

Gentium S.p.A.
Form SC 14D9
December 24, 2013

UNITED STATES

SECURITIES AND EXCHANGE COMMISSION

Washington, D.C. 20549

SCHEDULE 14D-9

**Solicitation/Recommendation Statement under Section 14(d)(4)
of the Securities Exchange Act of 1934**

GENTIUM S.P.A.

(Name of Subject Company)

GENTIUM S.P.A.

(Names of Persons Filing Statement)

Ordinary Shares, no par value

American Depositary Shares, each representing one Ordinary Share

(Title of Class of Securities)

37250B922 (Ordinary Shares, which are not traded on U.S. markets)

37250B104 (American Depositary Shares)

(CUSIP Number of Class of Securities)

Dr. Khalid Islam

Chairman & Chief Executive Officer

Gentium S.p.A.

Piazza XX Settembre 2

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(Name, address and telephone numbers of person authorized to receive notices and communications on behalf of the persons filing statement)

With copies to:

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** Check the box if the filing relates solely to preliminary communications made before the commencement of a tender offer.

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Annex A: Opinion of Jefferies LLC, dated December 18, 2013.

Item 1. Subject Company Information.

Name and Address.

The name of the subject company is Gentium S.p.A., a *società per azioni* organized under the laws of the Republic of Italy (the "**Company**"), and the address of the principal executive offices of the Company is Piazza XX Settembre 2, 22079, Villa Guardia (CO), Italy. The telephone number for the Company's principal executive offices is +39 031 537 3200.

Securities.

The title of the class of equity securities to which this Solicitation/Recommendation Statement on Schedule 14D-9 (together with any Exhibits or Annexes hereto, this "**Statement**") relates are (i) the ordinary shares, no par value, of the Company (the "**Common Stock**"), and (ii) the American Depositary Shares, each representing one share of Common Stock of the Company (the "**ADSs**" and, together with the Common Stock, the "**Company Shares**").

As of close of business on December 16, 2013, (i) 15,555,131 shares of Common Stock were issued and outstanding, of which 10,984,130 were represented by an equal amount of issued and outstanding ADSs and (ii) 2,194,494 shares of Common Stock were subject to issuance pursuant to outstanding options (the "**Company Options**") granted under (A) the Company's 2004 Equity Incentive Plan, including the Company's 2004 Italy Stock Award Sub-Plan, as amended and restated (the "**2004 Equity Incentive Plan**"), and (B) the Company's 2007 Equity Incentive Plan, as amended (the "**2007 Equity Incentive Plan**") (collectively, the "**Company Equity Plans**").

Item 2. Identity and Background of Filing Person.

Name and Address.

The name, business address and business telephone number of the Company, which is the person filing this Statement, are set forth above in "*Item 1. Subject Company Information – Name and Address.*"

Tender Offer.

This Statement relates to a tender offer by Jazz Pharmaceuticals Italy S.r.l. ("**Purchaser**"), an Italian *società a responsabilità limitata* and a wholly owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland ("**Parent**"), to purchase all of the outstanding Company Shares at a purchase price of \$57.00 per Company Share (without duplication for shares of Common Stock underlying ADSs) (the "**Offer Price**"), net to the seller in cash, without interest thereon and less any required withholding taxes, upon the terms and subject to the conditions set forth in the Offer to Purchase, dated December 23, 2013 (as amended or supplemented from time to time, the "**Offer to Purchase**"), and in the related Share Form of Acceptance (the "**Share Form of Acceptance**") and the ADS Letter of Transmittal (the "**ADS Letter of Transmittal**" and together with the Offer to Purchase and the Share Form of Acceptance, each as may be amended or supplemented from time to time, collectively constitute the "**Offer**") contained in the Tender Offer Statement on Schedule TO, dated December 23, 2013 (as amended or supplemented from time to time, the "**Schedule TO**") filed by Parent and Purchaser with the U.S. Securities and Exchange Commission (the "**SEC**") on December 23, 2013. The Offer to Purchase, the Share Form of Acceptance and the ADS Letter of Transmittal are filed as Exhibits (a)(1)(A), (a)(1)(F) and (a)(1)(B) to this Statement, respectively, and are incorporated herein by reference.

The Offer is being made in connection with, and subject to the terms and conditions of, the Tender Offer Agreement, dated as of December 19, 2013, by and among Parent, Purchaser and the Company (as such agreement may be amended from time to time, the "**Tender Offer Agreement**"). Consummation of the Offer is subject to various conditions set forth in the Tender Offer Agreement, including, but not limited to the valid tender (excluding ADSs tendered pursuant to guaranteed delivery procedures that are not yet delivered in settlement or satisfaction of such guarantee), without proper withdrawal, of a number of Company Shares that, together with any Company Shares owned by Parent or Purchaser or any of their subsidiaries, represents at least 66 2/3% of the sum of (A) the aggregate number of outstanding Company Shares immediately prior to the time that Purchaser first accepts any Company Shares for payment pursuant to the Offer (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 all cases without duplication for shares of Common Stock underlying the ADSs, provided that Parent may, in its sole discretion, elect to reduce such minimum tender condition to a majority of the aggregate number of outstanding Company Shares immediately prior to the Acceptance Time (without duplication for shares of Common Stock underlying ADSs).

The Offer is initially scheduled to expire at 12:00 midnight, New York time, on the evening of January 22, 2014.

Under the terms of the Tender Offer Agreement, Purchaser is also required to commence a subsequent offering period. A subsequent offering period will be an additional period of not less than three business days and up to 20 business days beginning on the next business day following the expiration of the Offer in accordance with Rule 14d-11 promulgated under the Exchange Act, during which any remaining holders of Company Shares may tender, but not withdraw, their Company Shares and receive the Offer Price. In a subsequent offering period, Purchaser will be required to immediately accept and promptly pay for all Company Shares that are validly tendered during the subsequent offering period. During a subsequent offering period, tendering holders will not have withdrawal rights.

As set forth in the Schedule TO, Parent's principal executive offices are located at Fourth Floor, Connaught House, One Burlington Road, Dublin 4, Ireland, and the telephone number of Parent at such principal offices is +011-353-1-634-7800. The registered office of Purchaser is at via Manzoni 30, Milano, Italy.

The Company has made information relating to the Offer available at www.gentium.com and the Company has filed this Schedule, and Parent and Purchaser have filed the Schedule TO, with the SEC at the website maintained by the SEC at www.sec.gov.

Item 3. Past Contacts, Transactions, Negotiations and Agreements.

Except as set forth in this Item 3, or as otherwise incorporated by reference herein, as of the date of this Statement, to the knowledge of the Company, there are no material agreements, arrangements or understandings or any actual or potential conflicts of interest between the Company or its affiliates and: (a) the Company's executive officers, directors or affiliates; or (b) Parent, Purchaser or their respective executive officers, directors or affiliates.

Agreements with Parent and Purchaser.

Tender Offer Agreement.

The summary of the Tender Offer Agreement contained in Section 12 of the Offer to Purchase and the description of the conditions of the Offer contained in Section 14 of the Offer to Purchase, which is filed as Exhibit (a)(1)(A) to this Statement, are incorporated by reference herein. Such summary and description do not purport to be complete and are qualified in their entirety by reference to the Tender Offer Agreement, a copy of which is filed as Exhibit (e)(1) to this Statement, which is incorporated by reference herein.

Confidentiality Agreement.

The Company and Parent are parties to a Confidentiality and Nondisclosure Agreement, effective as of September 30, 2013 (the "**Confidentiality Agreement**"). Under the terms of the Confidentiality Agreement, the Company and Parent each agreed to keep confidential certain information furnished to such party by the other in connection with the evaluation of a possible transaction pertaining to the pharmaceutical business (or part thereof) of the Company and/or its affiliates. The foregoing description of the Confidentiality Agreement does not purport to be complete and is qualified in its entirety by reference to the Confidentiality Agreement, a copy of which is filed as Exhibit (e)(3) to this Statement, which is incorporated by reference herein.

Support Agreements.

In connection with the Tender Offer Agreement, each of Dr. Khalid Islam, Chairman of the Company Board and Chief Executive Officer of the Company, Mr. Salvatore Calabrese, Senior VP Finance, Chief Financial Officer and Chief Operating Officer of the Company, Dr. Giorgio Mosconi, Vice President, Scientific Director of the Company, Mr. Adrian Haigh, Senior Vice President, Commercial Operations and Chief Operating Officer of Gentium GmbH, a wholly-owned subsidiary of the Company organized under the laws of Switzerland, each member of the Company Board (other than Mr. Marco Brughera), F3F S.r.l., the beneficial owner of approximately 15% of the outstanding Company Shares (and 13% of the Company Shares on a fully-diluted basis), and Dr. Carin Heringa, an employee of Gentium GmbH, entered into support agreements with Parent (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) exercise any options to acquire Company Shares held by them following the Acceptance Time and tender the underlying Company Shares into the Offer during the subsequent offering period; (iv) vote their Company Shares against the following actions (and, except in the case of F3F S.r.l., deliver a proxy or proxies to Parent if any Company shareholders' meeting is called during the term of the Support Agreement): (a) any Acquisition Transaction (as defined in the Tender Offer Agreement); (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 the Support Agreement; (v) not (a) 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 (as defined in the Tender Offer Agreement); (b) provide any non-public information concerning the Company or any of its subsidiaries to any individual or entity in connection with or in response to an Acquisition Proposal (as defined in the Tender Offer Agreement); (c) engage in any discussions or negotiations with any individual or entity with respect to any Acquisition Proposal (as defined in the Tender Offer Agreement); (d) approve, adopt, endorse or recommend any Acquisition Proposal (as defined in the Tender Offer Agreement); (e) enter into any letter of intent or similar document or any contract contemplating or otherwise relating to any Acquisition Transaction (as defined in the Tender Offer Agreement); or (f) continue any existing discussions with any individual or entity that relate to any Acquisition Proposal (as defined in the Tender Offer Agreement); and (vi) for a period of 18 months following the Acceptance Time, not, directly or indirectly, encourage, induce, attempt to induce, solicit or attempt to solicit (on their own behalf or on behalf of any other individual or entity) any current or former employee of the Company to leave his or her employment, consulting or independent contractor relationship with the Company or any of the Company's subsidiaries (subject to customary exceptions).

In addition, each of the Support Agreements, other than the Support Agreements with F3F S.r.l. and Dr. Heringa, contain a provision prohibiting the signatory, for a period of three years following the Acceptance Time, from: (i) having directly or indirectly, any financial or other interest in a business or company which engages in the field of: 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 (a "**Competitor**"); (ii) accepting any part or full time employment in a Competitor or to act as a consultant or representative or in any other form for a Competitor; (iii) directly or indirectly establishing a Competitor; (iv) enticing away or soliciting or trying to entice away or to solicit any customer of the Company or any of the Company's subsidiaries for whom each was active or with whom each was in contact during the last 12 months prior to the Acceptance Time (a "**Customer**"); or (v) directly or indirectly acting on behalf of or rendering services to a Customer, for example as an employee, consultant, agent, officer, director or submit or be involved in any offer for such activity; provided however, that Dr. Islam is permitted to perform certain consulting services.

The foregoing description of the Support Agreements does not purport to be complete and is qualified in its entirety by reference to the Support Agreements, a form of which is filed as Exhibit (e)(2) to this Statement and incorporated by reference herein.

Approximately 4,016,608 Company Shares (including 1,666,608 ADSs issuable upon the exercise of Company Options) are subject to Support Agreements (representing approximately 15% of the total outstanding Company Shares, and approximately 23% of the fully diluted number of Company Shares, as of December 16, 2013).

Arrangements with Current Executive Officers and Directors of the Company.

Certain executive officers and directors of the Company may be deemed to have interests in the transactions contemplated by the Tender Offer Agreement that are different from or in addition to those of the holders of the Company Shares generally. These interests may create potential conflicts of interest. The Company Board was aware of these interests and considered them, among other matters, in approving the Tender Offer Agreement and the transactions contemplated thereby. Moreover, the Company Board formed a special committee of independent directors (the "**Special Committee**") constituted by Mr. Elmar Schnee (Chairman), Ms. Joyce Victoria Bigio and Dr. Bobby Sandage, Jr. to review agreements and arrangements entered into or to be entered into with directors and employees of the Company in connection with or in the context of the Offer, and compensation proposed to be paid to employees as a result of the Offer, and the Special Committee approved such agreements, arrangements and compensation. Consummation of the Offer will constitute a change in control of the Company for the purposes of determining the entitlements due to executive officers and directors of the Company relating to certain benefits, as described below.

Consideration for Company Shares Tendered Pursuant to the Offer.

If the directors and executive officers of the Company who own Company Shares tender their Company Shares for purchase pursuant to the Offer, they will receive the same Offer Price on the same terms and conditions as the other holders of the Company Shares. As of December 16, 2013, the directors and executive officers of the Company beneficially owned in the aggregate 2,350,000 Company Shares, which for purposes of this subsection excludes any Company Shares issuable upon exercise or settlement of Company Options held by such individuals. If the directors and executive officers were to tender all of such Company Shares pursuant to the Offer and those Company Shares were accepted for purchase and purchased by Purchaser, the directors and executive officers would receive an aggregate of \$133,950,000 in cash, without interest, less any required withholding taxes. For a description of the treatment of Company Options held by the directors and executive officers of the Company, see below under the heading "Effect of the Offer on Company Options."

The following table sets forth, as of December 16, 2013, the cash consideration that each executive officer and director would be entitled to receive in respect of outstanding Company Shares beneficially owned by him or her (excluding shares underlying Company Options), assuming such individual were to tender all of his or her outstanding Company Shares pursuant to the Offer and those Company Shares were accepted for purchase and purchased by Purchaser.

| Name of Director or Executive Officer | Number of Company Shares | Payments in Respect of Company Shares |
|---------------------------------------|--------------------------|---------------------------------------|
| Laura Ferro (1) | 2,250,000 | \$ 128,250,000 |
| Khalid Islam | 100,000 | \$ 5,700,000 |

(1) Dr. Laura Ferro may be deemed to control F3F S.r.l. by virtue of her 66.67% ownership interest in F3F S.r.l. Accordingly, Dr. Ferro may be deemed to beneficially own the 2,250,000 Company Shares owned by F3F S.r.l.

Director Agreements with the Company.

See section below entitled "Director and Officer Indemnification and Insurance."

Executive Officer Agreements with the Company.

The Company is a party to a number of agreements with its Chairman and Chief Executive Officer, Dr. Khalid Islam, its Chief Financial Officer and Chief Operating Officer, Mr. Salvatore Calabrese, its Vice President, Scientific Director of the Company, Dr. Giorgio Mosconi, and its Senior Vice President, Commercial Operations and Chief Operating Officer of Gentium GmbH, Mr. Adrian Haigh.

The Company is party to employment agreements with each of the forgoing executive officers, which address the duties and responsibilities of such executive officers and specify the amounts payable to them in connection with certain terminations or change in control events.

Except as provided below, all of the employment agreements with the Company's executive officers contain substantially similar terms.

The employment agreements also provide each executive officer with certain severance benefits in the event his employment is terminated without Cause (as defined in the employment agreement), or, in the case of Dr. Islam, he resigns following a Change in Control (as defined in the employment agreement), or, in the case of Mr. Calabrese and Mr. Haigh, the executive officer resigns for Good Reason (as defined in the employment agreement) following a Change in Control (as defined in the employment agreement). Specifically, in the event such a termination or resignation (where applicable) occurs within 24 months following a Change in Control (as defined in the employment agreement), each executive officer would be eligible to receive cash severance, payment of certain fringe benefits, accelerated vesting of any unvested outstanding Company Options and, in the case of Dr. Islam and Mr. Calabrese, reimbursement of any legal expenses incurred in connection with enforcing payment of amounts due.

The foregoing description of the employment agreements with the Company's executive officers does not purport to be complete and is qualified in its entirety by reference to the employment agreements filed as Exhibits (e)(4) to (e)(7) to this Statement, respectively, all of which are incorporated by reference herein, subject to such amendments to such agreements as described below in this Item 3.

Concurrently with the execution of the Tender Offer Agreement, Dr. Islam entered into a transition, amendment and release agreement with Gentium GmbH (the "**Transition and Release Agreement**"), pursuant to which, among other things, (i) Dr. Islam has agreed to resign on the Closing Date (as defined in the Transition and Release Agreement), and (ii) Dr. Islam will only be eligible to receive certain change in control severance entitlements specified under his employment agreement if he successfully completes his Consultancy Agreement (as defined below), as determined by Gentium GmbH in its discretion, and signs a waiver and release substantially in the form attached as Exhibit A to the Transition and Release Agreement.

Concurrently with the execution of the Tender Offer Agreement, Dr. Islam also entered into a consultancy agreement with the Company (the "**Consultancy Agreement**"), whereby he has agreed to work as a consultant for the Company for the six (6) months following the Closing Date (as defined in the Transition and Release Agreement).

The foregoing descriptions of the Transition and Release Agreement and the Consultancy Agreement do not purport to be complete and are qualified in their entirety by reference to the Transition and Release Agreement and the Consultancy Agreement, which are filed as Exhibits (e)(8) and (e)(9) to this Statement, respectively, and are incorporated by reference herein.

Concurrently with the execution of the Tender Offer Agreement, each of Mr. Calabrese, Dr. Mosconi, and Mr. Haigh entered into a Retention and Amendment Agreement (collectively, the "**Retention Agreements**"), whereby, among other things, each individual has agreed not to resign and to continue working for the Company in accordance with the terms of the relevant individual's employment agreement for a period of six to nine months, as applicable, and receive a retention bonus for this period. Further, each agreed that he will not be entitled to receive certain change in control severance entitlements (and the retention bonus described above) if his employment is terminated by the Company during this period for Cause (as defined in the respective employment agreements), or in the case of Mr. Haigh and Mr. Calabrese, he resigns without Good Reason (as defined in the respective employment agreements), prior to the completion of the Retention Period (as defined in the Retention Agreements).

The foregoing description of the Retention Agreements does not purport to be complete and is qualified in its entirety by reference to the Retention Agreements, which are filed as Exhibits (e)(10) to (e)(12) to this Statement, all of which are incorporated by reference herein.

Effect of the Offer on Company Options.

Pursuant to the Tender Offer Agreement, as soon as practicable following the date of the Tender Offer Agreement (and, in any event, prior to the Acceptance Time), the Company Board is required to adopt such resolutions and take such other actions as may be required to (i) provide that all outstanding and unvested Company Options will 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, such holders, 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 the Tender Offer Agreement. The Company Board resolved to accelerate all unvested Company Options in connection with the Offer on December 18, 2013 and such resolution was ratified by the Special Committee on the same date.

The Tender Offer Agreement also provides that, as soon as practicable following the date of the Tender Offer Agreement, the Company must provide notice to each holder of outstanding Company Options (except directors, consultants, independent contractors, members of senior management or any executive officer of the Company as defined under Section 13(k) of the Securities Exchange Act of 1934, as amended (the "**Exchange 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 with 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 the Tender Offer Agreement.

The Company and its subsidiaries are required 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.

As of December 16, 2013, Company directors and executive officers held 1,625,108 Company Options. The table below sets forth information regarding the Company Options held by each of the Company's directors and executive officers as of December 16, 2013, including the net cash consideration payable in respect of Company Shares issued upon exercise of each individual's Company Options, assuming such individual were to tender all such Company Shares pursuant to the Offer, as extended by such subsequent offering period, and those Company Shares were accepted for purchase and purchased by Purchaser.

| Name of Director or Executive Officer | Number of Company Options Held (Vested) | Weighted Average Exercise Price per Company Option (Vested) | Number of Company Options Held (Unvested) | Weighted Average Exercise Price per Company Option (Unvested) | Total Number of Company Options Held (Whether Vested or Unvested) | Total Cash Spread Value of Company Options (Whether Vested or Unvested) |
|---------------------------------------|--|---|---|---|---|---|
| Khalid Islam | 460,414 | \$ 6.34 | 114,586 | \$ 8.93 | 575,000 | \$ 28,830,500 |
| Joyce Victoria Bigio | — | — | 31,500 | \$ 8.04 | 31,500 | \$ 1,542,240 |
| Gigliola Bertoglio | 123,000 | \$ 5.81 | 11,500 | \$ 8.04 | 134,500 | \$ 6,859,500 |
| Marco Brughera | 27,000 | \$ 9.30 | 11,500 | \$ 8.04 | 38,500 | \$ 1,851,080 |
| Laura Ferro | 73,000 | \$ 6.54 | 11,500 | \$ 8.04 | 84,500 | \$ 4,246,970 |
| Bobby W. Sandage, Jr. | 123,000 | \$ 5.81 | 11,500 | \$ 8.04 | 134,500 | \$ 6,859,500 |
| Elmar Schnee | 27,000 | \$ 9.30 | 11,500 | \$ 8.04 | 38,500 | \$ 1,851,080 |
| Salvatore Calabrese | 328,508 | \$ 7.90 | 89,600 | \$ 10.07 | 418,108 | \$ 20,336,773 |
| Adrian Haigh | 31,649 | \$ 9.11 | 58,351 | \$ 8.88 | 90,000 | \$ 4,323,600 |
| Giorgio Mosconi | — | — | 80,000 | \$ 12.73 | 80,000 | \$ 3,541,600 |

Employee Matters.

The Tender Offer Agreement provides that, unless a higher protection is given to the continuing employees of the Company under applicable law or any such continuing employee of the Company agrees otherwise, for a period of at least twelve months following the Acceptance Time, Parent will, and will cause its subsidiaries to, provide to all continuing employees of the Company located outside of the United States compensation and benefits (not including stock options or other equity compensation, retention or bonuses out of the ordinary course, with certain exceptions) that are, in the aggregate, no less favorable than the compensation and benefits being provided to continuing employees of the Company located outside of the United States immediately prior to the Acceptance Time.

Parent will 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 will recognize for such purposes continuous service with the Company or its subsidiaries, including predecessor employers, of participating continuing employees of the Company under such plans.

Nothing described above under "*Employee Matters*" or elsewhere in the Tender Offer Agreement prohibits Purchaser or its subsidiaries from amending or terminating any employment 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.

Furthermore, the Tender Offer Agreement also provides that the terms and conditions of employment after the Acceptance Time for continuing employees of the Company whose employment is subject to a collective bargaining agreement, works council agreement or similar contract with any labor organization, works council or employee association will be governed by such agreement or contract until its expiration, modification or termination in accordance with its terms and applicable law.

Unless otherwise requested by Parent in writing at least five days prior to the Acceptance Time, the Company has agreed 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 Acceptance Time, any Employee Benefit Plan that is intended to be qualified under Section 401(k) of the Code (the "**401(k) Plan**") and, 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.

As defined in the Tender Offer Agreement "continuing employees of the Company" means all employees of the Company (a) who are offered and timely accept employment by Parent or any subsidiary of Parent, (b) who continue their employment with the Company at the request of Parent, or (c) outside the U.S., who remain employees of the Company, or who become employees of Parent or any subsidiary of Parent, as required by applicable law.

Potential Payments Upon Change in Control.

See section above entitled "*Executive Officer Agreements with the Company.*"

Also, see section above entitled "*Effect of the Offer on Company Options.*"

On September 27, 2013, the Company Board resolved upon the adoption of a transaction bonus for selected senior managers of the Company and Gentium GmbH in consideration for the activities to be carried out by such senior managers in the event of consummation of a potential sale of the Company, subject to certain terms and conditions to be subsequently defined by the Compensation, Nominating and Corporate Governance Committee of the Company (the "**Transaction Bonus**"). On September 27, 2013, the Compensation, Nominating and Corporate Governance Committee of the Company proposed a Transaction Bonus ranging from 1.0% up to 1.5% of the total transaction value, depending on the premium over \$22.83, the volume weighted average closing price of the ADSs in the ten days prior to September 27, 2013, if any, achieved in any sale of the Company. Of the Transaction Bonus, 70% was allocated to Dr. Islam and the remaining 30% was to be allocated to selected senior managers of the Company and Gentium GmbH expressly identified by the Company Board following the recommendation of Dr. Islam. As the premium over \$22.83 exceeded 45%, the applicable Transaction Bonus is 1.5% of the transaction value, or approximately \$15.2 million. Dr. Islam proposed to the Company Board on December 23, 2013 that the unallocated 30% should be allocated as follows: 10% to Mr. Calabrese, 10% to Mr. Haigh, 5% to Dr. Mosconi, 2.5% to Mr. Terenzio Ignoni, VP Quality, 1.5% to Ms. Roberta Grandini, Finance, and 1% to Mr. Filippo Cerutti, Corporate Legal Manager, and the Company Board approved such allocation. The Transaction Bonus and proposed allocations were also approved by the Special Committee at meetings held on December 18, 2013 and December 23, 2013.

Director Compensation.

Under Italian law, shareholders determine director compensation relating to basic board service, such as annual fees for serving on the Company Board and fees for attending board meetings. For the term ending with the approval of the Company's financial statements as of December 31, 2013, each director receives an annual cash retainer of \$45,000. The chairman of the Company Board receives an additional annual retainer of \$25,000 and the chairs of the following committees of the Company Board receive the following additional annual retainers: (i) Audit Committee, \$25,000; (ii) Compensation, Nominating and Corporate Governance Committee, \$15,000; and (iii) Scientific Oversight Committee, \$15,000. In addition, the non-chair members of the following committees receive the following additional annual retainers: (i) Audit Committee, \$5,000; (ii) Compensation, Nominating and Corporate Governance Committee, \$5,000; and (iii) Scientific Oversight Committee, \$5,000.

Directors also receive reimbursement of their reasonable out-of-pocket expenses in connection with attendance at Company Board and committee meetings.

In addition, the Company's directors also receive equity incentive compensation in connection with their service on the Company Board. For the term ending with the approval of the Company's financial statements as of December 31, 2013, the Company has granted Company Options to purchase an aggregate of 11,500 Company Shares to each director at an exercise price of \$8.04, which Company Options terminate on March 26, 2022.

Please see the section above entitled "*Effect of the Offer on Company Options*" for more information on the treatment of equity held by directors in the Offer.

Director and Officer Indemnification and Insurance.

The Company has entered into indemnification agreements with its directors and officers (the "**Indemnification Agreements**"), which may require the Company to, among other things, indemnify its directors and officers against liabilities that may arise by reason of their status or service as directors or officers, other than liabilities arising on account of conduct which is an act or omission not in good faith or which involves intentional misconduct or a knowing violation of the law. The Indemnification Agreements also may require the Company to advance any expenses incurred by the directors or officers as a result of any proceeding against them as to which they could be indemnified. This summary of the Indemnification Agreements does not purport to be complete and is qualified in its entirety by reference to the Indemnification Agreements, the form of which is filed as Exhibit (e)(13) to this Statement and incorporated by reference herein.

The Tender Offer Agreement provides that, for five years after the Acceptance Time (the "**Indemnification Period**"), Parent will cause the Company and its subsidiaries and successors to honor and fulfill the obligations of the Company and its subsidiaries under its Indemnification Agreements in effect as of the date of the Tender Offer Agreement with any of the Company's or its subsidiaries' 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 Indemnified Party in his or her capacity as a director or officer of the Company or its subsidiary. In the event that any claim or claims are asserted or made within the Indemnification Period, all rights to indemnification in respect of any such claim or claims will continue until disposition of any and all such claims.

Further, during the Indemnification Period, Parent and the Company will 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 no less favorable, in the aggregate, than those of the D&O Insurance in effect on the date the Tender Offer Agreement is executed and with an insurance company with the same or better credit rating as the insurance company providing the current D&O Insurance.

The Company may, at its option, substitute the policies of Parent, Purchaser or any of their respective subsidiaries containing certain 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 request that the Company obtain such extended reporting period coverage under its existing insurance program (to be effective as of the Acceptance Time). Parent or Purchaser will not be obliged to pay annual premiums in excess of 250% of the annual premium for coverage under the D&O Insurance in effect on the date of the Tender Offer Agreement (such 250% amount, the "**Maximum Annual Premium**"), and if the annual premiums of such insurance coverage exceed such amount, Parent, Purchaser and the Company will be obligated to obtain a policy with the greatest coverage available for a cost not exceeding the Maximum Annual Premium. Parent, Purchaser or the Company may, prior to the Acceptance Time, purchase a five-year "tail" prepaid policy on the D&O Insurance containing certain terms no less favorable, in the aggregate, than the D&O Insurance in effect on the date of the Tender Offer Agreement 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, will 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 with respect to the D&O Insurance.

The Tender Offer Agreement provides that if Purchaser or the Company or any of its successors or assigns consolidates with or merges into any other person or entity and will not be the continuing or surviving corporation or entity of such consolidation or merger or transfers all or substantially all of its properties and assets to any person or entity, then, and in each such case, proper provisions will be made so that the successors and assigns of the Company will assume all of the obligations of Parent, Purchaser and the Company related to the rights and obligations described under "*Director and Officer Indemnification Insurance.*"

The obligations described under "*Director and Officer Indemnification Insurance*" may not be terminated, amended or otherwise modified in such a manner as to adversely affect any Indemnified Party (or certain other persons) 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. The rights of the Indemnified Parties (and certain other persons) are 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, or applicable law (whether at law or in equity).

Continuing Directors.

In the Tender Offer Agreement, the Company acknowledges that, pursuant to the Tender Offer Agreement, certain directors of the Company have delivered written resignations, respectively, to the Company and to the Purchaser, pursuant to which and subject to the Offer closing, the resignations of (a) three of such individuals will become effective as of the Acceptance Time (the "**Initial Director Resignations**"), and (b) up to three 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 will use reasonable efforts to obtain written resignations of all the Company's statutory auditors (including the alternate members), pursuant to which each statutory auditor will resign from its office effective as of the date of the first shareholders' meeting to be held following the Acceptance Time. The Company Board will (x) subject to the Offer closing, upon the Acceptance Time, make certain resolutions agreed to pursuant to the terms and subject to the conditions of the Tender Offer Agreement, including the appointment of certain of Parent's designees to the Company Board (the "**Acceptance Time Board Appointments**") and (y) no later than ten business days after the Acceptance Time, 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's 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: (A) Release of the resigning directors and (B) 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. However, if by February 7, 2014 closing of the Offer has not occurred, the Company Board will be free to revoke the notice of call of the shareholders' meeting. None of the parties guarantee that the resolutions set forth in this paragraph will, once voted, be valid and effective.

Arrangements with Affiliates of the Company.

On January 18, 2012, the Company entered into a commercial lease with F3F S.r.l. (formerly FinSirton S.p.A.), providing for the lease by the Company of office and laboratory space of approximately 4,800 square meters (the "**Lease Agreement**"). This summary of the Lease Agreement does not purport to be complete and is qualified in its entirety by reference to the Lease Agreement, incorporated by reference to Exhibit (e)(14) to this Statement, which is incorporated by reference herein. Dr. Laura Ferro, who serves as a member of the Company Board, together with members of her family, may be deemed to control F3F S.r.l., which beneficially owns approximately 15% of the outstanding Company Shares as of December 16, 2013.

On December 7, 2001, the Company entered into a license and supply agreement with Sigma-Tau Pharmaceuticals, Inc. ("**Sigma-Tau Pharmaceuticals**"), as assignee of Sigma-Tau Industrie Farmaceutiche Riunite S.p.A., for Sigma-Tau Pharmaceuticals to commercialize the intravenous formulation of defibrotide for the treatment of VOD in the United States upon FDA approval, if any (the "**License and Supply Agreement**"). In 2005, the Company expanded Sigma-Tau Pharmaceuticals's current license territory to the Americas. On October 12, 2007, the Company entered into a letter agreement with Sigma-Tau Pharmaceuticals, pursuant to which Sigma-Tau Pharmaceuticals

agreed to reimburse 50% of certain costs the Company incurred relating to its Phase III clinical trial of defibrotide to treat severe VOD (the "**Letter Agreement**"). The License and Supply Agreement and the Letter Agreement were amended effective January 7, 2010 to expand the license to include the intravenous formulation of defibrotide for the prevention of VOD in the Americas and to transfer the NDA to Sigma-Tau Pharmaceuticals post approval in the United States. On May 11, 2012, the Letter Agreement was amended to update those certain clinical trial costs that Sigma-Tau Pharmaceuticals will reimburse 50% of. This summary of the arrangements with Sigma-Tau Pharmaceuticals does not purport to be complete and is qualified in its entirety by reference to (a) the License and Supply Agreement, incorporated by reference to Exhibit (e)(15) to this Statement, (b) the Letter Agreement, incorporated by reference to Exhibit (e)(16) to this Statement, and (c) the amendments to the License and Supply Agreement and the Letter Agreement, incorporated by reference to Exhibit (e)(17) to this Statement, all of which are incorporated by reference herein. Sigma-Tau is an affiliate of Sigma-Tau Finanziaria S.p.A. ("**Sigma-Tau**"), which, together with its affiliates, beneficially owns approximately 17.4% of the outstanding Company Shares as of December 16, 2013. Mr. Marco Brughera, who serves as a member of the Company Board, also holds various senior-level positions within the Sigma-Tau Group.

Item 4. The Solicitation or Recommendation.

Recommendation of the Company Board.

The Company Board, during a meeting held on December 18, 2013, by unanimous vote of all directors of the Company, other than the vote of one director who abstained due to the potential conflicts of interest resulting from the security ownership of Sigma-Tau, of which he is a representative, (1) determined that the Tender Offer Agreement and the transactions contemplated thereby are fair to and in the best interests of the Company's shareholders and other Company stakeholders, and (2) approved and deemed advisable the Tender Offer Agreement and the transactions contemplated thereby.

Accordingly, and for the other reasons described in more detail below, the Company Board hereby recommends that the holders of Company Shares accept the Offer and tender their Company Shares to Purchaser pursuant to the Offer.

Background and Reasons for the Company Board's Recommendation.

Background of the Offer.

The Company Board and the Company's senior management regularly review the Company's business, operations, long-term strategic plans and prospects as an independent company, as well as trends in the industry in which the Company operates, all with the objective of managing the Company in the interest of all of its stockholders. Throughout calendar year 2013, the Company Board considered the Company's strategic alternatives, particularly in light of the uncertainty surrounding the outcome of the regulatory approval process for Defitelio® (defibrotide) in Europe, the U.S. and other parts of the world. Among the alternatives considered earlier in the year, the Company Board evaluated a capital raise, a sale to a strategic investor or remaining independent. In particular, the Company Board considered the benefits to the Company of being integrated into a larger, diversified pharmaceutical group that could support the commercialization of Defitelio® (defibrotide) as against the prospective risks to the Company as a stand-alone entity. Discussions surrounding the Company's strategic plans and prospects intensified as the outcome of the Company's process before the European Medicines Agency's ("EMA") Committee for Medicinal Products for Human Use ("CHMP") approached.

On February 14, 2013, a representative of Company A contacted Mr. Adrian J. Haigh, the Company's Senior VP, Commercial Operations, expressing a desire to discuss and explore strategic alternatives between Company A and the Company. Mr. Haigh responded that same day indicating interest in engaging in discussions, subject to execution of a confidentiality agreement.

In the second half of February 2013, in light of the expression of interest from Company A and uncertainty about the potential impact that a positive or negative opinion by the CHMP would have on the Company, the Company contacted a number of investment banks to discuss a potential capital raise or strategic transaction for the Company.

On March 4, 2013, Company A entered into a confidentiality agreement with a subsidiary of the Company, which did not include a standstill provision. On March 7, 2013, Mr. Haigh and Dr. Khalid Islam, Chairman and Chief Executive Officer of the Company, participated in a teleconference with representatives of Company A, at which Mr. Haigh and Dr. Islam presented certain confidential information relating to the Company. Following such discussion, and over the course of the next few months, the Company regularly updated Company A on the status of its EMA regulatory process.

On April 10, 2013, a representative of Company B contacted Mr. Haigh by telephone and expressed interest in a possible transaction involving the Company.

On April 18, 2013 Company B and the Company executed a confidentiality agreement, which did not contain a standstill provision and, on the following day, Mr. Haigh presented certain confidential information relating to the Company to representatives of Company B.

On June 5, 2013, Dr. Islam, Mr. Haigh and Mr. Salvatore Calabrese, the Company's Senior VP Finance, Chief Financial Officer, met Company B's Chief Executive Officer and other members of Company B's management team at Company B's offices in New York City, where Dr. Islam, Mr. Haigh and Mr. Calabrese were traveling for a pre-arranged and unrelated business trip, during which the parties engaged in discussions regarding a potential acquisition of the Company.

On June 19, 2013, Mr. Haigh received a non-binding offer from Company B to acquire the Company for \$10.06 per Company Share using part cash and part stock consideration. Dr. Islam informed the Company Board of the non-binding offer received from Company B at the July 18, 2013 Company Board meeting and the Company Board decided not to respond to Company B's offer until after the CHMP decision.

As the date for CHMP to render its opinion on Defitelio® (defibrotide) approached, the Company began to consider in greater depth its potential strategic alternatives and invited Jefferies LLC ("**Jefferies**") to discuss with the Company such alternatives and the Company's position and defense profile in the event of a positive or negative recommendation by the CHMP.

On July 18, 2013, Jefferies reviewed those matters with members of the Company Board and management.

On July 26, 2013, the Company announced that the CHMP had adopted a positive opinion for Defitelio® (defibrotide), recommending the granting of a Marketing Authorization ("**MA**") under exceptional circumstances, for

the treatment of severe hepatic veno-occlusive disease ("**VOD**") in adults and children undergoing hematopoietic stem cell transplantation therapy. The Company deemed this development to be significant as the European Commission, which has ultimate authority to grant marketing authorization, generally follows the recommendations of the CHMP and generally delivers its final decision within three months of the CHMP recommendation.

Following the CHMP's positive opinion, the Company's management believed that the Company had become too expensive for Company B, given Company B's relatively small market capitalization, and there were no further significant discussions between the parties.

On August 12, 2013, a representative of Company A contacted Mr. Haigh again, asking when the Company anticipated hearing from the European Commission on its MA and indicating that Company A was ready to commence additional, extensive due diligence following a favorable opinion from the European Commission.

On August 12, 2013, the Company Board held a regularly scheduled meeting and, among other things, discussed the potential engagement of Jefferies as the Company's financial advisor, the status of Defitelio® (defibrotide) in Europe and elsewhere and also discussed the Company's strategic plans. The Company Board decided to postpone engaging Jefferies as the Company's financial advisor until closer to the approval of Defitelio® by the European Commission.

On August 26, 2013, a representative of Parent, contacted Mr. Haigh by email, expressing Parent's interest in the Company and its strategic direction. During a subsequent call that day, Mr. Haigh indicated that the Company would be interested in learning more about Parent's interest in the Company and provided an update on the Company. In this connection, Mr. Haigh suggested that further information could be shared under a confidentiality agreement. Mr. Haigh sent the representative of Parent non-confidential information concerning the Company and VOD and a draft confidentiality agreement shortly after this conversation took place.

On September 4, 2013, Mr. Haigh and the representative of Parent held a call to discuss the Company's strategic goals and the possibility of holding a meeting between the companies' chief executive officers.

On September 5, 2013, Dr. Islam received an email from a representative of Parent requesting a meeting on September 12, 2013 to discuss and explore Parent's interest in the Company and Defitelio®. Dr. Islam confirmed his availability for such meeting.

On September 9, 2013, the Company Board held a regularly scheduled meeting for the purpose of reviewing the Company's strategic plan and the direction of the business.

On September 12, 2013, Dr. Islam and Mr. Haigh met with representatives of Parent, including Mr. Iain McGill, the Head of Parent's EUSA International division and Senior Vice President of Parent, in Milan, Italy. During the course of the meeting, the parties discussed various non-confidential aspects of the Company's and Parent's businesses and agreed to explore potential strategic opportunities between the Company and Parent, including a potential partnership and trade sale. Dr. Islam indicated that any decision regarding strategic options for the Company would likely be made in the fourth quarter, following the European Commissions' expected decision in respect of the Company's MA for Defitelio® in late September or October.

On September 27, 2013, at management's suggestion, in light of Parent's and Company A's stated interest in the Company and management's discussions with investment banks in the preceding months regarding the Company, the Company Board held a special meeting (partially by teleconference) at which the Company Board directed management to explore potential strategic alternatives that would enhance shareholder value and also provide the Company with the ability to be integrated into a larger, more diversified pharmaceutical group that could support the further development, marketing and distribution of Defitelio® (defibrotide). The Company Board and senior

management used the meeting as an opportunity to discuss the interest expressed by Company A and Parent. In this connection, the Company Board determined to engage Jefferies to serve as exclusive financial advisor to the Company. The Company Board also resolved to form a mergers and acquisitions committee (the "**M&A Committee**"), consisting of three directors (Mr. Elmar Schnee, Chairperson, Dr. Bobby Sandage, Jr. and Dr. Khalid Islam), to initiate and oversee a process to seek third-party interest in an acquisition of the Company, evaluate potential strategic alternatives, including a potential sale of the Company, and facilitate a timely response to developments as they arose. The Company Board resolved to nominate Mr. Calabrese (then Senior VP Finance, Chief Financial Officer) as the Company's Chief Operating Officer. The Company Board also resolved to adopt a transaction bonus, payable to selected senior managers of the Company and its subsidiary in the event of a successful sale of the Company. At this meeting, the Company informed its outside Italian legal counsel, Gianni Origoni Grippo Cappelli & Partners ("**GOP**"), that it was exploring potential strategic alternatives and requested GOP's assistance in connection with the Company's transaction process.

On September 30, 2013, Parent executed and delivered to the Company a Confidentiality and Nondisclosure Agreement, which did not include a standstill provision, and was countersigned by the Company on October 1, 2013.

On October 3, 2013, Dr. Islam met for dinner with Mr. Bruce Cozadd, Parent's Chief Executive Officer, in San Francisco, California, where Dr. Islam was traveling for a number of pre-arranged and unrelated business meetings. During the course of the dinner, the parties discussed potential strategic opportunities between the Company and Parent. Following the meeting, Mr. Cozadd sent Dr. Islam an email on October 4, 2013 expressing Parent's interest in evaluating a potential transaction with the Company.

On October 4, 2013, Parent sent an initial due diligence request list to the Company.

On October 16, 2013, a representative of Parent spoke with Dr. Islam by phone. During this call, the representative of Parent reiterated that Parent was interested in a business combination transaction with the Company. Dr. Islam advised the representative of Parent that the Company had engaged Jefferies as its financial advisor and suggested that Parent submit a written non-binding proposal describing the terms and conditions of its proposed transaction with the Company.

On October 17, 2013, the Company launched a virtual data room which contained corporate, financial, operating, regulatory, intellectual property, employment and other due diligence materials regarding the Company, and provided access to the virtual data room to representatives of Parent.

Beginning on October 21, 2013, in accordance with the Company's directives, Jefferies contacted 37 strategic parties (other than Parent) to solicit their potential interest in the Company. The parties contacted included large pharmaceutical companies and biotech and specialty pharmaceutical companies that the Company's management and Jefferies believed could have potential interest in the Company's process based on the profile of the Company. Financial sponsors were not approached given that the Company does not yet have sufficient cash flows to support the leverage and rates of return typically required by financial sponsors.

On October 22, 2013, representatives from Parent, including senior managers of its international division, visited members of Company management, together with representatives of Jefferies, in Como, Italy to discuss the companies' respective long-term technology plans and strategies. Additional representatives of Parent also participated in these discussions via telephone. Following the meeting, two representatives of Parent visited the Company's manufacturing facility in Villa Guardia (CO), Italy. On that same day, the Company announced that the European Commission had granted a Marketing Authorization for Defitelio® (defibrotide) for the treatment of severe hepatic veno-occlusive disease in adults and children undergoing hematopoietic stem cell transplantation therapy. Defitelio® is the first approved treatment in the European Union for this life-threatening condition, which the Company currently

plans to begin marketing in the European Union in the first quarter of 2014.

On October 23, 2013, Mr. Haigh contacted representatives of Company A by email to inquire whether they were interested in continuing discussions and advancing diligence on the Company in light of the Marketing Authorization received by the Company the prior day. The representatives of Company A indicated that they would be interested in engaging in discussions.

On October 24, 2013 and October 28, 2013, Dr. Islam updated the members of the Company Board via email on various matters, including the status of the Company's solicitation process and search for outside legal counsel in connection with such process.

On October 30, 2013, Parent sent a second due diligence request list to the Company.

During the week of November 4, 2013, representatives of Parent and the Company held multiple due diligence calls to cover a wide range of topics, including commercial, technical operations, clinical development, regulatory and intellectual property matters.

Between November 4, 2013 and November 6, 2013, Dr. Islam, Mr. Haigh, Mr. Calabrese, and Dr. Mosconi, together with representatives of Jefferies, attended the BIO-Europe 2013 conference in Vienna, Austria. A number of the companies that were previously contacted on behalf of the Company by Jefferies as part of the Company's solicitation process were present at the conference, and management met with 13 of those companies regarding the Company's business, operations and long-term strategic plans. None of the other companies previously contacted on behalf of the Company by Jefferies as a part of the Company's solicitation process accepted the Company's invitation to meet in Vienna.

On November 6, 2013, Company A indicated by email to representatives of Jefferies that it was not interested in pursuing a transaction with the Company.

On November 11, 2013, Dr. Islam provided an update to the members of the Company Board on the status of the Company's solicitation process and the meetings held in Vienna, and indicated that the Company's discussions with Parent, as well as Parent's due diligence review, were at a relatively advanced stage. Of the 13 companies that met with management at BIO-Europe, three of them (Company C, Company D and Company E) expressed interest in conducting more detailed due diligence on the Company and executed confidentiality agreements with the Company between November 11, 2013 and November 13, 2013. Seven other companies did not request access to more detailed due diligence on the Company, but continued a dialogue with Jefferies regarding their potential interest in the Company over the next several weeks. After consulting with the Company's advisors, the Company entered into a customary form of confidentiality agreement, which included a one-year standstill provision, with each of Company C, Company D and Company E. All of these standstill provisions terminated by their terms as a result of the execution of the tender offer agreement between the Company and Parent and, as of the date hereof, no longer have force or effect.

Each of Company C, Company D and Company E began receiving due diligence documents and information after the signing of the relevant confidentiality agreement.

On November 14, 2013, Mr. Cozadd called Dr. Islam to advise him that Parent would submit a non-binding proposal to the Company the following week. That same day, the Company launched a new electronic data room and provided access to Parent's representatives.

Beginning on November 15, 2013, each of, Company C, Company D and Company E was provided access to the Company's electronic data room.

On November 19, 2013, Dr. Islam received a letter from Parent setting forth a written non-binding proposal pursuant to which Parent proposed to acquire all outstanding Company Shares for cash consideration of \$53.00 per share, subject to due diligence and execution of definitive documentation. In its letter, Parent stated that its proposal took into consideration the medical and economic potential of Defitelio®, inclusive of the needed investment in commercial infrastructure, marketing, and post-approval development to support U.S. approval, additional indications and other requirements, and expressed Parent's desire to complete a transaction on an accelerated timeline, with a goal of completing due diligence and signing a definitive agreement by the middle of December, subject to due diligence and final approval by or on behalf of Parent's board of directors. Parent stated that its proposal would not be subject to financing conditions. Parent also requested a 30-day period of exclusivity to complete due diligence and negotiate and execute mutually acceptable definitive agreements, and requested that the Company respond to Parent's proposal by 5:00 P.M. (Pacific Standard Time) on November 26, 2013, after which time Parent's proposal would terminate.

On November 20, 2013, Jefferies and representatives of Barclays PLC ("**Barclays**"), financial advisor to Parent, discussed Parent's proposal by telephone. In accordance with the Company's directives, Jefferies informed Barclays that the Company had commenced a third-party solicitation process for the Company in which Parent was invited to participate.

Also on November 20, 2013, the M&A Committee held a telephonic meeting to evaluate Parent's proposal. Representatives of Jefferies also attended such telephonic meeting, and summarized the status of the bidding process for the M&A Committee. The M&A Committee determined that it was willing, on the basis of Parent's proposal, to continue discussions and allow Parent access to further due diligence information. However, the M&A Committee determined that it could not recommend a transaction to the Company Board at the price proposed by Parent in its November 19, 2013 letter and, therefore, would not grant exclusivity to Parent and would instead continue the ongoing process with Parent and other potential bidders.

On November 21, 2013, Jefferies delivered on behalf of the Company the Company's process letter outlining its procedures for soliciting interest in the Company to 10 companies (including Company C, Company D and Company E) with which there remained continued dialogue regarding their potential interest in the Company, instructing such parties to submit written preliminary, non-binding indications of interest no later than 5:00 P.M. (Eastern Standard Time) on December 20, 2013.

That same day, Barclays sent Jefferies a high level summary of Parent's confirmatory due diligence requests. From November 21, 2013 through the signing of the definitive agreements on December 19, 2013, Parent and its legal and financial advisors conducted a due diligence investigation of the Company in person, by phone and through document review.

On November 23, 2013, Skadden, Arps, Slate, Meagher & Flom LLP ("**Skadden**") was engaged as outside U.S. legal counsel to the Company.

On November 25, 2013, at the Company's request, Jefferies contacted representatives of Barclays and Parent by telephone and confirmed that the Company had commenced a third-party solicitation process and had requested that interested parties submit non-binding proposals by December 20, 2013. On behalf of the Company, Jefferies also communicated that the Company Board would not be interested in pursuing a transaction at \$53.00 per share, but would likely consider a transaction at a price of \$57.00 per share.

On November 27, 2013, Mr. Cozadd contacted Dr. Islam by telephone and indicated that Parent would consider increasing its offer price to \$57.00 per share in exchange for the Company agreeing to an exclusive negotiation period, subject to confirmatory due diligence, the execution of a definitive agreement containing strong deal protections satisfactory to Parent, including support agreements with the Company's principal shareholders, as well as the Company's management and directors.

Later that day, the M&A Committee held a conference call to evaluate Parent's increased non-binding indication of interest. Dr. Islam opened the meeting by providing the committee with an update on the status of the ongoing process and discussions. The M&A Committee determined that Parent's revised proposal represented a price that it could recommend to the Company Board, subject to agreeing mutually acceptable terms in definitive documentation. The M&A Committee again considered Parent's request for exclusivity and again decided not to enter into an exclusive arrangement in light of the ongoing process with other potential bidders.

After the meeting of the M&A Committee, Dr. Islam contacted Mr. Cozadd by telephone and expressed a desire to continue discussions with Parent relating to a potential sale of the Company. During this call, Dr. Islam verbally communicated to Parent that the Company could not grant a 30-day period of exclusivity given the Company's ongoing solicitation process, although the parties agreed to pursue further discussions on the basis of Parent's revised proposal made earlier that day.

On November 28, 2013, Weil, Gotshal & Manges LLP ("**Weil**"), outside U.S. legal counsel to Parent, sent a legal due diligence request list to Skadden.

On November 29, 2013, Mr. Calabrese, together with representatives of Skadden, GOP and Jefferies, held a telephonic meeting with representatives of Parent, its financial advisor Barclays, and Weil, to address a number of preliminary considerations relating to a potential transaction between the Company and Parent, including the due diligence process, timing of definitive agreements, and potential antitrust filings. Later that day, at the direction of the Company's senior management, Skadden sent a first draft transaction agreement to Parent and its legal and financial advisors, together with a marked version of Weil's due diligence request list.

Between November 29, 2013 and December 2, 2013, in accordance with the Company's directives, Jefferies contacted each of Company C, Company D and Company E, and informed them that December 13, 2013 had been set as the new deadline to submit a non-binding indication of interest for the Company. Between December 2, 2013 and December 6, 2013 the remaining parties that had received a process letter and had continued dialogue regarding their potential interest in the Company were contacted at the Company's request by Jefferies and informed of the December 13, 2013 deadline.

On December 2, 2013, Mr. Calabrese, together with representatives of Skadden, GOP and Jefferies, had a telephone call with representatives of Weil to review Weil's due diligence request list and provide primary contacts for further discussions related to various requested items. Later that day, representatives of Skadden, GOP and Weil held an additional telephonic meeting to discuss key open points relating to the draft transaction agreement.

On December 4, 2013, Weil submitted a markup to the draft transaction agreement to Skadden on behalf of Parent that, among other things: (i) contemplated a tender offer only (rather than a tender offer followed by a cross-border European merger); (ii) increased the minimum condition to the tender offer by requesting that there be validly tendered at least 66 2/3% of the Company Shares on a fully diluted basis (rather than a simple majority); (iii) requested a top-up option, which would allow Parent to purchase newly-issued shares of the Company following completion of the Offer as may be required to reach the proposed 66 2/3% minimum condition; (iv) added a number of conditions to the offer, including more extensive conditions surrounding the absence of pending or threatened government action challenging the transaction and a condition that six of seven directors resign from the Company Board prior to completion of the Offer; (v) requested certain changes to the parties' ability to terminate the agreement, and proposed an increased termination fee equal to 3% of the total consideration for Company Shares (rather than Parent's costs and expenses of up to 0.2% of the total consideration for Company Shares); (vi) requested additional representations and warranties from the Company; (vii) requested additional affirmative and negative obligations of the Company; (viii) requested changes to the Company's ability to respond to an unsolicited proposal and Parent's ability to match any such proposal; (ix) proposed a prohibition on the Company's ability to waive any standstill, confidentiality, non-solicitation, no hire or similar provisions in any contracts to which it is a party; and (x) requested changes to the Company Board's ability to change its recommendation with respect to the transaction with Parent. The draft further requested that certain significant and affiliated shareholders of the Company (including F3F S.r.l. and Sigma-Tau) and holders of Company Options, collectively representing approximately 38% of the fully diluted, outstanding share capital of the Company, enter into support agreements with Parent, containing a "tail" provision that would prohibit such holders from tendering Company Shares into a competing offer for a 12-month period notwithstanding a change of recommendation by the Company Board and granting Parent an option to acquire such shareholders' shares, as well as non-competition and employee non-solicitation agreements with Parent. The draft also requested that certain key Company employees enter into employment agreements with Parent.

On December 4, 2013, Mr. Cozadd met in New York with representatives of Jefferies that included a discussion of matters related to the potential transaction with the Company.

On December 5, 2013, Dr. Islam met with representatives of Sigma-Tau (Mr. Tugnoli, advisor, and Mr. Montevocchi, CEO and President of Sigma-Tau), one of the Company's largest shareholders, in Milan, Italy to discuss in general terms, on a no-names basis, the possibility of a potential bid for the Company and the approach Sigma-Tau would take in such event.

Later on December 5, 2013, the Company Board held a meeting to discuss the status of the increased non-binding indication of interest communicated by Parent on November 27, 2013, review the status of discussions with other potential bidders and discuss the ongoing process and timing for the solicitation of non-binding proposals. At the meeting, Dr. Islam informed the Company Board that Parent's due diligence review of the Company was at an advanced stage compared to the other participating bidders that had signed confidentiality agreements and been granted virtual data room access and that Parent had expressed its desire to sign a definitive transaction agreement with the Company no later than December 20, 2013. Dr. Islam explained that, as a result, the other bidders had been informed that the deadline for sending non-binding indications of interest had been accelerated. Jefferies updated the Company Board regarding Parent's most recent non-binding proposal and informed the Company Board that no other proposals had been submitted. Skadden delivered a presentation regarding transaction structure, terms and conditions of the tender offer agreement and related legal issues, while GOP advised the Company Board regarding its fiduciary duties under the circumstances.

Also on December 5, 2013, Mr. Cozadd and Dr. Islam discussed by phone the status of Parent's due diligence and agreement negotiations. Mr. Cozadd requested that the Company accelerate the process that the Company had commenced to seek non-binding proposals from other potential acquirers by December 20, 2013 so that the parties could execute definitive agreements during the week of December 16, 2013.

On December 6, 2013, the M&A Committee held a telephone conference, at which Dr. Islam briefed the committee on the ongoing process and the draft transaction agreement. Given that discussions with Parent were at an advanced stage, the M&A Committee requested Dr. Islam and his management team to undertake confidential discussions about a potential transaction with Parent with certain principal shareholders of the Company, namely F3F S.r.l. and Sigma-Tau, as soon as advanced drafts of Parent's proposed support agreements were received.

On December 6, 2013, Mr. Cozadd spoke with Dr. Islam by phone and proposed to increase Parent's offer price to \$57.00 per share in exchange for a commitment from the Company to work towards signing definitive agreements on or about December 18, 2013 and commencing the tender offer soon thereafter, and the Company's agreement to provide certain deal protections, including support agreements from certain key shareholders containing deal protection mechanisms satisfactory to Parent.

Also on December 6, 2013, the Company received a revised due diligence request list from Weil, and Mr. Calabrese, together with representatives of Skadden, GOP and Jefferies, held a telephonic meeting with representatives of Parent, Barclays, Weil, Cooley LLP ("**Cooley**"), Parent's outside financing and IP counsel, and the Italian outside legal co-counsel to Parent, Baker & McKenzie ("**Baker**") and Gattai Minoli & Partners ("**Gattai**").

Between December 6, 2013 and December 13, 2013, representatives of Parent engaged in administrative, tax and internal controls due diligence at the Company's headquarters and manufacturing facilities.

Between December 7, 2013 and December 19, 2013, Parent and its legal, financial, accounting and tax advisors conducted extensive due diligence, including additional due diligence calls with the Company and Skadden on December 11, 2013 and December 16, 2013, to discuss further revised due diligence questions and requests.

On December 8, 2013, Mr. Calabrese met with representatives of F3F S.r.l, one of the Company's principal shareholders, to inform it of Parent's offer.

On December 11, 2013, following meetings with the Company's management regarding the revised draft tender offer agreement submitted by Parent, Skadden sent a revised draft to Weil. This revised draft, among other things: (i) accepted the tender offer structure without a second step cross-border European merger; (ii) preserved the minimum condition at a majority of Company Shares on a fully diluted basis and rejected the concept of a top-up option; (iii) accepted some but not all of the additional conditions to offer; (v) proposed a termination fee equal to 1% of the total consideration for Company Shares (payable by a topping bidder rather than the Company under certain circumstances); (vi) accepted some but not all of the additional representations and warranties from the Company requested by Parent, including as to Parent's financing; (vii) proposed additional representations and warranties and covenants from Parent and Purchaser, including in respect of Parent and Purchaser's solvency and interests in the Company and in connection with the financing of the Offer; (viii) accepted some but not all of the interim operations covenants requested by Parent; (ix) proposed changes to the Company's ability to respond to an unsolicited proposal and Parent's ability to match any such proposal; (x) rejected Parent's request for a prohibition on the Company's ability to waive any standstill, confidentiality, non-solicitation, no hire or similar provisions in any contracts to which it is a party; and (xi) proposed changes to the provision relating to the Company Board's ability to change its recommendation with respect to the transaction with Parent. Skadden also communicated to Weil that the Company could not accept Parent's request that shareholders and option holders representing approximately 38% of the fully diluted share capital of the Company enter into support agreements containing a 12 month "tail" provision and granting Parent an option to acquire such shareholders' shares, as well as non-competition and employee non-solicitation agreements with Parent.

On December 11, 2013, Mr. Cozadd spoke with Dr. Islam by phone to discuss the status of the negotiations and the due diligence process. Mr. Cozadd expressed the importance to Parent of getting the requested support agreements from key shareholders in connection with the transaction.

On December 12, 2013, Dr. Islam called Mr. Tugnoli of Sigma-Tau, informing him of the possibility of a potential business combination with Parent and Parent's request that Sigma-Tau support the transaction by entering into a shareholder support agreement. The following day, Dr. Islam met with Mr. Tugnoli and Mr. Montevocchi of Sigma-Tau in Milan, Italy to further discuss the potential transaction. At the meeting, Sigma-Tau expressed general support for the proposed price, but expressed concerns relating to uncertainty as to how such new owner of the Company would approach the existing licensing arrangements between the Company and Sigma-Tau.

Also on December 12, 2013, Weil communicated to Skadden that Parent was renewing its request for support agreements from shareholders and option holders that together owned approximately 38% of the fully diluted capital stock of the Company, but would no longer request that such shareholders grant Parent an option to acquire their shares (although Parent would continue to request that such shareholders grant Parent a proxy over such shares) and was reducing its request for a "tail" from 12 to six months.

On December 13, 2013, Weil sent Skadden a form of support agreement for certain shareholders and option holders of the Company, including certain directors, officers and employees of the Company, to be entered into concurrently with the execution of the tender offer agreement. The draft support agreement included a six-month "tail" provision and required the shareholders to grant Parent a proxy over their Company Shares. Later that day, representatives of Skadden and Weil had a telephone call to negotiate the terms of the tender offer agreement.

Also on December 13, 2013, Skadden, GOP and Weil had a telephone call to discuss and negotiate portions of the draft tender offer agreement.

On or prior to December 13, 2013, each of the 10 contacted parties that received a process letter (including Company C, Company D and Company E) communicated to Jefferies that they would not be submitting a proposal for the Company. Reasons cited by parties included, among other factors, valuation considerations, the fact that the Company's product had not been approved in the U.S. and the fact the Company already had in Sigma-Tau an existing U.S. commercial partner for the product.

On December 14, 2013, Mr. Cozadd requested a telephone call with Dr. Islam, at which they discussed certain key aspects of the transaction documents, such as the minimum condition to the tender offer, the treatment of the outstanding options granted to a number of employees of the Company and its subsidiary, and the six-month "tail" provision included in the draft support agreements that Parent sought to enter into with shareholders and option holders representing approximately 38% of the Company's shares on a fully-diluted basis.

That same day, the M&A Committee held a telephone conference at which Dr. Islam updated the committee on the ongoing process, on the draft transaction agreement and on his conversation with Mr. Cozadd earlier that day. In

addition, he outlined the on-going discussions and positions of F3F S.r.l. and Sigma-Tau as regards Parent's offer and request for support agreements. After discussion, the M&A Committee resolved that the draft support agreement be forwarded to all board members and to Sigma-Tau and F3F S.r.l. The M&A committee also discussed the outstanding issue of Parent's request for a 6 month tail provision in the support agreements it was seeking from the Company's major shareholders and decided to instruct Skadden to discuss alternative scenarios with Parent's counsel and to push for the best outcome that could be achieved for the Company.

On December 15, 2013, Weil submitted a further revised tender offer agreement that, among other things: (i) reinstated a minimum condition at 66 2/3% of the Company Shares on a fully diluted basis, but did not include a request for a top-up option clause; (ii) proposed a termination fee equal to (a) 3% of the total consideration for Company Shares in the event the tender offer agreement was terminated due to a Superior Proposal (as defined in the tender offer agreement), and (b) 1% of the total consideration for the Company Shares, in the event the tender offer agreement was terminated for a different reason, it being understood that, if a Specified Acquisition Transaction (as defined in the tender offer agreement) were consummated within 12 months following the date of termination, an additional amount equal to 2% of the total consideration for the Company Shares would become due; and (iii) accepted some but not all changes to the representations and warranties from the Company. With this revised draft, Parent insisted that the support agreement would include a six-month "tail" provision and indicated that it would consider requiring certain ancillary agreements with a number of key employees of the Company and its subsidiary. Later that day, Skadden, GOP and Weil had a telephone call in which the terms of the tender offer agreement were further negotiated.

On December 16, 2013, Mr. Cozadd spoke with Dr. Islam by phone to discuss the status of the negotiations and the due diligence process and, again, underscored the importance of the support agreements, including the six-month tail provision, from F3F S.r.l., Sigma-Tau and each director and officer of the Company.

Later on December 16, 2013, Weil sent to Skadden proposed drafts of: (i) a transition and release agreement, and a consulting agreement to be executed by Dr. Islam; (ii) retention and amendment agreements to be executed by Mr. Calabrese and several other senior managers of the Company and its subsidiary; and (iii) a revised form of support agreement to be executed by certain directors, officers and employees of the Company and its subsidiary, including a non-competition covenant. The same day, representatives of Skadden and Weil had multiple telephone calls to further discuss, negotiate and agree upon portions of the tender offer agreement. During such call, representatives of Skadden again indicated to Weil that the Company Board would not support the transaction if shareholders and option holders representing 38% of the Company's fully diluted share capital were subject to support agreements containing a "tail" provision.

On December 17, 2013, Mr. Cozadd called Dr. Islam to inform him that Parent's board of directors had given Mr. Cozadd support to move forward with a transaction agreement with the Company. However, Mr. Cozadd confirmed that Parent continued to require support agreements in favor of the transaction from key shareholders of the Company.

Also on December 17, 2013, Skadden sent to Weil a draft of the disclosure letter, and a revised draft of the tender offer agreement.

Later on December 17, 2013, representatives of Sigma-Tau contacted the Company and Skadden to provide a mark-up of the support agreement proposed by Parent. In response, the Company informed Sigma-Tau's representatives that they should contact representatives of Parent to negotiate the terms of the support agreement directly.

Between December 17, 2013 and December 19, 2013, the final terms of the tender offer agreement and the disclosure letter were negotiated.

On December 18, 2013, the Company Board held a telephonic meeting, with representatives of Skadden, GOP and Jefferies present. Representatives of Skadden summarized the terms of the near-final Tender Offer Agreement. Jefferies reviewed with the Company Board its financial analysis of the \$57.00 per share cash consideration and delivered to the Company Board an oral opinion, confirmed by delivery of a written opinion dated December 18, 2013, to the effect that, as of that date and based on and subject to various assumptions, matters considered and limitations and qualifications described in its opinion, the \$57.00 per share cash consideration to be received in the Offer by holders of Company Shares (other than Parent, Purchaser and their respective affiliates) was fair, from a financial point of view, to such holders.

Following a discussion, the tender offer agreement was approved by the unanimous vote of the Company Board (other than for one director, Mr. Marco Brughera, who abstained due to the potential conflicts of interest resulting from the security ownership of Sigma-Tau, of which he is a representative), and the Company's management was authorized to enter into the tender offer agreement subject to such amendments as the Company's M&A Committee may deem advisable. The Company Board also discussed the acceleration of all outstanding Company Options in connection with, and contingent upon, the completion of the Offer, such that the holders of Company Options would be able to exercise their options and tender Company Shares in the Offer, or a subsequent offering period relating to the Offer, and resolved to form the Special Committee, composed of Mr. Schnee, Dr. Sandage and Ms. Bigio, to consider and approve such matter as well as the Transaction Bonus, the Transition and Release Agreement, the Consultancy Agreement and the Retention Agreements proposed by Parent.

Later that afternoon, the Special Committee of independent directors held a telephonic meeting at which they discussed and approved the acceleration of all outstanding Company Options in connection with the Offer, the Transition and Release Agreement, the Consultancy Agreement and the Retention Agreements proposed by Parent and approved the Transaction Bonus.

In the late evening of December 18, 2013, Parent communicated to the Company that it was prepared to enter into the tender offer agreement despite Sigma-Tau having communicated to Parent that it would not enter into a support agreement with Parent, and on the basis that F3F S.r.l. and certain directors and officers, together representing approximately 15% of the outstanding Company Shares and approximately 23% of the Company's fully diluted share capital, in each case as of that date, enter into support agreements with Parent.

On December 19, 2013, the M&A Committee held a telephonic meeting to discuss the status of the process and certain amendments proposed to the latest draft of the Tender Offer Agreement of the previous day, which had been discussed overnight between Skadden and Weil.

Later that day, Parent, Purchaser and the Company executed and delivered the Tender Offer Agreement, and certain directors and officers of the Company executed and delivered the Support Agreements, the Transition and Release Agreement, the Consultancy Agreement and the Retention Agreements. Later that day, the Company, Parent and Purchaser issued a joint press release announcing the transaction.

Reasons for the Recommendation.

At a meeting on December 18, 2013, the Company Board unanimously (other than for one director, Mr. Marco Brughera, who abstained due to the potential conflicts of interest resulting from the security ownership of Sigma-Tau, of which he is a representative): (1) determined that the Tender Offer Agreement is advisable and fair to, and in the

best interest of, the Company, its shareholders and other stakeholders; (2) determined that the Tender Offer Agreement and the transactions contemplated thereby, including the Offer, taken together, are at a price and on terms that are in the best interests of the Company, its shareholders and other stakeholders; and (3) approved the Tender Offer Agreement and the transactions contemplated thereby, including the Offer.

The Company Board unanimously (other than for one director, Mr. Marco Brughera, who deemed himself to be potentially conflicted) recommends that the Shareholders accept the Offer and tender their Company Shares pursuant to the Offer.

In evaluating the Tender Offer Agreement and the transactions contemplated thereby, the Company Board consulted with the Company's senior management, outside legal counsels and financial advisor. The Company Board concluded that entering into the Tender Offer Agreement with Parent and Purchaser is in the best interests of the Company, its shareholders and other stakeholders and recommends that the Company's shareholders accept the Offer and tender their Company Shares pursuant to the Offer.

The following discussion includes the material reasons and factors considered by the Company Board in making its recommendation, but is not, and is not intended to be, exhaustive:

Strategic Alternatives; Benefits of Being Integrated Into a Pharmaceutical Group with Greater Resources Weighed Against Risks of Remaining Independent. Throughout 2013 the Company Board considered the Company's strategic alternatives, particularly in light of the uncertainty surrounding the outcome of the regulatory approval process for Defitelio® (defibrotide) in Europe, the U.S. and other parts of the world. Among the alternatives considered, the Company Board considered a capital raise, a sale to a strategic investor or remaining independent. Following the CHMP's positive recommendation, as the date for approval of the European Commission approached, the Company Board weighed the benefits to the Company of being integrated into a larger, diversified pharmaceutical group that could support the commercialization of Defitelio® (defibrotide) against the prospective risks to the Company as a stand-alone entity, including, (A) risks associated with executing a successful commercial launch of Defitelio® in Europe in the first and second quarter of 2014, (B) uncertainty surrounding the regulatory clearance of Defitelio® in the U.S. and elsewhere, (C) the ability to maintain patent coverage for Defitelio® and risk associated with generic competition earlier than expected, (D) the Company's financial resources relative to competitors and (E) the introduction of new products by the Company in the future. In light of these and other factors, the Company Board believes that the value offered to the Company's shareholders in the Offer is more favorable to shareholders than the potential value that could be achieved if the Company remained an independent company.

Scope of Sales Process; Ability of Interested Parties to Make Superior Proposals. The Company Board considered the extensive scope of the process initiated by the Company beginning in September 2013 to explore possible strategic transactions with third parties, during which the Company, with the assistance of the M&A Committee and legal and financial advisors, contacted 38 potential counterparties (including Parent) regarding their potential interest in a strategic transaction with the Company, of which 13 attended a management presentation with the Company and four entered into confidentiality agreements with the Company (Parent and Companies C, D and E) and had access to substantial information on the Company. The Company Board considered the fact that during the process no other party had come forward and made a proposal for the Company. The Company Board likewise considered the relative likelihood that the most probable interested parties had been identified in the course of this process, while noting that the Tender Offer Agreement and related agreements would not foreclose the Company's ability to respond to superior proposals if made after the Tender Offer Agreement was signed.

Parent's Representations Relating to its Intentions with respect to the Business. The Company Board considered Parent's representation that Parent's current intent is that, following the Acceptance Time, the business of the Company would continue to be headquartered in Villa Guardia (CO), Italy, and that the production activities of the Business and substantially all of the other operations of the business would continue to be carried out substantially as they were on the date of the Tender Offer Agreement.

Future Availability of Parent Offer. The Company Board considered the risk that if the Company did not accept Parent's offer, there may not have been another opportunity to do so.

Cash Tender Offer; Certainty of Value. The Company Board noted that the form of consideration to be paid to Shareholders in the Offer was all cash and considered the certainty of value of such cash consideration.

Likelihood of Consummation. The Company Board considered the likelihood that the Offer will be consummated, based on, among other things, the business reputation of Parent and its management and the substantial financial resources of Parent and, by extension, Purchaser, the limited number of conditions to the Offer, the absence of a financing condition and the absence of a regulatory filings condition, Parent's representation that it has obtained a financing commitment from Barclays to provide funding for the Offer, and the fact that the consummation of the Offer is conditioned on 66 2/3% of Company Shares on a fully-diluted basis being validly tendered in the Offer and that such minimum tender condition could be waived by Parent to a majority of all outstanding shares.

Tender Offer Structure. The Company Board considered the fact that the transaction is structured as a tender offer and is not subject to financing or regulatory filings conditions, which means that it can be completed expeditiously and that cash consideration can be delivered to shareholders promptly, reducing the period of uncertainty during the pendency of the transaction on the shareholders, employees and business partners. The Company Board also considered the fact that the completion of the Offer will be followed by a subsequent offering period, in which shareholders who do not tender their shares in the Offer will receive the same cash price as is offered in the Offer.

Premium to Market Price. The Offer Price to be paid by Purchaser would provide the Shareholders with the opportunity to receive a significant premium over the market price of the Company Shares. The Company Board reviewed the historical market prices and trading information with respect to the ADSs, including the fact that the Offer Price represents:

a 22.2% premium over the \$46.66 per share closing price of the ADSs on November 17, 2013, approximately 30 days prior to the date of the Company Board's approval of the Offer;

a 72.7% premium over the \$33.00 per share closing price of the ADSs on October 23, 2013, the trading day following the Company's announcement that it had received marketing authorization from the EU for Defitelio; and

a 149.7% premium over the \$22.83 volume weighted average closing price of the ADSs for the ten days prior to September 27, 2013, the date that the Company Board decided to engage in a sale process.

Company Financial Advisor Opinion. The Company Board considered the financial presentation and opinion of Jefferies, dated December 18, 2013, to the Company Board as to the fairness, from a financial point of view and as of such date, of the \$57.00 per share cash consideration to be received in the Offer by holders of Company Shares (other than Parent, Purchaser and their respective affiliates), which opinion was based on and subject to the assumptions made, procedures followed, matters considered and limitations on the review undertaken by Jefferies as more fully

described under the caption "*Opinion of the Company's Financial Advisor.*" **Jefferies' opinion should not be construed as creating any fiduciary duty on Jefferies' part to any party and does not constitute a recommendation as to whether any shareholder should tender Company Shares in the Offer or how any shareholder should act with respect to the Offer or any related matter.**

Tender Offer Agreement. The Company Board considered the provisions of the Tender Offer Agreement, including the respective representations, warranties, covenants, conditions and termination rights of the parties and the termination fee payable by the Company. In particular:

No Financing Condition. The Company Board considered the representations of Parent and Purchaser that they will have access to sufficient cash resources to pay the amounts required to be paid under the Tender Offer Agreement and that the Offer is not subject to a financing condition, and that Parent had obtained a financing commitment from Barclays to provide funding for the Offer.

No Regulatory Filings Condition. The Company Board considered that the Offer is not subject to any waiting period or any regulatory clearance.

Ability to Respond to Certain Unsolicited Takeover Proposals. While the Company is prohibited from soliciting any Acquisition Proposal (as defined in the Tender Offer Agreement), the Tender Offer Agreement does permit the Company Board, subject to compliance with certain requirements (including that the Company Board determines that the Acquisition Proposal is reasonably likely to result in a Superior Proposal (as defined in the Tender Offer Agreement)) to (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.

Change in Recommendation in the Event of a Superior Proposal or an Intervening Event/Termination Right in the event of a Superior Proposal. The Company Board may withdraw or change its recommendation in favor of the Tender Offer Agreement and the Offer, if either a Superior Proposal is received or an Intervening Event (as defined in the Tender Offer Agreement) occurs, and may terminate the Tender Offer Agreement if a Superior Proposal is received, in the following circumstances.

In order for the Company Board to withdraw its recommendation in connection with a Superior Proposal, the Company (A) must not have breached its non-solicitation obligations or other specified obligations in such a way that resulted in the applicable Acquisition Proposal being made, (B) must give Parent notice of 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, (C) the Company Board determines in good faith (after consulting with the Company's financial advisor and outside legal counsel) that the Acquisition Proposal constitutes a Superior Proposal, (D) 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, among other things, that it intends to make a Company Board Recommendation Change or terminate the Tender Offer Agreement and of the material terms and conditions of such Superior Proposal (and any change to the consideration payable in connection with such Superior Proposal or any other material modification would require a new advance written notice by the Company (except that the four (4) Business Day period referred to above would be reduced to two (2) Business Days)), and (E) 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 the Tender Offer Agreement in such a manner that obviates the need for withdrawing the Company Board Recommendation or otherwise effecting a Company Board Recommendation Change.

In order for the Company Board to withdraw its recommendation in connection with an Intervening Event, the Company, among other things, must (A) give Parent notice of 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, (B) promptly notify Parent, in writing, at least four (4) Business Days before withdrawing the Company Board Recommendation or otherwise making any Company Board Recommendation Change, among other things, that it intends to make a Company Board Recommendation Change, and (C) during such notice period, if requested by Parent, engage in good faith negotiations with respect to any proposal made by Parent to amend the Tender Offer Agreement in such a manner that obviates the need for withdrawing the Company Board Recommendation or otherwise effecting a Company Board Recommendation Change.

Termination Fee.

The Company Board considered the fact that the Company must pay Parent a termination fee of \$25,300,000 if the Tender Offer Agreement is terminated in anticipation of or concurrently with entering into a Superior Proposal or an alternative termination fee of \$10,100,000 if the Tender Offer Agreement is terminated in certain other circumstances.

The Company Board also considered the fact that, if within one year following the termination of the Tender Offer Agreement, the Company consummates another transaction meeting certain criteria, the Company may be required to pay a termination fee of either \$15,200,000 or \$25,300,000 depending on the circumstances under which the Tender Offer Agreement was terminated (but that in no event would the Company be required to pay more than \$25,300,000 in termination fees).

The Company Board was of the view that the termination fees payable by the Company to Parent, if the Tender Offer Agreement is terminated for the reasons discussed in the Tender Offer Agreement, were reasonable, given the bidding and negotiation process that had been followed by the Company, the range of termination fees in precedent transactions, and the other terms of the Tender Offer Agreement, and would not likely deter competing bids and would not likely be required to be paid unless the Company Board entered into or intended to enter into a transaction more favorable to the Shareholders than the Offer.

Conditions to the Completion of the Offer. The Company Board considered the reasonable likelihood of the consummation of the transactions contemplated by the Tender Offer Agreement in light of the limited conditions to Purchaser's obligations to accept for payment and pay for the Company Shares tendered pursuant to the Offer.

Company Material Adverse Effect. The Company Board considered that the Tender Offer Agreement provides that any fact, circumstance, occurrence, event, change or effect ("**Effect**") related to the Company or its business, operations, assets (including intangible assets), financial condition or results of operations arising or resulting from the following items are all excluded as a Company Material Adverse Effect for determining Parent's obligation to complete the Offer:

general market, economic or political conditions in Italy or worldwide or conditions (or any changes therein) in the industries in which the Company conducts its business, in each case, including any acts of terrorism or war, weather conditions or other force majeure events, in each case, 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;

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 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;

o changes in GAAP or other accounting standards (or the interpretation thereof by a third party);

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 relative to other companies operating in the same industries in which the Company or any of its Subsidiaries conduct business;

- o changes in the trading price or trading volume of the Company ADSs, in and of themselves; or

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, in and of itself.

Reasonable Best Efforts to Close. The Company Board considered that Parent is required by the Tender Offer Agreement to use its reasonable best efforts to close the Offer.

Extension of Offer Period. The Company Board considered the Tender Offer Agreement's requirement that Purchaser is required to extend the Offer beyond the initial expiration date of the Offer or, if applicable, subsequent expiration dates, for up to April 19, 2014 if the conditions to the Offer are not satisfied on such expiration dates and up to September 19, 2014 if the condition that there be no pending or overtly threatened action by government entities is not satisfied (and certain other conditions are then satisfied or waived).

Enforcement. The Company Board considered the Company's ability to seek specific performance of Parent's and Purchaser's obligations under the Tender Offer Agreement, thereby ensuring that the Company has an appropriate remedy in the event that Parent and Purchaser were to decline to comply with their obligations under the Tender Offer Agreement.

The Company Board also considered potential risks or negative factors relating to the Offer, including the following:

Risk of Non-Consummation. The possibility that the transactions contemplated by the Tender Offer Agreement might not be consummated, the fact that if the Offer is not consummated, the Company's directors, senior management and other employees will have expended extensive time and effort and will have experienced significant distractions from their work during the pendency of the transaction, the fact that the Company will have incurred significant transaction costs, and the possibility that the Company's continuing business could potentially suffer a loss of customers, business partners and employees.

Termination Fee. The possibility that the \$25,300,000 termination fee, payable in circumstances where the Company recommends or accepts an alternative transaction as a Superior Offer, could potentially dissuade a potential acquirer from proposing a transaction that could be of greater value to the Company's shareholders than the Offer.

Impact of Announcement on the Company. The effect of the public announcement of the Tender Offer Agreement, including effects on the Company's sales, customer, reseller and other channel partner relationships, operating results and stock price, and the Company's ability to attract and retain key management and sales and marketing personnel.

Restrictions on the Company's Conduct of Business. The potential limitations on the Company's pursuit of business opportunities due to certain covenants in the Tender Offer Agreement, whereby the Company agreed that it will carry on its business in the ordinary course of business consistent with past practice and, subject to specified exceptions, will not take a number of actions related to the conduct of its business without the prior written consent of Parent. These restrictions could delay or prevent the Company from undertaking business opportunities that may arise prior to the consummation of the Offer and that may have a material and adverse effect on the Company's ability to respond to changing market and business conditions in a timely manner or at all.

Tax Treatment. The fact that the cash consideration to be received by the Company's shareholders in the Offer would be taxable to the Company's shareholders. The Company Board believed that this was mitigated by the fact that the entire consideration payable in the transaction would be cash, providing adequate cash for the payment of any taxes due.

Conflict of Interest. The interests of the executive officers and directors of the Company in the Offer, including the matters described under *Item 3—"Past Contacts, Transactions, Negotiations and Agreements"* of this Statement.

Control of the Company Board after Consummation of Offer. The provisions of the Tender Offer Agreement that provide Parent with the right to obtain representation on the Company Board following completion of the Offer.

No Shareholder Participation in Future Growth or Earnings. The fact that the nature of the Offer as a cash transaction means that the Company's shareholders will not participate in future earnings or growth of the Company and will not benefit from any appreciation in its value, unless they otherwise acquire common stock of Parent.

The foregoing discussion of the information and factors considered by the Company Board is intended to be illustrative and not exhaustive, but includes the material reasons and factors considered. In view of the wide variety of reasons and factors considered, the Company Board did not find it practical to, and did not, quantify or otherwise assign relative weights to the specified factors considered in reaching its determinations or the reasons for such determinations. Individual directors may have given differing weights to different factors or may have had different reasons for their ultimate determination. In addition, the Company Board did not reach any specific conclusion with respect to any of the factors or reasons considered. Instead, the Company Board conducted an overall analysis of the factors and reasons described above and determined that, in the aggregate, the potential benefits considered outweighed the potential risks or possible negative consequences of the Offer.

Intent to Tender.

To the knowledge of the Company after making reasonable inquiry, to the extent permitted by applicable securities laws, rules or regulations, each executive officer and director of the Company currently intends and, other than Mr. Marco Brughera, has committed pursuant to the Support Agreements to tender all Company Shares over which they

have sole dispositive power pursuant to the Offer, including any Company Shares issued upon exercise of any Company Options (and to exercise any Company Options, contingent upon the commencement of the subsequent offering period, and tender all Company Shares issued upon such exercise of Company Options into the subsequent offering period).

Opinion of the Company's Financial Advisor.

The Company has retained Jefferies as its financial advisor in connection with the Offer. In connection with this engagement, the Company requested that Jefferies evaluate the fairness, from a financial point of view, of the consideration to be received in the Offer by holders of Company Shares (other than Parent, Purchaser and their respective affiliates). On December 18, 2013, at a meeting of the Company Board held to evaluate the Offer, Jefferies delivered to the Company Board an oral opinion, confirmed by delivery of a written opinion dated December 18, 2013, to the effect that, as of that date and based on and subject to various assumptions, matters considered and limitations and qualifications described in its opinion, the \$57.00 per share cash consideration to be received in the Offer by holders of Company Shares (other than Parent, Purchaser and their respective affiliates) was fair, from a financial point of view, to such holders.

The full text of Jefferies' opinion describes the assumptions made, procedures followed, matters considered and limitations on the review undertaken by Jefferies. This opinion is attached as Annex A and is incorporated herein by reference. **Jefferies' opinion was provided for the use and benefit of the Company Board (in its capacity as such) in its evaluation of the per share cash consideration payable in the Offer from a financial point of view and did not address any other aspect of the Offer. The opinion did not address the relative merits of the Offer or other transactions contemplated by the Tender Offer Agreement as compared to any alternative transaction or opportunity that might be available to the Company, nor did it address the Company's underlying business decision to engage in the Offer or the terms of the Tender Offer Agreement or the documents referred to in the Tender Offer Agreement, including, any term, aspect or implication of any support agreement or other agreements or arrangements contemplated by or resulting from the Offer or otherwise. Jefferies' opinion should not be construed as creating any fiduciary duty on Jefferies' part to any party and does not constitute a recommendation as to whether any shareholder should tender Company Shares in the Offer or how any shareholder should act with respect to the Offer or any related matter.** The following summary is qualified in its entirety by reference to the full text of Jefferies' opinion.

In arriving at its opinion, Jefferies, among other things:

- reviewed a draft, dated December 17, 2013, of the Tender Offer Agreement;

- reviewed certain publicly available financial and other information about the Company;

- reviewed certain information furnished to Jefferies by the Company's management relating to the Company's business, operations and prospects, including financial forecasts and estimates prepared by the Company's management;

held discussions with members of the Company's senior management concerning the matters described in the two preceding bullets;

held discussions, at the direction of the Company Board, with third parties to solicit indications of interest in the possible acquisition of the Company;

reviewed the trading price history and implied multiples for Company Shares and compared them with those of certain publicly traded companies that Jefferies deemed relevant;

compared the financial terms of the Offer with publicly available financial terms of certain other transactions that Jefferies deemed relevant; and

conducted such other financial studies, analyses and investigations as Jefferies deemed appropriate.

In its review and analysis and in rendering its opinion, Jefferies assumed and relied upon, but did not assume any responsibility to independently investigate or verify, the accuracy and completeness of all financial and other information that was supplied or otherwise made available by the Company or that was publicly available to Jefferies (including, without limitation, the information described above), or that was otherwise reviewed by Jefferies. Jefferies relied on assurances of the Company's management that it was not aware of any facts or circumstances that would make such information inaccurate or misleading. In its review, Jefferies did not obtain any independent evaluation or appraisal of any of the assets or liabilities (contingent or otherwise) of the Company, nor did Jefferies conduct a physical inspection of any of the properties or facilities of the Company, and Jefferies was not furnished with, and assumed no responsibility to obtain, any such evaluations, appraisals or physical inspections.

With respect to the financial forecasts and estimates provided to and reviewed by Jefferies, Jefferies noted that projecting future results of any company is inherently subject to uncertainty. Jefferies was informed, however, and assumed, that such financial forecasts and estimates were reasonably prepared on bases reflecting the best currently available estimates and good faith judgments of the Company's management as to the future financial performance of the Company. Jefferies also was informed that the Euros to U.S. Dollars exchange rate reflected in such forecasts and estimates was reasonable to utilize for purposes of Jefferies' analyses and opinion. Jefferies expressed no opinion as to any such financial forecasts or estimates or the assumptions on which they were based (including, without limitation, any currency or exchange rate fluctuations in Euros and U.S. Dollars). Jefferies relied upon the assessments of the Company's management as to, among other things, (i) the potential impact on the Company of governmental and regulatory policies affecting the pharmaceutical industry and (ii) the validity and risks associated with the products, product candidates and intellectual property of the Company (including, without limitation, the timing and probability of successful development, testing and marketing of such products and product candidates, approval thereof by appropriate governmental authorities, the validity and life of relevant patents and the potential impact of generic competition).

Jefferies' opinion was based on economic, monetary, regulatory, market and other conditions existing and which could be evaluated as of the date of Jefferies' opinion. Jefferies did not assess or consider, for purposes of its analyses and opinion, foreign currency exchange risks associated with the Offer. Jefferies expressly disclaimed any undertaking or obligation to advise any person of any change in any fact or matter affecting its opinion of which Jefferies becomes aware after the date of the opinion.

Jefferies made no independent investigation of any legal, accounting or tax matters affecting the Company and assumed the correctness in all respects meaningful to its analysis of all legal, accounting and tax advice given to the Company or the Company Board, including, without limitation, advice as to the legal, accounting and tax consequences to the Company and its shareholders of the terms of, and transactions contemplated by, the Tender Offer

Agreement. In addition, in preparing its opinion, Jefferies did not take into account any tax consequences of the Offer to any holder of Company Shares. Jefferies assumed, at the direction of the Company Board, that the final form of the Tender Offer Agreement, when signed by the parties thereto, would not differ from the draft Tender Offer Agreement it reviewed in any respect meaningful to its analyses or opinion. Jefferies also assumed that, in the course of obtaining the necessary regulatory or third party approvals, consents and releases for the Offer, no delay, limitation, restriction or condition would be imposed that would have an adverse effect on the Company or the Offer.

Jefferies was not asked to address, and its opinion did not address, the fairness to, or any other consideration of, the holders of any class of securities, creditors or other constituencies of the Company, other than holders of Company Shares (excluding Parent, Purchaser and their respective affiliates) as expressly set forth in its opinion. Jefferies' opinion did not take into account different attributes of Company ADSs and Company Ordinary Shares which may distinguish such securities and Jefferies assumed, at the direction of the Company Board, that each Company ADS is convertible into, and equivalent for purposes of its analyses and opinion to, one Company Ordinary Share. Jefferies expressed no opinion as to the prices at which Company Shares would trade at any time. Furthermore, Jefferies did not express any view or opinion as to the fairness, financial or otherwise, of the amount or nature of any compensation payable to or to be received by any of the Company's officers, directors or employees, or any class of such persons, in connection with the Offer relative to the per share cash consideration payable in the Offer or otherwise. The issuance of Jefferies' opinion was authorized by Jefferies' fairness committee.

In connection with rendering its opinion to the Company Board, Jefferies performed a variety of financial and comparative analyses, which are summarized below. The following summary is not a complete description of all analyses performed and factors considered by Jefferies in connection with its opinion. The preparation of a financial opinion is a complex process involving subjective judgments and is not necessarily susceptible to partial analysis or summary description. With respect to the selected public companies and selected precedent transactions analyses summarized below, no company or transaction used as a comparison was identical or directly comparable to the Company or the Offer. These analyses necessarily involved complex considerations and judgments concerning financial and operating characteristics and other factors that could affect the public trading, acquisition or other values of the companies or transactions concerned.

Jefferies believes that its analyses and the summary below must be considered as a whole and that selecting portions of its analyses and factors or focusing on information presented in tabular format, without considering all analyses and factors or the narrative description of the analyses, could create a misleading or incomplete view of the processes underlying Jefferies' analyses and opinion. Jefferies did not draw, in isolation, conclusions from or with regard to any one factor or method of analysis for purposes of its opinion, but rather arrived at its ultimate opinion based on the results of all analyses undertaken by it and assessed as a whole.

The estimates of the future performance of the Company in or underlying Jefferies' analyses are not necessarily indicative of future results or values, which may be significantly more or less favorable than those estimates. In performing its analyses, Jefferies considered industry performance, general business and economic conditions and other matters, many of which were beyond the control of the Company. Estimates of the financial value of companies do not purport to be appraisals or necessarily reflect the prices at which companies or securities actually may be sold or acquired.

The per share cash consideration payable in the Offer was determined through negotiation between the Company and Parent, and the decision by the Company to enter into the Tender Offer Agreement was solely that of the Company Board. Jefferies' opinion and financial analyses were only one of many factors considered by the Company Board in its evaluation of the Offer and should not be viewed as determinative of the views of the Company Board or

management with respect to the Offer or the consideration payable in the Offer.

The following is a brief summary of the material financial analyses reviewed with the Company Board and performed by Jefferies in connection with its opinion. **The financial analyses summarized below include information presented in tabular format. In order to fully understand Jefferies' financial analyses, the tables must be read together with the text of each summary. The tables alone do not constitute a complete description of the financial analyses. Considering the data below without considering the full narrative description of the financial analyses, including the methodologies and assumptions underlying the analyses, could create a misleading or incomplete view of Jefferies' financial analyses.** For purposes of the analyses described below, implied per share equity value reference ranges derived for the Company were based on fully diluted shares using the treasury stock method.

Selected Public Companies Analysis. Jefferies reviewed publicly available financial and stock market information of the Company and the following nine selected companies which Jefferies in its professional judgment considered generally relevant for comparative purposes as publicly traded companies with operations in the biotech and specialty pharmaceuticals sectors, referred to as the selected companies:

| | |
|---|--------------------------------|
| . | Actelion Ltd. |
| . | Aegerion Pharmaceuticals, Inc. |
| . | Avanir Pharmaceuticals, Inc. |
| . | Hyperion Therapeutics, Inc. |
| . | Jazz Pharmaceuticals plc |
| . | Medivation, Inc. |
| . | NPS Pharmaceuticals, Inc. |
| . | Pacira Pharmaceuticals, Inc. |
| . | Raptor Pharmaceuticals Corp. |

Jefferies reviewed enterprise values, calculated as fully diluted equity value based on closing stock prices on December 17, 2013 plus total debt, preferred stock and non-controlling interests and less cash and cash equivalents, as a multiple of calendar year 2014 and calendar year 2015 estimated revenue. The overall low to high calendar year 2014 and calendar year 2015 estimated revenue multiples observed for the selected companies were 2.8x to 12.5x (with a mean of 8.1x and a median of 9.2x) and 1.9x to 8.1x (with a mean of 5.5x and median of 5.9x), respectively. Jefferies then applied a range of selected calendar year 2014 and calendar year 2015 estimated revenue multiples of 7.0x to 9.0x and 4.5x to 6.5x, respectively, derived from the selected companies to corresponding data of the Company. Financial data of the selected companies were based on publicly available research analysts' estimates, public filings and other publicly available information. Financial data of the Company was based on internal estimates of the Company's management. This analysis indicated the following approximate implied per share equity value reference ranges for the Company, as compared to the \$57.00 per share cash consideration in the Offer:

| Implied Per Share Equity Value | Per Share |
|--|-----------------------------|
| Reference Ranges Based on: | Cash Consideration in Offer |
| CY2014P Revenue CY2015P Revenue | |
| \$31.37 - \$39.90 \$29.71 - \$42.24 | \$ 57.00 |

Selected Precedent Transactions Analysis. Using publicly available information, Jefferies reviewed financial data relating to the following nine selected transactions publicly announced from March 12, 2009 to November 11, 2013 which Jefferies in its professional judgment considered generally relevant for comparative purposes as transactions involving target companies with operations in the pharmaceuticals and specialty pharmaceuticals sectors, referred to as the selected transactions:

| <u>Announcement Date</u> | <u>Acquiror</u> | <u>Target</u> |
|--------------------------|---|---------------------------------|
| November 11, 2013 | · Shire Pharmaceutical Holdings Ireland Limited | · ViroPharma Incorporated |
| August 25, 2013 | · Amgen Inc. | · Onyx Pharmaceuticals, Inc. |
| July 30, 2013 | · Cubist Pharmaceuticals, Inc. | · Optimer Pharmaceuticals, Inc. |
| July 16, 2012 | · GlaxoSmithKline plc | · Human Genome Sciences, Inc. |
| June 29, 2012 | · Bristol-Myers Squibb Company | · Amylin Pharmaceuticals, LLC |
| April 26, 2012 | · Jazz Pharmaceuticals plc | · EUSA Pharma Inc. |
| September 7, 2010 | · Bristol-Myers Squibb Company | · ZymoGenetics, Inc. |
| May 16, 2010 | · Astellas Pharma Inc. | · OSI Pharmaceuticals, Inc. |
| March 12, 2009 | · Gilead Sciences, Inc. | · CV Therapeutics, Inc. |

Jefferies reviewed transaction values of the selected transactions, calculated as the purchase prices paid (including contingent value right payments) for the target companies involved in such transactions plus debt, preferred stock, non-controlling interests and less cash and cash equivalents, as a multiple of such target companies' latest 12 months and one-year forward revenue multiples. The overall low to high latest 12 months and one-year forward revenue multiples observed for the selected transactions were 4.5x to 22.1x (with a mean of 10.4x and a median of 9.0x) and 4.1x to 16.6x (with a mean of 8.6x and a median of 7.1x), respectively. Jefferies then applied a range of selected latest 12 months and one-year forward revenue multiples of 10.0x to 12.0x and 8.0x to 10.0x, respectively, derived from such transactions to the Company's calendar year 2013 and calendar year 2014 estimated revenue, respectively. Financial data of the selected transactions were based on research analysts' research, public filings and other publicly available information. Financial data of the Company was based on internal estimates of the Company's management. This analysis indicated the following approximate implied per share equity value reference range for the Company, as compared to the \$57.00 per share cash consideration in the Offer:

| Implied Per Share Equity Value | | Per Share |
|--------------------------------|-------------------|-----------------------------|
| Reference Ranges Based on: | | Cash Consideration in Offer |
| CY2013E Revenue | CY2014P Revenue | |
| \$32.44 - \$38.62 | \$35.63 - \$44.16 | \$ 57.00 |

Discounted Cash Flow Analysis. Jefferies performed a discounted cash flow analysis of the Company by calculating the estimated present value of the standalone unlevered, after-tax free cash flows that the Company was forecasted to generate during fiscal years ending December 31, 2014 through December 31, 2023 based on internal forecasts and estimates of the Company's management. Jefferies calculated terminal values for the Company by applying to the Company's unlevered, after-tax free cash flows during such forecast period a range of perpetuity growth rates of (1.0)% to 1.0%. The present values (as of December 31, 2013) of the cash flows and terminal values were then calculated using discount rates ranging from 10.7% to 11.7%. This analysis indicated the following approximate implied per share equity value reference range for the Company, as compared to the \$57.00 per share cash consideration in the Offer:

| Implied Per Share | Per Share |
|-------------------|-----------|
|-------------------|-----------|

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| Equity Value Reference Range | Cash Consideration in Offer |
|------------------------------|-----------------------------|
| \$42.15 - \$51.91 | \$57.00 |

Other Information. Jefferies observed certain additional information that was not considered part of Jefferies' financial analysis with respect to its opinion but were noted for informational purposes, including:

premiums paid in 57 selected healthcare transactions announced from January 1, 2009 to December 17, 2013 with transaction values ranging from \$500 million to \$5 billion based on closing stock prices of the target companies involved in such transactions 30 days prior to public announcement of the relevant transaction; applying the 25th percentile to 75th percentile of the 30 day premiums derived from such transactions of approximately 27.5% to 71.2% to the closing price of Company Shares of approximately \$46.66 per share on November 17, 2013 (approximately 30 days prior to the date of the Company Board's approval of the Offer) indicated implied per share equity value reference ranges for the Company of approximately \$59.49 to \$79.86; and

stock price targets for Company Shares as reflected in publicly available Wall Street research analysts' reports, which indicated an overall target stock price range for the Company of \$68.00 to \$78.00 per share (with a mean and median of \$73.00 per share).

Miscellaneous

Jefferies maintains a market in the securities of the Company and Parent and, in the ordinary course of business, Jefferies and its affiliates may trade or hold securities of the Company, Parent and/or their respective affiliates for Jefferies' own account and for the accounts of Jefferies' customers and, accordingly, may at any time hold long or short positions in those securities. Jefferies and its affiliates may in the future seek to provide financial advisory and financing services to Parent, the Company or their respective affiliates, for which services Jefferies and its affiliates would expect to receive compensation.

Jefferies was selected to act as the Company's financial advisor in connection with the Offer because Jefferies is an internationally recognized investment banking firm with substantial experience in merger and acquisition transactions generally and in the healthcare and biotechnology sectors in particular. Jefferies is regularly engaged in the valuation of businesses and their securities in connection with mergers and acquisitions, leveraged buyouts, negotiated underwritings, competitive bids, secondary distributions of listed and unlisted securities and private placements.

For a description of the terms of Jefferies' engagement as the Company's financial advisor, see the discussion under "*Item 5. Persons/Assets Retained, Employed, Compensated or Used*" below.

Item 5. Persons/Assets, Retained, Employed, Compensated or Used.

Under the terms of Jefferies' engagement, the Company has agreed to pay Jefferies for its financial advisory services in connection with the Offer an aggregate fee currently estimated to be approximately \$25.6 million, a portion of which was payable upon delivery of Jefferies' opinion and approximately \$25.1 million of which is payable contingent upon completion of the Offer. In addition, the Company has agreed to reimburse Jefferies for its expenses, including

fees and expenses of counsel, and to indemnify Jefferies and related parties against liabilities, including liabilities under federal securities laws, arising out of or in connection with the services rendered and to be rendered by Jefferies under its engagement.

Neither the Company nor any person acting on its behalf has or currently intends to employ, retain or compensate any person to make solicitations or recommendations to the shareholders of the Company on its behalf with respect to the Offer or related matters.

Item 6. Interest in Securities of the Subject Company.

No transactions in Company Shares have been effected during the 60 days prior to the date of this Statement by the Company, or, to the Company's knowledge, any of the directors, executive officers, subsidiaries or affiliates of the Company, except for the entry into the Support Agreements by F3F S.r.l. and each executive officer and director of the Company other than Mr. Marco Brughera, as described under Item 3 above. During this period, no Company Options were granted to any executive officer, director, affiliate or subsidiary of the Company.

Item 7. Purposes of the Transaction and Plans or Proposals.

Except as set forth in this Statement, the Company is not undertaking or engaged in any negotiations in response to the Offer which relate to:

- a tender offer or other acquisition of the Company's securities by the Company, any subsidiary of the Company or any other person;
- any extraordinary transaction, such as a merger, reorganization or liquidation, involving the Company or any subsidiary of the Company;
- any purchase, sale or transfer of a material amount of assets of the Company or any subsidiary of the Company; or
- any material change in the present dividend rate or policy, or indebtedness or capitalization of the Company.

Except as set forth in this Statement, there are no transactions, resolutions of the Company Board, agreements in principle or signed contracts entered into in response to the Offer that relate to one or more of the events referred to in the preceding paragraph.

Item 8. Additional Information.

Purchaser's intentions following consummation of the Offer.

As set forth in the Schedule TO, Purchaser intends to cause the Company to seek to delist the ADSs from the Nasdaq Global Select Market and to cause the Company to terminate the existing deposit agreement between the Company and The Bank of New York, as ADS depository, dated as of June 15, 2005 (the "**Deposit Agreement**"), under which the ADSs were issued. If the Deposit Agreement is terminated, holders of ADSs will be entitled to receive one share of Common Stock in exchange for each ADS held by such holders upon surrender by them of their ADSs for at least one year following such termination. At any time after the expiration of one year from the date of termination, the ADS depository may sell the shares of Common Stock underlying ADSs that have not been surrendered and hold uninvested the proceeds on behalf of holders.

As set forth in the Schedule TO, Purchaser intends, to the extent possible following the consummation of the Offer, to seek to cause the Company to deregister the Common Stock and the ADSs under the Exchange Act, so that the Company will no longer be obligated to file any reports with the SEC or otherwise be required to comply with the SEC's rules relating to publicly held companies. Purchaser's intentions and the effects of the Offer are more fully described in the Offer to Purchase, which is filed as Exhibit (a)(1)(A) to this Statement, and incorporated herein by reference.

Financial Projections.

The Company does not, as a matter of course, publicly disclose long-term forecasts or projections as to the Company's future financial performance due to, among other reasons, the uncertainty of the underlying assumptions and estimates. However, financial projections prepared by senior management for fiscal years 2014 through 2023 (the "**Projections**") were made available to the Company Board in connection with its evaluation of the Offer. The Projections also were provided to Jefferies for use in connection with its analyses and opinion. The Projections were not provided to Parent in connection with its consideration of a transaction.

The Projections were not prepared with a view to public disclosure and are included in this Statement only because such information was considered by the Company Board in connection with its evaluation of the Offer and provided to the Company's financial advisor. Some of the Projections were not prepared in accordance with U.S. generally accepted accounting principles ("**GAAP**"), as applied by the Company at the time the Projections were made and were not prepared with a view to compliance with published guidelines of the SEC regarding projections or the guidelines established by the American Institute of Certified Public Accountants for preparation and presentation of prospective financial information. The Projections included in this Statement have been prepared by, and are the responsibility of, Company management. The Projections were prepared solely for internal use in support of strategic planning and are subjective in many respects and thus susceptible to interpretations and periodic revisions based on actual experience and business developments.

In compiling the Projections, Company management took into account historical performance, combined with estimates regarding revenues, expenses, operating income, and capital spending. Although the Projections are presented with numerical specificity, they reflect numerous assumptions and estimates as to future events made by Company management that Company management believed were reasonable at the time the Projections were prepared. However, this information is not fact and should not be relied upon as being necessarily indicative of actual future results. In addition, factors such as industry performance, the market for the Company's existing and new products and services, and general business, economic, regulatory, market and financial conditions, all of which are difficult to predict and beyond the control of Company management, may cause the Projections or the underlying assumptions not to be reflective of actual future results. In addition, the Projections do not take into account any circumstances or events occurring after the date that they were prepared and, accordingly, do not give effect to the Offer or any changes to the Company's operations or strategy that may be implemented after the Offer is consummated. Accordingly, there can be no assurance that the projections will be realized, and actual results may be materially greater or less than those contained in the Projections. The inclusion of this information should not be regarded as an indication that the Company, Parent, Purchaser or any of their respective affiliates, employees, advisors or other representatives or any other recipient of this information considered, or now considers, the information to be predictive of actual future results.

| PROJECTIONS (EUR in millions) | Fiscal Year Ending December 31 | | | | | | | | | |
|----------------------------------|--------------------------------|------|------|------|------|------|------|------|------|------|
| | 2014 | 2015 | 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | 2022 | 2023 |

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| | | | | | | | | | | |
|------------------------------|------|------|-------|-------|-------|-------|-------|-------|-------|-------|
| Total Revenues | 54.1 | 79.5 | 107.4 | 134.0 | 160.2 | 180.6 | 197.2 | 206.4 | 213.1 | 219.8 |
| Gross Profit | 47.5 | 72.0 | 98.6 | 124.0 | 149.2 | 168.8 | 185.0 | 193.9 | 200.5 | 207.0 |
| Operating Profit | 8.8 | 19.6 | 39.9 | 60.9 | 82.7 | 99.2 | 112.0 | 117.4 | 120.3 | 123.0 |
| Net Income | 6.6 | 15.6 | 34.9 | 48.6 | 62.7 | 75.5 | 85.4 | 89.7 | 91.9 | 94.0 |
| Unlevered Free Cash Flow (1) | 5.4 | 11.1 | 29.8 | 43.7 | 56.5 | 70.1 | 80.0 | 85.4 | 87.9 | 89.9 |

(1) Unlevered Free Cash Flow refers to operating profit plus depreciation & amortization and stock-based compensation, less tax expense, capital expenditures and changes in working capital.

The Projections, which do not give effect to the Offer or take into account any fees and expenses incurred in connection with the Offer, assume a commercial launch of Defitelio® (defibrotide) in Europe during the first quarter of 2014 and during 2015 in the United States following FDA approval, and they are risk adjusted to reflect multiple future scenarios and potential indications for Defitelio® (defibrotide) such as prevention of VOD and GvHD.

Holders of Company Shares are cautioned not to place undue reliance on the Projections or specific portions of the Projections set forth above. No one has made or makes any representation to any shareholder, nor has the Company made any representation to Parent or Purchaser, regarding the Projections. For the foregoing reasons, as well as the bases and assumptions on which the Projections were compiled, the inclusion of specific portions of the Projections in this Statement should not be regarded as an indication that such Projections will necessarily be predictive of actual future events, and they should not be relied on as such. The Company has prepared from time to time in the past, and may continue to prepare in the future, internal financial forecasts that reflect various estimates and assumptions that change from time to time. Accordingly, the Projections provided in conjunction with the Offer may differ from these projections.

Except as may be required by applicable securities laws, the Company does not intend to update or otherwise revise the Projections to reflect circumstances existing after the date when made or to reflect the occurrence of future events, even in the event that any or all of the assumptions underlying the Projections are shown to be in error or no longer appropriate.

Annual and Quarterly Reports.

For additional information regarding the business and the financial results of the Company, please see the Company's Annual Report on Form 20-F for the year ended December 31, 2012, as filed with the SEC on April 1, 2013.

Forward-Looking Statements.

Certain statements either contained in or incorporated by reference into this Schedule 14D-9, other than purely historical information, including estimates, projections and statements relating to the Company's business plans, objectives and expected operating results, and the assumptions upon which those statements are based, are "forward-looking statements." These forward-looking statements generally include statements that are predictive in nature and depend upon or refer to future events or conditions, and include words such as "believes," "plans," "anticipates," "projects," "estimates," "expects," "intends," "strategy," "future," "opportunity," "may," "will," "should," "could," "potential," or similar expressions. Such forward-looking statements include the ability of the Company, Parent and Purchaser to complete the transactions contemplated by the Tender Offer Agreement, including the parties' ability to satisfy the conditions to the consummation of the Offer and the other conditions set forth in the Tender Offer Agreement and the possibility of any termination of the Tender Offer Agreement, and statements related to the potential future financial performance of the Company. The forward-looking statements contained in this document are based on current expectations and assumptions that are subject to risks and uncertainties which may cause actual results to differ materially from the forward-looking statements. Actual results may differ materially from current expectations because of risks associated with uncertainties as to the timing of the Offer; uncertainties as to how many of the Company Shares will be tendered pursuant to the Offer; the risk that competing offers or acquisition proposals will be made; the possibility that various conditions to the consummation of the Offer may not be satisfied or waived,

including that a governmental entity may prohibit, delay or refuse to grant approval for the consummation of the Offer; the effects of disruption from the Offer on the Company's business and the fact that the announcement and pendency of the Offer may make it more difficult to establish or maintain relationships with employees, suppliers and other business partners; the risk that shareholder litigation in connection with the Offer may result in significant costs of defense, indemnification and liability; other uncertainties pertaining to the business of the Company, including the Company's unproven product sales and marketing; manufacturing and commercialization capabilities; strategic partnering activities; product safety; clinical trials and results; legislative and regulatory activity and oversight; intellectual property claims and litigation; the continuing global economic uncertainty and other risks detailed in the Company's public filings with the SEC from time to time, including the Company's most recent Annual Report on Form 20-F for the year ended December 31, 2012. The reader is cautioned not to unduly rely on these forward-looking statements. The Company expressly disclaims any intent or obligation to update or revise publicly these forward-looking statements except as required by law.

Item 9. Exhibits.

The following exhibits are filed with this Statement:

| Exhibit No. | Description |
|--------------------|--|
| (a)(1)(A) | Offer to Purchase, dated December 23, 2013 (incorporated by reference to Exhibit (a)(1)(A) to the Schedule TO). |
| (a)(1)(B) | Form of ADS Letter of Transmittal (incorporated by reference to Exhibit (a)(1)(B) to the Schedule TO). |
| (a)(1)(C) | Form of Notice of Guaranteed Delivery (incorporated by reference to Exhibit (a)(1)(C) to the Schedule TO). |
| (a)(1)(D) | Form of Letter to Brokers, Dealers, Commercial Banks, Trust Companies and Other Nominees (incorporated by reference to Exhibit (a)(1)(D) to the Schedule TO). |
| (a)(1)(E) | Form of Letter to Clients for use by Brokers, Dealers, Commercial Banks, Trust Companies and Other Nominees (incorporated by reference to Exhibit (a)(1)(E) to the Schedule TO). |
| (a)(1)(F) | Form of Share Form of Acceptance (including Guidelines for Certification of Taxpayer Identification Number on Substitute Form W-9) (incorporated by reference to Exhibits (a)(1)(F) and (a)(1)(G) to the Schedule TO). |
| (a)(5)(A)* | Opinion of Jefferies LLC, dated December 18, 2013 (included as Annex A to this Schedule 14D-9). |
| (a)(5)(B) | Summary Newspaper Advertisement published in <i>The Wall Street Journal</i> on December 23, 2013 (incorporated by reference to Exhibit (a)(5)(A) to the Schedule TO). |
| (a)(5)(C) | Joint Press Release issued by the Company and Parent, dated December 19, 2013 (incorporated by reference to Exhibit A to the Company's Schedule 14D-9/C, filed with the SEC on December 19, 2013). |
| (e)(1) | Tender Offer Agreement, dated December 19, 2013, by and between Parent, Purchaser, and the Company (incorporated by reference to Exhibit 2.1 to the Form 8-K/A filed by Parent with the SEC on December 20, 2013). |

- (e)(2) Form of Support Agreement (incorporated by reference to Exhibit 99.3 to the Form 8-K/A filed by Parent with the SEC on December 20, 2013).
- (e)(3) Confidentiality and Nondisclosure Agreement, effective as of September 30, 2013, by and between the Company and Parent (incorporated by reference to Exhibit (d)(6) to the Schedule TO).
- (e)(4)* Employment agreement by and between the Company and Dr. Khalid Islam, dated August 31, 2012, as amended on July 31, 2011 and November 11, 2013, and assigned to Gentium GmbH as of November 1, 2012.
- (e)(5)* Employment agreement by and between the Company and Salvatore Calabrese, dated September 7, 2007.
- (e)(6)* Employment agreement by and between Gentium GmbH and Giorgio Mosconi, dated May 22, 2013, as amended on October 28, 2013 and seconded to Gentium S.p.A. as of December 2, 2013.
- (e)(7)* Employment agreement by and between the Company and Adrian Haigh, dated March 1, 2011, as amended on July 20, 2011 and November 11, 2013, and assigned to Gentium GmbH as of October 1, 2011.
- (e)(8)* Transition, Amendment and Release Agreement, dated December 19, 2013, by and between Gentium GmbH and Dr. Khalid Islam.
- (e)(9)* Consultancy Agreement, dated December 19, 2013, by and between the Company and Dr. Khalid Islam.
- (e)(10)* Retention and Amendment Agreement (together with the Confidentiality, and IP Rights Agreement attached as Exhibit 1), dated December 19, 2013, by and between the Company and Salvatore Calabrese.
- (e)(11)* Retention and Amendment Agreement, dated December 19, 2013, by and between Gentium GmbH and Giorgio Mosconi.
- (e)(12)* Retention and Amendment Agreement, dated December 19, 2013, by and between Gentium GmbH and Adrian Haigh.
- (e)(13) Form of Indemnification Agreement between Gentium S.p.A. and each officer and director (incorporated by reference to Exhibit 10.34 to Amendment No. 2 to the Registration Statement on Form F-1, Registration No. 333-122233, previously filed with the SEC on May 10, 2005).
- (e)(14)* Lease Agreement by and between the Company and F3F S.r.l. (previously FinSirton S.p.A.) dated January 18, 2012.

(e)(15) License and Supply Agreement by and between the Company and Sigma-Tau Pharmaceuticals, Inc. (assignee of Sigma-Tau Industrie Farmaceutiche Riunite S.p.A.) dated December 7, 2001 (incorporated by reference to Exhibit 10.15 to the Registration Statement on Form F-1, Registration No. 333-122233, previously filed with the SEC on January 24, 2005).

(e)(16) Letter Agreement, dated October 12, 2007, by and between the Company and Sigma-Tau Pharmaceuticals, Inc. (incorporated by reference to Exhibit 99.4 to the report on Form 6-K, previously filed with the SEC on December 12, 2007).

(e)(17) Amendments to License and Supply Agreement and Letter Agreement, dated December 7, 2001 and October 12, 2007, respectively, effective January 7, 2010, by and between the Company and Sigma-Tau Pharmaceuticals, Inc. (incorporated by reference to Exhibit 2 to the Form 6-K, previously filed with the SEC on January 11, 2010).

(e)(18) Amended and Restated 2004 Equity Incentive Plan (incorporated by reference to Exhibit 10.1 to the Registration Statement on Form S-8, Registration No. 333-137534, previously filed with the SEC on September 22, 2006).

(e)(19) Amendment No. 1 to Amended and Restated 2004 Equity Incentive Plan, made as of March 26, 2007 (incorporated by reference to Exhibit 4.1.2 to the Annual Report on Form 20-F for the year ended December 31, 2007, previously filed with the SEC on April 30, 2007).

(e)(20) Amended and Restated Nonstatutory Share Option Plan and Agreement dated March 23, 2006 (incorporated by reference to Exhibit 4.2 to the Annual Report on Form 20-F for the year ended December 31, 2005, previously filed with the SEC on May 30, 2006).

(e)(21) Amendment No. 1 to Amended and Restated Nonstatutory Share Option Plan and Agreement, made as of March 26, 2007 (incorporated by reference to Exhibit 4.2.2 to the Annual Report on Form 20-F for the year ended December 31, 2007, previously filed with the SEC on April 30, 2007).

(e)(22) Amended 2007 Stock Option Plan dated May 9, 2011 (incorporated by reference to Exhibit 4.3 to the Annual Report on Form 20-F for the year ended December 31, 2011, previously filed with the SEC on March 30, 2012).

*

Filed herewith.

SIGNATURE

After due inquiry and to the best of its knowledge and belief, the undersigned certifies that the information set forth in this statement is true, complete and correct.

GENTIUM S.P.A.

By: /s/ Salvatore Calabrese

Name: Salvatore Calabrese

Title: Chief Financial Officer and Chief Operating Officer

Dated: December 24, 2013

ANNEX A

[Letterhead of Jefferies LLC]

December 18, 2013

The Board of Directors

Gentium S.p.A.
Piazza XX Settembre 2
22079 Villa Guardia (Como)

Italy

The Board of Directors:

We understand that Gentium S.p.A., an Italian corporation ("Gentium"), Jazz Pharmaceuticals Public Limited Company, an Irish public limited company ("Jazz"), and Jazz Pharmaceuticals Italy S.r.l., an Italian limited liability company and wholly-owned subsidiary of Jazz ("Purchaser"), propose to enter into a Tender Offer Agreement (the "Agreement") pursuant to which Purchaser will commence a tender offer (the "Tender Offer") to purchase all outstanding ordinary shares, no par value per share, of Gentium ("Gentium Ordinary Shares") and American Depositary Shares, each representing one Gentium Ordinary Share ("Gentium ADSs" and, together with Gentium Ordinary Shares, "Gentium Shares"), at a purchase price of \$57.00 per Gentium Share in cash (the "Consideration"). The terms and conditions of the Tender Offer are more fully set forth in the Agreement.

You have asked for our opinion as to whether the Consideration to be received in the Tender Offer by holders of Gentium Shares (other than Jazz, Purchaser and their respective affiliates) pursuant to the Agreement is fair, from a financial point of view, to such holders.

In arriving at our opinion, we have, among other things:

- (i) reviewed a draft, dated December 17, 2013, of the Agreement;

- (ii) reviewed certain publicly available financial and other information about Gentium;
- (iii) reviewed certain information furnished to us by the management of Gentium relating to the business, operations and prospects of Gentium, including financial forecasts and estimates prepared by the management of Gentium;
- (iv) held discussions with members of senior management of Gentium concerning the matters described in clauses (ii) and (iii) above;
- (v) held discussions, at the direction of the Board of Directors of Gentium (the "Board"), with third parties to solicit indications of interest in the possible acquisition of Gentium;
- (vi) reviewed the trading price history and implied multiples for Gentium Shares and compared them with those of certain publicly traded companies that we deemed relevant;
- (vii) compared the financial terms of the Tender Offer with publicly available financial terms of certain other transactions that we deemed relevant; and
- (viii) conducted such other financial studies, analyses and investigations as we deemed appropriate.

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The Board of Directors

Gentium S.p.A.

December 18, 2013

In our review and analysis and in rendering this opinion, we have assumed and relied upon, but have not assumed any responsibility to independently investigate or verify, the accuracy and completeness of all financial and other information that was supplied or otherwise made available by Gentium or that was publicly available to us (including, without limitation, the information described above), or that was otherwise reviewed by us. We have relied on assurances of the management of Gentium that it is not aware of any facts or circumstances that would make such information inaccurate or misleading. In our review, we did not obtain any independent evaluation or appraisal of any of the assets or liabilities (contingent or otherwise), nor did we conduct a physical inspection of any of the properties or facilities, of Gentium and we have not been furnished with, and assume no responsibility to obtain, any such evaluations, appraisals or physical inspections.

With respect to the financial forecasts and estimates provided to and reviewed by us, we note that projecting future results of any company is inherently subject to uncertainty. We have been informed, however, and we have assumed, that such financial forecasts and estimates were reasonably prepared on bases reflecting the best currently available estimates and good faith judgments of the management of Gentium as to the future financial performance of Gentium. We also have been informed that the Euros to U.S. Dollars exchange rate reflected in such forecasts and estimates is reasonable to utilize for purposes of our analyses and opinion. We express no opinion as to any such financial forecasts or estimates or the assumptions on which they are based (including, without limitation, any currency or exchange rate fluctuations in Euros and U.S. Dollars). We have relied upon the assessments of the management of Gentium as to, among other things, (i) the potential impact on Gentium of governmental and regulatory policies affecting the pharmaceutical industry and (ii) the validity and risks associated with the products, product candidates and intellectual property of Gentium (including, without limitation, the timing and probability of successful development, testing and marketing of such products and product candidates, approval thereof by appropriate governmental authorities, the validity and life of relevant patents and the potential impact of generic competition).

Our opinion is based on economic, monetary, regulatory, market and other conditions existing and which can be evaluated as of the date hereof. We have not assessed or considered, for purposes of our analyses and opinion, foreign currency exchange risks associated with the Tender Offer. We expressly disclaim any undertaking or obligation to advise any person of any change in any fact or matter affecting our opinion of which we become aware after the date hereof.

We have made no independent investigation of any legal, accounting or tax matters affecting Gentium, and we have assumed the correctness in all respects meaningful to our analysis of all legal, accounting and tax advice given to Gentium or the Board, including, without limitation, advice as to the legal, accounting and tax consequences to Gentium and its shareholders of the terms of, and transactions contemplated by, the Agreement. In addition, in

preparing this opinion, we have not taken into account any tax consequences of the Tender Offer to any holder of Gentium Shares. We have assumed, at the direction of the Board, that the final form of the Agreement, when signed by the parties thereto, will not differ from the draft Agreement reviewed by us in any respect meaningful to our analyses or opinion. We also have assumed that, in the course of obtaining the necessary regulatory or third party approvals, consents and releases for the Tender Offer, no delay, limitation, restriction or condition will be imposed that would have an adverse effect on Gentium or the Tender Offer.

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The Board of Directors

Gentium S.p.A.

December 18, 2013

It is understood that our opinion is for the use and benefit of the Board (in its capacity as such) in its evaluation of the Tender Offer, and our opinion does not address the relative merits of the Tender Offer or other transactions contemplated by the Agreement as compared to any alternative transaction or opportunity that might be available to Gentium, nor does it address the underlying business decision by Gentium to engage in the Tender Offer or the terms of the Agreement or the documents referred to therein, any term, aspect or implication of any support agreement or other agreements or arrangements contemplated by or resulting from the Tender Offer or otherwise. Our opinion does not constitute a recommendation as to whether any shareholder should tender Gentium Shares in the Tender Offer or how any shareholder should act with respect to the Tender Offer or any related matter. In addition, you have not asked us to address, and this opinion does not address, the fairness to, or any other consideration of, the holders of any class of securities, creditors or other constituencies of Gentium, other than holders of Gentium Shares (excluding Jazz, Purchaser and their respective affiliates) as expressly set forth herein. Our opinion does not take into account different attributes of Gentium ADSs and Gentium Ordinary Shares which may distinguish such securities and we have assumed, at the direction of the Board, that each Gentium ADS is convertible into, and equivalent for purposes of our analyses and opinion to, one Gentium Ordinary Share. We express no opinion as to the prices at which Gentium Shares will trade at any time. Furthermore, we do not express any view or opinion as to the fairness, financial or otherwise, of the amount or nature of any compensation payable to or to be received by any of Gentium's officers, directors or employees, or any class of such persons, in connection with the Tender Offer relative to the Consideration or otherwise. The issuance of our opinion has been authorized by the Fairness Committee of Jefferies LLC.

We have been engaged by Gentium to act as its financial advisor in connection with the Tender Offer and will receive a fee for our services, a portion of which is payable upon delivery of this opinion and the principal portion of which is payable contingent upon consummation of the Tender Offer. We also will be reimbursed for expenses incurred in connection with our engagement. In addition, Gentium has agreed to indemnify us against liabilities arising out of or in connection with the services rendered and to be rendered by us under such engagement. We maintain a market in the securities of Gentium and Jazz and, in the ordinary course of business, we and our affiliates may trade or hold securities of Gentium, Jazz and/or their respective affiliates for our own account and for the accounts of our customers and, accordingly, may at any time hold long or short positions in those securities. In addition, we and our affiliates may in the future seek to provide financial advisory and financing services to Gentium, Jazz and their respective affiliates, for which we and our affiliates would expect to receive compensation.

Based upon and subject to the foregoing, we are of the opinion that, as of the date hereof, the Consideration to be received in the Tender Offer by holders of Gentium Shares (other than Jazz, Purchaser and their respective affiliates) pursuant to the Agreement is fair, from a financial point of view, to such holders.

Very truly yours,

/s/ Jefferies LLC

Jefferies LLC

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