); provided, however, that notwithstanding the foregoing or anything to the contrary set forth in this Agreement, (i) Purchaser shall extend the Offer for any period required by any rule, regulation, interpretation or position of the SEC or its staff or NASDAQ that is applicable to the Offer and (ii) in the event that any of the conditions to the Offer set forth on Annex A hereto are not satisfied or waived as of any then scheduled expiration date of the Offer, Purchaser shall extend the Offer for successive extension periods of up to twenty (20) Business Days each (with Purchaser determining the length of any such extension) (or such longer period as the parties hereto may agree) in order to permit the satisfaction of each such condition to the Offer; provided, however, that notwithstanding the foregoing clause (ii) of this Section 2.1(c), in no event shall Purchaser be required to extend the Offer beyond the Termination Date or the Extended Termination Date, as applicable; and provided further, that the foregoing clause (i) or clause (ii) of this Section 2.1(c) shall not be deemed to impair, limit or otherwise restrict in any manner the right of Parent or the Company to terminate this

Agreement pursuant to Article VII hereof. Purchaser shall not terminate the Offer prior to the expiration date without the prior written consent of the Company, except in the event that this Agreement is terminated pursuant to Article VII hereof.

(d) Payment for Company Shares. On the terms set forth in this Agreement and subject to the conditions of Annex A, Purchaser shall accept for payment, and pay for, all Company Shares validly tendered and not withdrawn pursuant to the Offer, promptly after the applicable expiration date of the Offer (such date and time of payment, as it may be extended in accordance with Section 2.1(c) hereof, the “**Offer Closing**”); provided, however, that with regard to Company Shares tendered pursuant to guaranteed delivery procedures that have not yet been delivered in settlement or satisfaction of such guarantee, Purchaser shall be under no obligation to make any payment for such shares unless and until such shares are delivered in settlement or satisfaction of such guarantee. The Offer Price payable in respect of each Company Share validly tendered and not withdrawn pursuant to the Offer shall be paid net to the holder thereof in cash, without interest thereon, subject to reduction only for any applicable withholding Taxes or other Taxes payable by such holder.

(e) Subsequent Offering Period. Purchaser shall, and the Offer Documents shall reserve the right to, extend the Offer for a subsequent offering period (within the meaning of Rule 14d-11 under the Exchange Act) of not less than three (3) nor more than twenty (20) Business Days immediately following the expiration of the Offer; provided, however, that Purchaser shall be entitled, in its discretion and without the consent of the Company or any other Person, to provide for one or more extensions of such subsequent offering period. Subject to the terms and conditions of this Agreement and the Offer, Purchaser shall accept for payment, and pay for, all Company Shares validly tendered pursuant to the Offer as so extended by such subsequent offering period, promptly after any such Company Shares are tendered during such subsequent offering period; provided, however, that with regard to Company Shares tendered pursuant to guaranteed delivery procedures that have not yet been delivered in settlement or satisfaction of such guarantee, Purchaser shall be under no obligation to make any payment for such shares unless and until such shares are delivered in settlement or satisfaction of such guarantee. The Offer Price payable in respect of each Company Share validly tendered pursuant to the Offer, as so extended by such subsequent offering period, shall be paid net to the holder thereof in cash, subject to reduction only for any applicable withholding Taxes or other Taxes payable by such holder.

(f) Schedule TO; Offer Documents. Concurrently with the commencement of the Offer (within the meaning of Rule 14d-2 under the Exchange Act), Parent and Purchaser shall (i) prepare and file with the SEC a Tender Offer Statement on Schedule TO (together with all amendments and supplements thereto, and including all exhibits thereto, the “**Schedule TO**”) with respect to the Offer, which shall contain as an exhibit or incorporate by reference an Offer to Purchase, or portions thereof (the “**Offer to Purchase**”), and forms of the letters of transmittal and summary advertisement, if any, in respect of the Offer, and such other ancillary documents and instruments that are required to be filed in connection with the filing of the Schedule TO (collectively and together with any supplements or amendments thereto, the “**Offer Documents**”) and (ii) cause the Offer Documents to be disseminated to all Company Shareholders as and to the extent required by the Exchange Act or other applicable Law. The Company shall promptly furnish to Parent and Purchaser in writing all information concerning the Company that may be required by applicable securities Laws or reasonably requested by Parent for inclusion in the Schedule TO or the Offer Documents. Subject to Section 6.2 hereof, the Company agrees, to the inclusion of the Company Board Recommendation in the Offer Documents. Parent and

Purchaser shall use reasonable efforts to cause the Schedule TO and the Offer Documents to comply in all material respects with the Exchange Act and all other applicable Law. Each of Parent, Purchaser and the Company shall use reasonable efforts to promptly correct any information provided by it for use in the Schedule TO or the Offer Documents if and to the extent that such information shall have become false or misleading in any material respect. Parent and Purchaser shall take all steps necessary to cause the Schedule TO and the Offer Documents, as so corrected, to be filed with the SEC and the other Offer Documents, as so corrected, to be disseminated to the Company Shareholders, in each case, as and to the extent required by applicable federal securities Laws and all other applicable Law. Prior to filing the Schedule TO or Offer Documents with the SEC or dissemination thereof to the Company Shareholders, or responding to any comments of the SEC or its staff with respect to the Schedule TO or Offer Documents, Parent and Purchaser shall provide the Company and its counsel a reasonable opportunity to review and comment on the Schedule TO, Offer Documents or response and shall give reasonable consideration to any comments reasonably proposed by the Company in a timely manner. Parent and Purchaser shall promptly provide the Company and its counsel with a copy or a description of any comments received by Parent, Purchaser or their counsel from the SEC or its staff with respect to the Offer Documents, and Parent shall respond promptly to any such comments.

(g) Termination or Withdrawal of Offer. If (i) at any then-scheduled expiration of the Offer, (A) the conditions to the Offer set forth in Annex A hereto (other than the Minimum Condition) shall have been satisfied or waived, (B) the Minimum Condition shall not have been satisfied and (C) no further extensions or re-extensions of the Offer are permitted or required pursuant to Section 2.1(c) hereof or (ii) this Agreement is terminated pursuant to Article VII hereof, the Offer shall be deemed terminated and Purchaser shall promptly return, and shall cause any depository, acting on behalf of Purchaser to return, all tendered shares to the registered holders thereof. Notwithstanding anything to the contrary herein, if this Agreement is terminated pursuant to the terms of Article VII hereof, Purchaser shall promptly (and in any event within one (1) Business Day after the then-scheduled expiration date of the Offer) irrevocably and unconditionally terminate the Offer.

(h) Withholding. Notwithstanding anything to the contrary contained in this Agreement, each of Parent and Purchaser shall be entitled to deduct and withhold from any consideration payable pursuant to this Agreement to any security holder or former security holder of the Company (including any holder or former holder of Company ADSs) such amounts as are required to be deducted or withheld therefrom under any provision of applicable Tax Law. To the extent such amounts are so deducted or withheld, such amounts shall be treated for all purposes under this Agreement as having been paid to the Person to whom such amounts would otherwise have been paid.

Section 2.2 Company Actions.

(a) Schedule 14D-9. The Company shall (i) file with the SEC, on or promptly after the date of filing by Purchaser of the Schedule TO, a Solicitation/Recommendation Statement on Schedule 14D-9 (together with all amendments and supplements thereto, and including all exhibits thereto, the "Schedule 14D-9") which shall, subject to Section 6.2 hereof, describe and make the Company Board Recommendation with respect to the Offer and (ii) cause the Schedule 14D-9 to be disseminated to the Company Shareholders as and to the extent required by the Exchange Act or other applicable Law within ten (10) Business Days after the commencement of the Offer (within the meaning of Rule 14d-2 under the Exchange Act). To the extent requested by the Company, Parent shall cause the Schedule 14D-9 to be mailed or otherwise disseminated to the holders of Company Shares together with

the Offer Documents disseminated to the holders of Company Shares. Parent and Purchaser shall promptly furnish to the Company in writing all information concerning Parent or Purchaser that may be required by applicable securities Laws or reasonably requested by the Company for inclusion in the Schedule 14D-9. The Company shall use reasonable efforts to cause the Schedule 14D-9 to comply in all material respects with the Exchange Act and all other applicable Law. Each of the Company, Parent and Purchaser shall use reasonable efforts to promptly correct any information provided by it for use in the Schedule 14D-9 if and to the extent that such information shall have become false or misleading in any material respect. The Company shall take all steps necessary to cause the Schedule 14D-9, as so corrected, to be filed with the SEC and disseminated to the Company Shareholders, in each case as and to the extent required by applicable United States federal securities Laws and all other applicable Law. Prior to filing the Schedule 14D-9 with the SEC or dissemination thereof to the Company Shareholders, or responding to any comments of the SEC or its staff with respect to the Schedule 14D-9, the Company shall provide Parent and its counsel a reasonable opportunity to review and comment on the Schedule 14D-9 or response and shall give reasonable consideration to any comments reasonably proposed by Parent in a timely manner. The Company shall promptly provide Parent and its counsel with a copy or a description of any comments received by the Company or its counsel from the SEC or its staff with respect to the Schedule 14D-9, and the Company shall respond promptly to any such comments.

(b) Company Information. In connection with the Offer, the Company shall, or shall cause its transfer agent to, promptly following a request by Purchaser, furnish Parent and Purchaser with such information (to the extent reasonably available to the Company), including (i) a list of the Company Shareholders, mailing labels and any available listing or computer files containing the names and addresses of all record holders of Company Shares, lists of non-objecting beneficial owners of Company Shares and lists of security positions of Company Shares held in stock depositories, in each case as of the most recent practicable date, and (ii) such additional information (including updated lists of Company Shareholders, non-objecting beneficial owners, mailing labels, listings or files of securities positions), and with such assistance as Parent or its agents may reasonably request and as is reasonably available to the Company in order to disseminate and otherwise communicate the Offer to the record holders of Company Shares. Subject to any and all applicable Law, and except for such steps as are necessary to disseminate the Offer Documents, Parent, Purchaser and their respective Representatives and agents shall (i) hold in confidence the information contained in any such lists of Company Shareholders, mailing labels and listings or files of securities positions on the terms and conditions set forth in the Confidentiality Agreement, (ii) use such information only in connection with the Offer and (iii) if (A) this Agreement shall be terminated pursuant to Article VII hereof or (B) Purchaser shall withdraw the Offer in accordance with the terms hereof or the Offer shall otherwise expire or terminate in accordance with the terms hereof without Purchaser having accepted for payment any Company Shares pursuant to the Offer, deliver (and shall use their respective reasonable efforts to cause their agents to deliver) to the Company, or destroy any and all copies and any extracts or summaries from such information then in their possession or control.

ARTICLE III

REPRESENTATIONS AND WARRANTIES OF THE COMPANY

Except (i) as disclosed in the Company SEC Documents filed with, or furnished to, the SEC prior to the date of this Agreement and publicly available on EDGAR (excluding any disclosures set forth in any section of any Company SEC Document entitled "Risk Factors" or "Forward-Looking

Statements” or any other disclosures included in such document that are general cautionary, predictive or forward-looking in nature), or (ii) as set forth and/or qualified in the disclosure letter delivered by the Company to Parent and Purchaser immediately prior to the execution of this Agreement (the “**Company Disclosure Letter**”), the Company represents and warrants to Parent and Purchaser as follows:

Section 3.1 Organization, Standing and Power.

(a) The Company and each of its Subsidiaries (i) is an entity duly organized, validly existing and in good standing (with respect to jurisdictions that recognize such concept) under the Laws of its jurisdiction of incorporation, (ii) has all requisite corporate or similar power and authority and all necessary governmental approvals to own, lease and operate its properties and assets and to conduct its business as presently conducted, and (iii) is duly qualified or licensed to do business as a corporation and is in good standing (with respect to jurisdictions that recognize such concept) in each jurisdiction in which the nature of its business or the ownership, leasing or operation of its properties makes such qualification or licensing necessary, except as, individually or in the aggregate, has not had and would not reasonably be expected to have a Company Material Adverse Effect.

(b) The copies of the Company’s articles of association (the “**Company Charter**”) and the Company’s bylaws (*Statuto*) (the “**Company Bylaws**”) that are filed with the Italian Register of Enterprises (unofficial English translations of which are set forth on Section 3.1(b) of the Company Disclosure Letter) are complete and correct copies thereof as in effect on the Agreement Date. The Company is not in violation of any of the provisions of the Company Charter or Company Bylaws.

Section 3.2 Capital Stock.

(a) The authorized capital stock of the Company consists of 19,656,317 shares of Company Common Stock. As of the close of business on December 16, 2013, (i) 15,555,131 shares of Company Common Stock (excluding treasury shares) were issued and outstanding, of which 10,984,130 shares of Company Common Stock are represented by an equal amount of Company ADSs, (ii) there were no shares of Company Common Stock held by the Company in its treasury and (iii) 4,101,186 shares of Company Common Stock were reserved for issuance pursuant to Company Equity Plans (of which 806,645 shares were subject to outstanding Company Options under the Company’s 2004 Equity Incentive Plan (including the Company’s 2004 Italy Stock Award Sub Plan), as amended and restated and of which 1,387,849 shares were subject to outstanding Company Options under the Company’s 2007 Stock Option Plan, as amended).

(b) All the outstanding shares of capital stock of the Company are, and all shares reserved for issuance as noted in clause (iii) above will be, when issued in accordance with the terms thereof, duly authorized, validly issued and fully paid. No shares of capital stock of the Company are owned by any Subsidiary of the Company. All the outstanding shares of capital stock or other voting securities or equity interests of each Subsidiary of the Company have been duly authorized and validly issued and are fully paid. All of the shares of capital stock or other voting securities or equity interests of each such Subsidiary are owned, directly or indirectly, by the Company, free and clear of all Liens. Neither the Company nor any of its Subsidiaries has outstanding any bonds, debentures, notes or other obligations having the right to vote (or convertible into, or exchangeable or exercisable for, securities having the right to vote) with Company Shareholders or shareholders of such Subsidiary on any matter. There are no outstanding (i) shares of capital stock or other voting securities or equity

interests of the Company, (ii) securities of the Company or any of its Subsidiaries convertible into or exchangeable or exercisable for shares of capital stock of the Company or other voting securities or equity interests of the Company or any of its Subsidiaries, (iii) stock appreciation rights, “phantom” stock rights, performance units, interests in or rights to the ownership or earnings of the Company or any of its Subsidiaries or other equity equivalent or equity-based award or right, (iv) subscriptions, options, warrants, calls, commitments, Contracts or other rights to acquire from the Company or any of its Subsidiaries, or obligations of the Company or any of its Subsidiaries to issue, any shares of capital stock of the Company or any of its Subsidiaries, voting securities, equity interests or securities convertible into or exchangeable or exercisable for capital stock or other voting securities or equity interests of the Company or any of its Subsidiaries or rights or interests described in clause (iii), other than Company Options pursuant to the Company Equity Plans and Company Shares pursuant to the pre-emptive rights accorded to the Company’s shareholders pursuant to the Italian Civil Code, or (v) obligations of the Company or any of its Subsidiaries to repurchase, redeem or otherwise acquire any such securities.

(c) There are no shareholder agreements, voting trusts or other agreements or understandings to which the Company or any of its Subsidiaries is a party or on file with the Company with respect to the holding, voting, registration, redemption, repurchase or disposition of, or that restricts the transfer of, any capital stock or other equity interest of the Company or any of its Subsidiaries.

(d) By means of a resolution of the Company shareholders’ meeting dated September 30, 2004, the capital of the Company was increased in cash, on a separable basis, up to an amount equal to Euro 1,560,000 (one million five hundred and sixty thousand), divided into 1,560,000 (one million five hundred and sixty thousand) shares, to be issued in one or more installments and to be subscribed, within September 30, 2019, only by employees and operating consultants of the Company and/or its controlling/controlled companies, (including the current Chairman of the Company Board and managers) on the terms and conditions provided in the same resolution; as of December 16, 2013, such amount was reduced to Euro 1,122,479 (one million one hundred twenty two thousand four hundred seventy nine) as a consequence of the subscription of 245,905 (two hundred forty five thousand nine hundred and five) shares.

(e) By means of a resolution of the extraordinary shareholders’ meeting dated April 27, 2007, the capital of the Company was increased, with exclusion of the pre-emptive right pursuant to article 2441, fifth paragraph of the Italian Civil Code, in cash up to an amount equal to Euro 1,000,000 (one million) divided into 1,000,000 (one million) shares, reserved to the exercise of the available options to be granted under the Company’s 2007 Stock Option Plan, on the terms and conditions provided by the above said resolution of the Shareholders’ meeting; as of December 16, 2013, such amount was reduced to Euro 659,700 (six hundred fifty nine thousand and seven hundred) as a consequence of the subscription of 340,300 (three hundred forty thousand three hundred) shares.

(f) By means of a resolution of the extraordinary shareholders’ meeting dated May 9, 2011, the capital of the Company was increased, with exclusion of the pre-emptive right pursuant to article 2441, fifth paragraph of the Italian Civil Code, in cash up to an amount equal to Euro 2,200,000 (two million two hundred thousand) divided into 2,200,000 (two million two hundred thousand) shares, reserved to the exercise of the available options to be granted under the Company’s 2007 Stock Option Plan, on the terms and conditions provided by the above said resolution of the shareholders’ meeting; as of December 16, 2013, such amount was reduced to Euro 2,127,391 (two million one hundred twenty seven thousand three hundred ninety one) as a consequence of the subscription of 72,609 (seventy two thousand six hundred and nine) shares.

(g) The Company Board has the power, pursuant to article 2443 of the Italian Civil Code, within and not later than June 30, 2014, to increase the capital of the Company in cash, up to an amount equal to Euro 100,000,000 (including the possible share premium) on a separable basis, in one or more transactions, for a rights offering, (i) with the faculty to reserve all or part of such amount to the exercise of warrants issued by means of the same resolution of the Company Board providing for the relevant capital increase, (ii) with the faculty to reserve one quarter (1/4) of any such capital increase to employees under the Company's equity incentive plans in force from time to time; and (iii) to determine the modalities, terms and conditions of any such capital increase (including the measure of the possible share premium) within the limits and the context of the resolution taken by the shareholders' meeting.

(h) The Company's directors shall have the power, pursuant to articles 2443 and 2420-ter of the Italian Civil Code, for a five-year term starting from the date of the resolution of the extraordinary Shareholders' meeting dated May 9, 2011, to:

(i) increase the capital of the Company in cash up to Euro 90,000,000 (ninety million), in one or more transactions, divided into 90,000,000 (ninety million) shares, with the faculty to reserve all or part of such amount to the exercise of warrants issued by means of the same resolution of the Company Board providing for the relevant capital increase; as of December 16, 2013, such amount was reduced to Euro 85,450,565 (eight five million four hundred fifty thousand five hundred sixty five) as a consequence of the subscription of 4,549,435 (four million five hundred forty nine thousand four hundred thirty five) shares;

(ii) issue convertible bonds, (even subordinated) and increase the capital of the Company, in one or more transactions, where appropriate or in the interest of the Company up to Euro 10,000,000 (ten million), through the relevant issuance of shares reserved for the conversion of such convertible bonds, it being understood that such convertible bonds may be reserved, in whole or in part, to the exercise of warrants issued by means of the same resolution of the Company Board providing for issuance of the convertible bonds;

(iii) in each case, exclude or limit the option right of the shareholders if the Company Board determines that exclusion or limitation to be in the interest of the Company.

In particular, the Company Board may limit or exclude the option right of the shareholders, pursuant to article 2441, fifth paragraph, of the Italian Civil Code, for such reasons as the Company Board determines to be in the interest of the Company, including to reserving the capital increase to (i) strategic investors, such as biotechnology companies, pharmaceutical companies, healthcare companies, investment funds and private equity funds specializing in the biotechnology, pharmaceutical and/or healthcare industries, and other entities or individuals having a portfolio that includes stakes in biotechnology, pharmaceutical or healthcare companies and/or (ii) entities or individuals that are accredited investors or qualified institutional buyers under U.S. law (collectively, the "**Strategic Investors**"). The option right, in case of issuance of convertible bonds and/or warrants, may be excluded and/or limited in the event such financial instruments are granted to Strategic Investors and/or current owners of convertible bonds and/or warrants of the Company.

Section 3.3 Subsidiaries. Section 3.3 of the Company Disclosure Letter sets forth a true and complete list of each Subsidiary of the Company, including its jurisdiction of incorporation or formation, officers and directors. Except for the capital stock of, or other equity or voting interests in, its Subsidiaries, the Company does not own, directly or indirectly, any equity, membership interest, partnership interest, joint venture interest, or other equity or voting interest in, or any interest convertible into, exercisable or exchangeable for any of the foregoing, nor, other than in the ordinary course of business with respect to its wholly owned Subsidiaries, is the Company under any current or prospective obligation to provide funds to, make any loan, capital contribution, guarantee, credit enhancement or other investment in, or assume any liability or obligation of, any Person.

Section 3.4 Authority.

(a) The Company has all necessary corporate power and authority to execute and deliver this Agreement, to perform its obligations hereunder and to consummate the transactions contemplated hereby. The execution, delivery and performance of this Agreement by the Company and the consummation by the Company of the transactions contemplated hereby, including the Offer, have been duly authorized by all necessary corporate action on the part of the Company and no other corporate proceedings on the part of the Company are necessary to approve this Agreement or to consummate the transactions contemplated hereby. This Agreement has been duly executed and delivered by the Company and, assuming the due authorization, execution and delivery by Parent and Purchaser, constitutes a valid and binding obligation of the Company, enforceable against the Company in accordance with its terms, except as enforcement thereof may be limited against the Company by (i) bankruptcy, insolvency, reorganization, moratorium and similar Laws affecting the enforcement of creditors' rights or remedies in general as from time to time in effect or (ii) the exercise by courts of equity powers.

(b) The Company Board, at a meeting duly called and held at which all directors of the Company were present, and by the unanimous vote of all directors of the Company, other than one director that has deemed himself conflicted, duly adopted resolutions: (i) determining that the terms of this Agreement, and the transactions contemplated hereby are fair to and in the best interests of the Company Shareholders and other Company stakeholders; (ii) approving and declaring advisable this Agreement and the transactions contemplated hereby; (iii) resolving to recommend that the holders of Company Shares accept the Offer and tender their Company Shares to Purchaser pursuant to the Offer (the recommendation of the Company Board, adopted by the unanimous vote of all directors of the Company, other than one director that has deemed himself conflicted, that the holders of Company Shares accept the Offer and tender their Company Shares to Purchaser pursuant to the Offer being referred to as the "**Company Board Recommendation**"), which resolutions have not been subsequently rescinded, modified or withdrawn in any way, except as may be permitted by Section 6.2 hereof.

Section 3.5 No Conflict; Consents and Approvals.

(a) The execution, delivery and performance of this Agreement by the Company does not, and the consummation of the Offer and the other transactions contemplated hereby (including the purchase of the Company Shares tendered in the Offer) and compliance by the Company

with the provisions hereof will not (i) violate or conflict with the Company Charter or Company Bylaws or the charter or bylaws of any of the Company's Subsidiaries, (ii) result in any breach of, or default (with or without notice or lapse of time, or both) under, or give rise to a right of, or result in, termination, cancellation, modification or acceleration of any obligation or to the loss of a benefit under, or result in the creation of any Lien in or upon any of the properties, assets or rights of the Company or any of its Subsidiaries under, or require any consent, waiver or approval of any Person pursuant to, any provision of any Material Contract, (iii) result in the revocation, invalidation or termination of any material Permit; or (iv) subject to the governmental filings and other matters referred to in Section 3.5(b) hereof, materially violate or conflict with any Law or any rule or regulation of NASDAQ applicable to the Company or any of its Subsidiaries or by which the Company or any of its Subsidiaries or any of their respective properties or assets may be bound.

(b) No consent, approval, order or authorization of, or registration, declaration, filing with or notice to any Governmental Entity is required by or with respect to the Company or any of its Subsidiaries in connection with the execution, delivery and performance of this Agreement by the Company or the consummation of the Offer or compliance with the provisions hereof, except for (i) compliance with any applicable requirements of the Antitrust Laws, (ii) such filings and reports as required pursuant to the applicable requirements of the Securities Act, the Exchange Act the rules and regulations promulgated thereunder, and state securities Laws or "blue sky" Laws, (iii) any filings required under the rules and regulations of NASDAQ and (iv) such other filings the failure of which to be obtained or made, individually or in the aggregate, have not had and would not reasonably be expected to have a Company Material Adverse Effect or would not prevent, materially delay or materially impede the consummation of the Offer.

Section 3.6 Company SEC Documents; Financial Statements.

(a) The Company has filed or furnished (as applicable) on a timely basis with the SEC all forms, reports, schedules, statements and other documents required to be filed or furnished (as applicable) by the Company with the SEC under applicable Laws prior to the Agreement Date since January 1, 2011 (all such filed or furnished documents, together with all exhibits and schedules thereto and all information incorporated therein by reference, the "**Company SEC Documents**"). As of their respective filing dates (and, in the case of registration statements, as of the dates of effectiveness), or, if amended or superseded by a filing prior to the Agreement Date, on the date of the last such amendment or superseding filing prior to the Agreement Date, the Company SEC Documents complied in all material respects with the applicable requirements of the Securities Act and the Exchange Act, as the case may be, including, in each case, the rules and regulations promulgated thereunder, and none of the Company SEC Documents at the time they were filed, or, if amended or superseded by a filing prior to the Agreement Date, on the date of the last such amendment or superseding filing prior to the Agreement Date, contained any untrue statement of a material fact or omitted to state a material fact required to be stated therein or necessary in order to make the statements therein, in light of the circumstances under which they were made, not misleading. None of the Company's Subsidiaries is required to file any forms, reports or other documents with the SEC. No current or former executive officer of the Company has failed to make the certifications required of him or her under Rule 13a-14 or 15d-14 promulgated under the Exchange Act or Section 302 or 906 of the Sarbanes-Oxley Act with respect to any Company SEC Document since January 1, 2011, and such certifications are accurate and complete, and comply in all material respects as to form and content with all applicable Laws. The Company has Made Available to Parent or Purchaser true, correct and complete copies of all

correspondence, other than transmittal correspondence, between the SEC, on the one hand, and the Company, on the other, since January 1, 2011, including all SEC comment letter and responses to such comment letters and responses to such comment letters by or on behalf of the Company. To the Company's Knowledge, none of the Company SEC Documents is the subject of ongoing SEC review or outstanding SEC comment.

(b) The financial statements (including the related notes thereto) included (or incorporated by reference) in the Company SEC Documents comply as to form in all material respects with applicable accounting requirements and the published rules and regulations of the SEC with respect thereto, have been prepared in accordance with U.S. GAAP (except, in the case of unaudited statements, as may be permitted by the SEC) applied on a consistent basis during the periods involved (except as may be indicated in the notes thereto) and fairly present in all material respects the consolidated financial position of the Company and its consolidated Subsidiaries as of the dates thereof and their respective consolidated results of operations and cash flows for the periods indicated therein (subject, in the case of unaudited statements, to normal and year-end audit adjustments), all in accordance with U.S. GAAP and the applicable rules and regulations promulgated by the SEC. As of the Agreement Date, the Company does not intend to correct or restate, and to the Company's Knowledge there is not any basis to correct or restate, any of the audited consolidated financial statements or unaudited interim financial statements (including, in each case, the notes, if any, thereto) of the Company filed in or furnished with the Company SEC Documents.

(c) The Company and its Subsidiaries have implemented and maintain a system of "internal control over financial reporting" (as defined in Rules 13a-15(f) and 15d-15(f) of the Exchange Act) designed to provide reasonable assurances regarding the reliability of financial reporting and the preparation of financial statements in accordance with U.S. GAAP. The Company (i) has implemented and maintains "disclosure controls and procedures" (as defined in Rule 13a-15(e) of the Exchange Act) that are designed to ensure that material information relating to the Company, including its consolidated Subsidiaries, is made known to the Chief Executive Officer and the Chief Financial Officer of the Company by others within those entities and (ii) has disclosed, based on its most recent evaluation prior to the Agreement Date, to the Company's outside auditors and the audit committee of the Company Board (A) any significant deficiencies and material weaknesses in the design or operation of "internal control over financial reporting" that would be reasonably likely to adversely affect in any material way the Company's ability to record, process, summarize and report financial information and (B) any fraud, whether or not material, that involves management or other employees who have a significant role in the Company's "internal control over financial reporting." Any material change in internal control over financial reporting required to be disclosed in any Company SEC Document has been so disclosed.

(d) Neither the Company nor any of its Subsidiaries is a party to, or has entered into any Contract to become a party to, any joint venture, off-balance sheet partnership or any similar Contract or arrangement (including any Contract relating to any transaction or relationship between or among the Company or any of its Subsidiaries, on the one hand, and any unconsolidated Affiliate, including any structured finance, special purpose or limited purpose entity or person, on the other hand, or any "off-balance sheet arrangements" (as defined in Item 303(a) of Regulation S-K of the SEC), where the result, purpose or intended effect of such Contract is to avoid disclosure of any material transaction involving, or material liabilities of, the Company's or any of its Subsidiaries' in the Company's audited financial statements or other Company SEC Documents.

(e) The Company's and the Company's Subsidiary's financial statements from time to time approved by their respective corporate bodies were prepared in accordance with Italian GAAP and Swiss GAAP, respectively, in each case applied on a consistent basis and with the relevant books and records as at the date and for the period stated therein and present in all material respects the assets, liabilities and net worth of the Company and of the Company's Subsidiary (as the case may be) as at the reference date thereof, its revenues and expenses and the result of its operations for the period indicated.

Section 3.7 No Undisclosed Liabilities. Neither the Company nor any of its Subsidiaries has any liabilities or obligations of any nature, whether accrued, absolute, contingent or otherwise, that would be required to be recorded or reflected on a balance sheet under the respective GAAP, other than liabilities and obligations (a) accrued or reserved against in the consolidated balance sheet of the Company and its Subsidiaries as of June 30, 2013 included in the Company SEC Documents, in the Company's financial statements as of June 30, 2013 and/or in the Company's Subsidiary's financial statements as of June 30, 2013, (b) incurred in the ordinary course of business consistent with past practice since June 30, 2013, (c) that are executory performance obligations under any Contract to which the Company or any of its Subsidiaries is a party or bound and are not arising under or resulting from any breach or nonperformance of such Contract, or (d) under this Agreement or in connection with the transactions contemplated hereby (including legal and other advisors fees and expenses).

Section 3.8 Absence of Certain Changes or Events. Since June 30, 2013: (i) except for actions required to be taken by the terms of this Agreement or in connection with the transactions contemplated hereby (including legal and other advisors fees and expenses), the Company and its Subsidiaries have conducted their businesses in the ordinary course consistent with past practice in all material respects, (ii) no Company Material Adverse Effect has occurred, and there has not been, and there does not exist, any Effect that, individually or in the aggregate with other Effects, would reasonably be expected to have a Company Material Adverse Effect and (iii) neither the Company nor any of its Subsidiaries has suffered any material loss, damage, destruction or other casualty affecting any of its material properties or assets, whether or not covered by insurance. Since June 30, 2013 through the Agreement Date, except for actions required to be taken by the terms of this Agreement or in connection with the transactions contemplated hereby (including legal and other advisors fees and expenses):

(a) the Company has not (i) declared, accrued, set aside or paid any dividend or made any other distribution in respect of any shares of capital stock, or (ii) repurchased, redeemed or otherwise reacquired any shares of capital stock or other securities;

(b) the Company has not sold, issued or granted, or authorized the issuance of, (i) any capital stock or other security (except for Company Shares issued upon the valid exercise of outstanding Company Options), (ii) any option, warrant or right to acquire any capital stock or any other security (except for Company Options identified in Section 3.8(b) of the Company Disclosure Letter), or (iii) any instrument convertible into or exchangeable for any capital stock or other security;

(c) the Company has not effected or been a party to any merger, consolidation, share exchange, business combination, recapitalization, reclassification of shares, stock split, reverse stock split or similar transaction (other than the transactions contemplated by this Agreement);

(d) the Company has not made any capital expenditure which, when added to all other capital expenditures made on behalf of the Company since December 31, 2012, exceeds \$3,000,000 in the aggregate;

(e) the Company has not (i) acquired, leased or licensed any material right or other material asset from any other Person, (ii) sold or otherwise disposed of, or leased or licensed, any material right or other material asset to any other Person, or (iii) waived or relinquished any right, except, in each case, for rights or other assets acquired, leased, licensed or disposed of in the ordinary course of business and consistent with past practices;

(f) the Company has not made any pledge of any of its assets or otherwise permitted any of its assets to become subject to any Lien, except for pledges of immaterial assets made in the ordinary course of business and consistent with past practices;

(g) the Company has not (i) lent money to any Person (other than pursuant to reimbursement for expenses incurred in the ordinary course of business), or (ii) incurred or guaranteed any Indebtedness;

(h) the Company has not, unless it was required to do so by Law, outside the ordinary course of business consistent with past practice, unless required to do so by a collective bargaining agreement, or Contract with a works council or other labor organization, (i) adopted, established or entered into any Material Employee Benefit Plan, (ii) caused or permitted any Material Employee Benefit Plan to be amended in any Material respect or terminated, or (iii) paid any Material bonus or made any Material profit-sharing or similar payment to, or Materially increased the amount of the wages, salary, commissions, fringe benefits or other compensation or remuneration payable to, or granted any rights to receive severance, termination, retention or tax gross-up compensation or benefits to, any of its current or former directors, officers or employees whose base salary is greater than \$50,000;

(i) the Company has not made any material change to its methods of accounting or accounting practice, except as required by concurrent changes in GAAP;

(j) the Company has not made any material Tax election;

(k) the Company has not commenced or settled any Action; and

(l) the Company has not agreed or committed to take any of the actions referred to in clauses (a) through (k) above.

Section 3.9 Litigation. There is no Action pending or, to the Knowledge of the Company, threatened in writing against the Company or any of its Subsidiaries, any of their respective properties or assets, or any present or former officer or director of the Company or any of its Subsidiaries in such individual's capacity as such, other than as set forth on Section 3.9 of the Company Disclosure Letter. Neither the Company nor any of its Subsidiaries nor any of their respective properties or assets is subject to any outstanding judgment, order, injunction, rule or decree of any Governmental Entity. There is no Action pending or, to the Knowledge of the Company, threatened in writing seeking to impose any legal restraint on or prohibition against the Offer or that, if resolved adversely to the Company, would have the effect of preventing or materially delaying or making illegal, the Offer.

Section 3.10 Compliance with Laws and Regulations.

(a) Except with respect to Employee Benefit Plans, environmental matters and Taxes, which are the subject of Sections 3.11, 3.13 and 3.14, respectively, the Company and each of its Subsidiaries are and, at all times since January 1, 2011 have been, in compliance in all material respects with all Laws applicable to their businesses, operations, properties or assets, including Laws pertaining to the approval or marketing authorization, manufacture, processing, storage, testing, packaging, labeling, promotion or distribution of pharmaceuticals or medicinal products, antitrust, immigration, privacy and product safety and Legislative Decree 231/2001. The Company and each of its Subsidiaries have taken all required steps to ensure that all manufacturing and marketing authorizations and orphan designations granted by any Regulatory Authority or Governmental Entity to the Company or any of its Subsidiaries are currently valid and have not been suspended or withdrawn and, to the Knowledge of the Company, no Regulatory Authority or Governmental Entity has taken any steps that have resulted in, or may result in the suspension or withdrawal of any of these manufacturing and marketing authorizations or orphan designations. To the Knowledge of the Company, none of the Company or any of its Subsidiaries, has received, since January 1, 2011, a notice or other written communication alleging or relating to a possible material violation of any Law applicable to their businesses, operations, properties or assets. The Company and each of its Subsidiaries have in effect all material permits, licenses, variances, exemptions, authorizations, operating certificates, franchises, orders and approvals of all Governmental Entities and Regulatory Authorities (collectively, "**Permits**") necessary for them to own, lease or operate their properties and assets and to carry on their businesses and operations as now conducted, including the manufacture and marketing of medicinal products, and there has occurred no violation of, default (with or without notice or lapse of time or both) under or event giving to others any right of revocation, non-renewal, adverse modification or cancellation of, with or without notice or lapse of time or both, any such Permit, except for such lack of Permits, noncompliance, defaults, revocations, non-renewals, modifications or cancellations that, individually or in the aggregate, have not had and would not reasonably be expected to have, a Company Material Adverse Effect.

(b) The Company has Made Available to the Purchaser, with respect to Defibrotide as of the Agreement Date copies of (i) all Investigational New Drug applications, or Clinical Trial Applications submitted to the FDA or to any other Regulatory Authority, (ii) all Marketing Authorization Applications submitted to the EMA in accordance with Regulation 726/2004 and/or any national competent authorities in the European Union in accordance with Directive 2001/83/EC, (iii) where relevant, all documents required by Directive 93/42/EC to support CE-marking, (iv) all material supplements to and amendments of the items set forth in clauses (i) through (iii) and (v) all material written correspondence with, or notice, alert, or warning issued by, the FDA or any other Regulatory Authority with respect to Defibrotide, including communications between the Company and FDA regarding the acceptability for filing of a New Drug Application, and communications between the Company and FDA regarding clinical data or compliance with Good Clinical Practices.

(c) The manufacture of the Products by the Company and its Subsidiaries is being conducted in compliance with the applicable Laws regarding the manufacture of pharmaceuticals or medicinal products, including requirements set forth in 21 CFR Parts 210 and 211 and FDA guidances, Directive 2001/83/EC, Directive 2003/94/EC and similar applicable Laws, the European Commission's guidance in Volume 4 of the "Notice to Applicants," and EMA's guidance related to Good Manufacturing Practices, and the Company and its Subsidiaries are in compliance with all applicable Laws regarding registration and listing requirements, including those set forth in 21 USC 360 and 21

CFR Part 207, Directive 2001/83/EC and Directive 2003/94/EC and similar applicable Laws, except in each case where any non-compliance, individually or in the aggregate, has not had and would not reasonably be expected to have a Company Material Adverse Effect.

(d) All preclinical and clinical trials and all preclinical research, clinical development and manufacturing activities conducted by the Company and its Subsidiaries have been and are being conducted in compliance with all applicable Laws regarding such activities, including 21 CFR Parts 50, 54, 56, and 58, Directive 2001/20/EC, Commission Directive 2005/28/EC and similar applicable Laws, European Commission's guidance in Volume 10 of the "Notice to Applicants", the EMA's guidance related to Good Laboratory Practices or Good Clinical Practices, as applicable, and applicable Laws relating to protection of human subjects, and good laboratory practice requirements and standards, except in each case where any non-compliance, individually or in the aggregate, has not had and would not reasonably be expected to have a Company Material Adverse Effect.

(e) All required reports, including field notifications and alerts, adverse events, complaints, corrective or preventative actions, have been submitted by the Company or any of its Subsidiaries to the applicable Regulatory Authority to which such reports or related information must be submitted, in a manner and form that complies in all material respects with the applicable requirements of the Regulatory Authority.

(f) Other than non-material routine or periodic inspections or reviews, there are no pending, or to the Knowledge of the Company, threatened adverse inspections, including inspectional observations; findings of deficiency or non-compliance; warning letters or other regulatory letters or sanctions; compelled or voluntary recalls, field notifications or alerts; import alerts, holds, or detentions; or other compliance or enforcement action against the Company or any of its Subsidiaries. Neither the Company nor any of its Subsidiaries nor any of their respective directors, officers, employees, agents or subcontractors, has been convicted of any crime or engaged in any conduct that has resulted, or would be reasonably likely to result, in debarment or disqualification by any Regulatory Authority, including under 42 USC 1320a-7 or similar remedies under state Laws, or that is included in the United States Health and Human Services/Office of Inspector General List of Excluded Individuals Entities, the United States General Service Administration's List of Parties Excluded from Federal Programs, or the FDA Debarment List, and there are no proceedings pending or, to the Knowledge of the Company, threatened in writing that would be reasonably likely to result in criminal liability or debarment or disqualification by any Regulatory Authority.

Section 3.11 Benefit Plans.

(a) Section 3.11(a) of the Company Disclosure Letter sets forth a true and complete list of each Material Employee Benefit Plan as of the Agreement Date. The Company has Made Available to Parent correct and complete copies of all Material Employee Benefit Plans as of the Agreement Date. Neither the Company nor any of its Subsidiaries has made any plan or commitment, which plan or commitment is binding on the Company or its Subsidiaries, to establish any new Material Employee Benefit Plan or to modify any Material Employee Benefit Plan in any material respect (except as required by Law or to conform any such Employee Benefit Plan to the requirements of any applicable Law, in each case as previously disclosed to Parent or Purchaser in writing, or as required by this Agreement). Unless otherwise provided in the applicable Employee Benefit Plan, no binding contractual commitments, undertakings or representations have been made or given to any current or former

employees, directors, independent contractors, consultants or other persons engaged by the Company or any of its Subsidiaries regarding the continued operation, extension, material amendment or replacement of, or grants of awards or benefits under, any Employee Benefit Plan.

(b) As of the Agreement Date, the Company and each of its Subsidiaries has Made Available with respect to each Employee Benefit Plan, to the extent applicable, (i) correct and complete copies of all documents embodying each Material Employee Benefit Plan, including all amendments thereto and any related trust documents, (ii) if the Employee Benefit Plan is funded, the most recent annual and periodic accounting of its assets, (iii) copies of any tax rulings received by the Company or its Subsidiaries within the last three years in Italy, Switzerland or elsewhere for Company Options or Company Equity Plans, (iv) all Material written agreements and group contracts relating to each Material Employee Benefit Plan, including administrative service agreements and group or individual insurance contracts, (v) all correspondence received or sent by the Company or any of its Subsidiaries within the last three years to or from any Governmental Entity relating to any Employee Benefit Plan, (vi) all policies pertaining to fiduciary liability insurance covering the fiduciaries for each Employee Benefit Plan and (viii) any approvals, registrations or determinations issued by any Governmental Entity with respect to each Employee Benefit Plan within the last three years.

(c) Each Employee Benefit Plan complies in all material respects with applicable Law and has been maintained and operated in material compliance with the terms of such Employee Benefit Plan and applicable Law. There are no Material actions, suits or claims pending or, to the Knowledge of the Company, threatened or reasonably anticipated (other than routine claims for benefits) against any Employee Benefit Plan, the assets of any Employee Benefit Plan, or the plan sponsor, plan administrator or, to the Knowledge of the Company, any fiduciary of any Employee Benefit Plan relating to any Employee Benefit Plan. Each Employee Benefit Plan may be amended, terminated or otherwise discontinued after the Acceptance Time in accordance with its terms, without material liability to Parent, Purchaser, the Company, or any of their respective Subsidiaries or Affiliates (other than ordinary administration expenses or with respect to benefits, other than bonuses, commissions or amounts under other compensation plans, that were previously earned, vested or accrued under Employee Benefit Plans prior to the Acceptance Time), except as may be required by applicable Law. There are no audits, inquiries or proceedings pending or, to the Knowledge of the Company, threatened by any applicable Governmental Entity with respect to any Employee Benefit Plan. The Company and each of its Subsidiaries have timely made all contributions and other payments required by and due under the terms of each Employee Benefit Plan in accordance with applicable Law.

(d) No Employee Benefit Plan provides post-termination or retiree life insurance, health or other post-termination or retiree employee welfare benefits to any person for any reason, except (i) as may be required by applicable Law, or (ii) benefits under insured plans maintained by the Company or any of its Subsidiaries providing such benefits in the event an employee is disabled at the time of termination of the employee's employment with the Company or its Subsidiaries and the conversion privileges provided under such insured plans. The Company and its Subsidiaries have never maintained, sponsored, established, participated in or contributed to a defined benefit pension plan for its employees, except to the extent required by Law or by the applicable collective bargaining agreements.

(e) Unless otherwise contemplated by this Agreement, neither the execution or delivery of this Agreement nor the consummation of the transactions contemplated by this Agreement

will (either alone or upon the occurrence of any additional or subsequent event) (i) result in any material payment (including severance, bonus or otherwise) or benefit becoming due or payable, or required to be provided, to any current or former employee, director, independent contractor or consultant of the Company or any of its Subsidiaries, (ii) result in any forgiveness of material indebtedness, (iii) materially increase the amount or value of any benefit or compensation otherwise payable or required to be provided to any current or former employee, director, independent contractor or consultant of the Company or any of its Subsidiaries, or (iv) result in the acceleration or change of the time of payment, vesting or funding of any material benefit or material compensation.

(f) There is only one U.S.-based employee of the Company or its Subsidiaries, and there are no material financial, employment-related or other obligations to, or rights of, such employee under any Employee Benefit Plan or Employment Agreement.

Section 3.12 Employment Matters.

(a) Section 3.12(a) of the Company Disclosure Letter sets forth a true and complete list of all Employment Agreements and Consultant Agreements as of the Agreement Date. The Company has Made Available to Parent correct and complete copies of all Employment Agreements and Consultant Agreements as of the Agreement Date. Neither the Company nor any of its Subsidiaries has made any plan or commitment to enter into any new Employment Agreement or Consultant Agreement or to modify any existing Employment Agreement or Consultant Agreement (except as required by Law or to conform any such Employment Agreement or Consultant Agreement to the requirements of any applicable Law, in each case as previously disclosed to Parent or Purchaser in writing, or as required by this Agreement).

(b) To the Knowledge of the Company, as of the Agreement Date no employee, independent contractor or consultant intends to terminate his or her employment or services for any reason.

(c) The Company and its Subsidiaries are in compliance in all material respects with all applicable Laws, collective bargaining agreements and other Material agreements or arrangements with any employee, works council, employee representative or other labor organization or group of employees, extension orders and binding customs respecting labor and employment, including all Laws, its own policies, practices, handbooks, work rules and internal regulations relating to fair employment practices, terms and conditions of employment, discrimination, disability, fair labor standards, workers' compensation, wrongful discharge, immigration, occupational safety and health, family and medical leave, wage and hour (including overtime wages), worker classification, equal opportunity, pay equity, meal and rest periods, and employee terminations, and in each case, with respect to any employee (i) have withheld and reported and remitted all amounts required by Law or by agreement to be withheld and reported and remitted with respect to wages, salaries and other payments to employees in all material respects, (ii) are not liable for any: arrears of wages, compensation,

severance pay or any Taxes or any penalty for failure to comply with any of the foregoing, and (iii) are not liable for any payment to any trust or other fund governed by or maintained by or on behalf of any governmental authority, with respect to unemployment compensation benefits, social security or other benefits or obligations for employees (other than routine payments to be made in the normal course of business and consistent with past practice). The Company and its Subsidiaries are not a party to a conciliation agreement, consent decree or other agreement or order with any Governmental Entity with respect to employment practices. Neither the Company nor any of its Subsidiaries have current direct or indirect liability with respect to any misclassification of any person as an independent contractor or consultant rather than as an employee, or with respect to any employee leased from another employer or any employee currently or formerly classified as exempt from overtime wages or the equivalent under applicable Law.

(d) The Company and its Subsidiaries have properly paid all wages and salaries and are not liable for any penalties.

(e) Section 3.12(e) of the Company Disclosure Letter sets forth each collective bargaining or other similar agreement with any union, works council, employee representative or other labor organization or group of employees. As of the date of this Agreement, there is no labor dispute, strike, proceeding, work stoppage or lockout, or, to the Knowledge of the Company, threat thereof, by or with respect to any employees of the Company or any of its Subsidiaries, and there has been no such labor dispute, strike, work stoppage or lockout in the previous two (2) years. The Company has no Knowledge of any activities or proceedings of any labor union, works council, employee representative or other labor organization or group of employees to organize any employees. There are no actions, suits, claims, labor disputes or grievances pending or to the Knowledge of the Company, threatened relating to any labor matters involving any Company or Subsidiary employee, including charges of unfair labor practices or the equivalent under applicable Law. The Company and its Subsidiaries have not engaged in any unfair labor practices within the meaning of the laws of Italy, Switzerland or, to the Knowledge of the Company, other countries in which the Company or its Subsidiaries have employees.

(f) To the Knowledge of the Company, all of the employees and directors of the Company or its Subsidiaries are authorized and have appropriate documentation to work in the jurisdiction in which they are working. Further, to the Knowledge of the Company, to the extent that such knowledge is required by Law, all independent contractors or consultants engaged by the Company or its Subsidiaries are authorized and have appropriate documentation to work in the jurisdiction in which they are working.

Section 3.13 Environmental Matters. Except as set forth in Section 3.13 the Company Disclosure Letter: (a) no Hazardous Materials are present as a result of the actions of the Company, or, to the Knowledge of the Company, as a result of any actions of any third party or otherwise, in, on or under any property, including the land and the improvements, ground water and surface water thereof, that the Company has at any time owned, operated, occupied or leased or, to the Knowledge of the Company, on any surrounding site; and (b) the Company has not produced, used, transported, processed, manufactured, generated, treated, handled, stored or disposed of, released or exposed any employee or any third party to any Hazardous Materials, except in compliance with applicable Environmental Laws. Except as set forth in Section 3.13 of the Company Disclosure Letter: (a) the Company and its Subsidiaries are and have been at all times in material compliance with applicable Environmental Laws and the requirements of Permits required under such Environmental Laws for the operation of the

business of the Company, including with respect to any Company Property; and (b) the Company has not received written notice of, and is not a party to or the subject of any unresolved Action alleging any material liability or responsibility under or noncompliance with any Environmental Law or seeking to impose any financial responsibility for any investigation, cleanup, removal, containment or any other remediation or compliance under any Environmental Law and the Company is not subject to any Order or Contract by or with any Governmental Entity or third party imposing any material liability or obligation with respect to any of the foregoing.

Section 3.14 Taxes.

(a) The Company and each of its Subsidiaries have timely filed (taking into account any extensions) with the appropriate Governmental Entities all material Tax Returns that are required to have been filed and all such Tax Returns are correct and complete in all material respects. The Company and each of its Subsidiaries have timely paid all material Taxes required to have been paid by them, other than Taxes that are not yet due and payable or that are being contested in good faith by any appropriate Action (and for which adequate reserves have been established to the extent required by U.S. GAAP). No deficiency for any Tax has been asserted or assessed by a Governmental Entity against the Company or any of its Subsidiaries which deficiency has not been paid or is not being contested in good faith by any appropriate Action (and for which adequate reserves have been established to the extent required by U.S. GAAP).

(b) The Company has established, in the ordinary course of business and consistent with its past practices, reserves adequate for the payment of all Taxes for the period since that date. No material Taxes have been incurred since that date other than in the ordinary course of business of the Company and its Subsidiaries.

(c) No written claim has ever been made by a Governmental Entity in a jurisdiction where the Company or any Subsidiary does not file Tax Returns that the Company or any Subsidiary is or may be subject to taxation in that jurisdiction. There are no security interests or other Liens on any of the assets of the Company or any Subsidiary that arose in connection with any failure (or alleged failure) to pay any Tax, other than Liens for Taxes not yet due and payable or that are being contested in good faith by any appropriate Action and for which adequate reserves have been established to the extent required by U.S. GAAP.

(d) The Company and its Subsidiaries have timely withheld and paid to the appropriate Governmental Entity all material Taxes required to have been withheld and paid under applicable Tax Law, including withholdings with respect to amounts paid or owing to any employee, independent contractor, creditor, shareholder or other third party.

(e) No Tax Return of the Company or any Subsidiary is under audit or examination by any Governmental Entity, and no written (or, to the Knowledge of the Company, oral) notice of such an audit or examination has been received by the Company or any Subsidiary. No deficiencies for any Taxes have been proposed, asserted or assessed against the Company or any of its Subsidiaries, and no requests for waivers of the time to assess any such Taxes are pending. There are no outstanding waivers of any limitation periods or agreements providing for an extension of time for the filing of any Tax Return, the assessment or collection thereof by any relevant Governmental Entity or the payment of any Tax by the Company or any Subsidiary. No other procedure, proceeding or contest of any refund or deficiency in respect of Taxes is pending in or on appeal from any Governmental Entity.

(f) The Company has Made Available to Parent complete and accurate copies of all Tax Returns filed by the Company or any of its Subsidiaries on or prior to the Agreement Date for all tax periods beginning on or after January 1, 2008. There are no material Tax sharing, allocation or indemnification agreements to which the Company or any of its Subsidiaries is a party or by which the Company or any of its Subsidiaries is otherwise bound.

(g) To the Knowledge of the Company, neither the Company nor any of its Subsidiaries is or has ever been a party to a transaction or agreement that is in material conflict with the Tax rules on transfer pricing in any relevant jurisdiction or with OECD Guidelines. To the Knowledge of the Company, none of the transactions among any of the Company, any of its Subsidiaries or any related parties are subject to any material adjustment, apportionment, allocation or recharacterization under Section 482 of the Code or any similar federal, state or local or foreign rule or regulation, and all of such transactions have been effected in all material respects on an arm's length basis. The Company has Made Available to Parent all material intercompany agreements or descriptions thereof (whether or not in writing) relating to transfer pricing with respect to the Company and any of its Subsidiaries. The Company has also Made Available to Parent the Transfer Pricing Documentation drawn up according to the Regulation issued by Italian Tax Authorities (*Agenzia delle Entrate*) on September, 29, 2010 together with all the supporting documents demonstrating the data and information contained therein.

(h) Each of the Company and its Subsidiaries is in material compliance with all terms and conditions of any Tax exemptions, Tax holiday or other Tax reduction agreement, approval or order of any Governmental Body, and the consummation of the Offer and all other transactions contemplated in connection therewith will not have any adverse effect on the validity and effectiveness of any such Tax exemptions, Tax holiday or other Tax reduction agreement or order.

(i) The Company has Made Available to Parent complete and accurate set of original documents supporting the deductibility of incurred costs, with particular reference to costs incurred by the Company and paid to the Company Subsidiary formed under the laws of Switzerland and costs included in the scope of article 110 (10) of the Italian Decree of the President of Republic n. 917/1986.

(j) The Company has Made Available to Parent complete and accurate set of documents supporting its view that there is no ground for application of "CFC rules" according to Article 167 the Italian Decree of the President of Republic n. 917/1986.

(k) The Company has Made Available to Parent complete and accurate set of documents pertaining to (i) the qualifying R&D cost for obtaining the R&D credit, according to the Law 27 December 2006 no. 296 article 1, par from 280 to 283, as modified by Article 1, par 66 of the Law 24 December 2007, no. 244 and (ii) the fulfillments necessary for obtaining the tax credit (e.g. FRS form).

(l) To the Knowledge of the Company, Section 3.14(l) of the Company Disclosure Letter sets forth each country in which any of the Company or any of its Subsidiaries files, is required to file, or has been required to file an income Tax Return.

(m) Neither the Company nor any of its Subsidiaries (i) has ever been classified as a “passive foreign investment company” as defined in Section 1297 of the Code or (ii) as of the Agreement Date expects to be classified as a “passive foreign investment company” as defined in Section 1297 of the Code for the current Taxable year or the Taxable year that includes the Offer Closing.

Section 3.15 Contracts.

(a) For purposes of this Agreement, a “**Material Contract**” shall mean:

(i) any “material contract” (as such term is defined in Item 601(b)(10) of Regulation S-K of the Securities Act, other than those agreements and arrangements described in Item 601(b)(10)(iii)) with respect to the Company;

(ii) any Company Contract identified or required to be identified in Section 3.12(a) of the Company Disclosure Letter (pursuant to which the Company has continuing rights or obligations as of the Agreement Date) relating to the employment of, or the performance of service by, (A) any current or former executive officer or director of the Company or (B) any employee or consultant of the Company providing for an annual base salary or payment for services in excess of \$50,000;

(iii) any Company Contract: (A) limiting in any material respect the right of the Company or any of its Subsidiaries (1) to engage in any line of business or to compete with, or solicit any customer of, any Person in any line of business or in any geographic location, to acquire any product or other asset or any services from any other Person, (2) to solicit, hire or retain any Person as an employee, consultant or independent contractor, (3) to develop, manufacture, sell, supply, distribute, offer, support or service any product or any product, technology or other asset to or for any other Person, or (4) to perform services for any other Person; (B) which contemplates an exclusive relationship between the Company or any of its Subsidiaries and any other Person; (C) prohibiting the Company (or, after the Acceptance Time, Parent) from engaging in business with any Person or levying a fine, charge or other payment for doing so, other than any such Contracts that (1) may be cancelled without material liability to the Company upon notice of ninety (90) days or less or (2) are not, individually or in the aggregate, material to the Company and its Subsidiaries, taken as a whole; or (D) under which the Company or any of its Subsidiaries has granted or has obtained an option to purchase or acquire, or a right of first refusal or right of first negotiation with respect to the purchase or acquisition of, any assets of the Company or its Subsidiaries valued at an amount in excess of \$100,000;

(iv) any Company Contract (A) relating to the disposition or acquisition by the Company or any of its Subsidiaries after the date of this Agreement of assets with a fair market value in excess of \$100,000 outside of the ordinary course of business, or (B) pursuant to which the Company or any of its Subsidiaries will acquire any ownership interest in any other Person or other business enterprise outside of the ordinary course of business and with a value of greater than \$100,000;

(v) any mortgages, indentures, guarantees, loans or credit agreements, security agreements or other Company Contracts relating to the borrowing of money or extension of credit, in each case in excess of \$100,000, other than accounts receivables and payables and loans to Company Subsidiaries;

(vi) any Company Contract relating to the creation of any Lien with respect to any material asset of the Company or any of its Subsidiaries;

(vii) any settlement Contract or other Company Contract relating to the settlement of Actions other than (A) releases entered into with former employees or independent contractors of the Company in the ordinary course of business, or (B) settlement Contracts or other Company Contracts relating to the settlement of Actions only involving the payment of cash (which has been paid) in amounts that do not exceed \$100,000 in the aggregate;

(viii) any Company Contract that provides for indemnification, or for reimbursement of any legal fees or expenses, of any officer, director or employee of the Company or any of its Subsidiaries;

(ix) any Company Contract (other than Contracts evidencing Company Options) (A) contemplating the acquisition, issuance, voting, registration, sale or transfer of any securities, (B) providing any Person with any preemptive right, right of participation, right of maintenance or similar right with respect to any securities, or (C) providing the Company or any other Person with any right of first refusal with respect to, or right to repurchase, redeem, put or call, any securities;

(x) any Company Contract reasonably expected to involve payment or receipt of an amount in excess of \$200,000 per annum that relates to the research, development, testing, manufacturing or commercialization of any API or Product, including clinical trial agreements, consulting agreements, supply and contract manufacturing agreements (but excluding purchase orders under existing agreements and confidentiality agreements);

(xi) any Company Contract pursuant to which the Company or any of its Subsidiaries (A) is obligated to pay to any other person a percentage of sales, revenues or profits, or any development, regulatory or commercial or similar milestone payments, in each case, with respect to any API or Product; (B) is obligated to provide to any other person a percentage interest in the sales, profits or revenues of any API or Product; or (C) is obligated to supply any API or Product excluding pursuant to purchase orders under existing agreements, in each case whether as of the date of this Agreement or as of a future date, including upon the occurrence of any future event;

(xii) any Company Contract that obligates (including through the use of diligent or commercially reasonable efforts or similar undertaking) the Company or any of its Subsidiaries to develop and/or commercialize or manufacture any product, drug or compound, or to transfer any marketing authorization or orphan designation granted by the European Commission and held by the Company or any of its Subsidiaries;

(xiii) any Company Contract that is a partnership, collaboration or joint venture agreement that is material to the Company and its Subsidiaries as a whole;

(xiv) any Company Contract under which the consummation of the Offer or any of the other transactions contemplated by this Agreement would give rise to or expand any material rights in favor of, or any material obligations on the part of, the Company, any of its Subsidiaries or any other Person; and

(xv) any Company Contract identified or required to be identified in Section 3.12(a) of the Company Disclosure Letter (pursuant to which the Company has continuing obligations as of the Agreement Date) pursuant to which the Company or any Subsidiary of the Company is or may become obligated to make any severance, termination, tax gross-up, or similar payment;

(xvi) any other Company Contract not otherwise described in clauses (i)-(xv) above pursuant to which the Company or any of its Subsidiaries is obligated to make payments or incur costs in excess of \$100,000 in any year; or

(xvii) any other Company Contract, or group of other Company Contracts with a Person (or group of affiliated Persons), the breach or termination of which would reasonably be expected to have or result in a Company Material Adverse Effect.

(b) Section 3.15(b) of the Company Disclosure Letter sets forth a true and complete list of all Material Contracts as of the Agreement Date. The Company has Made Available to Parent correct and complete copies of all Material Contracts as of the Agreement Date.

(c) Each Material Contract is valid and binding on the Company and/or its Subsidiaries and, to the Knowledge of the Company, each other party thereto, and is in full force and effect, and the Company and/or its Subsidiaries have performed in all material respects all obligations required to be performed by them under each Material Contract and, to the Knowledge of the Company as of the Agreement Date, each other party to each Material Contract has performed in all material respects all obligations required to be performed by it under such Material Contract.

(d) None of the Company, any Subsidiary of the Company or, to the Knowledge of the Company, any other party thereto is in default under any Material Contract nor is there any event or circumstance that with notice or lapse of time, or both, could (i) constitute a material default or material breach by the Company and/or its Subsidiary or (to the Knowledge of the Company) any other party under any Material Contract, (ii) give any Person the right to receive or require a rebate, chargeback, penalty or change in delivery schedule under any Material Contract, (iii) give any Person the right to accelerate the maturity or performance of any Material Contract, or (iv) give any Person the right to cancel, terminate or modify any Material Contract.

(e) The Company has not received any notice or other communication (in writing or, to the Knowledge of the Company, otherwise) regarding any actual or possible material violation or material breach of, or material default under, any Material Contract; and the Company has not waived any of its material rights under any Material Contract.

(f) The Company completed the update of the “Dossier” (as such term is defined in that certain Master Agreement with EG SpA dated July 3, 2013 (the “*EG Master Agreement*”)) as set forth in the EG Master Agreement and delivered such updated Dossier to EG SpA no later than July 31, 2013.

Section 3.16 Insurance. The Company and each of its Subsidiaries is covered by valid and currently effective insurance policies issued in favor of the Company or one or more of its Subsidiaries that are customary and adequate for companies of similar size in the industries and locations in which the Company and its Subsidiaries operate. To the Knowledge of the Company, all material insurance policies and bonds with respect to the business and assets of the Company and its Subsidiaries are in full force and effect, no notice of cancellation has been received, and there is no existing material default or event which, with the giving of notice or lapse of time or both, would constitute a material default, by any of the insured parties thereunder. The Company and each of its Subsidiaries maintain mandatory insurance policies as required by applicable Law.

Section 3.17 Properties. Section 3.17 of the Company Disclosure Letter sets forth a true and complete list of all Owned Real Property and all Leased Real Property. To the Company’s Knowledge, no parcel of Owned Real Property or Leased Real Property is subject to any decree or order of any Governmental Entity to be sold or is being condemned, expropriated or otherwise taken by any Governmental Entity with or without payment of compensation therefore, nor, to the Knowledge of the Company, has any such condemnation, expropriation or taking been proposed. Each of the Company and its Subsidiaries has (a) good and marketable ownership title (*piena proprietà*) to all Owned Real Property, (b) good and marketable leasehold title to all Leased Real Property, and (c) good and marketable title to, or a valid leasehold interest in, all of its tangible personal properties and assets reflected in the Company 20-F or acquired after December 31, 2012 (other than assets disposed of since December 31, 2012 in the ordinary course of business consistent with past practice), in each case free and clear of all Liens. The tangible personal property and assets of the Company and its Subsidiaries are in good operating condition and in a state of good maintenance and repair, ordinary wear and tear excepted, are operated in accordance with all applicable licenses, Permits, consents and governmental authorizations, and are usable in the regular and ordinary course of business, except as would not reasonably be expected to have a Company Material Adverse Effect. Neither the Company nor any of its Subsidiaries has any legal obligation, absolute or contingent, to any other Person to sell or otherwise dispose of any of its tangible personal properties or assets (other than the sale of the Company’s products in the ordinary course of business consistent with past practice) with an individual value in excess of \$500,000 or an aggregate value in excess of \$2,000,000. This Section 3.17 does not relate to Intellectual Property Rights, which are the subject of Section 3.18.

Section 3.18 Intellectual Property.

(a) The Company and its Subsidiaries own, or are licensed or otherwise possess the rights to use, all Intellectual Property Rights used in the business of the Company and its Subsidiaries, as it is being conducted as of the Agreement Date.

(b) Part 1 of Section 3.18(b) of the Company Disclosure Letter contains a complete and accurate list (in all material respects), as of the Agreement Date, of the following Owned Company IP owned solely by the Company or any of its Subsidiaries (“**Company Solely Owned Registered IP**”): (i) all registered Trademarks; (ii) all Patents; and (iii) all registered Copyrights, in each case listing, as applicable, (A) the jurisdiction in which patents have been issued, patent applications have been filed, Trademarks or Copyrights have been registered and trademark applications have been filed and (B) the application, registration or filing number and date. Part 2 of Section 3.18(b) of the Company Disclosure Letter contains a complete and accurate list (in all material respects), as of the Agreement Date, of the following Owned Company IP in which the Company or any of its Subsidiaries has a joint, rather than a sole, ownership interest (“**Company Co-Owned Registered IP**” and together with the Company Solely Owned Registered IP, the “**Company Registered IP**”): (i) all registered Trademarks; (ii) all Patents; and (iii) all registered Copyrights, in each case listing, as applicable, (A) the jurisdiction in which patents have been issued, patent applications have been filed, Trademarks or Copyrights have been registered and trademark applications have been filed and (B) the application, registration or filing number and date. To the Knowledge of the Company, (i) all Company Registered IP rights are in effect and subsisting, and (ii) all material Company Registered IP Rights are valid and enforceable. All filings, payments and other actions required to be made or taken by the Company before the Agreement Date to maintain each item of material Company Registered IP, and, to the Knowledge of the Company, the other Company Registered IP, have been made and taken.

(c) The Company and its Subsidiaries own all right, title and interest in the Owned Company IP, including the Company Registered IP, in each case that is solely owned by the Company and its Subsidiaries, free and clear of all Liens (except for Liens permitted under Section 1.1(uu)(x)). To the Knowledge of the Company, all Material Company IP Rights that are not owned by the Company in whole or in part are valid.

(d) To the Knowledge of the Company, as of the Agreement Date, there is no material unauthorized use, disclosure, infringement or misappropriation of any Company Intellectual Property Rights by any third party, including any employee or former employee of the Company.

(e) Except as set forth in Section 3.18(e) of the Company Disclosure Letter, (i) neither the Company nor any of its Subsidiaries has been in the past six (6) years or currently is a party to any Action that involves a claim of infringement or misappropriation of any Intellectual Property Rights of any third party nor, to the Knowledge of the Company, is any such Action being threatened in writing against the Company or any of its Subsidiaries; (ii) to the Knowledge of the Company, neither the conduct of the business of the Company and its Subsidiaries as currently conducted nor the development, manufacture, sale or use of any of any API or Product by the Company or its Subsidiaries as now developed, manufactured, sold or used infringes or misappropriates, any Intellectual Property Rights of any third party; (iii) no third party has challenged in the past six (6) years or currently is challenging the ownership by the Company or its Subsidiaries, or the validity of, any material Owned Company IP (including the material Company Registered IP); (iv) neither the Company nor any of its Subsidiaries has brought in the past six (6) years or currently is bringing any Action for infringement or misappropriation of the Company Intellectual Property Rights. There are no pending or threatened in writing interference actions, re-examinations, oppositions or nullities involving any Company Registered IP, except such as may have been commenced by the Company or any of its Subsidiaries. There is no judgment outstanding against the Company or any of its Subsidiaries or any Owned Company IP or, to the Knowledge of the Company any Licensed Company IP, that limits the ability of the Company or any of its Subsidiaries to exploit any Owned Company IP or any Licensed Company IP.

(f) Except as set forth in [Section 3.18\(f\)](#) of the Company Disclosure Letter, the Company or any of its Subsidiaries owns, co-owns, is licensed or otherwise has the right to use all Material Company IP Rights, free and clear of all Liens (except for Liens permitted under Section 1.1(uu)(x)) and additionally in the case of Material Company IP Rights that are licensed to the Company or any of its Subsidiaries, other than Liens arising by virtue of the Contract pursuant to which such license is granted and any Liens granted or incurred by the licensor thereof), and, in the case of Material Company IP Rights owned in whole or in part by the Company or any of its Subsidiaries, except to the extent set forth in [Section 3.18\(f\)](#) of the Company Disclosure Letter, also free and clear of exclusive or non-exclusive licenses granted by the Company or any of its Subsidiaries to any third party. To the Knowledge of Company, except as set forth in [Section 3.18\(f\)](#) of the Company Disclosure Letter, no third party which is not the U.S. Patent Office or any foreign equivalent governmental administrative agency for patent matters (the “**Governmental Patent Authority**”) is overtly challenging in writing the right, title or interest of any of the Company or any of its Subsidiaries or, to the Knowledge of Company, any licensor of the Company or any of its Subsidiaries in, to or under the Material Company IP Rights, or the validity, enforceability or claim construction of any Patents within the Material Company IP Rights that are owned (in whole or in part) or exclusively licensed to the Company or any of its Subsidiaries. To the Knowledge of Company, except as set forth in [Section 3.18\(f\)](#) of the Company Disclosure Letter, there is no opposition, cancellation, proceeding, or objection involving a third party, other than a Governmental Patent Authority, pending with regard to any Material Company IP Rights. Except as set forth in [Section 3.18\(f\)](#) of the Company Disclosure Letter, none of the Material Company IP Rights owned solely by the Company or any of its Subsidiaries and, to the Knowledge of Company, none of the Material Company IP Rights jointly owned by or exclusively licensed to the Company or any of its Subsidiaries are subject to any outstanding order of, judgment of, decree of or agreement with any Governmental Entity other than a Governmental Patent Authority. To the Knowledge of Company, no valid basis exists for any of such challenges as set forth in this [Section 3.18\(f\)](#).

(g) [Section 3.18\(g\)](#) of the Company Disclosure Letter lists all Company IP Contracts. To the Knowledge of the Company, each Company IP Contract is valid and binding on the Company and/or its Subsidiaries and each other party thereto.

(h) [Section 3.18\(h\)](#) of the Company Disclosure Letter lists all written Contracts, licenses or other arrangements (excluding implied licenses granted by the Company or any of its Subsidiaries) in effect as of the date of this Agreement under which the Company or any of its Subsidiaries has licensed, granted or conveyed to any third party any right, title or interest in or to any Material Company IP Rights.

(i) Neither the Company nor any of its Subsidiaries is obligated to pay to any Person any royalties for the use by the Company or any of its Subsidiaries of any Company Intellectual Property Rights, other than as provided in a Contract listed in [Section 3.18\(g\)](#) of the Company Disclosure Letter.

(j) The Company takes reasonable measures to protect the confidentiality of its Trade Secrets, including, where the Company determines appropriate, by entering into assignments or other agreements for the protection of Intellectual Property Rights with its officers, employees, consultants and independent contractors or other third parties.

(k) Neither the execution, delivery or performance of this Agreement by the Company nor the consummation of any transactions contemplated hereunder will (i) result in any material limitation on the Company or any of its Subsidiaries' right, title or interest in or to any of the Material Company IP Rights or result in a breach of, default under or termination of any material Company IP Contract.

(l) To the Knowledge of the Company, (i) no Person employed or otherwise engaged by the Company who in the course of such engagement created, discovered, or developed any material invention connected to the Company Intellectual Property Rights has made or is entitled to make any claim for the benefit of that person to: (a) any right, title or interest in such Intellectual Property Right; or (b) any compensation or remuneration in relation to such Intellectual Property Right (including pursuant to Italian Article 64, Paragraph 2 of Legislative Decree no. 30/2005) and (ii) in the case of Patents, no further compensation (*equo premio*) is owed to any Person who at any time was employed or otherwise engaged by the Company.

(m) Notwithstanding any other provision of this Agreement, this Section 3.18, Sections 3.15(a)(i), (iii), (iv), (vi), (vii), (viii), (x), (xi), (xiii), (xiv), and (xvii), Sections 3.15(b), (c), (d), (e) and (f) and Section 3.5(a)(ii), shall be the exclusive section in the Agreement for representations and warranties with respect to Intellectual Property Rights and Contracts pertaining to or involving Intellectual Property Rights (including the Company IP Contracts), and no other representation or warranty in this Article III shall be made or deemed to be made by the Company or any of its Subsidiaries with respect to such subject matter or any matter related thereto or arising therefrom.

Section 3.19 Takeover Statutes. To the Knowledge of the Company, the Company Board has taken all actions necessary to ensure that no "moratorium," "fair price," "business combination," "control share acquisition" or similar provision of any state anti-takeover Law (collectively, "**Takeover Laws**") is, or at the Offer Closing will be, applicable to this Agreement or the Support Agreements.

Section 3.20 Related Party Transactions. Except for compensation or other employment arrangements in the ordinary course of business consistent with past practice, there are no transactions, agreements, arrangements or understandings between the Company or any of its Subsidiaries, on the one hand, and any Affiliate (including any director or officer) thereof, but not including any wholly-owned Subsidiary of the Company, on the other hand, that would be required to be disclosed pursuant to Item 404 of Regulation S-K of the SEC in the Company's Annual Report on Form 20-F.

Section 3.21 Brokers. No broker, investment banker, financial advisor or other Person, other than Jefferies LLC ("**Jefferies**"), the fees and expenses of which will be paid by the Company, is entitled to any broker's, finder's, financial advisor's or other similar fee or commission in connection with the transactions contemplated by this Agreement based upon Contracts with the Company. The Company has Made Available to Parent a true and complete copy of the Contract between the Company and Jefferies pursuant to which Jefferies is entitled to payment from the Company relating to the transactions contemplated hereby.

Section 3.22 No Vote Required. No vote of the holders of any class or series of Company Shares is necessary to approve this Agreement or to consummate the transactions contemplated by this Agreement.

Section 3.23 Opinion of Financial Advisor. The Company Board has received the opinion of Jefferies to the effect that, as of the date of such opinion and based upon and subject to the assumptions and qualifications stated therein, the consideration to be received in the Offer by holders of Company Shares (other than Parent, Purchaser and their respective Subsidiaries) pursuant to this Agreement is fair, from a financial point of view, to such holders.

Section 3.24 Information Supplied. None of the documents required to be filed by the Company with the SEC after the date hereof in connection with the Offer or the other transactions contemplated by this Agreement will, on the date of such filing, contain any untrue statement of a material fact or omit to state any material fact required to be stated therein or necessary in order to make the statements therein, in light of the circumstances under which they are made, not misleading. In furtherance and not in limitation of the foregoing, none of the information supplied by or on behalf of the Company specifically for inclusion or incorporation by reference in the Offer Documents or the Schedule 14D-9 will, at the time the Offer Documents or the Schedule 14D-9, as applicable, is filed with the SEC, at any time it is amended or supplemented, or at the time it is first distributed or otherwise disseminated to the stockholders of the Company, contain any untrue statement of a material fact or omit to state any material fact required to be stated therein or necessary in order to make the statements therein, in the light of the circumstances under which they were made, not misleading. Notwithstanding the foregoing, no representation or warranty is made by the Company or its Subsidiaries with respect to statements made or incorporated by reference in the Offer Documents or the Schedule 14D-9 based on information supplied by or on behalf of Parent in writing specifically for inclusion or incorporation by reference therein.

Section 3.25 Rule 14d-10. As of the Acceptance Time, a committee of the independent members of the Company Board (each member of which the Company Board has determined is an “independent director” in accordance with the requirements of Rule 14d-10(d)(2) under the Exchange Act) shall have: (i) at a meeting duly called and held, duly adopted resolutions approving each employment compensation, severance and employee benefit agreement, arrangement or understanding entered into or established on or before the Agreement Date by the Company or its Subsidiaries with or for the benefit of current or future directors, officers or employees of the Company and its Subsidiaries as an “employment compensation, severance or other employee benefit arrangement” within the meaning of Rule 14d-10(d)(2) under the Exchange Act; and (ii) taken all other actions and made all other determinations necessary or advisable to ensure that any such arrangements fall within the safe harbor provisions of Rule 14d-10(d).

ARTICLE IV

REPRESENTATIONS AND WARRANTIES OF PARENT AND PURCHASER

Except as set forth in the corresponding section or subsection of the Disclosure Letter delivered by Parent and Purchaser to the Company immediately prior to the execution of this Agreement (the “**Purchaser Disclosure Letter**”), if any, each of Parent and Purchaser jointly and severally represent and warrant to the Company as follows:

Section 4.1 Organization, Standing and Power. Each of Parent and Purchaser (a) is a corporation duly organized, validly existing and in good standing (with respect to jurisdictions that recognize such concept) under the Laws of its jurisdiction of incorporation and (b) has all requisite corporate power and authority and all necessary government approvals to own, lease and operate its properties and assets and to conduct its business as presently conducted, except as, individually or in the aggregate, has not had and would not reasonably be expected to have a Purchaser Material Adverse Effect. Neither Parent nor Purchaser is in violation of its articles of incorporation, bylaws or other equivalent charter documents, as applicable.

Section 4.2 Authority. Each of Parent and Purchaser has all necessary corporate power and authority to execute and deliver this Agreement, to perform its obligations hereunder and to consummate the transactions contemplated hereby. The execution, delivery and performance of this Agreement by Parent and Purchaser and the consummation by Parent and Purchaser of the transactions contemplated hereby have been duly authorized by all necessary corporate action on the part of Parent and Purchaser and no other corporate proceedings on the part of Parent or Purchaser are necessary to approve this Agreement or to consummate the transactions contemplated hereby. This Agreement has been duly executed and delivered by Parent and Purchaser and, assuming the due authorization, execution and delivery by the Company, constitutes a valid and binding obligation of Parent and Purchaser, enforceable against Parent and Purchaser in accordance with its terms, except as enforcement thereof may be limited against Parent or Purchaser by (a) bankruptcy, insolvency, reorganization, moratorium and similar Laws affecting the enforcement of creditors’ rights or remedies in general as from time to time in effect or (b) the exercise by courts of equity powers.

Section 4.3 No Conflict; Consents and Approvals.

(a) The execution, delivery and performance of this Agreement by Parent and Purchaser does not, and the consummation of the Offer and the other transactions contemplated hereby and compliance by Parent and Purchaser with the provisions hereof will not (i) violate or conflict with the articles of incorporation or bylaws (or similar organizational documents) of Parent or Purchaser, (ii) result in any breach of, or default (with or without notice or lapse of time, or both) under, or give rise to a right of, or result in, termination, cancellation, modification or acceleration of any obligation or to the loss of a benefit under, or result in the creation of any Lien in or upon any of the properties, assets or rights of Parent or Purchaser under, or require any consent, waiver or approval of any Person pursuant to, any provision of any Contract to which Parent or Purchaser is a party by which Parent or Purchaser or any of their respective properties or assets maybe bound or (iii) subject to the governmental filings and other matters referred to in Section 4.3(b) hereof, violate or conflict with any Law applicable to Parent or Purchaser or by which Parent or Purchaser, or any of its respective properties or assets may be bound.

(b) No consent, approval, order or authorization of, or registration, declaration, filing with or notice to, any Governmental Entity is required by or with respect to Parent or Purchaser in connection with the execution, delivery and performance of this Agreement by Parent and Purchaser or the consummation by Purchaser of the Offer or compliance with the provisions hereof, except for (i) compliance with any applicable requirements of the Antitrust Laws, (ii) such filings and reports as

required pursuant to the applicable requirements of the Securities Act, the Exchange Act the rules and regulations promulgated thereunder, and state securities Laws or “blue sky” Laws, and (iii) such other filings the failure of which to be obtained or made, individually or in the aggregate, have not had and would not reasonably be expected to have a Purchaser Material Adverse Effect.

Section 4.4 Brokers. No broker, investment banker, financial advisor or other Person, other than Barclays Capital, the fees and expenses of which will be paid by Parent or Purchaser, is entitled to any broker’s, finder’s, financial advisor’s or other similar fee or commission in connection with the transactions contemplated by this Agreement based upon arrangements made by or on behalf of Parent or Purchaser.

Section 4.5 Financing.

(a) Purchaser will have, as of the respective dates of consummation of the Offer (including any subsequent offering period), access to sufficient funds to consummate the Offer (including any subsequent offering period) on the terms and subject to the conditions contemplated hereby, and for the payment to the Company of funds sufficient to pay holders of Company Options in accordance with the provisions of Section 6.7.

(b) Parent has delivered to the Company a complete and correct copy of the executed commitment letter, dated as of the date hereof, from Barclays Bank PLC (collectively, the “**Financing Commitment**”) pursuant to which the lender party thereto has committed to provide, subject to the terms and conditions set forth therein and the conditions set forth in the Purchaser Credit Facility, debt financing in an aggregate amount set forth therein (the “**Financing**”). Parent has also delivered to the Company complete and correct copies of any fee letters (redacted to exclude fees and certain other information at the request of the Financing Sources party thereto) (the “**Fee Letters**”) in connection with the Financing Commitment and the Purchaser Credit Facility and, as of the Agreement Date, except as would not reasonably be expected to materially impair the validity of the Financing Commitment, materially impact the availability of the Financing or materially decrease the amount of financing that could be expected to be provided under the Financing Commitment, there are no Contracts, agreements, side letters or arrangements to which Parent or Purchaser is a party relating to the funding or investing, as applicable, of the amount of the Financing other than as expressly set forth in the Financing Commitment, Fee Letters and Purchaser Credit Facility.

(c) As of the date hereof, the Financing Commitment has not been amended or modified, no such amendment or modification is presently contemplated, and the respective obligations and commitments contained in the Financing Commitment have not been withdrawn or rescinded in any respect. Parent or Purchaser has fully paid any and all commitment fees or other fees in connection with the Financing Commitment that are payable on or prior to the date hereof, and, as of the date hereof, the Financing Commitment is in full force and effect and are the valid, binding and enforceable obligations of Parent and Purchaser (except as such enforceability may be (i) limited by bankruptcy, insolvency, reorganization, moratorium or similar Laws affecting the enforcement of creditors’ rights generally and (ii) the exercise by courts of equity powers) and, to the knowledge of Parent and Purchaser, the other parties thereto.

(d) Assuming the satisfaction or waiver of the conditions to Parent’s and Purchaser’s obligation to consummate the Offer (including any subsequent offering period), the net

proceeds of the Financing if funded in accordance with the Financing Commitments, together with Parent's consolidated cash and cash equivalents and borrowings under Purchaser Credit Facility are, in the aggregate, sufficient for Purchaser to pay the aggregate Offer Price, consummate the transactions contemplated by this Agreement and pay all fees and expenses required to be paid by Parent or Purchaser in connection with the Offer and the Financing.

(e) As of the date of this Agreement, no event has occurred which, with or without notice, lapse of time or both, would constitute a default or breach on the part of Parent or Purchaser under the Financing Commitments or, to the knowledge of Parent and Purchaser, any other party thereto. As of the date of this Agreement, neither Parent nor Purchaser has any reason to believe that any of the conditions to the Financing will not be satisfied or that the full amount of the Financing will not be available to Parent or Purchaser on the date of the consummation of the Offer.

Section 4.6 Information Supplied. None of the documents required to be filed by Parent or Purchaser with the SEC in connection with the Offer or the other transactions contemplated by this Agreement after the date hereof will, on the date of such filing, contain any untrue statement of a material fact or omit to state any material fact required to be stated therein or necessary in order to make the statements therein, in light of the circumstances under which they are made, not misleading. In furtherance and not in limitation of the foregoing, none of the information supplied by or on behalf of Parent or Purchaser specifically for inclusion or incorporation by reference in the Offer Documents or the Schedule 14D-9 will, at the time the Offer Documents or the Schedule 14D-9, as applicable, is filed with the SEC, at any time it is amended or supplemented, or at the time it is first distributed or otherwise disseminated to the stockholders of the Company, contain any untrue statement of a material fact or omit to state any material fact required to be stated therein or necessary in order to make the statements therein, in the light of the circumstances under which they were made, not misleading. Notwithstanding the foregoing, no representation or warranty is made by Parent or Purchaser with respect to statements made or incorporated by reference in the Offer Documents or the Schedule 14D-9 based on information supplied by or on behalf of the Company in writing specifically for inclusion or incorporation by reference therein.

Section 4.7 Litigation. None of Parent, Purchaser or any of their respective properties or assets is subject to any outstanding judgment, order, injunction, rule or decree of any Governmental Entity that, individually or in the aggregate, would reasonably be expected to have a Purchaser Material Adverse Effect. There is no Action pending or, to the knowledge of Parent or Purchaser, threatened in writing seeking to impose any legal restraint on or prohibition against the Offer.

Section 4.8 Ownership of Purchaser; No Prior Activities. Parent and its wholly-owned Subsidiaries collectively own one hundred percent (100%) of the issued and outstanding capital stock of Purchaser. Purchaser was formed solely for the purpose of engaging in the transactions contemplated by this Agreement. Except for obligations or liabilities incurred in connection with its formation and the transactions contemplated by this Agreement, including the Financing, Purchaser has not and will not have incurred, directly or indirectly, through any Subsidiary or Affiliate or otherwise, any obligations or liabilities or engaged in any business activities of any type or kind whatsoever or entered into any agreements or arrangements with any Person (other than Parent or a wholly-owned Subsidiary of Parent).

Section 4.9 Certain Agreements. There are no Contracts between Parent or Purchaser, on the one hand, and any executive officer or director of the Company or any Affiliate of the Company (collectively, "**Company Senior Management**"), on the other hand, as of the Agreement Date that relate in any way to the Company, the transactions contemplated by this Agreement or any financial benefits to be received by Company Senior Management as a result of such transactions, except for the Support Agreements and the other agreements referred to in the Recitals to this Agreement (each of which have been made available to the Company or its Representatives by Parent prior to the execution and delivery of this Agreement).

Section 4.10 Solvency. Assuming that the Company's consolidated financial statements as set forth in the Company 20-F fairly present the consolidated financial condition of the Company and its Subsidiaries as at the end of the periods covered thereby and the consolidated results of operations of the Company and its Subsidiaries for the periods covered thereby, then immediately following the Acceptance Time and after giving effect to all of the transactions contemplated by this Agreement, including the Financing, and the payment of the aggregate consideration to which the shareholders of the Company are entitled hereunder in the Offer, (a) the fair value of the assets of Parent and its Subsidiaries, on a consolidated basis, exceeds, on a consolidated basis, their debts and liabilities, subordinated, contingent or otherwise, (b) the present fair saleable value of the property of Parent and its Subsidiaries, on a consolidated basis, is greater than the amount that will be required to pay the probable liability, on a consolidated basis, of their debts and other liabilities, subordinated, contingent or otherwise, as such debts and other liabilities become absolute and matured; (c) Parent and its Subsidiaries, on a consolidated basis, will be able to pay their debts and liabilities, subordinated, contingent or otherwise, as such liabilities become absolute and matured; and (d) Parent and its Subsidiaries, on a consolidated basis, are not engaged in, and are not about to engage in, business for which they have unreasonably small capital.

Section 4.11 No Other Information. Parent and Purchaser acknowledge that the Company makes no representations or warranties as to any matter whatsoever except as expressly set forth in Article III. The representations and warranties set forth in Article III are made solely by the Company, and no Representative of the Company shall have any responsibility or liability related thereto. Notwithstanding anything to the contrary contained in this Section 4.11 or elsewhere in this Agreement, nothing in this Section 4.11 will operate to limit any right or remedy that Parent, Purchaser or any of their respective Representatives may have against any Person for fraud committed by the Company or any of the Company's Affiliates or Representatives.

Section 4.12 Interest in Company Shares. Neither Parent, Purchaser nor any of their Affiliates owns (directly or indirectly, or as a beneficial owner or constructive owner or of record) any Company Shares (other than passive investments for cash management purposes), and neither Parent, Purchaser nor any of their Affiliates holds any rights to acquire or vote any Company Shares, in each case except pursuant to the Support Agreements and the transactions contemplated by this Agreement.

Section 4.13 Access to Information. Parent and Purchaser each acknowledge and agree that it has not relied on any representation, warranty or other statement by any person on behalf of the Company or any of its Subsidiaries, other than the representations and warranties of the Company expressly contained in Article III and that all other representations and warranties are specifically disclaimed. Each of Parent and Purchaser further acknowledges and agrees that none of the Company or any of its shareholders, directors, officers, employees, Affiliates, advisors, agents or other

Representatives has made any representation or warranty concerning any estimates, projections, forecasts, business plans or other forward-looking information regarding the Company, its subsidiaries or their respective businesses and operations. Notwithstanding anything to the contrary contained in this Section 4.13 or elsewhere in this Agreement, nothing in this Section 4.13 will operate to limit any right or remedy that Parent, Purchaser or any of their respective Representatives may have against any Person for fraud committed by the Company or any of the Company's Affiliates or Representatives.

Section 4.14 Operations Following the Offer. Parent currently intends that, following the Acceptance Time, the business of the Company (the "**Business**") shall continue to be headquartered in Villa Guardia (CO), Italy, and Parent currently intends that the production activities of the Business and substantially all of the other operations of the Business shall continue to be carried out substantially as they are on the Agreement Date.

ARTICLE V

INTERIM CONDUCT OF BUSINESS

Section 5.1 Affirmative Obligations of the Company. Except (a) as contemplated or permitted by this Agreement, (b) as set forth in Section 5.1 of the Company Disclosure Letter or (c) as approved in advance by Parent in writing (such approval not to be unreasonably withheld, conditioned or delayed), at all times during the Pre-Closing Period, the Company shall (and shall cause each of its Subsidiaries to):

- (a) carry on its business in all material respects in the ordinary course in substantially the same manner as heretofore conducted and in material compliance with all applicable Laws;
- (b) pay its debts and Taxes when due, in each case subject to good faith disputes over such debts or Taxes contested in good faith by any appropriate Action and for which adequate reserves are established to the extent required by applicable GAAP;
- (c) use commercially reasonable efforts to (i) preserve intact its present business organization, (ii) pay or perform all material obligations when due, subject to good faith disputes over such obligations, (iii) keep available the services of its present officers and employees and (iv) preserve its relationships with material customers, suppliers, investigators, distributors, licensors, licensees and others with which it has material business dealings;
- (d) use its reasonable best efforts to (i) prepare and prosecute the approval by the FDA of the Defibrotide NDA consistent with past practice and to develop Defibrotide; (ii) obtain and maintain on a commercially reasonable basis quantities of finished Defibrotide drug product and related raw materials and components that the Company reasonably expects to be required for the anticipated commercial launch of Defibrotide; and
- (e) use commercially reasonable efforts to (i) preserve and maintain the validity of all authorizations for the manufacture of and marketing of medicinal products and orphan designations for medicinal products granted by any Regulatory Authority to the Company or any of its Subsidiaries and (ii) ensure that such authorizations or orphan designations are not varied, suspended or withdrawn.

Section 5.2 Negative Obligations of the Company. Except (i) as contemplated or permitted by this Agreement, (ii) as set forth in Section 5.2 of the Company Disclosure Letter or (iii) as approved in advance by Parent in writing (such approval not to be unreasonably withheld, conditioned or delayed with respect to the matters described in clauses “(i),” “(j),” “(p),” “(r),” “(s),” “(t)” and “(u)” of this Section 5.2), at all times during the Pre-Closing Period, the Company and its Subsidiaries shall not do any of the following and shall not permit its Subsidiaries to do any of the following:

(a)(i) declare, set aside or pay any dividends on, or make any other distributions (whether in cash, stock or property) in respect of, any of its capital stock or other equity interests, except for dividends by a wholly owned Subsidiary of the Company to the Company, (ii) except in connection with the exercise of Company Options outstanding prior to the Agreement Date, purchase, redeem or otherwise acquire shares of capital stock or other equity interests of the Company or its Subsidiaries or any options, warrants, or rights to acquire any such shares or other equity interests, or (iii) split, combine, reclassify or otherwise amend the terms of any of its capital stock or other equity interests;

(b) issue, sell, deliver or agree or commit to issue, sell or deliver (whether through the issuance or granting of options, warrants, commitments, subscriptions, rights to purchase or otherwise) any Company Shares or any securities of any Subsidiary, except for the issuance of Company Shares pursuant to Company Options outstanding prior to the date hereof;

(c) adopt, amend, authorize or propose to amend its articles of incorporation or bylaws (or similar organizational documents), except for amendments as required by Law in connection with the exercise of any outstanding Company Options;

(d) create or form any Subsidiary;

(e) directly or indirectly acquire or agree to acquire (i) by merging or consolidating with, purchasing a substantial equity interest in or a substantial portion of the assets of, making an investment in or loan or capital contribution to or in any other manner, any corporation, partnership, association or other business organization or division thereof, except any such transactions that are for cash consideration not in excess of \$1,000,000 individually, or \$2,500,000 for all such transactions by the Company and its Subsidiaries in the aggregate, and except for loans, advances, contributions or investments between or among the Company and any direct or indirect wholly owned Subsidiaries, or (ii) any assets that are otherwise material to the Company and its Subsidiaries, other than in the ordinary course of business consistent with past practice;

(f) directly or indirectly sell, lease, license, sell and leaseback, abandon, mortgage or otherwise encumber or subject to any Lien or otherwise dispose in whole or in part of any of its material properties, assets or rights or any interest therein, except (i) the abandonment or non-exclusive licenses under Intellectual Property Rights in the ordinary course of business consistent with past practice, (ii) sales of inventory of API and Products in the ordinary course of business consistent with past practice, and (iii) disposition of equipment and property no longer used in the operation of business;

(g) propose or adopt a plan of complete or partial liquidation, dissolution, merger, consolidation, restructuring, recapitalization or other reorganization of the Company or any of its Subsidiaries, except for the transactions contemplated by this Agreement;

(h)(i) incur, create, assume or otherwise become liable for, any indebtedness for borrowed money, any obligations under conditional or installment sale Contracts or other retention Contracts relating to purchased property, any capital lease obligations or any guarantee or any such indebtedness of any other Person, issue or sell any debt securities, options, warrants, calls or other rights to acquire any debt securities of the Company or any of its Subsidiaries, guarantee any debt securities of any other Person, enter into any “keepwell” or other agreement to maintain any financial statement condition of any other Person or enter into any arrangement having the economic effect of any of the foregoing except accounts payable to trade creditors (collectively, “Indebtedness”), or amend, modify or refinance any Indebtedness, (ii) make any loans, advances or capital contributions to, or investments in, any other Person, other than the Company or any direct or indirect wholly owned Subsidiary of the Company, other than in the ordinary course of business consistent with past practice, or (iii) syndicate or issue, attempt to syndicate or issue or announce the syndication or issuance of any debt facility or debt security that competes with Parent’s financing activities in connection with the Offer (including any renewal or refinancing of any of the Company’s existing debt facilities or securities);

(i) incur or commit to incur any capital expenditure or authorization or commitment with respect thereto in excess of \$500,000 individually or \$1,000,000 in the aggregate, except those provided for in the capital expenditure budget previously Made Available to Parent;

(j)(i) pay, discharge, settle or satisfy any claims, liabilities or obligations (whether absolute, accrued, asserted or unasserted, contingent or otherwise), in an excess of \$500,00 individually or \$1,000,000 in the aggregate, other than the payment, discharge or satisfaction of claims, liabilities or obligations reflected or reserved against in the most recent audited financial statements (or the notes thereto) of the Company included in the Company SEC Documents (for amounts not in excess of such reserves) or incurred since the date of such financial statements in the ordinary course of business consistent with past practice or incurred in connection with this Agreement and the transactions contemplated hereby (including legal and financial advisors fees and expenses), (ii) cancel any material Indebtedness or (iii) waive, release, grant or transfer any right of material value;

(k) compromise, settle or agree to settle any Action (including any Action relating to this Agreement or the transactions contemplated hereby) other than compromises, settlements or agreements that involve only the payment of money damages in an amount not greater than \$500,000 individually or \$1,000,000 in the aggregate, in any case without the imposition of any equitable relief on, or the admission of wrongdoing by, the Company;

(l) change its financial or tax accounting methods, principles or practices, except insofar as may have been required by a change in GAAP, Regulation S-X promulgated under the Exchange Act or applicable Law;

(m) settle, compromise or enter into any closing agreement with respect to any liability for Taxes, amend any material Tax Return, enter into any material Contract with or request any material ruling from any Governmental Entity relating to Taxes, make, change or revoke any material Tax election, change any method of accounting for Tax purposes, take any material position on a Tax

Return inconsistent with a position taken on a Tax Return previously filed, extend or waive any statute of limitations with respect to Taxes, surrender any claim for a material refund of Taxes or fail to timely file correct and complete Tax Returns as required by applicable Tax law;

(n) change its fiscal year;

(o) except as may be required by applicable Law or the terms of any applicable Contract or Employee Benefit Plan as in effect on the Agreement Date, and except in the context of new hires whose base salary is less than \$50,000, in each case in the ordinary course of business consistent with past practice, (i) grant any new, current or former director, officer, employee or independent contractor of the Company or any of its Subsidiaries any material increase in compensation, bonus or other benefits, or grant any type of material compensation, bonus or benefits to any new, current or former director, officer, employee or independent contractor of the Company or any of its Subsidiaries not previously receiving or entitled to receive such type of compensation, bonus or benefit, or pay any material bonus to any new, current or former director, officer, employee or independent contractor of the Company or any of its Subsidiaries, (ii) grant or pay to any new, current or former director, officer, employee or independent contractor of the Company or any of its Subsidiaries or Affiliates any severance, change in control or termination pay, or modifications thereto or increases therein, (iii) pay any benefit or grant or amend any award (including in respect of stock options, restricted stock units, restricted stock or other stock-based or stock-related awards or the removal or modification of any restrictions in any Company Equity Plan or awards made thereunder) except the issuance of Company Shares upon the exercise of Company Options outstanding on the Agreement Date, (iv) adopt or enter into any collective bargaining agreement or other similar labor union contract except as required by Law, (v) take any action to accelerate the vesting or payment of any compensation or benefit under any Company Equity Plan or other Employee Benefit Plan or other Contract except as required pursuant to Section 6.7 or Section 6.8(f) hereof, (vi) adopt any new employee benefit plan (that would be an Employee Benefit Plan upon adoption) or arrangement or amend, modify or terminate any existing Company Equity Plan or other Employee Benefit Plan (except pursuant to Section 6.7 or Section 6.8(f) hereof), in each case, for the benefit of any new, current or former director, officer, employee or independent contractor of the Company or any of its Subsidiaries, or (vii) contribute to any trust or other arrangement funding any Employee Benefit Plan, except to the extent required by the existing terms of such Employee Benefit Plan, trust or other funding arrangement as in effect on the Agreement Date, by any collective bargaining agreement or other labor union contract, or by any written Employment Agreement as in effect as of the Agreement Date (or which may be entered into pursuant to the terms of this Agreement); provided, however, that notwithstanding anything to the contrary herein, the Company may pay out bonuses under the terms of any Employee Benefit Plan or Contract as in effect on the Agreement Date for any completed fiscal year (or any portion thereof completed prior to the Acceptance Time) to its employees based on the achievement of performance targets adopted prior to the Agreement Date;

(p) fail to use commercially reasonable efforts to keep in force insurance policies or replacement or revised provisions regarding insurance coverage with respect to material assets, operations and activities of the Company and its Subsidiaries as currently in effect;

(q) renew or enter into any non-compete, exclusivity or similar agreement that would restrict or limit, in any material respect, the Company or any of its Subsidiaries from engaging or competing in any line of business or geographic area;

(r) enter into any new material lease of real property or materially amend the terms of any existing material lease of real property, other than in the ordinary course of business;

(s) forgive any loans to any employees, officers or directors of the Company and its Subsidiaries, or any of their respective Affiliates or Associates;

(t) enter into or become bound by, or permit any of the assets owned or used by it to become bound by, any Material Contract, or seek to amend, terminate or waive, or exercise any material right or remedy under, any Material Contract or enter into or become bound by, or seek to amend, terminate or waive, or exercise any material right or remedy under, any Contract pursuant to which the Company or any of its Subsidiaries grants or has granted to any Person an exclusive license to practice or exploit the Material Company IP Rights;

(u) other than in the ordinary course of business consistent with past practices or as required by GAAP, write off as uncollectible, or establish any extraordinary reserve with respect to, any receivable or other indebtedness;

(v) take any actions with the intention or for the purpose of undermining the (i) continued validity of the manufacturing and marketing authorizations and orphan designation held by the Company or its Subsidiaries; or (ii) the Company's ability to transfer any authorization for the manufacturing or marketing of medicinal products or any orphan designations to Parent or Purchaser after the Acceptance Time;

(w) enter into or become bound by any distribution agreement or commercial tendering contract;

(x) other than an agreement substantially in the form identified in Section 5.2 of the Company Disclosure Letter, enter into or become bound by or seek to amend, terminate or waive any Contract with Patheon UK Limited or any of its Affiliates regarding the manufacture of any Product;

(y) grant any exclusive rights with respect to any Owned Company IP or divest any Owned Company IP, other than in the ordinary course of business consistent with past practices; or

(z) authorize any of, or commit, resolve or agree to take any of, the foregoing actions unless approved in advance by Parent in writing (such approval not to be unreasonably withheld, conditioned or delayed with respect to the matters described in clauses "(i)," "(j)," "(p)," "(r)," "(s)," "(t)" and "(u)" of this Section 5.2).

Notwithstanding the foregoing, nothing contained in this Agreement shall give Parent or Purchaser, directly or indirectly, the right to control or direct the operations of the Company or its Subsidiaries prior to the Acceptance Time. Prior to the Acceptance Time, the Company shall exercise, consistent with the terms and conditions of this Agreement, complete control and supervision over its and its Subsidiaries' respective businesses, assets and operations.

ARTICLE VI
ADDITIONAL AGREEMENTS

Section 6.1 No Solicitation.

(a) Subject to Section 6.1(b) hereof, from the Agreement Date until the Acceptance Time, or, if earlier, the valid termination of this Agreement in accordance with Section 7.1 hereof, the Company shall not, and the Company shall cause its Subsidiaries and their respective officers, directors, employees, investment bankers, attorneys, accountants, consultants and other agents and advisors (collectively, "Representatives") not to, directly or indirectly:

- (i) solicit, initiate or knowingly encourage (including by way of providing non-public information) the submission or announcement of any inquiries, proposals or offers that constitute or would reasonably be expected to result in an Acquisition Proposal;
- (ii) provide any non-public information concerning the Company or any of its Subsidiaries to any Person in connection with or in response to an Acquisition Proposal;
- (iii) engage in any discussions or negotiations with any Person with respect to any Acquisition Proposal;
- (iv) approve, adopt, endorse or recommend any Acquisition Proposal; or
- (v) enter into any agreement, letter of intent or similar document or any Contract contemplating or otherwise relating to any Acquisition Transaction or enter into any agreement, letter of intent or similar document requiring the Company to exempt any third party from the restrictions contained in any state takeover or similar Laws or otherwise cause such restrictions not to apply to such third party or to any Acquisition Proposal.

(b) Notwithstanding anything to the contrary contained in Section 6.1(a) hereof or any other provisions of this Agreement, if at any time prior to the Acceptance Time, (i) the Company has received a bona fide written Acquisition Proposal (other than an Acquisition Proposal that arises as a result of a breach of any of the provisions set forth in Section 6.1(a) hereof) that is reasonably likely to result in a Superior Proposal, (ii) the Company Board determines in good faith, after having consulted with outside legal counsel, that the failure to take such action would be inconsistent with its fiduciary obligations to the Company Shareholders and other stakeholders of the Company under applicable Law, (iii) prior to furnishing or making available any non-public information to, or entering into discussions or negotiations with such Person, the Company gives Parent written notice of the identity of the Person making the Acquisition Proposal and of the Company's intention to furnish or make available non-public information to, or enter into discussions or negotiations with, such Person, and the Company receives from such Person an executed confidentiality agreement containing provisions (including nondisclosure provisions and use restrictions) at least as favorable to the Company as the provisions of the Confidentiality Agreement, and (iv) simultaneously with furnishing or making available any non-

public information to such Person, the Company makes available such non-public information to Parent (to the extent the Company has not previously made available such non-public information to Parent), then the Company may (A) furnish information with respect to the Company and its Subsidiaries to the Person making such Acquisition Proposal and (B) engage in discussions or negotiations with the Person making such Acquisition Proposal regarding such Acquisition Proposal.

(c) The Company shall promptly (and, in any event, within one (1) Business Day) notify Parent in the event that the Company, any Subsidiary of the Company or any of their respective Representatives receives any Acquisition Proposal, including the identity of the Person making such Acquisition Proposal, and the material terms and conditions thereof and, if applicable, shall provide Parent with copies of any written requests, proposals or offers from the Person making such Acquisition Proposal promptly after receipt thereof. The Company shall keep Parent informed with respect to: (i) the status of any such Acquisition Proposal; and (ii) the status and material terms of any material modification thereto.

(d) The Company shall, and shall cause each of its Subsidiaries and its and their Representatives to, immediately cease and cause to be terminated any solicitations or discussions with any Person ongoing as of the date of this Agreement with respect to any actual or potential Acquisition Proposal.

(e) The Company agrees not to release or permit the release of any Person from, or to waive or permit the waiver of any provision of, any confidentiality, non-solicitation, no hire, "standstill" or similar Contract to which the Company is a party or under which the Company has any rights to the extent permitted thereunder, and will use commercially reasonable efforts to cause each such agreement to be enforced (it being understood that any automatic termination of any such provision resulting from the execution and delivery of this Agreement shall not constitute a breach of this Section 6.1(e)). The Company also shall promptly request each Person that has executed a confidentiality agreement with the Company or its Subsidiaries within the last 18 months in connection with its consideration of a possible Acquisition Transaction or a possible equity investment in the Company to return to the Company or destroy all confidential information heretofore furnished to such Person by or on behalf of the Company.

Section 6.2 Company Board Recommendation.

(a) Neither the Company Board nor any committee thereof shall (i) withdraw (or modify in a manner adverse to Parent or Purchaser), or propose (other than proposals made at any meeting of the Company Board solely among the participants at such meeting) or adopt any resolution to withdraw (or modify in a manner adverse to Parent or Purchaser), the Company Board Recommendation (it being understood that the Company Board Recommendation shall be deemed to have been modified in a manner adverse to Parent or Purchaser if it shall no longer be by the unanimous vote of all directors of the Company, other than those directors that have deemed themselves conflicted), (ii) resolve in favor of or recommend publicly or to any third party the adoption of any Acquisition Proposal, or (iii) cause or permit the Company or any of its Subsidiaries to execute or enter into, any letter of intent, memorandum of understanding, agreement in principle, merger agreement, acquisition agreement or other similar agreement providing for any Acquisition Proposal, other than any confidentiality agreement referred to in Section 6.1(b) hereof (an "**Acquisition Agreement**") (any action described in clause (i) being referred to as a "**Company Board Recommendation Change**").

(b) Notwithstanding anything to the contrary contained in this Agreement, at any time prior to the Acceptance Time, the Company Board may effect a Company Board Recommendation Change if: (i) an unsolicited, bona fide, written Acquisition Proposal that did not otherwise result from a breach of any provision of this Agreement or any “standstill” or similar agreement or provision under which the Company or any of its Subsidiaries has any rights is made to the Company and is not withdrawn, (ii) the Company provides to Parent written notice at least twenty-four (24) hours (or simultaneous with the Company Board if lesser notice is given to the Company Board) before any meeting of the Company Board at which the Company Board will consider the possibility of withdrawing the Company Board Recommendation or otherwise effecting a Company Board Recommendation Change in connection with such Acquisition Proposal, which notice specifies the reasons for holding such meeting; (iii) at such meeting, the Company Board determines in good faith (after consulting with the Company’s financial advisor and outside legal counsel) that such Acquisition Proposal constitutes a Superior Proposal, (iv) at such meeting, the Company Board determines in good faith, after consulting with its outside legal counsel, that, in light of such Superior Proposal, the failure to withdraw the Company Board Recommendation or otherwise effect a Company Board Recommendation Change would be inconsistent with its fiduciary obligations to the Company Shareholders and other stakeholders of the Company under applicable Law, (v) the Company promptly notifies Parent, in writing, at least four (4) Business Days before withdrawing the Company Board Recommendation or otherwise making any Company Board Recommendation Change: (A) that it has received a Superior Proposal that did not result from a breach of Section 6.1; (B) that the Company Board intends to withdraw the Company Board Recommendation or otherwise make an Company Board Recommendation Change and/or take action pursuant to Section 7.1(d)(ii) hereof as a result of such Superior Proposal; and (C) specifying the material terms and conditions of such Superior Proposal, including the identity of the Person making such Superior Proposal (and attaching the current version of any written agreement relating thereto) (it being understood and agreed that any change to the consideration payable in connection with such Superior Proposal or any other material modification thereto shall require a new advance written notice by the Company (except that the four (4) Business Day period referred to above shall be reduced to two (2) Business Days)), (vi) during such notice period, if requested by Parent, the Company engages in good faith negotiations with respect to any proposal made by Parent to amend this Agreement in such a manner that obviates the need for withdrawing the Company Board Recommendation or otherwise effecting a Company Board Recommendation Change, and (vii) at the time of any Company Board Recommendation Change, the Company Board confirms the determinations referred to in clauses (iii) and (iv) above (taking into account any changes to the terms of this Agreement proposed by Parent as a result of the negotiations required by clause (vi) above or otherwise). The Company shall notify Parent promptly upon any withdrawal of the Company Board Recommendation or other Company Board Recommendation Change.

(c) Notwithstanding anything to the contrary contained in this Agreement, at any time prior to the Acceptance Time, the Company Board may effect a Company Board Recommendation Change if: (i) there occurs or arises after the date of this Agreement a material event, material development or material change in circumstances that does not relate to any Acquisition Proposal and that was not known by or, if known, the consequences of which (A) were not known by the Company or any of its Subsidiaries on the Agreement Date, and (B) would not have been reasonably expected to have occurred, which event, development or change in circumstance, or any material consequences thereof, becomes known to the Company Board prior to the Acceptance Time (any such material event, material development or material change in circumstances unrelated to an Acquisition Proposal being referred to as an “*Intervening Event*”), (ii) the Company provides to Parent written

notice at least twenty-four (24) hours (or simultaneous with the Company Board if lesser notice is given to the Company Board) before any meeting of the Company Board at which the Company Board will consider the possibility of withdrawing the Company Board Recommendation or otherwise effecting a Company Board Recommendation Change in connection with such Intervening Event, which written notice specifies the reasons for holding such meeting and a reasonably detailed description of such Intervening Event; (iii) at such meeting, the Company Board determines in good faith, after consulting with the Company's outside legal counsel, that, in light of such Intervening Event, the failure to withdraw the Company Board Recommendation or otherwise effect a Company Board Recommendation Change would be inconsistent with its fiduciary obligations to the Company Shareholders and other stakeholders of the Company under applicable Law, (iv) the Company promptly notifies Parent, in writing, at least four (4) Business Days before making any Company Board Recommendation Change that the Company Board has determined that, in light of such Intervening Event, the failure to withdraw the Company Board Recommendation or otherwise effect a Company Board Recommendation Change would be inconsistent with its fiduciary obligations to the Company Shareholders and other stakeholders of the Company under applicable Law and that it intends to make an Company Board Recommendation Change as a result of such Intervening Event, (v) during such notice period, if requested by Parent, the Company engages in good faith negotiations with respect to any proposal made by Parent to amend this Agreement in such a manner that obviates the need for withdrawing the Company Board Recommendation or otherwise effecting a Company Board Recommendation Change, and (vi) at the time of any Company Board Recommendation Change, the Company Board confirms the determination referred to in clause (iii) above (taking into account any changes to the terms of this Agreement proposed by Parent as a result of the negotiations required by clause (v) or otherwise).

(d) Nothing contained in this Agreement shall prohibit the Company from (i) taking and disclosing to its shareholders a position contemplated by Rule 14d-9 or Rule 14e-2(a) promulgated under the Exchange Act or Item 1012(a) of Regulation M-A under the Exchange Act, or (ii) making any "stop-look-and-listen" communication to the Company Shareholders pursuant to Section 14d-9(f) promulgated under the Exchange Act (or any similar communications to the Company Shareholders), in the case of each of clauses (i) and (ii), in which the Company indicates that it has not made a Company Board Recommendation Change; provided, however, that this Section 6.2(d) shall not be deemed to permit the Company Board to make a Company Board Recommendation Change (it being understood that any such Company Board Recommendation Change shall be permitted only to the extent provided by Section 6.2(b) or Section 6.2(c) hereof).

Section 6.3 Reasonable Best Efforts to Complete.

(a) Subject to Section 6.2(b) and Section 6.2(c) hereof, upon the terms and subject to the conditions set forth in this Agreement, each of Parent, Purchaser and the Company shall use their reasonable best efforts to take, or cause to be taken, all actions, and to do, or cause to be done, and to assist and cooperate with the other party or parties hereto in doing, all things reasonably necessary, proper or advisable to consummate and make effective, in the most expeditious manner practicable, the transactions contemplated by this Agreement, including using reasonable best efforts to: (i) cause the conditions to the Offer set forth on Annex A hereto to be satisfied or fulfilled; and (ii) obtain all necessary actions or material non-actions, waivers, consents, approvals, Orders and authorizations from Governmental Entities, the expiration or termination of any applicable waiting periods, making all necessary registrations, declarations and filings (including registrations, declarations and filings with Governmental Entities, if any).

(b) Without limiting the generality of the foregoing provisions of Section 6.3(a) hereof, in the event that any Takeover Law is or becomes applicable to this Agreement or any of the transactions contemplated by this Agreement, each party shall use reasonable best efforts to ensure that the transactions contemplated by this Agreement may be consummated in the most expeditious manner practicable on the terms and subject to the conditions set forth in this Agreement, and otherwise to minimize the effect of such Takeover Law on this Agreement and the transactions contemplated hereby.

(c) Each of Parent, Purchaser and the Company shall file comparable pre-merger notification filings, forms and submissions with any Governmental Entity that may be required by any Antitrust Laws. Each of Parent, Purchaser and the Company shall use reasonable best efforts to promptly (i) cooperate and coordinate with the other in the making of such filings, (ii) supply the other with any information that may be required in order to effectuate such filings and (iii) supply any additional information that reasonably may be required or requested by any Governmental Entity charged with enforcing, applying, administering, or investigating any Law designed to prohibit, restrict or regulate actions for the purpose or effect of monopolization, restraining trade or abusing a dominant position (collectively, "**Antitrust Laws**"), including the FTC, the Antitrust Division of the DOJ, any attorney general of any state of the United States, the European Commission or any other competition authority of any jurisdiction ("**Antitrust Authority**"). Each party hereto shall use reasonable best efforts to promptly inform the other party or parties hereto, as the case may be, of any communication from any Governmental Entity regarding any of the transactions contemplated by this Agreement. If any party hereto or Affiliate thereof receives a request for additional information or documentary material from any such Governmental Entity with respect to the transactions contemplated by this Agreement, then such party shall use reasonable best efforts to make, or cause to be made, as soon as reasonably practicable and after consultation with the other party, an appropriate response in compliance with such request. None of Parent, Purchaser or the Company shall extend, directly or indirectly, any waiting periods required in connection with the Antitrust Laws, or enter into any agreement with any Governmental Entity to delay the consummation of the transactions contemplated hereby unless such action is consented to by the other party (which consent shall not be unreasonably withheld, delayed or conditioned).

Section 6.4 Access ; Financing.

(a) At all times during the period commencing with the execution and delivery of this Agreement and continuing until the earlier to occur of the termination of this Agreement pursuant to Article VII hereof and the Acceptance Time (the "**Pre-Closing Period**"), the Company shall afford Parent and its accountants, legal counsel and other representatives (including Parent's financing sources or prospective financing sources and their respective advisors and representatives) reasonable access during normal business hours, upon reasonable notice, to the properties, books and records and personnel of the Company to enable Parent to obtain all information concerning the business, including the status of product development efforts, properties, results of operations, commercialization of API and Products and personnel of the Company, as Parent may reasonably request; provided, however, that the terms and conditions of the Confidentiality Agreement shall apply to any information provided to Parent or Purchaser pursuant to this Section 6.4. Notwithstanding anything to the contrary set forth herein, the Company shall not be required to provide access to, or to disclose information, where such access or disclosure would reasonably be expected to jeopardize the attorney-client privilege (or similar protections) of the Company or its Subsidiaries or contravene any applicable Law or Contract entered into prior to the Agreement Date; provided, however, that, in such circumstance, the Company shall

cooperate with Parent to implement a procedure to permit access to or disclosure of such information in a manner that would not jeopardize the attorney-client privilege (or similar protections) or contravene such applicable Law or Contract.

(b) Each of Parent and Purchaser shall not permit any amendment or modification to be made to, or any waiver of any provision or remedy pursuant to, the Financing Commitment if such amendment, modification or waiver would or would reasonably be expected to delay or prevent in any material respect the ability of Purchaser to consummate the Offer.

(c) Unless Parent and/or Purchaser have otherwise obtained financing sufficient to consummate the Offer, each of Parent and Purchaser shall keep the Company informed on a reasonable basis and in reasonable detail of the status of its efforts to arrange the Financing and shall use commercially reasonable efforts to take or cause to be taken all actions necessary, proper and advisable to maintain in effect the Financing Commitment in accordance with the terms and subject to the conditions thereof and comply in all material respects with its obligations pursuant to the Financing Commitment; provided, however, that neither Parent nor Purchaser shall be required to, and shall not be required to cause any Person to, commence, participate in, pursue or defend any Action against or involving any Financing Sources. In the event that any portion of the Financing becomes unavailable on the terms and conditions contemplated in the Financing Commitment or otherwise reasonably acceptable to the Parent, each of Parent and Purchaser shall use commercially reasonable efforts to obtain alternative financing promptly following the occurrence of such event. Parent shall deliver to the Company a copy of any amendment, modification, waiver or supplement to the Financing Commitment that would reasonably be expected to materially impact the availability of the Financing or materially decrease the amount of financing that could be expected to be provided under the Financing Commitment as promptly as practicable after the execution thereof; provided that any such amendments, modification, waiver or supplement may be redacted to the extent requested by the applicable Financing Source. Parent and Purchaser acknowledge and agree that their obligations under this Agreement, including their obligations to consummate the Offer, are not subject to, or conditioned on, receipt of the Financing or any other financing.

(d) During the Pre-Closing Period, the Company shall use commercially reasonable efforts to provide Parent with all cooperation reasonably requested by Parent in connection with Parent's financing of the Offer and the other transactions contemplated by this Agreement, including by (i) providing to Parent information reasonably requested by Parent required for purposes of the financing and participate and make senior management of the Company available in meetings to discuss any such information, (ii) preparing and providing to Parent financial statements and projections (including providing audited financial statements for the year ending December 31, 2013 no later than March 31, 2014) reasonably requested by Parent for purposes of the financing, (iii) assisting in the preparation of those sections of any customary offering memoranda and other customary presentation materials, private placement memoranda, prospectuses and similar documents (including, if applicable, the delivery of one or more customary representation letters) that relate to the Company and its business, and (iv) obtaining the consent of, and customary comfort letters from, Reconta Ernst & Young S.p.A. (including by providing customary management letters and requesting legal letters to obtain such consent) if necessary or desirable for Parent's use of the Company's financial statements. Notwithstanding the foregoing, nothing herein shall require such cooperation to the extent it would (A) unreasonably disrupt the conduct of the business or operations of the Company or its Subsidiaries (in the reasonable judgment of the Company), (B) require the Company or any of its Subsidiaries to agree to

pay any fees, reimburse any expenses or otherwise incur any liability or give any indemnities prior to the Acceptance Time for which it is not promptly reimbursed or simultaneously indemnified or (C) require the Company or any of its Subsidiaries to take any action that would reasonably be expected to conflict with, or result in any violation or breach of, or default (with or without notice or lapse of time, or both) under, the Company Charter, the Company Bylaws, any applicable Laws, or any Contract. Parent may reasonably use logos of the Company solely in connection with the financing of the Offer; provided, that (1) such logos are used solely in a manner that is not intended to nor reasonably likely to harm or disparage any of the Company and its Subsidiaries or the reputation or goodwill of any of the Company and its Subsidiaries and its or their Intellectual Property Rights, (2) the logos shall be used in accordance with any written guidelines provided by the Company to Parent; and (3) Parent must immediately terminate use of such logos upon termination of this Agreement, and provided further that the Company shall have an opportunity to review and reasonably object to any use of the Company's and its Subsidiaries' logos prior to the dissemination of any materials in which such logos are used. Any goodwill resulting from any such use of the logos shall inure to the benefit of the Company or its Subsidiaries, as applicable.

Section 6.5 Certain Litigation. The Company shall promptly advise Parent in writing of any litigation commenced after the Agreement Date against the Company or any of its directors by any Company Shareholders (on their own behalf or on behalf of the Company), relating to this Agreement or the transactions contemplated hereby and shall keep Parent reasonably informed regarding any such litigation. The Company shall give Parent the opportunity to consult with the Company regarding the defense or settlement of any such shareholder litigation and shall consider in good faith Parent's views with respect to such litigation and shall not settle any such litigation without the prior written consent of Parent (which consent shall not be unreasonably withheld, conditioned or delayed).

Section 6.6 Public Disclosure. Parent and the Company shall consult with each other before issuing any press release or otherwise making any public statement with respect to this Agreement and the transactions contemplated hereby, and shall not issue any such press release or make any such public statement prior to such consultation, except as may be required by applicable Law or any listing agreement with a national securities exchange, in which case commercially reasonable efforts to consult with the other party hereto shall be made prior to any such release or public statement; provided, however, that, (a) each party may, without such consultation or consent, make any public statement in response to questions from the press, analysts, investors or those attending industry conferences and make internal announcements to employees, so long as such statements are consistent with previous press releases, public disclosures or public statements made jointly by the parties (or individually, if approved by the other party), and (b) the Company shall not be required to consult or agree with Parent in connection with any public disclosure, including issuing any factually accurate public statement by the Company that describes the Company's receipt of an Acquisition Proposal and the operation of this Agreement with respect thereto or "stop-look-and listen communication" pursuant to, or otherwise complying with, the provisions of Rule 14d-9 promulgated under the Exchange Act or any similar communication to the Company Shareholders, in connection with any Acquisition Proposal or Superior Proposal, except as expressly required by Section 6.1 and Section 6.2 hereof. The Company shall consult with Parent and consider the views and comments of Parent before the Company, any Subsidiary of the Company or any of their respective Representatives sends any emails or other documents to the employees of the Company generally or otherwise communicates with such employees generally, with respect to the Offer or any of the other transactions contemplated by this Agreement.

Section 6.7 Treatment of Equity Awards.

(a) As soon as practicable following the Agreement Date (and, in any event, prior to the Acceptance Time), the Company Board shall adopt such resolutions and take such other actions (including obtaining any required consents from holders of Company Options and amending the Company Equity Plans and the applicable award agreements) as may be required to (i) provide that all outstanding and unvested Company Options shall become fully vested immediately prior to the Acceptance Time, and (ii) provide notice to holders of Company Options that all Company Options that are outstanding as of the Acceptance Time may be exercised for Company Shares immediately following the Acceptance Time and, prior to the expiration date of the subsequent offering period with respect to the Offer, as it may be extended, may tender such Company Shares in exchange for the Offer Price pursuant to the Offer, as extended by such subsequent offering period, and the terms and conditions set forth in this Agreement.

(b) As soon as practicable following the Agreement Date, the Company shall provide notice to each holder of outstanding Company Options (except directors and the consultants, independent contractors, member of senior management or any executive officer of the Company as defined under the Sarbanes-Oxley Act or Italian Law or any officer of the Company or a Subsidiary for whom such an arrangement would violate any applicable Law) that such holder may elect to pay the aggregate exercise price for the exercise of such holder's outstanding vested Company Options (including Company Options that vested pursuant to Section 6.7(a)(i)) with the funds made available by the Company (on terms and conditions that conform to applicable Law) in consideration for such holder's irrevocable instructions to the Company or its broker to (i) exercise such Company Options for Company Shares after the Acceptance Time and, (ii) prior to the expiration date of the subsequent offering period with respect to the Offer, as it may be extended, tender each such Company Share in exchange for the Offer Price pursuant to the Offer, as extended by such subsequent offering period, and the terms and conditions set forth in this Agreement.

(c) The Company and its Subsidiaries agree to comply with applicable Law requiring withholding, reporting or remittance of any Taxes due with respect to any Company Options that are exercised following the Acceptance Time.

Section 6.8 Employee Matters.

(a) The Company shall, within fifteen (15) days following the Agreement Date, deliver to Parent an accurate and complete list as of the Agreement Date of all employees and independent contractors or consultants currently providing services to the Company or any of its Subsidiaries and indicate as of the Agreement Date for each: employing / engaging entity; classification (including notation of executive officer status with the Company); date of hire / engagement or date of seniority if different; location where services are regularly performed; current salary, wage, actual bonus and/or target bonus opportunity, and/or commission rate, as applicable; accrued vacation/paid-time off; and any bonuses declared, but not paid.

(b) Unless a higher protection is given to the Continuing Employees under applicable Law or any such Continuing Employee agrees otherwise, for a period of at least twelve (12) months following the Acceptance Time, Parent shall, and shall cause its Subsidiaries to, provide to all Continuing Employees outside of the United States compensation and benefits (not including stock

options or other equity compensation, retention or bonuses out of the ordinary not otherwise agreed to in Section 6.8(c)) that are in the aggregate, no less favorable than the compensation and benefits being provided to Continuing Employees outside of the United States by the Company immediately prior to the Acceptance Time.

(c) Parent shall cause its Subsidiaries to honor in accordance with their terms as in effect immediately prior to the Acceptance Time all existing employment, change of control, severance and retention arrangements between the Company or any of its Subsidiaries, on the one hand, and any current or former employee, director or consultant of the Company or any of its Subsidiaries, on the other hand, and shall recognize for such purposes continuous service with the Company or its Subsidiaries, including predecessor employers, of participating Continuing Employees under such plans; provided, however, that nothing in this Agreement shall prohibit Parent or its Subsidiaries from amending or terminating any such plans, programs, agreements or arrangements, so long as such amendment or termination complies with the terms of any such plans, programs, agreements or arrangements and applicable Law, including specifically obtaining any necessary or required consents

(d) Notwithstanding anything in this Agreement to the contrary, the terms and conditions of employment after the Acceptance Time for Continuing Employees whose employment is subject to a collective bargaining agreement, works council agreement or similar Contract with any labor organization, works council or employee association shall be governed by such agreement or Contract until its expiration, modification or termination in accordance with its terms and applicable Law.

(e) This Section 6.8 shall be binding upon and inure solely to the benefit of each of the parties to this Agreement, and nothing in this Section 6.8, express or implied, is intended to confer upon any other Person any rights or remedies of any nature whatsoever under or by reason of this Section 6.8.

(f) Unless otherwise requested by Parent in writing at least five days prior to the Acceptance Time, the Company agrees to take (or cause to be taken) all actions necessary or appropriate to terminate, effective as of the day immediately prior to, and conditioned upon the occurrence of, the Acceptance Time, any Employee Benefit Plan that is intended to be qualified under Section 401(k) of the Code (the "**401(k) Plan**") and shall, prior to and conditioned upon such termination, fully vest any and all unvested amounts of the accounts of all individuals who are participants under the 401(k) Plan at the time of such termination. The form and substance of any resolutions providing for the termination of the 401(k) Plan shall be subject to the review and approval of Parent, which shall not be unreasonably withheld, conditioned or delayed. The Company shall deliver to Parent an executed copy of such resolutions as soon as practicable following adoption of such resolutions and shall fully comply with such resolutions.

Section 6.9 Directors' and Officers' Indemnification and Insurance.

(a) For five (5) years after the Acceptance Time, Parent and Purchaser shall cause the Company and its Subsidiaries and successors to honor and fulfill in all respects the obligations of the Company and its Subsidiaries under any and all indemnification agreements in effect as of the Agreement Date between the Company or any of its Subsidiaries and any of their respective current or former directors and officers and any Person who becomes a director or officer of the Company or any

of its Subsidiaries prior to the Acceptance Time (the “*Indemnified Parties*”) with respect to actions taken by such Person in his or her capacity as a director or officer of the Company or its Subsidiary; provided, however, that in the event any claim or claims are asserted or made within such period, all rights to indemnification in respect of any such claim or claims shall continue until disposition of any and all such claims.

(b) For a period of five (5) years after the Acceptance Time, Parent and the Company shall maintain in effect the Company’s current directors’ and officers’ liability insurance (“*D&O Insurance*”) in respect of acts or omissions occurring at or prior to the Acceptance Time, covering each person covered by the D&O Insurance immediately prior to the Acceptance Time, on terms with respect to the coverage, retention, limitations of liability and amounts no less favorable, in the aggregate, than those of the D&O Insurance in effect on the Agreement Date and with an insurance company with the same or better credit rating as the insurance company providing the current D&O Insurance; provided, however, that the Company may, at its option, (i) substitute therefor policies of Parent or Purchaser or any of their respective Subsidiaries containing terms with respect to coverage, retention, limitations of liability and amounts no less favorable, in the aggregate, to such persons than the D&O Insurance or (ii) request that the Company obtain such extended reporting period coverage under its existing insurance program (to be effective as of the Acceptance Time); provided further, however, that in satisfying its obligations under this Section 6.9(b), none of Parent, Purchaser or the Company shall be obligated to pay annual premiums in excess of two hundred fifty percent (250%) of the annual premium for coverage under the D&O Insurance in effect at the Agreement Date (such two hundred fifty percent (250%) amount, the “*Maximum Annual Premium*”); provided, further, that if the annual premiums of such insurance coverage exceed such amount, Parent, Purchaser and the Company shall be obligated to obtain a policy with the greatest coverage available for a cost not exceeding the Maximum Annual Premium. Prior to the Acceptance Time, notwithstanding anything to the contrary set forth in this Agreement, Parent, Purchaser or the Company may purchase a five-year “tail” prepaid policy on the D&O Insurance containing terms with respect to coverage, retention, limitations of liability and amounts no less favorable, in the aggregate, than the D&O Insurance in effect on the Agreement Date and with an insurance company with the same or better credit rating as the insurance company providing the current D&O Insurance. In the event that Parent, Purchaser or the Company purchases such a “tail” policy prior to the Acceptance Time, Parent, Purchaser and the Company, as applicable, shall maintain such “tail” policy in full force and effect through such five-year period, in lieu of all other obligations of Parent, Purchaser and the Company under the first sentence of this Section 6.9(b) for so long as such “tail” policy shall be maintained in full force and effect.

(c) If Purchaser or the Company or any of its successors or assigns shall (i) consolidate with or merge into any other Person and shall not be the continuing or surviving corporation or entity of such consolidation or merger or (ii) transfer all or substantially all of its properties and assets to any Person, then, and in each such case, proper provisions shall be made so that the successors and assigns of the Company shall assume all of the obligations of Parent, Purchaser and the Company set forth in this Section 6.9.

(d) The obligations under this Section 6.9 shall not be terminated, amended or otherwise modified in such a manner as to adversely affect any Indemnified Party (or any other person who is a beneficiary under the D&O Insurance or the “tail” policy referred to in Section 6.9(b) hereof (and their heirs and representatives)) without the prior written consent of such affected Indemnified Party or other person who is a beneficiary under the D&O Insurance or the “tail” policy referred to in

Section 6.9(b) hereof (and their heirs and representatives). Each of the Indemnified Parties or other persons who are beneficiaries under the D&O Insurance or the “tail” policy referred to in Section 6.9(b) hereof are intended to be third party beneficiaries of this Section 6.9, with full rights of enforcement as if a party thereto. The rights of the Indemnified Parties (and other persons who are beneficiaries under the D&O Insurance or the “tail” policy referred to in Section 6.9(b) hereof (and their heirs and representatives)) under this Section 6.9 shall be in addition to, and not in substitution for, any other rights that such persons may have under the certificate or articles of incorporation, bylaws or other equivalent organizational documents, any and all indemnification agreements of or entered into by the Company or any of its Subsidiaries, or applicable Law (whether at law or in equity).

Section 6.10 Rule 14d-10 Matters; Employment Compensation Approval. Prior to the expiration of the Offer, the Company will take all such steps as may be required to cause to be exempt under Rule 14d-10(d) promulgated under the Exchange Act any employment compensation, severance or employee benefit arrangements that have been or will be entered into after the date of this Agreement by the Company or its subsidiaries with current or future directors, officers or employees of the Company or its subsidiaries and to ensure that any such arrangements fall within the safe harbor provisions of such rule.

Section 6.11 Director and Statutory Auditors Resignations; Appointment of Directors and Statutory Auditors; Call of the Ordinary Shareholders’ Meeting. The Company acknowledge that certain directors of the Company have delivered written resignations, respectively, to the Company (in connection with the resignations under Section 6.11(a)) and to the Purchaser (in connection with the resignations under Section 6.11(b)), pursuant to which, subject to Offer Closing, the resignations of (a) three (3) of such individuals will become effective as of the Acceptance Time (the “**Initial Director Resignations**”), and (b) up to three (3) of the other such individuals will become effective as of immediately following the effectiveness of the Acceptance Time Board Appointments (as defined below). Furthermore, the Company shall use reasonable efforts to obtain written resignations of all the Company’s Statutory Auditors (including the alternate members), pursuant to which each Statutory Auditor resigns from its office effective as of the date of the first shareholders’ meeting to be held following the Acceptance Time. The Company Board shall (x) subject to the Offer Closing, upon the Acceptance Time, take the resolution attached as Schedule 2 (the “**Acceptance Time Board Appointments**”) and (y) no later than ten (10) Business Days after the Acceptance Time, the Company Board shall issue the notice of call and take all reasonable actions to cause an ordinary shareholders’ meeting of the Company to be convened in compliance with the Company Bylaws, applicable Laws and Company past practice at the offices of the Company at Piazza XX settembre 2, Villa Guardia (Como), Italy, on February 24, 2014 at 10:00 a.m. (Milan time), on first call, and on February 25, 2014 at 10:00 a.m. (Milan time), on second call, with record date on February 7, 2014, with the following agenda: 1. Release of the resigning directors and 2. Appointment of new members of the Company’s corporate bodies to fill the vacancies created by the resigning members and any related and consequent resolutions, it being understood that if by February 7, 2014 the Offer Closing has not occurred, the Company Board will be free to revoke the notice of call of the shareholders’ meeting. The Company, Parent and Purchaser do not guarantee that the resolutions to be taken under this Section 6.11 and Section 6.12 below, once voted, will be valid and effective.

Section 6.12 Release of the Resigning Directors. At the ordinary shareholders meeting of the Company set forth under Section 6.11, Parent and Purchaser agree to propose, and to vote in favor of, the resolution set forth in Schedule 1.

Section 6.13 Obligations of Purchaser. Parent shall take all actions necessary to cause Purchaser and, following the Acceptance Time, the Company, to perform their respective obligations under this Agreement and to consummate the transactions contemplated by this Agreement including the Offer, upon the terms and subject to the conditions set forth in this Agreement.

Section 6.14 Confidentiality Agreements. The Company shall use commercially reasonable efforts to ensure that, prior to the Acceptance Time, each Person identified on Schedule 3, and each other individual as reasonably requested by Parent, enters into a confidentiality and intellectual property assignment agreement in favor of the Company or its Subsidiary, as applicable, in form and substance reasonably satisfactory to Parent.

ARTICLE VII

TERMINATION, AMENDMENT AND WAIVER

Section 7.1 Termination Prior to the Acceptance Time. This Agreement may be terminated and the Offer may be abandoned at any time prior to the Acceptance Time, provided that the party desiring to terminate this Agreement pursuant to this Section 7.1 (other than pursuant to Section 7.1(a)) hereof shall give notice of such termination to the other party or parties hereto, as the case may be:

(a) by the mutual written consent of the Company and Parent;

(b) by either Parent or the Company, if the Offer shall have expired or been terminated in accordance with the terms hereof without Purchaser having accepted for payment any Company Shares pursuant to the Offer on or before April 19, 2014 (the “**Termination Date**”); provided, however, that in the event that, on the Termination Date, the only conditions to the Offer set forth in Annex A hereto that have not been satisfied or waived on or prior to the Termination Date are the conditions set forth in (x) clause (B)(7) of Annex A or (y) clause (B)(7) of Annex A and either or both of clauses (A) and (B)(5) of Annex A hereto, then either Parent or the Company may elect to extend the Termination Date, by written notice to the other prior to or on the Termination Date, until September 19, 2014 (the “**Extended Termination Date**”) so long as the condition set forth in clause (B)(7) would be reasonably likely to be satisfied by such Extended Termination Date if the parties used their reasonable best efforts to do so; provided, further, that the right to terminate this Agreement under this Section 7.1(b) shall not be available to any party hereto whose action or failure to fulfill any obligation under this Agreement has been a principal cause of or resulted in (i) any of the conditions to the Offer set forth in Annex A hereto having failed to be satisfied on or before the Termination Date or the Extended Termination Date, as applicable, or (ii) the expiration or termination of the Offer in accordance with the terms hereof without Purchaser having accepted for payment any Company Shares pursuant to the Offer;

(c) by either Parent or the Company, if any Governmental Entity of competent jurisdiction shall have (i) enacted, issued, promulgated, entered, enforced or deemed applicable to the Offer any Law that is in effect and has the effect of making the Offer illegal or that has the effect of prohibiting or otherwise preventing the Offer or (ii) issued or granted any Order that is in effect and has the effect of making the Offer illegal or that has the effect of prohibiting or otherwise preventing the Offer, and such Order has become final and non-appealable;

(d) by the Company:

(i) in the event (A) of a breach of any covenant or agreement on the part of Parent or Purchaser set forth in this Agreement or (B) that any of the representations and warranties of Parent or Purchaser set forth in this Agreement shall have been inaccurate when made or shall have become inaccurate, in the case of each of clauses (A) and (B), so as to prevent Purchaser from consummating the Offer in accordance with the terms hereof; provided, however, that notwithstanding the foregoing, in the event that such breach by Parent or Purchaser or such inaccuracies in the representations and warranties of Parent or Purchaser are curable by Parent or Purchaser through the exercise of commercially reasonable efforts prior to the date that is thirty (30) days after the date on which Parent is notified by the Company in writing of such breach or inaccuracy, or, if earlier, the Termination Date (or Extended Termination Date, as applicable), then the Company shall not be permitted to terminate this Agreement pursuant to this Section 7.1(d)(i) on account of such breach or inaccuracy until the earlier of: (1) the end of such thirty (30) day cure period, or, if earlier, the Termination Date (or Extended Termination Date, as applicable), or (2) Parent ceasing to exercise commercially reasonable efforts to cure such breach or inaccuracy following delivery of written notice from the Company to Parent of such breach or inaccuracy (it being understood that the Company may not terminate this Agreement pursuant to this Section 7.1(d)(i) if such breach or inaccuracy by Parent or Purchaser is cured within the applicable time period); or

(ii) immediately prior to or concurrently with entering into a binding, written, definitive Acquisition Agreement providing for the consummation of a Superior Proposal (a "**Specified Definitive Acquisition Agreement**") if: (A) the Company has not materially breached the terms of Section 6.1 or Section 6.2 hereof, (B) subject to complying with the terms of Section 6.2(b) hereof, the Company Board has effected a Company Board Recommendation Change and authorized the Company to enter into the Specified Definitive Acquisition Agreement; and (C) the Company, immediately prior to or concurrently with such termination, pays (or causes to be paid) to Parent the payment required to be made to Parent pursuant to Section 7.3(b)(i) hereof; or

(e) by Parent:

(i) in the event (A) of a breach of any covenant or agreement on the part of the Company set forth in this Agreement such that the condition to the Offer set forth in clause (B)(3) of Annex A hereto would not be satisfied, or (B) that any representation or warranty of the Company set forth in this Agreement shall not have been true and correct when made or shall have thereafter become untrue or incorrect (in each case determined after giving effect to the provisos set forth in clause (B)(1) or clause (B)(2) of Annex A hereto, as applicable) such that any condition to the Offer set forth in clause (B)(1) or clause (B)(2) of Annex A hereto would not be satisfied; provided, however, that notwithstanding the foregoing, in the event that such breach by the Company or such inaccuracies in the representations and warranties of the Company are curable by the Company through the exercise of commercially reasonable efforts prior to the date that is thirty (30) days after the date on which the Company is notified by

Parent in writing of such breach or inaccuracy, or, if earlier, the Termination Date (or Extended Termination Date, as applicable), then Parent shall not be permitted to terminate this Agreement pursuant to this Section 7.1(e)(i) until the earlier of: (1) the end of such thirty (30) day cure period, or, if earlier, the Termination Date (or Extended Termination Date, as applicable), or (2) the Company ceasing to exercise commercially reasonable efforts to cure such breach or inaccuracy following delivery of written notice from Parent to the Company of such breach or inaccuracy (it being understood that Parent may not terminate this Agreement pursuant to this Section 7.1(e)(i) if such breach or inaccuracy by the Company is cured within the applicable time period); or

(ii) in the event that any of the following shall have occurred: (A) the Company Board or any committee thereof shall have for any reason effected a Company Board Recommendation Change; (B) the Company shall have failed to include the Company Board Recommendation in the Schedule 14D-9 or to permit Purchaser to include the Company Board Recommendation in the Offer Documents; (C) the Company Board or any committee thereof shall have for any reason approved, or recommended that the Company Shareholders approve, any Acquisition Proposal or Acquisition Transaction (whether or not a Superior Proposal); (D) an Acquisition Transaction Tender Offer shall have been made by a Person unaffiliated with Parent or Purchaser and, within ten (10) Business Days after notice of such Acquisition Transaction Tender Offer is first published, sent or given to the Company Shareholders, the Company shall not have made, pursuant to Rule 14e-2 under the Exchange Act, a statement that the Company recommends rejection of such offer and unconditionally reaffirming the Company Board Recommendation; (E) an Acquisition Proposal shall have been publicly disclosed, announced, commenced, submitted or made, and the Company Board fails to (1) publicly and unconditionally reaffirm the Company Board Recommendation and (2) either unconditionally reject or recommend, by the unanimous vote of all directors of the Company, other than those directors that have deemed themselves conflicted, that the Company Shareholders reject such Acquisition Proposal, as applicable, within ten (10) Business Days after Parent requests in writing that such recommendation be reaffirmed publicly; or (F) the Company, any Subsidiary of the Company or any of their respective Representatives shall have breached any of the provisions set forth in Section 6.1 hereof and such breach shall have resulted in or facilitated an Acquisition Proposal being announced, submitted or made.

The party desiring to terminate this Agreement pursuant to subsection (b), (c), (d) or (e) of this Section 7.1 shall give written notice of such termination to the other party in accordance with Section 8.2, specifying the provision or provisions hereof pursuant to which such termination is effected.

Section 7.2 Notice of Termination; Effect of Termination. Any proper termination of this Agreement pursuant to Section 7.1 hereof shall be effective immediately upon the delivery of written notice of the terminating party to the other party or parties hereto, as applicable. In the event of the termination of this Agreement pursuant to Section 7.1 hereof, this Agreement shall be of no further force or effect without liability of any party or parties hereto, as applicable (or any shareholder, director, officer, employee, agent, consultant or representative of such party or parties) to the other party or parties hereto, as applicable, except (a) for the terms of this Section 7.2, and Section 7.3 and Article VIII hereof, each of which shall survive the termination of this Agreement and (b) that nothing herein shall

relieve any party or parties hereto, as applicable, from liability for any willful and material breach of, or fraud in connection with, this Agreement. In addition to the foregoing, no termination of this Agreement shall affect the obligations of the parties hereto set forth in the Confidentiality Agreement, all of which obligations shall survive termination of this Agreement in accordance with their terms.

Section 7.3 Termination Fees and Expenses.

(a) General. All fees and expenses incurred in connection with this Agreement and the transactions contemplated hereby (including the Offer) shall be paid by the party or parties, as applicable, incurring such expenses whether or not the Offer is consummated.

(b) Termination Fees.

(i) In the event that this Agreement is terminated by the Company pursuant to Section 7.1(d)(ii) hereof, the Company shall pay (or cause to be paid) to Parent a non-refundable fee in the amount of \$25,300,000, by wire transfer of immediately available funds to an account or accounts designated in writing by Parent, immediately prior to or concurrent with such termination.

(ii) In the event that this Agreement is terminated (A) by Parent pursuant to Section 7.1(e) hereof, or (B) by Parent or the Company pursuant to any subsection of Section 7.1 hereof following the occurrence of any of the events referred to in Section 7.1(e)(ii) hereof, the Company shall pay (or cause to be paid) to Parent, by wire transfer of immediately available funds to an account or accounts designated in writing by Parent: (1) within two (2) Business Days of such termination, a non-refundable fee in the amount of \$10,100,000, and (2) if, within twelve (12) months after the date of any such termination, a Specified Acquisition Transaction (other than a Specified Acquisition Transaction in the form of a license of assets of the Company and/or its Subsidiaries) is consummated or a definitive agreement contemplating such a Specified Acquisition Transaction is executed, then a non-refundable fee in the amount of \$15,200,000 at the time such Specified Acquisition Transaction is consummated. A “**Specified Acquisition Transaction**” shall mean any Acquisition Transaction; provided, however, that for purposes of this definition, all references to “15%” in the definition of Acquisition Transaction shall be deemed to be references to “40%,” and all references to “85%” in the definition of Acquisition Transaction shall be deemed to be references to “60%.”

(iii) In the event that this Agreement is terminated by Parent or the Company pursuant to Section 7.1(b) hereof and: (A) at or prior to the time of such termination an Acquisition Proposal shall have been disclosed, announced, commenced, submitted or made; and (B) within twelve (12) months after the date of any such termination, a Specified Acquisition Transaction (whether or not relating to such Acquisition Proposal) is consummated or a definitive agreement contemplating a Specified Acquisition Transaction (whether or not relating to such Acquisition Proposal) is executed, then the Company shall pay (or cause to be paid) to Parent a non-refundable fee in the amount of \$25,300,000 (such non-refundable fee, and each other non-refundable fee set forth in clauses (i) and (ii) of this Section 7.3(b), a “**Termination**”

Fee”), by wire transfer of immediately available funds to an account or accounts designated in writing by Parent, at the time such Specified Acquisition Transaction is consummated; provided, however, that no Termination Fee shall be payable under this Section 7.3(b)(iii) if such Specified Acquisition Transaction is in the form of a license of assets of the Company and/or its Subsidiaries unless such Specified Acquisition Transaction is consummated by, or a definitive agreement contemplating such Acquisition Transaction is entered into with, any Person that disclosed, announced, commenced, submitted or made an Acquisition Proposal prior to the date of such termination, or any of such Person’s Affiliates.

(iv) The parties hereto acknowledge and agree that in no event shall the Company be required to pay any Termination Fee(s) pursuant to more than one of clauses (i), (ii) and (iii) of this Section 7.3(b), whether or not such Termination Fee(s) may be payable under more than one of such clauses, at the same or at different times and upon the occurrence of different events.

(v) Each of the Company and Parent acknowledges and agrees that the agreements contained in this Section 7.3 are an integral part of the transactions contemplated by this Agreement, and that, without these agreements, Parent would not have entered into this Agreement. Accordingly, if the Company fails to pay when due any amount payable under this Section 7.3, then: (A) the Company shall reimburse Parent for all costs and expenses (including fees and disbursements of counsel) incurred in connection with the collection of such overdue amount and the enforcement by Parent of its rights under this Section 7.3; and (B) the Company shall pay to Parent interest on such overdue amount (for the period commencing as of the date such overdue amount was originally required to be paid and ending on the date such overdue amount is actually paid to Parent in full) at a rate per annum equal to 300 basis points over the “prime rate” (as announced by Bank of America or any successor thereto) in effect on the date such overdue amount was originally required to be paid.

(vi) Upon payment of the applicable Termination Fee(s) in accordance with this Section 7.3(b), the Company will have no further liability or obligation to Purchaser under this Agreement, at Law or in equity or otherwise. Each of the parties acknowledge that a Termination Fee is not a penalty, but rather is liquidated damages in a reasonable amount that will compensate the Purchaser in the circumstances in which such Termination Fee is payable. Notwithstanding anything to the contrary contained in this Section 7.3(b)(vi) or elsewhere in this Agreement, this Section 7.3(b)(vi) shall not apply in the event of any breach of this Agreement by the Company, including any breach that results in the termination of this Agreement by Parent pursuant to Section 7.1(e)(i) hereof, or in the event of fraud.

Section 7.4 Amendment. Subject to applicable Law and subject to the other provisions of this Agreement, this Agreement may be amended by the parties hereto at any time by execution of an instrument in writing signed on behalf of each of Parent and the Company. To the extent that any amendment or modification to Section 7.4, Section 8.5, Section 8.10(b), Section 8.11 or Section 8.12 hereof is sought which is adverse to the rights of the Financing Sources, the prior written consent of the Financing Sources party to the Financing Commitment shall be required before such amendment or modification is rendered effective.

Section 7.5 Extension; Waiver. At any time and from time to time prior to the Acceptance Time, any party or parties hereto may, to the extent legally allowed and except as otherwise set forth herein, (a) extend the time for the performance of any of the obligations or other acts of the other party or parties hereto, as applicable, (b) waive any inaccuracies in the representations and warranties made to such party or parties hereto contained herein or in any document delivered pursuant hereto and (c) waive compliance with any of the agreements or conditions for the benefit of such party or parties hereto contained herein. Any agreement on the part of a party or parties hereto to any such extension or waiver shall be valid only if set forth in an instrument in writing signed on behalf of such party or parties, as applicable. Any delay in exercising any right under this Agreement shall not constitute a waiver of such right.

ARTICLE VIII

GENERAL PROVISIONS

Section 8.1 Survival of Representations, Warranties and Covenants. The representations, warranties and covenants of the Company, Parent and Purchaser contained in this Agreement shall terminate at the Offer Closing; provided that this Section 8.1 shall not limit any covenants that by their terms contemplate performance after the Offer Closing or after termination of this Agreement.

Section 8.2 Notices. All notices and other communications hereunder shall be in writing and shall be deemed given if delivered and received hereunder (i) immediately upon delivery personally, (ii) four (4) Business Days after being sent by registered or certified mail, return receipt requested, postage prepaid, (iii) one Business Day after being sent for next Business Day delivery, fees prepaid, via a reputable international overnight commercial delivery service, or (iv) immediately if sent via facsimile (if electronic receipt is confirmed), in case to the parties at the following addresses or facsimile numbers (or at such other address or facsimile numbers for a party as shall be specified by like notice):

(a) if to Parent or Purchaser, to:

Jazz Pharmaceuticals Public Limited Company
Fourth Floor, Connaught House,
One Burlington Road, Dublin 4, Ireland
Attention: Executive Vice President
Facsimile No.: +353 1 634 7850

with copies (which shall not constitute notice) to:

jazz_notices@jazzpharma.com

and to

Jazz Pharmaceuticals, Inc.
3180 Porter Drive
Palo Alto, CA 94304
Attention: General Counsel
Facsimile No.: (650) 496-3781

and to

Weil Gotshal & Manges LLP
201 Redwood Shores Parkway
Redwood Shores, California 94065
Attention: Keith A. Flaum
Jane Ross
Facsimile No.: 650-802-3100

(b) if to the Company, to:

Gentium S.p.A.
Piazza XX Settembre, 2
22079 Villa Guardia (CO)
Attention: General Counsel
Facsimile No.: +39-031-481784

with copies (which shall not constitute notice) to:

Skadden, Arps, Slate, Meagher & Flom (UK) LLP
40 Bank Street
London E14 5DS
United Kingdom
Attention: Lorenzo Corte
Facsimile No.: +44.20.7072.7025

and

Gianni, Origoni, Grippo, Cappelli & Partners
Piazza Belgioioso, 2
20100 Milano, Italy
Attention: Raimondo Premonte
Facsimile No.: 064871101

Section 8.3 Assignment. No party may assign either this Agreement or any of its rights, interests, or obligations hereunder without the prior written approval of the other parties; provided, however, that each of Parent and Purchaser may transfer or assign its rights and obligations hereunder, in whole or in part from time to time, to one or more wholly-owned Subsidiary of Parent; provided, further, however, that any such transfer or assignment shall not relieve either Parent or Purchaser of its obligations hereunder. Subject to the preceding sentence, this Agreement shall be binding upon and shall inure to the benefit of the parties hereto and their respective successors and permitted assigns.

Section 8.4 Entire Agreement. This Agreement and the documents and instruments and other agreements among the parties hereto as contemplated by or referred to herein, including the Company Disclosure Letter, constitute the entire agreement among the parties with respect to the subject matter hereof and supersede all prior agreements and understandings, both written and oral, among the parties with respect to the subject matter hereof; provided, however, the Confidentiality Agreement shall not be superseded, shall survive any termination of this Agreement and shall continue in full force and effect until the earlier to occur of (a) the Offer Closing and (b) the date on which the Confidentiality Agreement is terminated in accordance with its terms.

Section 8.5 Third Party Beneficiaries. Except (a) as set forth in or contemplated by the provisions of Section 6.9 hereof, and (b) following the Acceptance Time, the rights of the holders of Company Shares to receive cash in an amount equal to the Offer Price, this Agreement is not intended to confer upon any Person other than the parties hereto any rights or remedies hereunder; provided that that the Financing Sources shall be third party beneficiaries of Sections 8.9, 8.10, 8.11 and 8.12. The representations and warranties in this Agreement are the product of negotiations among the parties hereto and are for the sole benefit of the parties. Any inaccuracies in such representations and warranties are subject to waiver by the parties in accordance with Section 7.5 without notice or liability to any other Person. The representations and warranties in this Agreement may represent an allocation among the Parties of risks associated with particular matters.

Section 8.6 Severability. In the event that any provision of this Agreement, or the application thereof, becomes or is declared by a court of competent jurisdiction to be illegal, void or unenforceable, the remainder of this Agreement will continue in full force and effect and the application of such provision to other persons or circumstances will be interpreted so as reasonably to effect the intent of the parties hereto. The parties further agree to replace such void or unenforceable provision of this Agreement with a valid and enforceable provision that will achieve, to the extent possible, the economic, business and other purposes of such void or unenforceable provision.

Section 8.7 Other Remedies. Except as otherwise provided herein, any and all remedies herein expressly conferred upon a party will be deemed cumulative with and not exclusive of any other remedy conferred hereby, or by Law or equity upon such party, and the exercise by a party of any one remedy will not preclude the exercise of any other remedy.

Section 8.8 Specific Performance. The parties hereto agree that irreparable damage for which monetary damages, even if available, would not be an adequate remedy would occur in the event that any of the provisions of this Agreement were not performed in accordance with their specific terms or were otherwise breached. It is accordingly agreed that the parties shall be entitled to an injunction or injunctions, specific performance and other equitable relief to prevent or restrain breaches or threatened breaches of this Agreement and to enforce specifically the terms and provisions hereof in any court of competent jurisdiction, this being in addition to any other remedy to which they are entitled at Law or in equity. Each of the parties agrees that it will not oppose the granting of an injunction, specific performance and other equitable relief on the basis that any other party has an adequate remedy at law or in equity.

Section 8.9 Governing Law. This Agreement shall be governed by and construed in accordance with the Laws of the State of Delaware applicable to agreements made and to be performed entirely within such State, without regard to the conflict of laws principles of such State to the extent that the laws of another jurisdiction would be required thereby.

Section 8.10 Consent to Jurisdiction; Service of Process.

(a) With respect to any Action resulting from, relating to or arising out of this Agreement, or the rights and obligations arising hereunder, brought by any party hereto or its successors or assigns, each of the parties hereto irrevocably and unconditionally submits to the exclusive jurisdiction of the Delaware Court of Chancery and any state appellate court therefrom within the State of Delaware or, only if the Delaware Court of Chancery declines to accept jurisdiction over a particular Action, to the exclusive jurisdiction of any state court in the State of Delaware or the United States District Court for Delaware (the "**Designated Courts**"). In any such Action, each of the parties hereto irrevocably and unconditionally waives and agrees not to assert by way of motion, as a defense or otherwise (i) that its property is exempt or immune from attachment or execution, (ii) that such Action is brought in an inconvenient forum, (iii) that the venue of such Action is improper, (iv) that such Action should be transferred or removed to any court other than the Designated Courts, or should be stayed by reason of the pendency of some other proceeding in any other court other than the Designated Courts, or (v) that this Agreement or the subject matter hereof may not be enforced in or by the Designated Courts. Each of the parties hereto hereby agrees not to commence any such Action other than before the Designated Courts. Each of the parties hereto also hereby agrees that any final and unappealable judgment against a party in connection with any such Action shall be conclusive and binding on such party and that such judgment may be enforced in any court of competent jurisdiction, either within or outside of the U.S. A certified or exemplified copy of such judgment shall be conclusive evidence of the fact and amount of such judgment. With respect to any Action for which it has submitted to jurisdiction pursuant to this Section 8.10(a), each party irrevocably consents to service of process in the manner provided for the giving of notices pursuant to Section 8.2 hereof. Nothing in this Section 8.10(a) shall affect the right of any party to serve process in any other manner permitted by Law. The foregoing consent to jurisdiction shall not (a) constitute submission to jurisdiction or general consent to service of process in the State of Delaware for any purpose except with respect to any Action resulting from, relating to or arising out of this Agreement or (b) be deemed to confer rights on any Person other than the respective parties to this Agreement.

(b) Notwithstanding anything herein to the contrary, each of the parties hereto agrees, on behalf of itself and its Subsidiaries, that it will not bring or support any Action (whether at law, in equity, in contract, in tort or otherwise) against any Financing Source in any way relating to this Agreement or any of the transactions contemplated by this Agreement, including any dispute arising out of or relating in any way to the Financing Commitment, the Purchaser Credit Agreement or the performance thereof, in any forum other than the Supreme Court of the State of New York, County of New York, or, if under applicable Law exclusive jurisdiction is vested in the Federal courts, the United States District Court for the Southern District of New York (and appellate courts thereof). The provisions of this Section 8.10(b) shall be enforceable by each Financing Source.

Section 8.11 WAIVER OF JURY TRIAL. EACH OF PARENT, PURCHASER AND THE COMPANY HEREBY IRREVOCABLY WAIVES ALL RIGHT TO TRIAL BY JURY IN ANY ACTION, PROCEEDING OR COUNTERCLAIM (WHETHER BASED ON CONTRACT, TORT OR OTHERWISE) ARISING OUT OF OR RELATING TO THIS AGREEMENT, THE FINANCING COMMITMENT, PURCHASER CREDIT AGREEMENT OR THE ACTIONS OF PARENT, PURCHASER, THE COMPANY OR ANY FINANCING SOURCE IN THE NEGOTIATION, ADMINISTRATION, PERFORMANCE AND ENFORCEMENT HEREOF.

Section 8.12 Financing Sources. Notwithstanding anything to the contrary contained in this Agreement, the Company agrees, on behalf of itself and its Affiliates, that it will not bring or support any legal action, suit or proceeding (whether at law, in equity, in contract, in tort or otherwise) against any Financing Source in any way relating to this Agreement or any of the transactions contemplated by this Agreement (including the Debt Financing), including any dispute arising out of or relating in any way to the Debt Financing or the performance thereof. The provisions of this Section 8.12 shall enure to the benefit of, and be enforceable by, each Financing Source, each of which is hereby intended to be an express third party beneficiary of this Section 8.12.

Section 8.13 Counterparts. This Agreement may be executed in one or more counterparts, all of which shall be considered one and the same agreement and shall become effective when one or more counterparts have been signed by each of the parties and delivered to the other party, it being understood that all parties need not sign the same counterpart.

[Remainder of Page Intentionally Left Blank]

IN WITNESS WHEREOF, the undersigned have caused this **TENDER OFFER AGREEMENT** to be executed by their respective duly authorized officers to be effective as of the date first above written.

**JAZZ PHARMACEUTICALS PUBLIC LIMITED
COMPANY**

By: /s/ Fintan Keegan
Name: Fintan Keegan
Title: Executive Vice President

JAZZ PHARMACEUTICALS ITALY S.R.L.

By: /s/ Fintan Keegan
Name: Fintan Keegan
Title: Director

[Signature Page to Tender Offer Agreement]

IN WITNESS WHEREOF, the undersigned has caused this **TENDER OFFER AGREEMENT** to be executed by its duly authorized officer to be effective as of the date first above written.

GENTIUM S.P.A.

By: /s/ Khalid Islam

Name: Khalid Islam

Title: President & CEO

[Signature Page to Tender Offer Agreement]

ANNEX A

CONDITIONS TO THE OFFER

Notwithstanding any other provisions of the Offer, but subject to compliance with Section 2.1 of that certain Tender Offer Agreement, dated as of December 19, 2013 (the "**Agreement**"), by and among Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland ("**Parent**"), Jazz Pharmaceuticals Italy S.r.l., an Italian *società a responsabilità limitata* and a wholly owned subsidiary of Parent ("**Purchaser**") and Gentium S.p.A., a *società per azioni* incorporated in Italy (the "**Company**") (capitalized terms that are used but not otherwise defined in this Annex A shall have the respective meanings ascribed thereto in the Agreement), and in addition to (and not in limitation of) the rights and obligations of Purchaser to extend and/or amend the Offer at any time in its sole discretion (subject to the terms and conditions of the Agreement), Purchaser (i) shall not be required to accept for payment or, subject to any applicable rules and regulations of the SEC (including Rule 14e-1(c) under the Exchange Act (relating to the obligation of Purchaser to pay for or return tendered Company Shares promptly after termination or withdrawal of the Offer)), pay for, and (ii) may delay the acceptance for payment of or, subject to the restriction referred to above, the payment for, any tendered Company Shares, in the event that at or prior to the scheduled Offer Closing (as it may be extended pursuant to Section 2.1(c) of the Agreement):

(A) the Minimum Condition (as it may have been modified by Purchaser electing to effect a Permitted Minimum Condition Modification) shall not have been satisfied,

(B) any of the following shall have occurred and be continuing as of the scheduled expiration of the Offer:

(1) any of the representations and warranties of the Company set forth in this Agreement, other than the Specified Representations, shall not have been true and correct as of the date of this Agreement or shall not be true and correct as of the scheduled expiration of the Offer with the same force and effect as if made as of the scheduled expiration of the Offer (except to the extent expressly made as of an earlier date, in which case as of such date), except that any failure of any of such representations and warranties to be true and correct shall be disregarded if the circumstances giving rise to such failures to be true and correct would not, individually or in the aggregate, reasonably be expected to have or result in a Company Material Adverse Effect; provided, however, that, for purposes of determining whether such representations and warranties are true and correct: (x) all materiality or "Company Material Adverse Effect" qualifications limiting the scope of such representations and warranties shall be disregarded; and (y) any update of or modification to the Company Disclosure Letter made or purported to have been made after the execution and delivery of the Agreement shall be disregarded;

(2) any of the Specified Representations shall not have been true and correct in all material respects as of the date of this Agreement or shall not be true and correct in all material respects as of the scheduled expiration of the Offer with the same force and effect as if made as of the scheduled expiration of the Offer (except to the extent

expressly made as of an earlier date, in which case as of such date); provided, however, that, for purposes of determining whether such representations and warranties are true and correct any update of or modification to the Company Disclosure Letter made or purported to have been made after the execution and delivery of this Agreement shall be disregarded;

(3) the Company shall have failed to perform or comply in all material respects with its obligations required to be performed or complied with by it under the Agreement;

(4) since the date of the Agreement, any Company Material Adverse Effect shall have occurred;

(5) Parent shall not have received a certificate executed by the Company's Chief Executive Officer and Chief Financial Officer confirming that the conditions set forth in clauses "(B)(1)," "(B)(2)" and "(B)(3)" of this Annex A have been duly satisfied as of the scheduled expiration of the Offer;

(6) any temporary restraining order, preliminary or permanent injunction or other Order preventing the acquisition of or payment for Company Shares pursuant to the Offer shall have been issued by any court of competent jurisdiction or other Governmental Entity and remain in effect, or any Law is enacted or deemed applicable to the Offer that makes the acquisition of or payment for Company Shares pursuant to the Offer illegal or violative of any applicable Law;

(7) there shall be pending or overtly threatened any Action by any Governmental Entity of competent jurisdiction against Parent, Purchaser, the Company or any Subsidiary of the Company: (x) challenging or seeking to restrain or prohibit the acquisition of or payment for Company Shares pursuant to the Offer or any of the other transactions contemplated by the Agreement; (y) seeking to prohibit or limit in any material respect Parent or Purchaser's ability to vote, transfer, receive dividends with respect to or otherwise exercise ownership rights with respect to the stock of the Company; or (z) seeking to compel Parent, Purchaser, the Company or any of their respective Subsidiaries to dispose of or hold separate any Company Shares or any material assets as a result of the Offer or any of the other transactions contemplated by the Agreement;

(8) Parent shall not have received a resolution approved by the Company Board in the form set forth in Schedule 2; or

(9) the Agreement shall have been terminated in accordance with its terms.

The foregoing conditions (other than the Minimum Condition, as it may be modified by Purchaser electing to effect a Permitted Minimum Condition Modification) are for the sole benefit of Parent and Purchaser and, subject to the terms and conditions of the Agreement, may be waived by Parent or Purchaser, in whole or in part at any time and from time to time in the sole discretion of Parent and Purchaser. The failure by Parent or Purchaser at any time to exercise any of the foregoing rights shall not be deemed a waiver of any such right and each such right shall be deemed an ongoing right which may be asserted at any time and from time to time.

* * * * *

SUPPORT AGREEMENT

THIS SUPPORT AGREEMENT (“Agreement”) is entered into as of December 19, 2013, by and between **JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY**, a public limited company formed under the laws of Ireland (“**Parent**”), and (“**Securityholder**”).

RECITALS

A. Securityholder is (i) a holder of record and the “beneficial owner” (within the meaning of Rule 13d-3 under the Securities Exchange Act of 1934) of certain ordinary shares (“**Ordinary Shares**”) and/or American Depositary Shares, each representing one Ordinary Share (“**ADSs**” and, together with the Ordinary Shares, the “**Company Shares**”) and/or (ii) a holder of options to purchase Company Shares, of Gentium S.p.A., a *società per azioni* incorporated in Italy (the “**Company**”).

B. Parent, **JAZZ PHARMACEUTICALS S.R.L.**, a *società a responsabilità limitata* incorporated in Italy (“**Purchaser**”), and the Company are entering into a Tender Offer Agreement of even date herewith (the “**Tender Offer Agreement**”) that provides (subject to the conditions set forth therein) for Parent to acquire the Company by causing Purchaser to make a tender offer as contemplated by the Tender Offer Agreement (the “**Offer**”) for all of the issued and outstanding Company Shares.

C. Securityholder is entering into this Agreement in order to induce Parent to enter into the Tender Offer Agreement.

AGREEMENT

The parties to this Agreement, intending to be legally bound, agree as follows:

SECTION 1. CERTAIN DEFINITIONS

For purposes of this Agreement:

(a) Capitalized terms used but not otherwise defined in this Agreement have the meanings assigned to such terms in the Tender Offer Agreement.

(b) Securityholder shall be deemed to “**Own**” or to have acquired “**Ownership**” of a security if Securityholder: (i) is the record owner of such security; or (ii) is the “beneficial owner” (within the meaning of Rule 13d-3 under the Securities Exchange Act of 1934) of such security.

(c) “**Specified Individual**” shall mean any individual who is or was an employee (including any temporary or leased employees), consultant or independent contractor of the Company or any of its Subsidiaries at, or during the 90 days prior to, the Acceptance Time.

(d) “**Subject Securities**” shall mean: (i) all securities of the Company (including all Company Shares and all options, warrants and other rights to acquire Company Shares) Owned by Securityholder as of the date of this Agreement; and (ii) all additional securities of the Company (including all additional Company Shares and all additional options, warrants and other rights to acquire Company Shares) of which Securityholder acquires Ownership during the period from the date of this Agreement through the Support Termination Date.

(e) “**Subject Shares**” shall mean: (i) all Company Shares Owned by Securityholder as of the date of this Agreement; (ii) all additional Company Shares of which Securityholder acquires Ownership during the period from the date of this Agreement through the Support Termination Date; and (iii) all securities into which any Company Shares described in clause “(i)” or clause “(ii)” above are exchanged or converted.

(f) “**Support Period**” shall mean the period commencing on (and including) the date of this Agreement and ending on (and including) the Support Termination Date.

(g) “**Support Termination Date**” shall mean the earlier of: (A) the date on which the Acceptance Time occurs; and (B) the date upon which the Tender Offer Agreement is validly terminated; *provided, however*, that if the Tender Offer Agreement is terminated (1) pursuant to Section 7.1(b) thereof and an Acquisition Proposal shall have been disclosed, announced, commenced, submitted or made, at or prior to the time of such termination, (2) pursuant to Section 7.1(d)(ii) thereof, (3) pursuant to Section 7.1(e)(ii) thereof, or (4) pursuant to any subsection of Section 7.1 thereof following the occurrence of any of the events referred to in Section 7.1(e)(ii) thereof, then the Support Termination Date shall be the _____ month anniversary of the date on which the Tender Offer Agreement is so terminated.

(h) A Person shall be deemed to have effected a “**Transfer**” of a security if such Person directly or indirectly: (i) sells, pledges, encumbers, grants an option with respect to, transfers or disposes of such security or any interest in such security to any Person other than Parent; (ii) enters into an agreement or commitment contemplating the possible sale of, pledge of, encumbrance of, grant of an option with respect to, transfer of or disposition of such security or any interest therein to any Person other than Parent; or (iii) reduces such Person’s beneficial ownership of, interest in or risk relating to such security.

SECTION 2. TRANSFER OF SUBJECT SECURITIES AND VOTING RIGHTS

2.1 Restriction on Transfer of Subject Securities. Subject to Section 2.3, during the Support Period, Securityholder shall not, directly or indirectly: (a) cause or permit any Transfer of any of the Subject Securities to be effected; or (b) enter into or become bound by any letter of intent, memorandum of understanding, agreement in principle, agreement to tender, merger agreement, acquisition agreement or other Contract contemplating any Transfer of any Subject Securities.

2.2 Restriction on Transfer of Voting Rights. During the Support Period, Securityholder shall ensure that: (a) none of the Subject Securities is deposited into a voting trust; and (b) no proxy or power of attorney is granted that is inconsistent with this Agreement, and no tender, voting or similar agreement is entered into, with respect to any of the Subject Securities; and (c) no action is otherwise taken with respect to the Subject Securities that would in any way restrict, limit or interfere with the performance of Securityholder’s obligations hereunder or the transactions contemplated hereby.

2.3 Permitted Transfers. Section 2.1 shall not prohibit a transfer of Subject Securities by Securityholder: (a) if Securityholder is an individual: (i) to any member of Securityholder’s immediate family, or to a trust for the benefit of Securityholder or any member of Securityholder’s immediate family, or (ii) upon the death of Securityholder; or (b) if Securityholder is a partnership or limited liability company, to one or more partners or members of Securityholder or to an affiliated corporation under common control with Securityholder; *provided, however*, that a transfer referred to in this sentence shall be permitted only if, as a precondition to such transfer, the transferee agrees in a writing, reasonably satisfactory in form and substance to Parent, to be bound by all of the terms of this Agreement.

SECTION 3. AGREEMENT TO TENDER

3.1 Tender of Subject Shares. Securityholder agrees to promptly (and, in any event, not later than five (5) Business Days after commencement of the Offer) validly tender or cause to be validly tendered into the Offer, pursuant to and in accordance with the terms of the Offer and Rule 14d-2 under the Securities Exchange Act of 1934, all of the Subject Shares Owned by Securityholder as of the date of this Agreement (free and clear of any encumbrances or restrictions), and if Securityholder acquires Ownership of any additional Subject Shares after the date of this Agreement, to promptly (and, in any event, not later than two (2) Business Days after Securityholder acquires Ownership of such additional Subject Shares) validly tender or cause to be validly tendered into the Offer, pursuant to and in accordance with the terms of the Offer, all of such additional Subject Shares (free and clear of any encumbrances or restrictions).

3.2 No Withdrawal. Securityholder agrees not to withdraw or cause to be withdrawn any of the Subject Shares from the Offer unless and until the Offer expires without Purchaser having accepted for payment Company Shares validly tendered in the Offer.

3.3 Conditional Obligation. Securityholder acknowledges and agrees that Purchaser's obligation to accept for payment Company Shares in the Offer, including any Subject Shares tendered by Securityholder, is subject to the terms and conditions of the Tender Offer Agreement and the Offer.

3.4 Exercise of Options. If the Offer is extended for a subsequent offering period (within the meaning of Section 14d-11 under the Securities Exchange Act of 1934), then Securityholder hereby agrees to exercise, contingent upon the commencement of such subsequent offering period, and with effect as of immediately following the commencement thereof, all of Securityholder's options to purchase Company Shares that are vested and outstanding as of the Acceptance Time. Securityholder agrees to validly tender or cause to be validly tendered into the subsequent offering period all of the Subject Shares issued to Securityholder upon the exercise of such options (free and clear of any encumbrances or restrictions) promptly following the date of such exercise.

SECTION 4. VOTING OF SHARES

4.1 Voting Covenant. Securityholder hereby agrees that, during the Support Period, at any ordinary or extraordinary meeting of the shareholders of the Company, however called, unless otherwise directed in writing by Parent, Securityholder shall cause the Subject Shares to be voted against the following actions (each, a "**Specified Action**"): (a) any Acquisition Transaction; (b) any change in a majority of the board of directors of the Company; (c) any amendment to the Company's articles of association or bylaws; (d) any material change in the capitalization of the Company or the Company's corporate structure; and (e) any other action which is intended, or could reasonably be expected, to impede, interfere with, delay, postpone, discourage or adversely affect the Offer or any of the other transactions contemplated by the Tender Offer Agreement or this Agreement. During the Support Period, Securityholder shall not enter into any agreement or understanding with any Person to vote or give instructions in any manner inconsistent with the preceding sentence.

4.2 Proxies.

(a) If any ordinary or extraordinary meeting of the shareholders of the Company is called during the Support Period, the Securityholder shall:

(i) promptly (and in no event less than ten (10) Business Days prior to the date of the first call for such meeting) notify Parent in writing that such meeting has been called and deliver to Parent a copy of any written notice, information circular or other materials received by Securityholder in connection with such meeting;

(ii) deliver to Parent, at least three (3) Business Days prior to the date of the meeting for the first call of such meeting, a proxy, in form and substance reasonably satisfactory to Parent, executed by Securityholder, which shall be irrevocable to the fullest extent permitted by applicable Law, with respect to any outstanding Company Shares that are owned of record by Securityholder; and

(iii) cause to be delivered to Parent, at least three (3) Business Days prior to the date of meeting for the first call of such meeting, additional proxies, in form and substance reasonably satisfactory to Parent, executed on behalf of the record owner of any outstanding Company Shares that are owned beneficially (within the meaning of Rule 13d-3 under the Securities Exchange Act of 1934), but not of record, by Securityholder, which shall be irrevocable to the fullest extent permitted by applicable Law.

(b) The Securityholder hereby revokes (and agrees to cause to be revoked) all proxies, if any, that it has heretofore granted with respect to its Subject Shares, to the extent such proxies relate to, or purport to grant any voting rights with respect to, any Specified Action.

SECTION 5. NO SOLICITATION

Securityholder agrees that, from the date of this Agreement through the earlier of (a) the Acceptance Time or (b) the date upon which the Tender Offer Agreement is validly terminated, Securityholder shall not, and Securityholder shall cause its Subsidiaries (if applicable) and their respective Representatives not to, directly or indirectly: (i) solicit, initiate or knowingly encourage (including by way of providing non-public information) the submission or announcement of any inquiries, proposals or offers that constitute or would reasonably be expected to result in an Acquisition Proposal; (ii) provide any non-public information concerning the Company or any of its Subsidiaries to any Person in connection with or in response to an Acquisition Proposal; (iii) engage in any discussions or negotiations with any Person with respect to any Acquisition Proposal; (iv) approve, adopt, endorse or recommend any Acquisition Proposal; or (v) enter into any letter of intent or similar document or any Contract contemplating or otherwise relating to any Acquisition Transaction. Securityholder shall immediately cease and discontinue, and Securityholder shall ensure that its Subsidiaries (if applicable) and their respective Representatives immediately cease and discontinue, any existing discussions with any Person that relate to any Acquisition Proposal.

SECTION 6. NON-SOLICITATION AND NON-COMPETITION COVENANTS

6.1 Non-Solicitation. Securityholder agrees that, for a period of _____ months following the Acceptance Time, Securityholder shall not, and Securityholder shall ensure that Securityholder's Affiliates do not, directly or indirectly, personally or through others, encourage, induce, attempt to induce, solicit or attempt to solicit (on Securityholder's own behalf or on behalf of any other Person) any

Specified Individual to leave his or her employment, consulting or independent contractor relationship with the Company or any of the Company's Subsidiaries; *provided, however*, that Securityholder shall not be prohibited from conducting generalized solicitations for employees, including through the use of media advertisements and over the internet, provided that such solicitations are not specifically targeted at any Specified Individual(s).

6.2 Non-Competition. Securityholder agrees that, without prejudice for the loyalty obligations of the Securityholder under article 2105 of the Italian civil code, that substantiate the prohibition to carry out competing activities during employment, during any notice period and in any case in the period elapsing from the date of termination of the Securityholder's employment with the Company (whenever this will occur) and his confidentiality obligations both during employment and after its termination, as an essential condition to this Support Agreement, the Securityholder shall also be bound for a period of years following the Acceptance Time not to engage, directly or indirectly, even if for free, as proprietor, partner, shareholder, director, employee, agent, consultant, or in any other capacity or manner, in any activity, also for free, consisting of the research, development, manufacture, production, promotion, sale and commercialization of (a) any agent for the treatment or prevention of veno-occlusive disease (sinusoidal obstruction syndrome) or graft versus host disease and (b) any polydisperse agent comprising deoxyribonucleic acid fragments. This non-competition covenant will be enforceable within the territory of any country within Europe, Canada, Japan, or the United States.

SECTION 7. REPRESENTATIONS AND WARRANTIES OF SECURITYHOLDER

Securityholder hereby represents and warrants to Parent as follows:

7.1 Authorization, etc. Securityholder has the absolute and unrestricted right, power, authority and capacity to execute and deliver this Agreement and to perform Securityholder's obligations hereunder and thereunder. This Agreement has been duly executed and delivered by Securityholder and constitutes the legal, valid and binding obligation of Securityholder, enforceable against Securityholder in accordance with its terms, subject to (a) bankruptcy, insolvency, reorganization, moratorium and similar Laws affecting the enforcement of creditors' rights or remedies in general as from time to time in effect or (b) the exercise by courts of equity powers. If Securityholder is a corporation, then Securityholder is a corporation duly organized, validly existing and in good standing under the laws of the jurisdiction in which it was organized. If Securityholder is a general or limited partnership, then Securityholder is a partnership duly organized, validly existing and in good standing under the laws of the jurisdiction in which it was organized. If Securityholder is a limited liability company, then Securityholder is a limited liability company duly organized, validly existing and in good standing under the laws of the jurisdiction in which it was organized.

7.2 No Conflicts or Consents.

(a) The execution and delivery of this Agreement by Securityholder do not, and the performance of this Agreement by Securityholder will not: (i) conflict with or violate any law, rule, regulation, order, decree or judgment applicable to Securityholder or by which Securityholder or any of Securityholder's properties is or may be bound or affected; or (ii) result in or constitute (with or without notice or lapse of time) any breach of or default under, or give to any other Person (with or without notice or lapse of time) any right of termination, amendment, acceleration or cancellation of, or result (with or without notice or lapse of time) in the creation of any encumbrance or restriction on any of the Subject Securities pursuant to, any Contract to which Securityholder is a party or by which Securityholder or any of Securityholder's Affiliates or properties is or may be bound or affected.

(b) The execution and delivery of this Agreement by Securityholder does not, and the performance of this Agreement by Securityholder will not, require any consent or approval of any Person. The execution, delivery and performance of any proxy pursuant to Section 4.2 with respect to any Company Shares will not require any consent or approval of any Person.

7.3 Title to Securities. As of the date of this Agreement: (a) Securityholder holds of record (free and clear of any encumbrances or restrictions) the number of outstanding Ordinary Shares set forth under the heading “Ordinary Shares Held of Record” on the signature page hereof; (b) Securityholder holds of record (free and clear of any encumbrances or restrictions) the number of outstanding ADSs set forth under the heading “ADSs Held of Record” on the signature page hereof; (c) Securityholder holds (free and clear of any encumbrances or restrictions) the options, warrants and other rights to acquire Company Shares set forth under the heading “Options and Other Rights” on the signature page hereof; (d) Securityholder Owns the additional securities of the Company set forth under the heading “Additional Securities Beneficially Owned” on the signature page hereof; and (e) Securityholder does not directly or indirectly Own any shares of capital stock or other securities of the Company, or any option, warrant or other right to acquire (by purchase, conversion or otherwise) any shares of capital stock or other securities of the Company, other than the shares and options, warrants and other rights set forth on the signature page hereof.

7.4 No Other Proxies. None of Securityholder’s Subject Shares are subject to any voting agreement, trust or other agreement or arrangement with respect to voting or to any proxy, on the date of this Agreement, except pursuant to this Agreement.

7.5 Absence of Litigation. As of the date hereof, there is no Action pending against, or, to the knowledge of Securityholder, threatened against or otherwise affecting, Securityholder or any of its or his properties or assets (including Securityholder’s Subject Shares) that would reasonably be expected to impair in any material respect the ability of such Securityholder to perform its or his obligations hereunder or to consummate the transactions contemplated hereby on a timely basis.

7.6 Accuracy of Representations. The representations and warranties contained in this Agreement are accurate in all respects as of the date of this Agreement, and will be accurate in all respects at all times and dates throughout the Support Period as if made as of any such time or date.

SECTION 8. ADDITIONAL COVENANTS OF SECURITYHOLDER

8.1 Securityholder Information. Securityholder hereby agrees to permit Parent and Purchaser to publish and disclose in the Schedule TO or other publicly-filed documents relating to the Offer Securityholder’s identity and ownership of Company Shares and the nature of Securityholder’s commitments, arrangements and understandings under this Agreement.

8.2 Further Assurances. From time to time and without additional consideration, Securityholder shall execute and deliver, or cause to be executed and delivered, such additional transfers, assignments, endorsements, proxies, consents and other instruments, and shall take such further actions, as Parent may request for the purpose of carrying out and furthering the intent of this Agreement.

SECTION 9. MISCELLANEOUS

9.1 Survival of Representations, Warranties and Agreements. All representations, warranties, covenants and agreements made by Securityholder in this Agreement shall survive until the Support Termination Date.

9.2 Expenses. All costs and expenses incurred in connection with the transactions contemplated by this Agreement shall be paid by the party incurring such costs and expenses.

9.3 Notices. Any notice or other communication required or permitted to be delivered to either party under this Agreement shall be in writing and shall be deemed properly delivered, given and received when received at the address or facsimile telephone number set forth beneath the name of such party below (or at such other address or facsimile telephone number as such party shall have specified in a written notice given to the other party):

if to Securityholder:

at the address set forth on the signature page hereof; and

if to Parent:

Jazz Pharmaceuticals Public Limited Company
Fourth Floor, Connaught House
One Burlington Road
Dublin 4, Ireland
Attn: Executive Vice President
Fax: +353.1.634.7850

With copies to:

Jazz Pharmaceuticals
3180 Porter Drive
Palo Alto, CA 94304
Attn: General Counsel
Fax: +1.650.496.3781

and:

Jazz_Notices@jazzpharma.com

9.4 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. If the final judgment of a court of competent jurisdiction declares that any term or provision hereof is invalid or unenforceable, the parties hereto agree that the court making such determination shall have the power to limit the term or provision, to delete specific words or phrases, or to replace any invalid or unenforceable term or provision with a term or provision that is valid and enforceable and that comes closest to expressing the intention of the invalid or unenforceable term or provision, and this Agreement shall be enforceable as so modified. In the event such court does not exercise the power granted to it in the prior sentence, the parties hereto agree to replace such invalid or unenforceable term or provision with a valid and enforceable term or provision that will achieve, to the extent possible, the economic, business and other purposes of such invalid or unenforceable term.

9.5 Entire Agreement. This Agreement and any other documents delivered by the parties in connection herewith constitute the entire agreement between the parties with respect to the subject matter hereof and thereof and supersede all prior agreements and understandings between the parties with respect thereto. No addition to or modification of any provision of this Agreement shall be binding upon either party unless made in writing and signed by both parties.

9.6 Assignment; Binding Effect. Except as provided herein, neither this Agreement nor any of the interests or obligations hereunder may be assigned or delegated by Securityholder, and any attempted or purported assignment or delegation of any of such interests or obligations shall be void. Subject to the preceding sentence, this Agreement shall be binding upon Securityholder and Securityholder's heirs, estate, executors and personal representatives (if Securityholder is an individual), and Securityholder's successors and assigns, and shall inure to the benefit of Parent and its successors and assigns. Without limiting any of the restrictions set forth in Section 2 or elsewhere in this Agreement, this Agreement shall be binding upon any Person to whom any Subject Securities are transferred. Nothing in this Agreement is intended to confer on any Person (other than Parent and its successors and assigns) any rights or remedies of any nature.

9.7 Specific Performance. The parties agree that irreparable damage would occur in the event that any of the provisions of this Agreement were not performed in accordance with its specific terms or were otherwise breached. Securityholder agrees that, in the event of any breach or threatened breach by Securityholder of any covenant or obligation contained in this Agreement, Parent shall be entitled (in addition to any other remedy that may be available to it, including monetary damages) to seek and obtain (a) a decree or order of specific performance to enforce the observance and performance of such covenant or obligation, and (b) an injunction restraining such breach or threatened breach. Securityholder further agrees that neither Parent nor any other Person shall be required to obtain, furnish or post any bond or similar instrument in connection with or as a condition to obtaining any remedy referred to in this Section 9.7, and Securityholder irrevocably waives any right he or it may have to require the obtaining, furnishing or posting of any such bond or similar instrument.

9.8 Non-Exclusivity. The rights and remedies of Parent under this Agreement are not exclusive of or limited by any other rights or remedies which it may have, whether at law, in equity, by contract or otherwise, all of which shall be cumulative (and not alternative). Without limiting the generality of the foregoing, the rights and remedies of Parent under this Agreement, and the obligations and liabilities of Securityholder under this Agreement, are in addition to their respective rights, remedies, obligations and liabilities under common law requirements and under all applicable statutes, rules and regulations. Nothing in this Agreement shall limit any of Securityholder's obligations, or the rights or remedies of Parent, under any other agreement between Parent and Securityholder; and nothing in any such agreement shall limit any of Securityholder's obligations, or any of the rights or remedies of Parent, under this Agreement.

9.9 Governing Law; Jurisdiction; Waiver of Jury Trial.

(a) This Agreement shall be governed by, and construed in accordance with, the laws of the State of Delaware, regardless of the laws that might otherwise govern under applicable principles of conflicts of laws thereof; *provided, however*, that the laws of Italy shall apply to the formalities required with respect to the transfer of any Subject Securities, and the laws of the jurisdiction of the residence of Securityholder shall govern Section 6.2. In any action between the parties arising out of or relating to this Agreement or any of the transactions contemplated by this Agreement each of the parties irrevocably and unconditionally consents and submits to the jurisdiction and venue of the Designated Courts.

(b) SECURITYHOLDER AND PARENT IRREVOCABLY WAIVES THE RIGHT TO A JURY TRIAL IN CONNECTION WITH ANY LEGAL PROCEEDING RELATING TO THIS AGREEMENT OR THE ENFORCEMENT OF ANY PROVISION OF THIS AGREEMENT.

9.10 Counterparts. This Agreement may be executed in separate counterparts, each of which when so executed and delivered shall be an original, but all such counterparts shall together constitute one and the same instrument.

9.11 Captions. The captions contained in this Agreement are for convenience of reference only, shall not be deemed to be a part of this Agreement and shall not be referred to in connection with the construction or interpretation of this Agreement.

9.12 Attorneys' Fees. If any legal action or other legal proceeding relating to this Agreement or the enforcement of any provision of this Agreement is brought against Securityholder, the prevailing party shall be entitled to recover reasonable attorneys' fees, costs and disbursements (in addition to any other relief to which the prevailing party may be entitled).

9.13 Waiver. No failure on the part of Parent to exercise any power, right, privilege or remedy under this Agreement, and no delay on the part of Parent in exercising any power, right, privilege or remedy under this Agreement, shall operate as a waiver of such power, right, privilege or remedy; and no single or partial exercise of any such power, right, privilege or remedy shall preclude any other or further exercise thereof or of any other power, right, privilege or remedy. Parent shall not be deemed to have waived any claim available to Parent arising out of this Agreement, or any power, right, privilege or remedy of Parent under this Agreement, unless the waiver of such claim, power, right, privilege or remedy is expressly set forth in a written instrument duly executed and delivered on behalf of Parent; and any such waiver shall not be applicable or have any effect except in the specific instance in which it is given.

9.14 Construction.

(a) For purposes of this Agreement, whenever the context requires: the singular number shall include the plural, and vice versa; the masculine gender shall include the feminine and neuter genders; the feminine gender shall include the masculine and neuter genders; and the neuter gender shall include masculine and feminine genders.

(b) The parties agree that any rule of construction to the effect that ambiguities are to be resolved against the drafting party shall not be applied in the construction or interpretation of this Agreement.

(c) As used in this Agreement, the words "include" and "including," and variations thereof, shall not be deemed to be terms of limitation, but rather shall be deemed to be followed by the words "without limitation."

(d) Except as otherwise indicated, all references in this Agreement to "Sections" and "Exhibits" are intended to refer to Sections of this Agreement and Exhibits to this Agreement.

[Remainder of page intentionally left blank.]

IN WITNESS WHEREOF, Parent and Securityholder have caused this Agreement to be executed as of the date first written above.

JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY

By _____
Title

Signature Page to Support Agreement

SECURITYHOLDER

Signature _____

Printed Name _____

Address: _____

Facsimile: _____

Ordinary Shares
Held of Record

ADs Held of
Record

Options and Other
Rights

Additional Securities
Beneficially Owned

Signature Page to Support Agreement