Gentium S.p.A.
Form 20-F
April 01, 2014

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

_____

FORM 20-F

[ ]	REGISTRATION STATEMENT PURSUANT TO SECTION 12(b) OR (g) OF THE SECURITIES EXCHANGE ACT OF 1934
OR	
[x]	ANNUAL REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934 For the Fiscal Year Ended: December 31, 2013
OR	
[ ]	TRANSITION REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934
OR	
[ ]	SHELL COMPANY REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934

001-32529
(Commission file number)

GENTIUM S.p.A.
(Exact Name of Registrant as Specified in its Charter)
NOT APPLICABLE
(Translation of Registrant’s Name into English)

_____

Italy
(Jurisdiction of incorporation or organization)

Piazza XX Settembre 2
22079 Villa Guardia (Como), Italy
+39 031 5373200
(Address, including zip code, and telephone number,
including area code, of Registrant’s principal executive offices)

Securities registered or to be registered pursuant to Section 12(b) of the Act: None

Securities registered or to be registered pursuant to Section 12(g) of the Act: American depositary shares, as evidenced by American depositary receipts, each representing one ordinary share

Securities for which there is a reporting obligation pursuant to Section 15(d) of the Act: None

Indicate the number of outstanding shares of each of the issuer’s classes of capital or common stock as of the close of the period covered by the annual report.

15,555,131 ordinary shares, as of December 31, 2013

Indicate by check mark if the registrant is a well-known seasoned issuer, as defined in Rule 405 of the Securities Act.

Yes [ ] No [x]

If this report is an annual or transition report, indicate by check mark if the registrant is not required to file reports pursuant to Section 13 or 15(d) of the Securities Exchange Act of 1934.

Yes [ ] No [x]

Note – Checking the box above will not relieve any registrant required to file reports pursuant to Section 13 or 15(d) of the Securities Exchange Act of 1934 from their obligations under those Sections.

Indicate by check mark whether the registrant (1) has filed all reports required to be filed by Section 13 or 15(d) of the Securities Exchange Act of 1934 during the preceding 12 months (or for such shorter period that the registrant was required to file such reports), and (2) has been subject to such filing requirements for the past 90 days.

Yes [x] No [ ]

Indicate by check mark whether the registrant has submitted electronically and posted on its corporate Web site, if any, every Interactive Data File required to be submitted and posted pursuant to Rule 405 of Regulation S-T (§232.405 of this chapter) during the preceding 12 months (or for such shorter period that the registrant was required to submit and post such files).

Yes [x] No [ ]

Indicate by check mark whether the registrant is a large accelerated filer, an accelerated filer, or a non-accelerated filer. See definition of “accelerated filer and large accelerated filer” in rule 12b-2 of the Exchange Act. (Check one):

Large accelerated filer [ ] Accelerated filer [x] Non-accelerated filer [ ]

Indicate by check mark which basis of accounting the registrant has used to prepare the consolidated financial statements included in this filing:

U.S. GAAP [x] International Financial Reporting Standards as issued by the International Accounting Standards Board [ ] Other [ ]

If “Other" has been checked in response to the previous question, indicated by check mark which financial item the registrant has elected to follow.

Item 17 [ ] Item 18 [ ]

If this is an annual report, indicate by check mark whether the registrant is a shell company (as defined in Rule 12b-2 of the Exchange Act).

Yes [ ] No [x]

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We own or have rights to various copyrights, trademarks, and trade names used in our business in Italy, the United States and/or other countries, including Gentium® and Defitelio®. This annual report also includes trademarks, service marks, and trade names of other companies and organizations.

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CAUTIONARY NOTE REGARDING FORWARD-LOOKING STATEMENTS

This annual report contains forward-looking statements within the meaning of Section 27A of the Securities Act of 1933, as amended, and Section 21E of the Securities Exchange Act of 1934, as amended, which are subject to the "safe harbor" created by those sections. Forward-looking statements are based on our management's beliefs and assumptions and on information currently available to our management. In some cases, you can identify forward-looking statements by terms such as "may," "will," "should," "could," "would," "expect," "plan," "anticipate," "believe," "estimate," "predict," "intend," "continue," "potential," "possible," "foreseeable," "likely" and similar expressions intended to identify forward-looking statements. These statements involve known and unknown risks, uncertainties and other factors which may cause our actual results, performance, time frames or achievements to be materially different from any future results, performance, time frames or achievements expressed or implied by the forward-looking statements. We discuss many of these risks, uncertainties and other factors in this annual report in greater detail under the heading "Risk Factors." Given these risks, uncertainties and other factors, you should not place undue reliance on these forward-looking statements. Also, these forward-looking statements represent our estimates and assumptions only as of the date of this filing. You should read this annual report completely and with the understanding that our actual future results may be materially different from what we expect. We hereby qualify our forward-looking statements by our cautionary statements. Except as required by law, we assume no obligation to update our forward-looking statements publicly, or to update the reasons that actual results could differ materially from those anticipated in these forward-looking statements, even if new information becomes available in the future.

You should rely only on the information contained in this annual report. We have not authorized anyone to provide you with information different from that contained in this annual report.

PART I

ITEM 1. IDENTITY OF DIRECTORS, SENIOR MANAGEMENT AND ADVISERS

Not applicable.

ITEM 2. OFFER STATISTICS AND EXPECTED TIMETABLE

Not applicable.

ITEM 3. KEY INFORMATION

GENTIUM S.P.A.

We are a biopharmaceutical company primarily focused on the development of defibrotide, a sodium salt of a complex mixture of single- and double-stranded oligodeoxyribonucleotides derived from porcine DNA. Our development of defibrotide has been directed to the treatment and prevention of hepatic veno-occlusive disease, or VOD, a potentially life-threatening complication of hematopoietic stem cell transplantation, or HSCT.

On December 19, 2013, we entered into a definitive tender offer agreement with Jazz Pharmaceuticals Public Limited Company, or Jazz, and Jazz Pharmaceuticals Italy S.p.A., a wholly-owned subsidiary of Jazz, or Jazz Italy, pursuant to which Jazz Italy made an offer to purchase all our outstanding ordinary shares and American Depositary Shares, or ADSs, each representing one ordinary share, at a purchase price of $57.00 per ordinary share and per ADS (without duplication for ordinary shares underlying ADSs). On February 21, 2014, Jazz Italy completed its tender offer, having acquired approximately 98 percent of our combined ordinary shares and ADSs. Jazz is now our indirect majority shareholder. We filed a Notice of Voluntary Delisting with The NASDAQ Stock Market on March 5, 2014, and trading in our ADSs on The NASDAQ Global Market, or NASDAQ, was suspended on March 7, 2014. We filed a Form 25 with the Securities and Exchange Commission, or the SEC, on March 17, 2014, to terminate registration of our ordinary shares and ADSs under Section 12(b) of the Securities Exchange Act of 1934, as amended, or the Exchange Act, and intend to file a Form 15 with the SEC promptly after this annual report is filed with the SEC to

terminate registration of our ordinary shares and ADSs under Section 12(g) of the Exchange Act and to suspend our duty to file reports under Sections 13(a) and 15(d) of the Exchange Act. We expect that this will be the final report we will be required to file under Sections 13(a) and 15(d) of the Exchange Act.

We and The Bank of New York Mellon, or the ADS depositary, entered into an amendment dated March 21, 2014 to the deposit agreement dated as of June 15, 2005, or the deposit agreement, which amendment will be effective on April 13, 2014. Upon effectiveness, the amendment to the deposit agreement will reduce from one year to 60 days the period that must elapse between the date of termination of the deposit agreement and the date on which the ADS depositary may sell the ordinary shares of the Company it then holds and provides for the ADS depositary to accept any offer by a subsidiary of Jazz to purchase

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those shares at a price of no less than $57.00 per ordinary share, unless the ADS depositary has received what it considers to be a superior bona fide offer from another party. The ADS depositary has given notice such that the deposit agreement will terminate at 5:00 pm (Eastern time) on April 13, 2014. While the date or dates on which the ADS depositary will sell the remaining shares have not been determined, the sale will not occur before June 13, 2014. Jazz Italy has indicated that it intends to offer to purchase ordinary shares from the ADS depositary at a per share price equal to $57.00.

Defibrotide has been granted orphan drug designation to treat and prevent VOD by the United States Food and Drug Administration, or FDA, by the European Medicines Agency, or EMA, and by the Korean Ministry of Food and Drug Safety. The Commonwealth of Australia-Department of Health has granted defibrotide orphan drug designation for the treatment of VOD. In November 2013, the EMA also granted orphan drug designation to defibrotide for the prevention of acute graft versus host disease, or GvHD, another potentially fatal complication of hematopoietic stem cell transplantation, or HSCT, that afflicts up to 50% of all donor transplant patients.

In October 2013, the European Commission, or EC, granted marketing authorization under exceptional circumstances for our defibrotide product, Defitelio® (defibrotide), for the treatment of severe VOD in adults and children undergoing HSCT therapy. Stem cell transplantation is a frequently used treatment modality for hematologic cancers and other conditions in both adults and children. Certain high-dose conditioning regimens used as part of HSCT can damage the lining cells of hepatic vessels, which is thought to lead to the development of VOD, a blockage of the small vessels in the liver, that leads to liver failure and can result in significant dysfunction in other organs such as the kidneys and lungs. The condition is also referred to as “sinusoidal obstruction syndrome.” Severe VOD is the most extreme form of VOD and is associated with multi-organ failure and high rates of morbidity and mortality. Defitelio is the first approved treatment in the European Union, or EU, for this potentially life-threatening condition. Defitelio has generally been well-tolerated; the most frequent adverse reactions observed during pre-marketing use of the product are hemorrhage, hypotension and coagulopathy.

Our wholly-owned subsidiary, Gentium GmbH, together with other subsidiaries of Jazz, commenced the launch of Defitelio in Europe in March 2014, with an initial launch in Germany and Austria. We expect to launch Defitelio in additional European countries on a rolling basis during 2014 and 2015 and are engaged in pricing and reimbursement submissions as applicable in preparation for planned launches in those countries. We intend eventually to promote Defitelio in all European markets where it has marketing authorization. In addition, we expect to continue to give patients access to defibrotide in countries where it is not commercially available through our expanded access program in the United States, as discussed below, and on a named patient basis throughout the rest of the world, which we refer to as our named patient program.

Under a license and supply agreement, we have licensed to Sigma-Tau Pharmaceuticals, Inc., or Sigma-Tau, the rights to commercialize defibrotide for the treatment and prevention of VOD in North America, Central America and South America, subject to receipt of marketing authorization, if any, in the applicable territory. In connection with the license and supply agreement, we also entered into a cost sharing agreement under which Sigma-Tau has agreed to reimburse us 50% of certain costs associated with the development of defibrotide.

There are currently no approved treatments for VOD in the United States. Defibrotide is being distributed to patients diagnosed with VOD in the United States through an expanded access program pursuant to a treatment investigational new drug, or IND, protocol, which we call our expanded access program. Defibrotide also received Fast Track designation by the FDA to treat severe VOD. The Fast Track program is designed to enable more frequent interactions with the FDA during drug development and to expedite the FDA’s review of a new drug candidate. In 2011, we voluntarily withdrew from consideration a new drug application, or NDA, we submitted to the FDA in July 2011 seeking approval in the United States for defibrotide for the treatment of VOD in order to address issues raised by the FDA. We are currently assessing what we believe would be the optimal path for potential approval of defibrotide in the United States, which may include filing a new application with existing clinical data or generating additional clinical data before a new application is ready for submission and FDA review.

We have manufacturing facilities in Italy where we produce active pharmaceutical ingredients, or APIs, including the defibrotide compound, sodium heparin, urokinase and sulglicotide. These APIs are subsequently used to make the

finished forms of various drugs. With respect to defibrotide, we have contracted with Patheon UK Limited, or Patheon, to process the defibrotide compound into its finished form at its Italian manufacturing plant. Consistent with our long-term manufacturing strategy, and in order to secure an additional source to manufacture commercial supply of Defitelio, we have initiated work with Fresenius Kabi, a global health care company specialized in lifesaving medicines and technologies for infusion, to conduct a technology transfer of our manufacturing process for the finished form of Defitelio to their manufacturing site in Graz, Austria. Subject to a successful technology transfer, including manufacture of process validation batches, and receipt of all necessary regulatory approvals, we intend to qualify Fresenius Kabi as a second source of Defitelio.

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SELECTED CONSOLIDATED FINANCIAL DATA

The following selected consolidated financial data should be read in conjunction with Part 1, Item 5 of this annual report entitled "Operating and Financial Review and Prospects" and our consolidated financial statements and the related notes appearing elsewhere in this annual report. The selected consolidated financial data as of December 31, 2012 and 2013 and for each of the three years in the period ended December 31, 2013 are derived from our audited consolidated financial statements that are included in this annual report. The selected financial data as of December 31, 2009, 2010 and 2011 and for each of the two years in the period ended December 31, 2010 are derived from our audited consolidated financial statements that are not included in this annual report. Our historical results are not necessarily indicative of results to be expected in any future period.

	For the Year Ended December 31,				
	2009	2010	2011	2012	2013
	(in thousands, except share and per share data)				
Consolidated Statements of Operations Data:					
Revenues:					
Product sales to related party	€195	€—	€—	€—	€—
API product sales	4,603	6,533	4,848	4,856	6,172
NPP product sales	4,904	13,182	16,886	22,774	33,653
Total product sales	9,702	19,715	21,734	27,630	39,825
Other revenues	129	289	123	152	490
Other revenues from related party	337	4,547	2,026	1,257	2,602
Total revenues	10,168	24,551	23,883	29,039	42,917
Operating costs and expenses:					
Cost of goods sold	4,002	5,786	6,035	5,778	6,055
Research and development	3,512	6,104	5,533	10,531	15,672
Selling, general and administrative	6,036	5,835	7,727	10,829	12,883
Charges from related parties	279	346	222	186	189
Depreciation and amortization	916	908	870	1,003	1,031
Restructuring charges	—	1,101	—	—	—
Total costs and expenses	14,745	20,080	20,387	28,327	35,830
Operating income/(loss)	(4,577)	4,471	3,496	712	7,087
Foreign currency exchange gain/(loss), net	162	90	46	(67)	55
Interest income/(expense), net	(110)	(87)	(21)	155	237
Income/(loss) before income tax provision/(benefit)	(4,525)	4,474	3,521	800	7,379
Income tax provision/(benefit)	—	397	811	26	(17,222)
Net income/(loss)	€(4,525)	€4,077	€2,710	€774	€24,601
Net income/(loss) per share:					
Basic	€(0.30)	€0.27	€0.18	€0.05	€1.61
Diluted	€(0.30)	€0.27	€0.18	€0.05	€1.48
Weighted-average shares used to compute net income per share:					
Basic	14,956,317	14,956,317	14,964,021	15,014,411	15,261,799
Diluted	14,956,317	14,956,317	15,340,859	15,639,890	16,602,743

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	As of December 31,				
	2009	2010	2011	2012	2013
	(in thousands, except share data)				
Consolidated Balance Sheet Data:					
Cash and cash equivalents	€1,392	€8,742	€9,990	€12,485	€22,038
Working capital	1,041	6,555	10,730	14,220	27,981
Property, manufacturing facility and equipment, net	9,717	8,598	8,508	7,449	7,581
Net assets	7,330	12,930	17,383	20,350	49,216
Total assets	18,167	24,674	27,412	28,638	65,471
Long-term debt, net of current maturities	3,098	1,759	1,545	1,135	1,386
Shareholders' equity	7,330	12,930	17,383	20,350	49,216
Share capital	106,962	108,485	110,228	112,421	116,686
Number of shares	14,956,317	14,956,317	14,969,150	15,038,483	15,555,131

Exchange Rate Information

The following table sets forth information regarding the exchange rates of U.S. dollars per Euro for the years indicated, calculated by using the average of the noon buying rates for the Euro as reported by the Federal Reserve Bank of New York on its website (Statistical Release G.5), or the noon buying rates, on the last day of each month during the years presented.

	U.S. Dollar per Euro
Year	Average
2009	1.3935
2010	1.3261
2011	1.3931
2012	1.2859
2013	1.3281
2014 (through March 21, 2014)	1.3696

The following table sets forth information regarding the high and low exchange rates of U.S. dollars per Euro for the months indicated as reported by the Federal Reserve Bank of New York on its website (Statistical Release H.10) based on the noon buying rate on each day of such month. On March 21, 2014, the noon buying rate was U.S.$1.3783 per Euro.

	U.S. Dollar per Euro	
Month	High	Low
October 2013	1.3810	1.3490
November 2013	1.3606	1.3357
December 2013	1.3816	1.3552
January 2014	1.3682	1.3500
February 2014	1.3806	1.3507
March 2014 (through March 21, 2014)	1.3927	1.3731

We use the Euro as our functional currency for financial reporting. This annual report contains translations of Euros into U.S. dollars at specified rates solely for the convenience of the reader. No representation is made that the Euro amounts referred to in this annual report could have been or could be converted into U.S. dollars at any particular rate or at all.

CAPITALIZATION AND INDEBTEDNESS

Not applicable.

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REASONS FOR THE OFFER AND USE OF PROCEEDS

Not applicable.

RISK FACTORS

We have identified the following risks and uncertainties that may have a material adverse effect on our business, financial condition or results of operations. The risks described below are not the only ones we face. Additional risks not presently known to us or that we currently believe are immaterial may also significantly impair our business operations. Our business could be harmed by any of these risks. You should carefully consider the risks described below, in conjunction with the other information and consolidated financial statements and related notes included elsewhere in this annual report when considering an investment in us. You should pay particular attention to the fact that we conduct our operations in Italy and are governed by a legal and regulatory environment that in some respects differs significantly from the environment that prevails in other countries with which you may be familiar. The value of our ADSs or ordinary shares could decline due to any of these risks, and you may lose all or part of your investment.

We may not be able to successfully launch and market Defitelio in the EU, or obtain regulatory approval in other countries, including the United States, which could have a material adverse effect on our business, financial condition, results of operations and growth prospects.

In October 2013, the EC granted marketing authorization under exceptional circumstances for Defitelio for the treatment of severe VOD in adults and children undergoing HSCT therapy. Our wholly-owned subsidiary, Gentium GmbH, together with other subsidiaries of Jazz, commenced the launch of Defitelio in Europe in March 2014, with an initial launch in Germany and Austria. We expect to launch in additional European countries on a rolling basis during 2014 and 2015, subject to receipt of pricing and reimbursement approvals as applicable. Any delay in the planned timing of the launch of Defitelio would negatively affect anticipated revenue from Defitelio in 2014 and could negatively affect our growth prospects.

We are making pricing and reimbursement submissions with respect to Defitelio in those European countries where pricing and reimbursement approvals are required for launch. We cannot predict the timing of Defitelio's launch in countries in which we are awaiting pricing and reimbursement guidelines. If we experience delays and unforeseen difficulties in obtaining pricing and reimbursement approvals, the planned launch would be delayed in the applicable countries and our anticipated revenue from Defitelio in 2014 and our growth prospects could be negatively affected.

We have developed estimates of anticipated pricing, which are based on our research and understanding of the product and target market. However, due to efforts to provide for containment of health care costs, one or more countries may not support our estimated level of governmental pricing and reimbursement for Defitelio, particularly in light of the budget crises faced by a number of countries in Europe, which would negatively impact anticipated revenue from Defitelio. In addition, until 2008, we sold forms of defibrotide in Italy to treat vascular disease with risk of thrombosis at a price that was substantially lower than the anticipated commercial price for Defitelio. The regulators in Italy may use the price of the past sales by us as a reference price for Defitelio, which may make it more difficult for us to justify our requested higher commercial price, which would also negatively impact anticipated revenue from Defitelio in Italy.

Furthermore, after initial price and reimbursement approvals, reductions in prices and changes in reimbursement levels can be triggered by multiple factors, including reference pricing systems and publication of discounts by third party payors or authorities in other countries. In the EU, prices can be reduced further by parallel distribution and parallel trade, or arbitrage between low-priced and high-priced countries. If any of these events occurs, our anticipated revenue from Defitelio would be negatively affected.

We also cannot predict the level of sales of Defitelio in Europe after its planned launch. If sales of Defitelio do not reach the levels we expect, our anticipated revenue from Defitelio would be negatively affected which could have a material adverse effect on our business, financial condition, results of operations and growth prospects.

Defitelio was authorized under "exceptional circumstances" because it was not possible to obtain complete information about the product due to the rarity of the disease and because ethical considerations prevented conducting a study

directly comparing Defitelio with a placebo. A marketing authorization granted under exceptional circumstances is subject to approval conditions and an annual reassessment of the risk-benefit balance by the EMA. As such, if we fail to meet the approval condition for Defitelio, which requires that we set up a patient registry to investigate the long term safety, health outcomes and patterns of utilization of defibrotide during normal use, or if it is determined that the balance of risk and benefit changes materially, the EMA could suspend, impose variation or withdraw the marketing authorization for Defitelio. This could negatively impact our anticipated revenue from Defitelio and could have a material adverse effect on our business, financial condition, results of operations and growth prospects.

Although Defitelio has been approved in Europe under exceptional circumstances, a prior NDA submission seeking

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approval in the United States for defibrotide for the treatment of VOD was voluntarily withdrawn from consideration in 2011 in order to address issues raised by the FDA. We are currently assessing what we believe would be the optimal path for potential approval of defibrotide in the United States, which may include filing a new application with existing clinical data or generating additional clinical data before a new application is ready for submission and FDA review. We are also assessing the potential for approval of defibrotide in other countries and for additional development of defibrotide in other indications. We cannot know when, if ever, defibrotide will be approved in the United States or in any other country or under what circumstances, and what, if any, additional clinical or other development activities will be required in order to potentially obtain such regulatory approval and the cost associated with such required activities if any. If we fail to obtain approval for defibrotide in other countries or for new indications, our anticipated revenue from defibrotide and our growth prospects would be negatively affected.

The Marketing Authorization Application, or MAA, we initially filed with the EMA in 2011 sought approval for defibrotide for the treatment and prevention of VOD in adults and children. The approval we received from the EC in October 2013 was for a narrower indication – the treatment of severe VOD in adults and children undergoing HSCT therapy. The scope of any future approvals we receive may negatively affect our growth prospects.

While we have limited revenue from sales of defibrotide on a named patient basis, we cannot predict whether historical revenues from named patient programs will continue or whether we will be able to continue to distribute defibrotide on a named patient basis.

Defibrotide is currently available in approximately 40 countries on a named patient basis and is being distributed to patients diagnosed with VOD in the United States through an expanded access program pursuant to a treatment IND protocol. In certain European countries, reimbursement for products that have not yet received marketing authorization is provided through national named patient or compassionate use programs. Such reimbursement may cease to be available if authorization for named patient or compassionate use programs expires or is terminated. While we have generated and we continue to generate revenue on the distribution of defibrotide through named patient programs, we cannot predict whether historical revenues from these programs will continue, whether we will be able to continue to distribute defibrotide on a named patient basis in these countries, or whether the launch of Defitelio in Europe will proceed as planned and commercial revenues will replace revenues historically generated from sales on a named patient basis. Any failure to maintain revenues from sales of defibrotide on a named patient basis and/or to generate higher revenues following the launch of Defitelio would have a material adverse effect on our business, financial condition, results of operations and growth prospects.

We currently rely upon a sole processor, Patheon, to process defibrotide into its finished form, and we may not be able to quickly replace Patheon if it is unable to perform this service.

With respect to defibrotide, we have contracted with Patheon to process the defibrotide compound into its finished form at Patheon’s manufacturing facility in Italy. Patheon is currently the sole processor of finished Defitelio. If Patheon does not or is not able to perform these services for any reason, it may take time and resources to implement and execute the necessary technology transfer to another processor, and such a delay could potentially cause us to breach contractual obligations with distributors or other customers which we have entered into or may in the future enter into, violate local laws requiring us to deliver the product to those in need, and negatively impact our product launch and anticipated revenues.

Consistent with our long-term manufacturing strategy, and in order to secure an additional source to manufacture commercial supply of Defitelio, we have initiated work with Fresenius Kabi, a global health care company specialized in lifesaving medicines and technologies for infusion, to conduct a technology transfer of our manufacturing process for the finished form of Defitelio to their manufacturing site in Graz, Austria. Subject to a successful technology transfer, including manufacture of process validation batches, and receipt of all necessary regulatory approvals, we intend to qualify Fresenius Kabi as a second source of Defitelio. The process of technology transfer is complicated, and Fresenius Kabi may not be able to successfully obtain regulatory approval to produce Defitelio commercially, which could negatively impact our anticipated revenues if Patheon for any reason does not or cannot provide us with sufficient quantities of finished product to meet our clinical and commercial needs.

Our manufacturing activities and those of our defibrotide processor and any future defibrotide processor are subject to significant ongoing regulatory obligations and oversight, which may result in significant additional expense and limit our ability to commercialize Defitelio and to provide the other APIs we manufacture.

We manufacture certain APIs, including the defibrotide drug substance, at our manufacturing facilities. In addition, we have engaged Patheon to process defibrotide into the finished product at its Italian manufacturing plant. These facilities are subject to continuing regulation by the Italian Health Authority and other Italian regulatory authorities with respect to the manufacturing of APIs, including the defibrotide drug substance, and defibrotide in its finished form. These facilities are also

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subject to inspection and regulation by the EMA with respect to the manufacturing of the defibrotide drug substance and its finished form and the other APIs we manufacture. Also, part of the process to obtain FDA and EMA approval for defibrotide is to obtain certification from those authorities that these facilities are in compliance with current Good Manufacturing Practices, or cGMP. Following initial approval, if any, regulatory authorities will continue to inspect our manufacturing facilities, in some cases unannounced, to confirm ongoing compliance with cGMP. These regulators may deny approval to manufacture our APIs or otherwise require us to stop manufacturing our APIs, including defibrotide, if they determine that either our facilities or our third party processor's facility does not meet the standards of compliance required under applicable regulations. In addition, these regulators may require us to complete costly alterations to our facilities.

We use hazardous materials in our manufacturing facilities, and any claims relating to the improper handling, storage, release or disposal of these materials could be time-consuming and expensive.

Our API manufacturing involves the controlled storage, use and disposal of chemicals and solvents. We are subject to Italian laws, which implement EU directives and regulations governing the use, transportation, treatment, storage, handling and disposal of solid and hazardous materials, wastewater discharges and air emissions. The environmental management system for our plant was certified under the UNI EN ISO 14001 Standard on April 20, 2007 and under the Eco-management and Audit Scheme, or EMAS, on July 26, 2007. These certifications were valid for a three-year period. Both certifications have been renewed into 2016. Our environmental policy is designed to comply with current regulations on environmental protection, to provide for continuous improvement of our manufacturing performance, to protect our employees' health and safety and to respect the safety of people living or working close to our plant and in the surrounding community. Although we believe that our safety procedures for handling and disposing of these hazardous materials comply with the standards prescribed by these laws and regulations, we cannot completely eliminate the risk of contamination or injury from hazardous materials. If an accident occurs, an injured party could seek to hold us liable for any damages that result and any liability could exceed the limits or fall outside the coverage of our insurance. We may not be able to maintain insurance on acceptable terms, or at all. We may incur significant costs to comply with current or future environmental laws and regulations.

We may have difficulty obtaining raw material for defibrotide.

Defibrotide is produced from DNA extracted from swine intestinal mucosa. La.bu.nat. S.r.l., or La.bu.nat., is our sole supplier of swine intestinal mucosa. La.bu.nat.'s processing centers may be evaluated and inspected by regulatory authorities in connection with regulatory approvals and the ongoing manufacture of our products. If La.bu.nat. experiences safety or other issues that impact its ability to supply swine intestinal mucosa to us as needed, we may not be able to find alternative suppliers in a timely fashion. In that case, we would have to slow or cease our manufacture of defibrotide which could negatively impact our product launch and anticipated revenues.

Historically, there has been no significant price volatility for any of our raw materials. However, given the demand for swine intestinal mucosa for heparin, we may experience volatility in the price of swine intestinal mucosa. In addition, the widespread illness or destruction of pigs could result in volatility in the price of swine intestinal mucosa, which may negatively impact our anticipated revenues.

It is difficult and costly to protect our proprietary rights relating to defibrotide, and we may not be able to ensure their protection.

Our commercial success depends in part on obtaining and maintaining patent protection and/or trade secret protection of defibrotide and its use and the methods used to manufacture it, as well as successfully defending these patents against third party challenges, and successfully protecting our trade secrets. Our ability to protect defibrotide from unauthorized making, using, selling, offering to sell or importation by third parties depends on the extent to which we have rights under valid and enforceable patents, or have trade secrets that cover these activities.

We have a portfolio of U.S. and non-U.S. patents and patent applications relating to various compositions of defibrotide, methods of use and methods of characterization, which will expire at various times between April 2017 and June 2032. Changes in patent laws could increase the uncertainties and costs surrounding the enforcement or defense of our issued patents. Any patent may be challenged, invalidated, held unenforceable or circumvented. In addition, our patents may not prevent other companies from developing similar or therapeutically equivalent products.

In recent years, several companies have been extremely aggressive in challenging patents covering pharmaceutical products, and the challenges have often been successful. We cannot assure you that our patents will not be challenged by third parties or that we will be successful in any defense we undertake. Failure to successfully defend a patent challenge could materially and adversely affect our business.

We cannot ensure that others will not be issued patents that may prevent the sale of defibrotide or require licensing and the payment of significant fees or royalties. We may be unable to avoid infringement of third party patents and may have to

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obtain a license, defend an infringement action, or challenge the validity of the patents in court. A license may be unavailable on terms and conditions acceptable to us, if at all. Patent litigation is costly and time-consuming, and we may be unable to prevail in any such patent litigation or devote sufficient resources to pursue such litigation. If we do not obtain a license under necessary patents, are found liable for infringement, or are not able to have such patents declared invalid, we may be liable for significant money damages or be precluded from the manufacture, use or sale of defibrotide in a manner requiring such licenses.

Trade secrets are difficult to protect. Although we use reasonable efforts to protect our trade secrets and other unpatented proprietary information, our employees, consultants, advisors and partners may unintentionally or willfully disclose our proprietary information to competitors, and we may not have adequate remedies for such disclosures. Protracted and costly litigation could be necessary to enforce and determine the scope of our proprietary rights. Courts in certain jurisdictions are sometimes less willing to protect trade secrets. Failure to obtain or maintain patent and trade secret protection, for any reason, could have a material adverse effect on our business.

We face substantial competition from other companies, including companies with greater resources and larger sales organizations than we have.

Defitelio is the first approved treatment in the EU for the treatment of severe VOD in HSCT. Various anti-clotting strategies have been tried by researchers with mixed results, including Activase (Alteplase), a recombinant tissue plasminogen activator, marketed by Genentech, Inc., generic heparin sodium injection, and Thrombate III (antithrombin III (human)), marketed by Grifols Therapeutics, Inc. While there is currently no direct competition to Defitelio to treat severe VOD, changes in the types of conditioning regimens used as part of HSCT may affect the incidence rate of VOD and demand for Defitelio.

In addition, there may be organizations, including large pharmaceutical and biopharmaceutical companies, as well as academic and research organizations and government agencies, who are interested in, or are currently, pursuing the research and development of drug therapies that target the blood vessel wall. The commercial potential of Defitelio may be reduced or eliminated if our competitors develop or acquire and commercialize products that are safer or more effective, have fewer side effects, are easier to administer or are less expensive than Defitelio. Many of our competitors, particularly large pharmaceutical and life sciences companies, have substantially greater research and development capabilities and financial, operational and human resources than we do.

Many of our competitors are able to deploy more personnel to market and sell their products than we do. We currently have a relatively small number of sales representatives compared with the number of sales representatives of most other pharmaceutical companies with marketed products. Each of our sales representatives is responsible for a territory of significant size. The continued growth of Defitelio may require expansion of our sales force and sales support organization, and we may need to commit significant additional funds, management and other resources to the growth of our sales organization. We also have to compete with other pharmaceutical and life sciences companies to recruit, hire, train and retain sales and marketing personnel, and turnover in our sales force and marketing personnel could negatively affect sales. If our specialty sales force and sales organization are not appropriately sized to adequately promote Defitelio, the commercial potential of Defitelio may be negatively affected.

Conducting clinical trials is costly and time-consuming, and the outcomes are uncertain. A failure to prove safety and efficacy in clinical trials would require us to discontinue development, which could materially and adversely affect our business, financial condition, results of operations and growth prospects.

We are evaluating potential development of defibrotide in indications in addition to the treatment of severe VOD in adults and children undergoing HSCT therapy. These development efforts may not be successful, and any adverse events or other information generated during the course of our studies related to defibrotide could result in action by the EMA or other regulatory agencies, which may restrict our ability to sell, or sales of, Defitelio. Clinical testing can take many years to complete and failure can occur any time during the clinical trial process. Any failure or delay in completing clinical trials for additional indications or the generation of additional clinical data could materially and adversely affect the maintenance and growth of the markets for defibrotide, which could adversely affect our business, financial condition, results of operations and overall growth prospects.

Although Defitelio has been approved in Europe, a prior NDA submission for defibrotide in the United States was voluntarily withdrawn from consideration in 2011 before an FDA decision on accepting the application for filing, based on issues raised by the FDA. We are currently assessing what we believe would be the optimal path for potential approval of defibrotide in the United States, which may include filing a new application with existing clinical data or generating additional clinical data before a new application is ready for submission and FDA review. We cannot know when, if ever, defibrotide will be approved in the United States or under what circumstances, and what, if any, additional clinical or other development

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activities will be required in order to potentially obtain regulatory approval in the United States and the cost associated with such required activities, if any. These development efforts may not be successful, which could adversely affect our potential future revenue from defibrotide and our growth prospects.

Clinical trials can be delayed or halted for a variety of reasons, including delays or failures in obtaining regulatory authorization to commence a trial because of safety concerns of regulators or failure to follow regulatory guidelines; delays or failures in obtaining clinical materials and manufacturing sufficient quantities for use in trials; delays or failures in obtaining approval of our clinical trial protocols from Ethics Committees to conduct a clinical trial at a prospective study site; delays in recruiting patients to participate in a clinical trial; failure of our clinical trials and clinical investigators to be in compliance with regulations applicable to the conduct of clinical trials; unforeseen safety issues; inability to monitor patients adequately during or after treatment or at multiple study sites; failure of our third party clinical trial managers to satisfactorily perform their contractual duties, comply with regulations or meet expected deadlines; or insufficient funds to complete the trials.

The results from early clinical trials may not be predictive of results obtained in later and larger clinical trials, and later clinical trials may fail to show the desired safety and efficacy despite having progressed successfully through initial clinical testing. In that case, the EMA or the equivalent in jurisdictions outside of the EU may determine our data is not sufficiently compelling to warrant marketing approval and may require us to engage in additional clinical trials or provide further analysis which may be costly and time-consuming. A number of companies in the pharmaceutical industry, including us, have suffered significant setbacks in clinical trials, even in advanced clinical trials after showing positive results in earlier clinical trials.

We rely on contract research organizations, or CROs, and other third parties to assist us in designing, managing, monitoring and otherwise carrying out our clinical trials, including with respect to site selection, contract negotiation and data management. We do not control these third parties and, as a result, they may not treat our clinical studies as a high priority, or in the manner in which we would prefer, which could result in delays. If third parties do not successfully carry out their duties under their agreements with us, if the quality or accuracy of the data they obtain is compromised due to failure to adhere to our clinical protocols or regulatory requirements, or if they otherwise fail to comply with clinical trial protocols or meet expected deadlines, our clinical trials may not meet regulatory requirements. If our clinical trials do not meet regulatory requirements or if these third parties need to be replaced, our clinical trials may be extended, delayed, suspended or terminated. If any of these events occur, we may not be able to obtain regulatory approval of defibrotide outside the EU or in additional indications or otherwise generate additional useful clinical data in support of defibrotide.

We may be required to suspend or discontinue any current and future clinical trials due to adverse events or other safety issues that could negatively affect our anticipated revenues from and future growth of defibrotide.

If we choose to, or are required to, conduct any future clinical trials for defibrotide, the trials may be suspended at any time for a number of safety-related reasons. For example, we may voluntarily suspend or terminate such clinical trials if, at any time, we believe that defibrotide presents an unacceptable risk to the clinical trial patients. In addition, Ethics Committees, institutional review boards or regulatory agencies may order the temporary or permanent discontinuation of a clinical trial at any time if they believe that the clinical trial is not being conducted in accordance with applicable regulatory requirements, including if it presents an unacceptable safety risk to patients.

Administering any product candidate to humans may produce undesirable side effects. VOD is a condition associated with high dose chemotherapy and HSCT. Adverse events involving vascular disorders, coagulation and potentially life-threatening bleeding have been reported in VOD patients treated with defibrotide, which could potentially be related to treatment with defibrotide. Hypotension has been reported in patients participating in clinical trials of defibrotide to treat severe VOD, which may also be related to defibrotide. For example, we discontinued a 69-patient Phase I/II clinical trial of defibrotide to prevent deep vein thrombosis after hip surgery in Denmark in 2002, when three patients experienced hypotension after receiving defibrotide intravenously.

It is possible that new adverse events or safety issues will emerge from future clinical trials and the data generated in those trials, which could impact conclusions relating to the safety of defibrotide. Any complications associated with the use of defibrotide would severely harm our anticipated revenue and potential future growth.

If we fail to comply with the significant ongoing regulatory obligations and requirements, we could face increased costs and penalties.

Our activities, and the activities of our collaborators and third-party providers, are subject to extensive government regulation and oversight by the EC, the competent authorities of the EU member states, the FDA and other regulatory agencies. These regulatory authorities directly regulate many of our most critical business activities, including the conduct of preclinical and clinical studies, product manufacturing, advertising and promotion, product distribution, adverse event reporting and product risk management. Failure by us or any of our third party partners, including suppliers, manufacturers and distributors, to comply with applicable requirements could subject us to administrative or judicial sanctions or other negative consequences,

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such as delays in approval or refusal to approve a product candidate, withdrawal, suspension or variation of product approval, untitled letters, warning letters, fines and other monetary penalties, unanticipated expenditures, product recall, withdrawal or seizure, total or partial suspension of production or distribution, interruption of manufacturing or clinical trials, operating restrictions, injunctions; suspension of licenses, civil penalties and/or criminal prosecution, any of which could have a significant impact on our sales, business and financial condition.

If we receive regulatory approvals to sell our products, as we have in the EU for Defitelio for the treatment of severe hepatic VOD in adults and children undergoing HSCT therapy, the EC, the competent authorities of the EU member states, the FDA and other regulatory agencies in Europe or other countries where defibrotide may possibly be approved in the future may impose significant restrictions on the indicated uses or marketing of defibrotide, or impose requirements for burdensome post-approval study commitments. The terms of any product approval, including labeling, may be more restrictive than we desire and could affect the commercial potential of the product. If we become aware of problems with defibrotide, a regulatory agency may impose restrictions on the product, our contract manufacturers or on us. In such an instance, we could experience a significant drop in the sales of defibrotide, our product revenues and reputation in the marketplace may suffer, and we could become the target of lawsuits. Under regulations in the EU related to pharmacovigilance, or the assessment and monitoring of the safety of drugs, we may be required to conduct a labor intensive collection of data regarding the risks and benefits of defibrotide and may be required to engage in ongoing assessments of those risks and benefits, including the possible requirement to conduct additional clinical studies, which may be time consuming and expensive and could impact our profitability. Non-compliance with such obligations can lead to the imposition of financial penalties or other enforcement measures.

The marketing authorization in the EU for Defitelio requires us to comply with a number of post-marketing obligations. These include obligations relating to the establishment of a patient registry. We may be unable to comply with the post-marketing obligations imposed as part of the marketing authorization for Defitelio. Failure to comply with these requirements may lead to the suspension, variation or withdrawal of the marketing authorization for Defitelio in the EU.

Defitelio has been provided to patients in some EU countries on a named patient basis and, in certain of these countries, reimbursement is provided for unauthorized products provided through national named patient or compassionate use programs. Such reimbursement may no longer be available if authorization for named patient or compassionate use programs expire or are terminated. While we believe we have satisfied the regulations regarding our communications and medical affairs activities in those countries, if any such country's regulatory authorities determine that we are promoting Defitelio without a marketing authorization in place, we could be found to be in violation of pharmaceutical advertising law or the regulations permitting sales under named patient programs. In that case, we may be subject to financial or other penalties.

In the EU, the advertising and promotion of medicinal products are subject to EU member states' laws governing promotion of medicinal products, interactions with physicians, misleading and comparative advertising and unfair commercial practices. In addition, other legislation adopted by individual EU member states may apply to the advertising and promotion of medicinal products. These laws require that promotional materials and advertising in relation to medicinal products comply with the product's Summary of Product Characteristics, or SmPC, as approved by the competent authorities. The SmPC is the document that provides information to physicians concerning the safe and effective use of the medicinal product. It forms an intrinsic and integral part of the marketing authorization granted for the medicinal product. Promotion of a medicinal product that does not comply with the SmPC is considered to constitute off-label promotion. The off-label promotion of medicinal products is prohibited in the EU. The applicable laws at the EU level and in the individual EU member states also prohibit the direct-to-consumer advertising of prescription-only medicinal products. Violations of the rules governing the promotion of medicinal products in the EU could be penalized by administrative measures, fines and imprisonment. These laws may further limit or restrict the advertising and promotion of Defitelio to the general public and may also impose limitations on our promotional activities with health care professionals.

The FDA, the competent authorities of the EU member states and other governmental authorities also actively enforce regulations prohibiting off-label promotion, and the government has levied large civil and criminal fines against companies for alleged improper promotion. The government has also required companies to enter into complex corporate integrity agreements and/or non-prosecution agreements that impose significant reporting and other burdens on the affected companies. Failure to maintain a comprehensive and effective compliance program could subject us to a range of regulatory actions that could affect our ability to commercialize Defitelio and could harm or prevent sales of Defitelio, or could substantially increase the costs and expenses of commercializing and marketing Defitelio.

We are subject to strict laws, including anti-bribery and anti-corruption laws, that regulate our relationship with physicians and our other business activities, and any violation of those laws can result in significant adverse consequences.

Interactions between pharmaceutical companies and physicians are also governed by strict laws, regulations, industry self-regulation codes of conduct and physicians' codes of professional conduct in the individual EU member states. The

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provision of benefits or advantages to physicians to induce or encourage the prescription, recommendation, endorsement, purchase, supply, order or use of medicinal products is prohibited in the EU. The provision of benefits or advantages to physicians is also governed by the national anti-bribery laws of the EU member states as described below.

Payments made to physicians in certain EU member states must be publicly disclosed. Moreover, agreements with physicians must often be the subject of prior notification and approval by the physician's employer, his/her competent professional organization, and/or the competent authorities of the individual EU member states. These requirements are provided in the national laws, industry codes, or professional codes of conduct applicable in the EU member states. Failure to comply with these requirements could result in reputational risk, public reprimands, administrative penalties, fines or imprisonment.

Our business activities are also subject to the the Bribery Act 2010 of the United Kingdom, or UK Bribery Act, the Foreign Corrupt Practices Act of 1977, or FCPA, and similar anti-bribery or anti-corruption laws, regulations or rules of other countries in which we operate. The FCPA and similar anti-corruption laws generally prohibit the offering, promising, giving, or authorizing others to give anything of value, either directly or indirectly, to non-U.S. government officials in order to improperly influence any act or decision, secure any other improper advantage, or obtain or retain business. The FCPA also requires public companies to make and keep books and records that accurately and fairly reflect the transactions of the company and to devise and maintain an adequate system of internal accounting controls. The UK Bribery Act prohibits giving, offering, or promising bribes to any person, including non-UK government officials and private persons, as well as requesting, agreeing to receive, or accepting bribes from any person. In addition, under the UK Bribery Act, companies which carry on a business or part of a business in the UK may be held liable for bribes given, offered or promised to any person, including non-UK government officials and private persons, by employees and persons associated with the company in order to obtain or retain business or a business advantage for the company. Liability is strict, with no element of a corrupt state of mind, but a defense of having in place adequate procedures designed to prevent bribery is available. Furthermore, under the UK Bribery Act, there is no exception for facilitation payments. As described above, our business is heavily regulated and therefore involves significant interaction with public officials, particularly officials of non-U.S. governments. Additionally, in many other countries, the health care providers who prescribe pharmaceuticals are employed by their government, and the purchasers of pharmaceuticals are government entities; therefore, our dealings with these prescribers and purchasers may be subject to regulation under the FCPA and the UK Bribery Act. We are engaged in ongoing efforts that are designed to ensure our compliance with these laws. However, there is no certainty that all employees and third party business partners will comply with anti-bribery laws. In particular, we do not control the actions of manufacturers and other third party agents, although we may be liable for their actions. Violation of these laws may result in civil or criminal sanctions, which could include monetary fines, criminal penalties, and disgorgement of past profits, which could have a material adverse impact on our business and financial condition.

Product liability and product recalls could harm our business.

The development, manufacture, testing, marketing and sale of pharmaceutical products are associated with significant risks of product liability claims or recalls. Side effects of, or manufacturing defects in, defibrotide could exacerbate a patient's condition, or could result in serious injury or impairments or even death. This could result in product liability claims and/or recalls of defibrotide or the other APIs we manufacture. In many countries, including in EU member states, national laws provide for strict (no-fault) liability which applies even where damages are caused both by a defect in a product and by the act or omission of a third party.

We face an inherent risk of product liability exposure in connection with human clinical trial testing of defibrotide and the distribution of defibrotide through our named patient and expanded access programs and commercially after launch in the EU. An individual may bring a product liability claim against us if defibrotide causes, or merely appears to have caused, an injury. Product liability claims may be brought by individuals seeking relief for themselves, or by groups seeking to represent a class of injured patients. Further, third party payors, either individually or as a putative class, may bring actions seeking to recover monies spent on one of products. While we have not had to defend against any product liability claims to date, as sales of defibrotide increase, we believe it is likely product liability claims will

be made against us. The risk of product liability claims may also increase if a company receives a warning letter from a regulatory agency. We cannot predict the frequency, outcome or cost to defend any such claims.

Product liability insurance coverage is expensive, can be difficult to obtain and may not be available in the future on acceptable terms, if at all. Our product liability insurance may not cover all of the future liabilities we might incur in connection with the development, manufacture or sale of our products. In addition, we may not continue to be able to obtain insurance on satisfactory terms or in adequate amounts.

A successful claim or claims brought against us in excess of available insurance coverage could subject us to significant liabilities and could have a material adverse effect on our business, financial condition, results of operations and growth

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prospects. Such claims could also harm our reputation and the reputation of defibrotide, adversely affecting our ability to market defibrotide successfully. In addition, defending a product liability lawsuit is expensive and can divert the attention of key employees from operating our business.

Product recalls may be issued at our discretion or at the discretion of our suppliers, government agencies and other entities that have regulatory authority for pharmaceutical sales. Any recall of defibrotide could materially adversely affect our business by rendering us unable to sell defibrotide for some time and by adversely affecting our reputation. A recall could also result in product liability claims by individuals and third party payors. In addition, product liability claims could result in an investigation of the safety or efficacy of defibrotide, our manufacturing processes and facilities, or our marketing programs conducted by the FDA, the EMA, or the competent authorities of the EU member states, which could lead to product liability lawsuits as well.

The Public Company Accounting Oversight Board, or PCAOB, is unable to enforce its review of the audits conducted by our independent registered public accounting firm operating in Italy; therefore, investors may be or may have been deprived of the benefits of such inspection.

Our independent registered public accounting firm, Reconta Ernst and Young S.p.A., or Reconta, issues the audit reports included in our annual reports filed with the SEC. Reconta is required by the laws of the United States to undergo regular inspections by the PCAOB to assess its compliance with SEC rules and PCAOB professional standards. While our audits are performed in accordance with the standards of PCAOB, our auditors are a registered public accounting firm in Italy, a jurisdiction where the PCAOB is currently unable, under Italian law, to conduct inspection of our audits by Reconta. As a result, Reconta, like other independent registered public accounting firms in Italy, is currently not inspected by the PCAOB.

Inspections of audit firms that the PCAOB has conducted have identified deficiencies in those firms' audit procedures and quality control procedures, which may be addressed as part of the inspection process to improve future audit quality. The lack of PCAOB inspections in Italy prevents the PCAOB from regularly evaluating our auditor's audits and quality control procedures. As a result, the inability of the PCAOB to conduct inspections of auditors in Italy may deprive or may have deprived investors of the benefits of PCAOB inspections.

If we are unable to attract and retain qualified personnel and key relationships, our business could be seriously harmed.

We are highly dependent on our senior management, whose services are critical to the successful implementation of research and development and manufacturing and regulatory strategies, and our ability to maintain relationships with key opinion leaders. If we lose one or more of the members of our senior management or other key opinion leaders, our business could be seriously harmed.

Replacing key employees may be difficult and may take an extended period of time because of the limited number of individuals in our industry with the breadth of specific skills and experience required to further develop and commercialize defibrotide and to handle regulatory matters relating to defibrotide. Competition to hire from this limited pool is intense, and we may be unable to hire, train, retain or motivate additional key personnel, if needed.

All of our defibrotide drug compound and most of our other API manufacturing capability is located in one facility that is vulnerable to natural disasters, telecommunication and information system failures, terrorism and similar problems, and we are not insured against losses that may be caused by any of these occurrences or events.

We conduct all of our manufacturing operations for the defibrotide drug compound and two of our other APIs in a single facility located in Villa Guardia, near Como, Italy. This facility could be damaged by fire, flood, earthquake, power loss, telecommunication and information system failure, terrorism or similar events. Our insurance covers damages to the facility, including the buildings, machinery, electronic equipment and goods, of up to approximately €23 million, but does not cover damages caused by any of the events listed above, including terrorism and some types of flooding. Although we believe that our insurance coverage is adequate for our current and proposed operations, there can be no guarantee that it will adequately compensate us for any losses that may occur. We are not insured against business interruption and we do not have a replacement manufacturing facility readily available.

Our ADSs have been delisted from NASDAQ. There is no public market for our ordinary shares and only a limited over-the-counter market for our ADSs. We will make no reports publicly available in the future about our financial

position or results of operations.

On March 5, 2014, we notified The NASDAQ Stock Market of our intention to voluntarily delist our ADSs from NASDAQ. We determined to withdraw our ADSs from listing because of the small number of ADSs that remained outstanding following the completion of the tender offer by Jazz and because delisting is a necessary step in the process of

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terminating our obligations to file reports with the SEC. We have not and do not intend to arrange for the listing of the ADSs on another securities exchange or for any other quotation of the ADSs or our ordinary shares to be made available. Trading of our ADSs on NASDAQ was suspended on March 7, 2014, and there is currently no public market for our ordinary shares and only a limited over-the-counter market for our ADSs. Holders of our ADSs will have only limited price and trading information available and holders of our ordinary shares will have no price and trading information available in making investment decisions. The absence of an active public market will make it difficult for holders of our ADSs or ordinary shares to sell these securities and may adversely affect the value of these securities. In addition, as a result of the suspension and termination of our duty to file reports with the SEC, we will cease providing annual reports and current or periodic information, including information about our financial position or results of operations. The lack of publicly available reports with financial information and other information about us will make it more difficult for an investor to value our ADSs and ordinary shares, further complicating the possibility of a market being available for them.

We have amended the deposit agreement with our ADS depositary governing our ADSs to reduce the period after which the ADS depositary may sell the remaining ordinary shares it holds and to provide for the ADS depositary to accept any offer by a subsidiary of Jazz to purchase those shares at a per share price of no less than $57.00. Holders of ADSs who wish to retain an ownership interest in us will need to surrender their ADSs and request delivery of ordinary shares before any sale occurs.

Our ADS depositary gave notice to the holders of the ADSs on March 14, 2014 that the deposit agreement will terminate at 5:00 pm (Eastern time) on April 13, 2014, and of a proposed amendment to the deposit agreement governing the ADSs. The amendment is dated March 21, 2014, will be effective April 13, 2014, and will reduce from one year to 60 days the period that must elapse between the date of termination of the deposit agreement and the date on which the ADS depositary may sell the ordinary shares of the Company held by it. The amendment also provides for the ADS depositary to accept any offer by a subsidiary of Jazz to purchase those shares at a price of no less than $57.00 per ordinary share, unless the ADS depositary has received what it considers to be a superior bona fide offer from another party. The deposit agreement contemplates that ordinary shares held after termination of the deposit agreement are to be sold and the proceeds made available to the holders of the ADSs upon surrender of their ADSs. While the date or dates on which the ADS depositary will sell the remaining ordinary shares has not been determined, it will not occur before June 13, 2014. Jazz Italy has indicated that it intends to offer to purchase ordinary shares from the ADS depositary at a per share price equal to $57.00. Affirmative action on the part of the holders of ADSs will be necessary if they are to retain an ownership interest in shares, including compliance with any Italian requirements for holding our ordinary shares directly.

Jazz controls our company and the outcome of all shareholder actions of our company.

Approximately 98% of our ordinary shares are owned by Jazz Italy, which is a wholly-owned subsidiary of Jazz. This means that Jazz has the voting power to elect our entire board of directors and to control all actions that require shareholder approval under Italian law and our articles of association and bylaws, including the election of our board of directors, significant mergers and acquisitions and other business combinations, and changes to our articles of association. As a result, Jazz has effective control over our company, our management and our operations, including control over our decisions to enter into any corporate transaction and the ability to prevent any transaction that requires shareholder approval regardless of whether our minority shareholders believe that the transaction is in our best interests. Although our minority shareholders will continue to be entitled to exercise limited rights as shareholders provided to them under the Italian Civil Code, Jazz could cause corporate or other actions to be taken even if the interests of Jazz conflict with the interests of our other shareholders. In addition, three of our five directors are employed by Jazz, which means that we do not have a board of directors making business decisions on our behalf independent from Jazz. Jazz's voting control may cause transactions to occur that might not be beneficial to the holders of our ordinary shares, and may prevent transactions that would be beneficial to those holders.

Our business relationships may be subject to disruptions due to uncertainty associated with the acquisition of control of our company by Jazz.

Parties with which we currently do business or may do business in the future, including our suppliers and customers, may experience uncertainty associated with the acquisition of control of our company by Jazz, including with respect to current or future business relationships with Jazz or us. As a result, our business relationships may be subject to disruptions if customers, suppliers and others attempt to negotiate changes in existing business relationships or consider entering into business relationships with parties other than Jazz or us. In addition, our success will depend, in part, upon our ability to attract and retain qualified personnel and key relationships, especially during Jazz's integration efforts. Our current and prospective employees of might experience uncertainty about their future roles with us now that Jazz controls our company, which might adversely affect our ability to retain qualified personnel and key relationships. These disruptions could have an adverse effect on our business, financial condition, results of operations or prospects.

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Due to the differences between Italian and U.S. law, our ordinary shareholders may have fewer shareholder rights than they would have as a shareholder of a U.S. company.

We are incorporated under the laws of the Republic of Italy. As a result, the rights and obligations of our ordinary shareholders are governed by Italian law and our bylaws, and are in some ways different from those that apply to U.S. corporations. Some of these differences may result in our shareholders having fewer shareholder rights than they would have as a shareholder of a U.S. corporation. We have presented a detailed comparison of the Italian laws applicable to our company versus Delaware law in Part I, Item 10 of this annual report in the section entitled "Differences in Corporate Law." We compare the Italian laws applicable to our company against Delaware law because Delaware is the most common state of incorporation for U.S. public companies.

Italian labor laws could impair our flexibility to restructure our business.

In Italy, our employees are protected by various laws which afford them consultation rights with respect to specific matters regarding their employers' business and operations, including the downsizing or closure of facilities and employee terminations. In particular: (i) Law no. 604/1966 regulates the individual dismissals; (ii) Law no. 223/1991 concerns the collective dismissal procedure; (iii) Law no. 428/1990, as amended by legislative decree no. 18/2001, provides for an information and consultation procedure in case of a transfer of the undertaking or a part thereof; and (iv) Legislative decree no. 25/2007 introduces a general right to information and consultation for employees. These laws and the collective bargaining agreements to which we are subject could impair our flexibility if we need to restructure our business.

ITEM 4. INFORMATION ON THE COMPANY

HISTORY AND DEVELOPMENT OF THE COMPANY

We started as a group of pharmaceutical businesses founded in Italy in 1944 and have been involved in the research and development of drugs derived from DNA and DNA molecules since the 1970s. In 1993, we were formed by F3F S.r.l. (formerly known as FinSirton S.p.A.) as Pharma Research S.r.L., an Italian società a responsabilità limitata, for the purpose of pursuing research and development of prospective pharmaceutical specialty products. In July 2001, we were reformed as an Italian società per azioni and changed our name to Gentium S.p.A. Under our current bylaws, our company's term of existence will expire on December 31, 2050. We are governed by the Italian Civil Code.

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On December 19, 2013, we entered into a definitive tender offer agreement with Jazz and Jazz Italy, pursuant to which Jazz Italy made an offer to purchase all our outstanding ordinary shares and ADSs at a purchase price of $57.00 per ordinary share and per ADS (without duplication for ordinary shares underlying ADSs). On February 21, 2014, Jazz Italy completed its tender offer, having acquired approximately 98 percent of our combined ordinary shares and ADSs. Jazz is now our indirect majority shareholder. We filed a Notice of Voluntary Delisting with The NASDAQ Stock Market on March 5, 2014, and trading in our ADSs on NASDAQ was suspended on March 7, 2014. We filed a Form 25 with the SEC on March 17, 2014 to terminate registration of our ordinary shares and ADSs under Section 12(b) of the Exchange Act and intend to file a Form 15 with the SEC promptly after this annual report is filed with the SEC to terminate registration of our ordinary shares and ADSs under Section 12(g) of the Exchange Act and to suspend our duty to file reports under Sections 13(a) and 15(d) of the Exchange Act. We expect that this will be the final report we will be required to file under Sections 13(a) and 15(d) of the Exchange Act.

We and the ADS depositary entered into an amendment dated March 21, 2014 to the deposit agreement dated as of June 15, 2005, or the deposit agreement, which amendment will be effective on April 13, 2014. Upon effectiveness, the amendment to the deposit agreement will reduce from one year to 60 days the period that must elapse between the date of termination of the deposit agreement and the date on which the ADS depositary may sell the ordinary shares of the Company it then holds and provides for the ADS depositary to accept any offer by a subsidiary of Jazz to purchase those shares at a price of no less than $57.00 per ordinary share, unless the ADS depositary has received what it considers to be a superior bona fide offer from another party. The ADS depositary has given notice such that the deposit agreement will terminate at 5:00 pm (Eastern time) on April 13, 2014. While the date or dates on which the ADS depositary will sell the remaining shares have not been determined, the sale will not occur before June 13, 2014. Jazz Italy has indicated that it intends to offer to purchase ordinary shares from the ADS depositary at a per share price equal to $57.00.

Our principal executive offices are located at Piazza XX Settembre 2, 22079 Villa Guardia (Como), Italy. Our telephone number is +39 031 5373200. Our website is located at www.gentium.it. The information contained on our website is not part of this annual report. Our registered agent for service of process is CT Corporation System, located at 111 Eighth Avenue, 13th Floor, New York, New York 10011, telephone number (212) 894-8940.

This annual report contains market data and industry forecasts that were obtained from industry publications and third parties.

Capital Expenditures

The following table sets forth our capital expenditures (on an accrual basis) for each year in the three-year period ended December 31, 2013 (in thousands):

	For the Year Ended December 31,		
	2011	2012	2013
Land and buildings	€8	€4	€165
Plant and machinery	84	43	606
Industrial equipment	4	27	36
Furniture and fixtures	213	52	63
Leasehold improvements	718	97	11
Internally developed software	23	11	15
Construction in progress	228	159	711
Total	€1,278	€393	€1,607

All of these capital expenditures were in Italy with the exception of a few computer purchases. We financed these expenditures with revenue and cash-flow generated from the distribution and sale of defibrotide, on a pre-approval basis, under an expanded access program pursuant to a treatment IND protocol in the United States and named patient programs elsewhere.

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BUSINESS OVERVIEW

Defibrotide

Overview

We are a biopharmaceutical company primarily focused on the development and manufacture of defibrotide, a sodium salt of a complex mixture of single- and double-stranded oligodeoxyribonucleotides derived from porcine DNA. Our development of defibrotide has been directed to the treatment and prevention of VOD, a potentially life-threatening complication of HSCT. Stem cell transplantation is a frequently used treatment modality for hematologic cancers and other conditions in both adults and children. Certain high-dose conditioning regimens used as part of HSCT can damage the lining cells of hepatic vessels which is thought to lead to the development of VOD, a blockage of the small vessels in the liver, that leads to liver failure and can result in significant dysfunction in other organs such as the kidneys and lungs. The condition is also referred to as “sinusoidal obstruction syndrome.” Severe VOD is the most extreme form of VOD and is associated with multi-organ failure and high rates of morbidity and mortality.

Defibrotide has been granted orphan drug designation to treat and prevent VOD by the FDA, by the EMA and by the Korean Ministry of Food and Drug Safety. The Commonwealth of Australia-Department of Health has granted defibrotide orphan drug designation for the treatment of VOD. In November 2013, the EMA also granted orphan drug designation to defibrotide for the prevention of acute graft versus host disease, or GvHD, another potentially fatal complication of HSCT that afflicts up to 50% of all donor transplant patients.

In October 2013, the EC granted marketing authorization under exceptional circumstances for our defibrotide product, Defitelio, for the treatment of severe VOD in adults and children undergoing HSCT therapy. Defitelio is the first approved treatment in the EU for this potentially life-threatening condition. Defitelio has generally been well-tolerated; the most frequent adverse reactions observed during pre-marketing use of the product are hemorrhage, hypotension and coagulopathy.

Our wholly-owned subsidiary, Gentium GmbH, together with other subsidiaries of Jazz, commenced the launch of Defitelio in Europe in March 2014, with an initial launch in Germany and Austria. We expect to launch in additional European countries on a rolling basis during 2014 and 2015 and are engaged in pricing and reimbursement submissions as applicable in preparation for planned launches in those countries. We intend eventually to promote Defitelio in all European markets where it has marketing authorization. In addition, we expect to continue to give patients access to defibrotide in countries where it is not commercially available through our expanded access program in the United States and through our named patient program throughout the rest of the world.

Defibrotide has received Fast Track designation by the FDA to treat severe VOD. The Fast Track program is designed to enable more frequent interactions with the FDA during drug development and to expedite the FDA’s review of a new drug candidate. In 2011, we voluntarily withdrew from consideration an NDA we submitted to the FDA in July 2011 seeking approval in the United States for defibrotide for the treatment of VOD in order to address issues raised by the FDA. We are currently assessing what we believe would be the optimal path for potential approval of defibrotide in the United States, which may include filing a new application with existing clinical data or generating additional clinical data before a new application is ready for submission and FDA review.

We have completed two clinical trials of defibrotide, a Phase III trial for the treatment of severe VOD with multiple organ failure, conducted in the United States, Canada and Israel, and a Phase II/III pediatric trial conducted in Europe for the prevention of VOD. We also have an ongoing study of defibrotide for the treatment of VOD through our IND protocol. We expect to collect additional usage tolerability and safety data from patients receiving defibrotide through our expanded access and named patient programs.

Commercialization and Distribution

We expect to continue to distribute Defitelio commercially after launch in certain major European countries with our current logistics partner, IDIS Ltd, a U.K.-based company. We have entered into license and/or supply and distribution agreements with specialized regional partners to distribute defibrotide, including on a named patient basis, in the following territories: the Asian Pacific, the Middle East and North Africa, other countries in Europe, the Nordics and Baltics, Turkey, Israel and the Palestinian Authority. Certain of these regional partners have also agreed

to assist us with local registration, marketing authorization, reimbursement, marketing, sales, distribution and medical affairs activities.

Under a license and supply agreement, we have licensed to Sigma-Tau the rights to commercialize defibrotide for the treatment and prevention of VOD in North America, Central America and South America, subject to receipt of marketing authorization, if any, in the applicable territory. Pursuant to the license and supply agreement, between 2001 and 2010, we

received milestone and other payments in the amount of $11.35 million, and we are entitled to a payment of $6 million following regulatory approval from the FDA to market defibrotide in the United States, a further $2 million payment following the transfer of the approved NDA to Sigma-Tau, royalty payments equal to 7% of Sigma-Tau's net sales of defibrotide and a supply price equal to the greater of 31% of net sales or €50 (approximately $68) per unit of defibrotide finished product. In addition, in connection with the license and supply agreement, we entered into a cost sharing agreement under which Sigma-Tau agreed to reimburse us 50% of certain costs associated with the development of defibrotide. We also agreed that $1 million in costs reimbursed by Sigma-Tau will be deductible from royalty payments that we are entitled to receive in the future under the license and supply agreement.

There are currently no approved treatments for VOD in the United States. As a result, defibrotide is being distributed to patients diagnosed with VOD in the United States through an expanded access program pursuant to a treatment IND protocol. In addition, we expect to continue to give patients access to defibrotide in other countries where it is not commercially available through our named patient program. We have received €16.89 million, €22.77 million, and €33.65 million through our named patient and expanded access programs in 2011, 2012 and 2013, respectively. Information on our revenues by geographic markets for the last three fiscal years is included in the "Operating Results" section under Item 5 of this annual report.

Market Overview

We historically focused the development of defibrotide on the treatment and prevention of VOD, a potentially life-threatening complication of HSCT. Severe VOD is the most extreme form of VOD and is associated with multi-organ failure and high rates of morbidity and mortality. An analysis of retrospective data, prospective cohort studies and clinical trials published between 1979 and 2007 found that the 100-day mortality rate in severe VOD cases is greater than 80%. Based on data from published surveys and our market research, we estimate that of the approximately 35,000 patients undergoing HSCT annually in the EU, approximately 6,300 are considered at high risk for the development of VOD, and the incidence of VOD is approximately 3,600 patients. Our review of relevant literature and market research also suggests that about one-third to two-thirds of VOD patients may be eligible for treatment using defibrotide.

Competition

To our knowledge, there are no approved treatments in the EU for VOD at this time, other than Defitelio, which was granted marketing authorization under exceptional circumstances by the EC for the treatment of severe VOD in adults and children undergoing HSCT therapy. There are currently no approved treatments for VOD in the United States. Various anti-clotting strategies have been tried by researchers with mixed results, including Activase (Alteplase), a recombinant tissue plasminogen activator, marketed by Genentech, Inc., generic heparin sodium injection, and Thrombate III (antithrombin III (human)), marketed by Grifols Therapeutics, Inc. While there is currently no direct competition to Defitelio to treat severe VOD, changes in the types of conditioning regimens used as part of HSCT may affect the incidence rate of VOD and demand for Defitelio.

In addition, there may be organizations, including large pharmaceutical and biopharmaceutical companies, as well as academic and research organizations and government agencies, who are interested in, or are currently, pursuing the research and development of drug therapies that target the blood vessel wall. While we are unaware of any other products or product candidates that treat or prevent VOD, we believe that other companies have products or are currently developing products to treat some of the same disorders and diseases that defibrotide is designed to treat.

Manufacturing of Active Pharmaceutical Ingredients

We have a manufacturing plant in Italy where we produce APIs, including the defibrotide compound. We believe that we are currently the sole worldwide producer of defibrotide. With respect to defibrotide, we entered into a manufacturing services agreement with Patheon, effective September 15, 2013, pursuant to which Patheon will process the defibrotide compound into finished form at its Italian manufacturing plant and supply the finished product to us. The initial term of the agreement ends on December 31, 2018, subject to automatic extension for additional two-year terms unless either party provides notice to the other of its intent to terminate the agreement at least 18 months before the end of the then-current term. Either party has the right to terminate the agreement in the event of the other party's uncured material breach or insolvency.

Consistent with our long-term manufacturing strategy, and in order to secure an additional source to manufacture commercial supply of Defitelio, we have initiated work with Fresenius Kabi, a global health care company specialized in lifesaving medicines and technologies for infusion, to conduct a technology transfer of our manufacturing process for the finished form of Defitelio to their manufacturing site in Graz, Austria. Subject to a successful technology transfer, including

manufacture of process validation batches, and receipt of all necessary regulatory approvals, we intend to qualify Fresenius Kabi as a second source of Defitelio.

In addition to the defibrotide compound, we manufacture sodium heparin, urokinase and sulglicotide for third parties. These APIs are subsequently used to make the finished forms of various drugs. Sodium heparin and sulglicotide are manufactured in our plant. We sell sulglicotide primarily to a South Korean partner, which uses this API to finish a drug that it markets in South Korea. We currently manufacture urokinase in a small leased laboratory and manufacturing space. Two companies purchase this API to create a finished drug that treats various vascular disorders such as deep vein thrombosis and pulmonary embolisms.

Our revenues from the sales of sodium heparin, urokinase and sulglicotide amounted to €4.85 million, €4.86 million and €6.17 million in 2011, 2012 and 2013, respectively. We did not sell any sodium heparin in 2011 and 2013 and do not know if there will be any demand for us to supply sodium heparin in the future.

On July 3, 2013, we entered into an agreement with EG S.p.A., a subsidiary of the Stada Group, whereby we (i) sold the Italian marketing authorization for our former urokinase product, Genkinase, and (ii) agreed to supply EG S.p.A. and Crinos S.p.A., or Crinos, another subsidiary of the Stada Group, urokinase (the active pharmaceutical ingredient of Genkinase) for a period of five years. In connection with this agreement, Crinos has agreed to waive its right to royalty payments equal to 1.5% of the net sales of defibrotide in Europe for seven years following our receipt of a marketing authorization for defibrotide from the EMA, the rights of which Crinos had previously acquired pursuant to an agreement with us in 2006.

Seasonality does not affect our business, although the timing of manufacturer orders can cause variability in sales.

Raw Materials

Defibrotide and sulglicotide are produced from DNA extracted from pig intestines, using well-established processes that are used by others to manufacture various drugs. In particular, defibrotide is derived from swine intestinal mucosa and sulglicotide is derived from swine duodenum. On May 31, 2013 and on October 1, 2013 we amended our existing supply agreements with La.bu.nat. to supply us, respectively, with the swine duodenum and swine intestinal mucosa we need to produce sulglicotide and defibrotide.

The contract term of the swine intestinal mucosa supply agreement expires on September 30, 2018, with automatic renewal for a five-year period, unless either party notifies the other party in writing 90 days prior to the expiration date. The contract term of the swine duodenum supply agreement expires on December 31, 2015, with no automatic renewal. We entered into an agreement on March 1, 2013 with a backup supplier named Far-Pet S.r.l. for the supply of swine duodenum. This agreement will expire on December 31, 2014, with no automatic renewal.

Urokinase is derived from human urine. We currently purchase the urine from one supplier, Polyamine (Taiwan Corporation), with whom we signed a supply agreement on March 8, 2013 which has a five year term.

The FDA and other regulatory bodies may evaluate the processing centers of La.bu.nat., Polyamine or any other raw material supplier in connection with regulatory approvals and the ongoing manufacture of our products.

Historically, there has been no significant price volatility for any of our raw materials. However, given the demand for swine mucosa for heparin, we may experience volatility in the price of pig intestines. In addition, the widespread illness or destruction of pigs could result in volatility of the price of pig intestines.

Regulatory Matters

Overview

The preclinical and clinical testing, manufacturing, labeling, storage, distribution, promotion, sale, import and export, reporting and record-keeping of our product candidates are subject to extensive regulation by governmental authorities in the United States, principally the FDA and corresponding state agencies, as well as regulatory agencies in foreign countries.

Non-compliance with applicable regulatory requirements can result in, among other things, injunctions, seizure of products, total or partial suspension of product manufacturing and marketing, denial of approval by the government, the withdrawal of marketing approvals, civil penalty actions and criminal prosecution. Except as discussed below, we believe that we are in substantial compliance in all material respects with each of the currently applicable laws, rules and regulations mentioned in this section. During the most recent inspections of our manufacturing facility, for the

drug product and drug substances, by the Italian Health Authority in April and September 2012, the Italian Health Authority observed certain

deficiencies which had been corrected. In order to obtain FDA approval for the sale of any of our product candidates, the FDA must determine that this facility meets their cGMPs, including requirements for equipment, and we must receive verification and validation of our manufacturing and cleaning processes. The FDA has not yet inspected our facility, but since 2004 we have spent over €10 million in upgrades to our facility in anticipation of such an inspection. We are not aware of any other situation that could be characterized as an incidence of non-compliance in the last three years.

United States Regulatory Approval

Drug Approval Process

FDA regulations require us to endure a long and rigorous process before any of our product candidates may be marketed or sold in the United States. This regulatory process generally requires:

The steps required before a product candidate may be approved for marketing in the United States generally include:

- performance of satisfactory preclinical laboratory and animal studies under the FDA's good laboratory practices regulations;
- submission to and acceptance by the FDA of an IND for human clinical testing, which must become effective before human clinical trials may commence;
- adequate and well-controlled human clinical trials to establish the safety and efficacy of any product candidate for each intended indication under the FDA's good clinical practices regulations;
- submission to the FDA of a marketing application;
- satisfactory completion of an FDA inspection of the manufacturing facilities at which the product is made, analyzed and stored to assess compliance with cGMPs;
- potential FDA audit of the nonclinical and clinical trial sites that generated the data in support of the application; and
- FDA review and approval of the application.

This process requires a substantial amount of time and financial resources. In 2002, the FDA announced a reorganization that resulted in a change in the oversight and approval process for certain therapeutic biologic drugs and a reassignment of responsibility for the process from the Center for Biologics Evaluation and Research to the Center for Drug Evaluation and Research. Our initial product candidate, defibrotide to treat severe VOD, is now being regulated through the latter.

Assuming successful completion of all required testing in accordance with all applicable regulatory requirements, detailed information on the product candidate is submitted to the FDA in the form of an NDA requesting approval to market the product for one or more indications, together with payment of a user fee, unless waived. An NDA includes all relevant data available from pertinent nonclinical and clinical studies, including negative or ambiguous results as well as positive findings, together with detailed information on the chemistry, manufacture, controls (CMC) and proposed labeling, among other things. To support marketing approval, the data submitted must be sufficient in quality and quantity to establish the safety and efficacy of the product candidate for its intended use to the satisfaction of the FDA.

If an NDA submission is accepted for filing, the FDA begins an in-depth review of the NDA. Under the Prescription Drug User Fee Act, or PDUFA, the FDA's goal is to complete its initial review and respond to the applicant within 12 months of submission, unless the application relates to an unmet medical need in a serious or life-threatening indication and is designated for priority review, in which case the goal may be within eight months of NDA submission. However, PDUFA goal dates are not legal mandates and FDA response often occurs several months beyond the original PDUFA goal date. Further, the review process and the target response date under PDUFA may be extended if the FDA requests, or the NDA sponsor otherwise provides, additional information or clarification regarding information already provided in the NDA. The NDA review process can, accordingly, be very lengthy. During its review of an NDA, the FDA may refer the application to an advisory committee for review, evaluation and recommendation as to whether the application should be approved. The FDA is not bound by the recommendation of an advisory committee, but it typically follows such recommendations. Data from clinical studies are not always conclusive and the FDA and/or any advisory committee it appoints may interpret data differently than the NDA sponsor.

After the FDA evaluates the NDA and inspects manufacturing facilities where the drug product and/or its API will be produced, it will either approve commercial marketing of the drug product with prescribing information for specific indications or issue a complete response letter indicating that the application is not ready for approval and stating the conditions that must be met in order to secure approval of the NDA. If the complete response letter requires additional data and the applicant subsequently submits that data, the FDA nevertheless may ultimately decide that the NDA does not satisfy its criteria for approval. The FDA could also approve the NDA with a Risk Evaluation and Mitigation Strategies, or REMS, plan to mitigate

risks, which could include medication guides, physician communication plans, or elements to assure safe use, such as restricted distribution methods, patient registries and other risk minimization tools. The FDA also may condition approval on, among other things, changes to proposed labeling, development of adequate controls and specifications, or a commitment to conduct post-marketing testing. Such post-marketing testing may include phase 4 clinical studies and surveillance to further assess and monitor the product's safety and efficacy after approval. Regulatory approval of products for serious or life-threatening indications may require that participants in clinical studies be followed for long periods to determine the overall survival benefit of the product.

Post-Approval Regulations

Any approval of a product candidate is limited to specific clinical uses. Subsequent discovery of previously unknown side effects or other problems relating to a product may result in additional restrictions on its use or even the complete withdrawal of the product from the market. Any FDA-approved product that we manufacture or distribute will be subject to continuing regulation by the FDA, which requires record-keeping and reporting of adverse events or experiences. Drug manufacturers are required to register their establishments with the FDA and state agencies, and are subject to periodic inspections by the FDA and state agencies to ensure compliance with cGMPs, which impose rigorous procedural and documentation requirements upon us and our contract manufacturers. Failure to comply with these requirements may result in, among other things, total or partial suspension of production activities, a denial by the FDA of marketing approvals, or the withdrawal, suspension, or revocation of marketing approvals.

If the FDA approves one or more of our product candidates, we, along with our contract manufacturers, must provide certain safety and effectiveness information while the drug is being marketed. Changes in the product, as well as changes in the manufacturing process or facilities, or other post-approval changes, may necessitate additional FDA review and approval. The labeling, advertising, promotion, marketing and distribution of a drug must also be in compliance with FDA requirements relating to, among others, disclosure of risk information, standards and regulations for communication of information relating to off-label uses, industry sponsored scientific and educational activities and other promotional activities, including those involving the Internet. The FDA has very broad enforcement authority, and failure to abide by these requirements can result in a warning letter mandating the correction of deviations from regulatory standards, or enforcement actions that can include seizures, fines, injunctions and criminal prosecution.

Fast track and orphan drug designation

The FDA has a "fast track" program which allows for expedited review of an application. However, there is no assurance that the FDA will, in fact, accelerate the review process for a fast track product candidate. Fast track status is provided only for new and novel therapies that are intended to treat persons with life-threatening and severely debilitating diseases, and where there is a defined unmet medical need, particularly when no satisfactory alternative therapy exists or the new therapy is found to be significantly superior to alternative therapies. During the development of product candidates that qualify for this status, the FDA may expedite consultations and reviews of these experimental therapies. Approval of an application for a fast track review can be based on an effect on a clinical endpoint, or on a surrogate endpoint that is reasonably likely to predict a clinical benefit. The FDA may condition the approval of an application for fast track review on additional post-approval studies that validate the surrogate endpoint or confirm the effect on the clinical endpoint. Fast track status also provides the potential for a product candidate to obtain a "priority review." A priority review allows for a portion of the application to be submitted to the FDA for review prior to the completion of the entire application, which could result in a reduction of the length of time it would otherwise take the FDA to complete its review of the application. Fast track status may be revoked by the FDA at any time after approval if the clinical trial results do not continue to support that the product candidate has the potential to address an unmet medical need. A product approved under a "fast track" designation is subject to expedited withdrawal procedures and to enhanced FDA scrutiny of promotional materials.

The FDA may grant orphan drug status to drugs intended to treat a "rare disease or condition," which is generally characterized as a disease or condition that affects fewer than 200,000 individuals in the United States. If and when the FDA grants orphan drug status, the generic name and trade name of the therapeutic agent and its potential orphan use are publicly disclosed by the FDA. Aside from guidance concerning the non-clinical laboratory studies and

clinical investigations necessary for approval of the application, orphan drug status does not convey any advantage in, or shorten the duration of, the regulatory review and approval process. The FDA may grant orphan drug designations to competing product candidates targeting the same uses. A product that has been designated as an orphan drug and subsequently receives the first FDA approval for the designated orphan use is entitled to orphan drug exclusivity, which means that, except in limited circumstances, the FDA may not approve any other applications for the same indication for seven years from the date of FDA approval. Orphan drug status may also provide certain tax benefits. Finally, the FDA may fund the development of orphan drugs through its grant program for clinical studies.

The FDA has designated defibrotide as an orphan drug for the treatment of VOD and the prevention of VOD and has provided funding for clinical studies for defibrotide to treat VOD. The FDA has approved our application for "fast track" designation for defibrotide to treat severe VOD occurring after stem cell transplantation by means of injection. If our other product candidates meet the criteria, we may apply for orphan drug status and fast track status for these other products.

Market Exclusivity

In addition to orphan drug exclusivity, a product regulated by the FDA as a "new drug" may be entitled to non-patent and/or patent exclusivity under the Federal Food, Drug and Cosmetic Act, or FFDCA, over a third party obtaining an abbreviated approval of a generic product during the exclusivity period. An abbreviated approval allows an applicant to obtain FDA approval without generating, or obtaining a right of reference to, the basic safety and effectiveness data necessary to support the initial approval of the drug product or active ingredient. In the case of a new chemical entity (an active ingredient which has not been previously approved with respect to any drug product) non-patent exclusivity precludes an applicant for abbreviated approval from submitting an abbreviated application until five years after the date of approval of the new chemical entity. With regard to any drug substance (active ingredient), drug product (formulation and composition) and method of use patent listed with the FDA, patent exclusivity under the FFDCA precludes the FDA from granting effective approval of an abbreviated application of a generic product until the relevant patent(s) expire, unless the abbreviated applicant certifies that the relevant listed patents are invalid, not infringed or unenforceable and either the NDA/patent holder does not file an infringement action within 45 days of receipt of notification of the certification, or an infringement action is filed within 45 days and a court determines that the relevant patent(s) are invalid, not infringed or unenforceable or 30 months have elapsed without a court decision of infringement.

User Fees

A New Drug Application for a prescription drug product that has been designated as an orphan drug is not subject to the payment of user fees to the FDA unless the application includes an indication other than the orphan indication.
A supplement proposing to include a new indication for a designated orphan disease or condition in an application is also not subject to a user fee if the drug has been designated an orphan drug with regard to the indication proposed in such supplement.
There is no specific exemption for orphan drug products from annual product and establishment fees. However, sponsors of orphan drugs can request a waiver of such fees due to hardship or on other grounds.

HIPAA

Certain federal and state legislation may affect our ability to obtain certain health information in conjunction with our research activities. Specifically, the Health Insurance Portability and Accountability Act of 1996, or HIPAA, and the Health Information Technology for Economic and Clinical Health Act, or the HITECH Act, mandate, among other things, that covered entities and business associates safeguard the privacy and security of individually identifiable health information in specific ways. In relevant part, the U.S. Department of Health and Human Services, or HHS, has released two rules to date, which mandate how the confidentiality and integrity of such information must be protected. The Privacy Rule imposes standards relating to the privacy of individually identifiable health information. These standards restrict the manner and circumstances under which covered entities and their business associates may use and disclose protected health information so as to protect the privacy of that information. The second rule released by HHS establishes minimum standards for the security of electronic health information. HIPAA and the HITECH Act impose requirements on covered entities and business associates, including those covered entities and/or their business associates that conduct research activities regarding the use and disclosure of individually identifiable health information. As a result, unless covered entities conducting clinical trials for us obtain an effective authorization from each research subject for the release of the subject's individually identifiable health information meeting applicable requirements, such covered entities or their business associates may not be able to share with us all of the results from clinical trials that include such health information.

Non-U.S. Regulatory Approval

Outside of the United States, our ability to market our product candidates will also be contingent upon our receipt of marketing authorizations from the appropriate foreign regulatory authorities, regardless of whether or not FDA approval has been obtained. The foreign regulatory approval process in most industrialized countries generally involves risks similar to those associated with the FDA approval process, as described herein. The requirements governing the conduct of clinical trials and marketing authorizations, and the time required to obtain requisite approvals may widely vary from country to country and may differ from that required for FDA approval.

European Union Regulatory Approval

Under the current EU regulatory system, applications for marketing authorizations may be submitted under a centralized, mutual recognition, or decentralized procedure.

The centralized procedure

An applicant under the centralized procedure must be a person who is domiciled in the EU or an entity established in the EU. The applicant must file a preliminary request containing information regarding the product candidate, including a description of the product applicant and the location of the production plant, along with payment of the application fees. The EMA formally evaluates the preliminary request and either indicates initial approval or a rejection of the preliminary request. If the EMA indicates an initial approval of the preliminary request, the applicant must then submit a full application to the EMA for review. This application must indicate certain specific information regarding the product candidate, including the composition (quality and quantity) of all the substances contained in the product, therapeutic indications and adverse events, modalities of use, the results of physical, chemical, biological and microbiological tests, pharmacological and toxicity tests, clinical tests, a description of production and related control procedures, a summary of the characteristics of the product as required by the European legislation and samples of labels and information to consumers. The applicant must also file copies of marketing authorizations obtained, applications filed and denials received for the same product in other countries, and must prove that the manufacturer of the product candidate is duly authorized to produce it in its country.

The EMA (through its CHMP) examines the documents and information filed and may carry out technical tests regarding the product, request information from the member state concerned with regard to the manufacturer of the product candidate and, when it deems necessary, inspect the manufacturing facility in order to verify that the manufacturing facility is consistent with the specifications of the product candidate, as indicated in the application. The Committee generates and submits its final opinion to the European Commission, the member states and the applicant. The Commission then issues its decision, which is binding on all member states. However, if the Commission approves the application, member states still have authority to determine the pricing of the product in their territories before it can be actually marketed.

The EMA may reject the application if the Agency decides that the quality, safety and effectiveness of the product candidate have not been adequately and sufficiently proven by the applicant, or if the information and documents filed are incomplete, or where the labeling and packaging information proposed by the applicant does not comply with the relevant European rules.

The EMA has also established an accelerated evaluation procedure applicable to product candidates intended to treat or prevent serious diseases or conditions for which no suitable therapy exists, and for which substantial beneficial effects on patients can be predicted.

The marketing authorization granted under the centralized procedure is valid in all EU countries, as well as in Iceland, Liechtenstein and Norway. The marketing authorization is valid for five years and may be renewed, upon application, for additional five year terms. After the issue of the authorization, the holder must constantly take into consideration scientific and technical progress so that the product is manufactured and controlled in accordance with generally accepted scientific methods. In addition, products for which the applicant can demonstrate that comprehensive data on the efficacy and safety under normal conditions of use cannot be provided as a result of certain specified reasons may be eligible for marketing authorization under exceptional circumstances. A marketing authorization granted under exceptional circumstances is also valid for five years, but is subject to an annual reassessment of the risk-benefit balance.

We filed our MAA for defibrotide under the centralized procedure. In October 2013, the EC granted marketing authorization under exceptional circumstances for Defitelio for the treatment of severe VOD in adults and children undergoing HSCT therapy.

The mutual recognition procedure

Under the mutual recognition procedure, the holder of a national marketing authorization, obtained in accordance with the procedure and requirements applicable in the member state concerned, is entitled to submit an application to the

remaining member states in which it seeks a marketing authorization. Within 90 days of receipt of the application and assessment report, each member state must decide whether or not to recognize approval. The mutual recognition process results in separate national marketing authorizations in the reference member state and each concerned member state.

The decentralized procedure

The decentralized procedure should be used for products that have not yet received authorization in a EU member state. The applicant may request one or more concerned member state(s) to approve a draft assessment report, a summary of product characteristics, the product labeling and a package leaflet as proposed by the chosen reference member state. If a member state cannot approve the assessment report, the summary of product characteristics, the product labeling and the package leaflet on grounds of potential serious risk to human and animal health or to the environment, a pre-referral procedure should be commenced by the relevant coordination group. If the member states fail to reach an agreement during the 60-day pre-referral procedure, the matter is deferred to an arbitration proceeding.

Post-approval issues

There are many national legislative instruments (implementing EU rules) governing controls on drugs in the post-authorization phase. For instance, the holder of the national marketing authorization must promptly record in detail any adverse reaction to the drug of which it becomes aware, regardless of the country in which the reaction occurs, and prepare periodic update reports on these adverse events. The holder of the authorization must hire and retain for its organization an expert who will be responsible for drug controlling and reporting activities.

Moreover, any form of information and advertising aimed at promoting the sale of drugs is governed by specific national legislation (also implementing EU rules), which sets the standards for and limitations on advertising messages generally, and specific promotional activities, such as the organization of conferences regarding certain drugs and the distribution of free samples.

The export of drugs (other than plasma and blood-related products) from Italy is not subject to authorization, but the import of drugs into Italy from non-EU countries is subject to authorization by the Ministry of Health, on the basis of the adequacy of the quality controls to be carried out on the imported drugs.

Pediatric Investigation Plan

The pediatric investigation plan, or PIP, is a key element in the European pediatric regulations, which came into effect in January 2007. The PIP is a plan for defining the use of a medicinal product across all age groups of the pediatric population and across all indications. The pediatric committee, or PDCO, is a body within the EMA responsible for overseeing the requirements of the pediatric regulation. The PDCO may issue a waiver with respect to the use of a medicinal product in certain (or all) indications and/or certain (or all) pediatric age groups, or it may issue a deferral of the start or completion dates of all or some of the studies in the PIP. If a sponsor complies with a PIP agreed by PDCO, the sponsor may be eligible for a six-month extension on patents covering the product described in the plan. If the product has been designated an orphan drug by the EMA, it may be eligible for an additional two years of market exclusivity even if a pediatric indication is not approved.

European orphan drug status

European legislation provides for a particular procedure for the designation of medicinal products as orphan drugs. Such a designation may include incentives for the research, development and marketing of these drugs, and allows for an extended period of market exclusivity in the event of a later successful application for a marketing authorization regarding the therapeutic indications for which orphan status was awarded.

A medicinal product, during any stage of its development but, in any case, prior to the filing of any application for the marketing authorization, may be designated as an orphan drug if the person/entity that has applied for the designation can establish that it is intended for the diagnosis, prevention or treatment of a life-threatening or chronically debilitating condition affecting no more than five persons out of every ten thousand persons in the EU, or that it is intended for the diagnosis, prevention or treatment of a life-threatening, seriously debilitating or serious and chronic condition in the EU and, without incentives, it is unlikely that the marketing of the medicinal product within the EU would generate sufficient income to justify the necessary investments in the relevant medicinal product. Moreover, the sponsor must prove that no satisfactory method of diagnosis, prevention or treatment of the condition in question has been authorized in the EU or, if a satisfactory method exists and has been authorized, that the medicinal product will be of significant benefit to those affected by that condition.

In order to obtain the designation of a medicinal product as an orphan drug, the sponsor must submit an application to the EMA for the Evaluation of Medical Products, which must describe the indication of the active ingredients of the medicinal product, the proposed therapeutic indications and proof that the criteria established by the relevant European legislation are met.

The EMA reviews the application and prepares a summary report to a special Committee for Orphan Medicinal Products, which issues an opinion within 90 days of the receipt of the application. The European Commission must adopt a decision within 30 days of receipt of the Committee's opinion. If the European Commission approves the application, the designated medicinal product is entered in the European Register of Orphan Medicinal Products and the product becomes eligible for incentives made available by the EU, and by member states, to support research into, and development and availability of, orphan drugs.

After registration, the product sponsor must submit an annual report to the EMA describing the state of development of the designated orphan drug. A designated orphan drug may be removed from the Register of Orphan Medicinal Products in three cases:

- at the request of the sponsor;
- before the market authorization is granted, if it is established that the requirements provided for in the European orphan drug legislation are no longer being met; or
- at the end of the period of market exclusivity (as explained below).

Orphan drug market exclusivity means that the EU shall not, for a period of 10 years from the grant of the marketing authorization for an orphan drug, accept any other application for a marketing authorization, grant a marketing authorization or accept an application to extend an existing marketing authorization, for the same product. This period, however, may be reduced to six years if at the end of the fifth year it is established that the criteria set forth in the legislation are no longer met by the orphan drug, or if the available evidence shows that the orphan drug is sufficiently profitable, so that market exclusivity is no longer justified.

However, as an exception to orphan drug market exclusivity, a marketing authorization may be granted for the same therapeutic indications as a similar medicinal product if:

- the holder of the marketing authorization for the orphan drug has given his consent to the second applicant;
- the holder of the marketing authorization for the orphan drug is unable to supply sufficient quantities of the latter; or
- the second applicant can establish in its application that the second medicinal product, although similar to the authorized orphan drug, is safer, more effective or otherwise clinically superior to the orphan drug.

Legal Proceedings

In January 2014, we became aware of a purported class action lawsuit filed in the Southern District of New York in connection with the tender offer conducted by Jazz Italy to acquire our ordinary shares and ADSs. The lawsuit, captioned Xavion Jyles, Individually and on Behalf of All Others Similarly Situated v. Gentium S.P.A. et al., names us, each of our directors, Jazz and Jazz Italy as defendants. The lawsuit alleges, among other things, that our directors breached their fiduciary duties to our shareholders in connection with the tender offer agreement that we entered into with the Jazz entities valuing our ordinary shares and ADSs at $57.00 per share, and that the Jazz entities violated Sections 14(e) and 20(a) of the Exchange Act, by allegedly overseeing our preparation of an allegedly false and misleading Section 14D-9 Solicitation/Recommendation Statement. The lawsuit seeks, among other relief, class action status, rescission, and unspecified costs, attorneys' fees and other expenses. We cannot predict the timing or outcome of this matter.

From time to time we are involved in legal proceedings arising in the ordinary course of business. We believe there is no other litigation pending that could have, individually or in the aggregate, a material adverse effect on our results of operations or financial condition.

ORGANIZATIONAL STRUCTURE

Approximately 98% of our ordinary shares are owned by Jazz Italy, a wholly-owned subsidiary of Jazz. Jazz, a specialty biopharmaceutical company organized under the laws of Ireland, is focused on improving patients' lives by identifying, developing and commercializing differentiated products that address unmet medical needs.

In August 2011, we formed a wholly-owned subsidiary, Gentium GmbH, organized under the laws of Switzerland.

PROPERTY, PLANTS AND EQUIPMENT

Manufacturing and Facilities

We own a manufacturing plant near Como, Italy which, at December 31, 2013, was subject to a mortgage securing repayment of an aggregate of €1.08 million of debt owed to Banca Nazionale del Lavoro. The manufacturing facility is 2,350 square meters in size. Historically, we relied on a third party to generate steam used in the manufacturing process. We are currently completing the installation of two steam generators at an estimated cost of approximately €0.40 million in order to generate steam without reliance on any third party. We are also installing a reactor and a dryer at an estimated cost of approximately €0.30 million. In 2013, we completed a back entrance to our manufacturing plant to facilitate vehicle access and installed a new and independent fire ring.

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We produce defibrotide and sulglicotide at this facility and have the capability to produce sodium heparin. We did not sell any sodium heparin in 2011 and 2013 and do not know if there will be any demand for us to supply sodium heparin in the future.

In addition, we lease 100 square meters of laboratory and manufacturing space in Villa Guardia (Como) from Sirton S.p.A., where we currently manufacture urokinase. The capacity and utilization of our manufacturing facilities are such that we can satisfy the currently anticipated requirements for production of defibrotide, sodium heparin and urokinase.

Our manufacturing plant is subject to the regulation of regional agencies regarding worker health and safety, the fire department, and Azienda Sanitaria Locale and Agenzia Regionale Prevenzione e Ambiente with respect to water, air, noise and environmental pollution protection. We have engaged Lariana Depur, a consortium that specializes in the treatment of waste water, to treat our waste water. We monitor our waste water to control the levels of nitrogen, chlorides and chemical oxygen before delivering the waste water for additional treatment. We have also installed two scrubbers to reduce the odors and chemicals released into the air by the facility in order to comply with Italian regulations.

The environmental management system for the plant was certified under the UNI EN ISO 14001 Standard on April 20, 2007 and under the EMAS on July 26, 2007. These certifications were valid for a three-year period. Both certifications have been renewed into 2016. Our environmental policy is designed to comply with current regulations on environmental protection, to provide for continuous improvement of our manufacturing performance, to protect our employees' health and safety and to respect the safety of people living close to our plant and the surrounding community.

We also lease approximately 4,800 square meters of office and laboratory space from F3F S.r.l. (formerly known as FinSirton, S.p.A.).

ITEM 4A. UNRESOLVED STAFF COMMENTS

Not applicable.

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ITEM 5. OPERATING AND FINANCIAL REVIEW AND PROSPECTS

You should read the following discussion together with the consolidated financial statements, related notes and other financial information included elsewhere in this annual report. This discussion may contain predictions, estimates and other forward-looking statements that involve risks and uncertainties, including those discussed under "Risk Factors" and elsewhere in this annual report. These risks could cause our actual results to differ materially from any future performance suggested below.

OPERATING RESULTS

Our product sales are generated through customers located outside of Italy and amounted to 97%, 97% and 99% of total product sales for the years ended December 31, 2011, 2012 and 2013, respectively.

We have manufacturing and laboratory facilities where we produce APIs, including the defibrotide compound. These APIs are subsequently used to make the finished forms of various drugs. We generate revenues from the sale of the APIs sodium heparin, urokinase and sulglicotide, which are used by other companies to make the finished form of various drugs. For the years ended December 31, 2011, 2012 and 2013, sales of these APIs amounted to approximately 22%, 18% and 15% of our total product sales, respectively. Sulglicotide is sold primarily to a partner in South Korea and urokinase is sold primarily in Spain. We did not sell any sodium heparin in 2011 and 2013 and do not know if there will be any demand for us to supply sodium heparin in the future.

Defibrotide was sold in the United States and the rest of the world through our expanded access and named patient programs, respectively, and accounted for 78%, 82% and 85% of our total product sales for the years ended December 31, 2011, 2012 and 2013, respectively.

Under a license and supply agreement, we have licensed to Sigma-Tau the rights to commercialize defibrotide for the treatment and prevention of VOD in North America, Central America and South America, subject to receipt of marketing authorization, if any, in the applicable territory. Pursuant to the license and supply agreement, between 2001 and 2010, we received milestone and other payments in the amount of $11.35 million, and we are entitled to a payment of $6 million following regulatory approval from the FDA to market defibrotide in the United States, a further $2 million payment following the transfer of the approved NDA to Sigma-Tau, royalty payments equal to 7% of Sigma-Tau's net sales of defibrotide and a supply price equal to the greater of 31% of net sales or €50 (approximately $68) per unit of defibrotide finished product. In addition, in connection with the license and supply agreement, we entered into a cost sharing agreement with Sigma-Tau, under which it agreed to reimburse us 50% of certain costs associated with the development of defibrotide. We also agreed that $1 million in costs reimbursed by Sigma-Tau will be deductible from royalty payments that we are entitled to receive in the future under the license and supply agreement.

We recognized other revenue of €2.03 million, €1.26 million and €2.56 million for the years ended December 31, 2011, 2012 and 2013, respectively, in connection with our agreements with Sigma-Tau. In 2013, we received €1.40 million ($1.80 million) as reimbursement of research and development expenses, and in 2014, Sigma-Tau agreed to reimburse us an estimated total of approximately $4.77 million as incurred.

In 2013, we were cash flow positive, primarily due to revenue generated from our expanded access and named patient programs. We expect that existing cash and cash equivalents together with the anticipated cash flow from product sales will be sufficient to support our current operations for at least the next twelve months. However, if we are unable to successfully commercialize defibrotide, unable to generate sufficient revenue and cash flow through our expanded access and named patient programs, or if we increase expenditures above our current expectations, we may need to obtain additional funding either through arrangements with Jazz or its other subsidiaries, or through debt financings or collaborative agreements with unrelated parties, which may not be available to us on favorable terms, if at all.

As of December 31, 2013, substantially all of our cash and cash equivalents were held in accounts at financial institutions located in the Republic of Italy, Switzerland and the United States, which we believe are of acceptable credit quality. We invest our cash in liquid instruments that meet high credit quality standards and generally mature within three to six months of the purchase date. We are exposed to exchange rate risk with respect to certain of our cash balances, accounts receivable and

accounts payable that are denominated in U.S. dollars. As of December 31, 2013, we held a cash balance of $3.99 million, receivables of $2.95 million and payables of $7.28 million that were denominated in U.S. dollars. These dollar-based balances are available to be used for future purchases and other liquidity requirements that may be denominated in such currency. We are exposed to unfavorable and potentially volatile fluctuations of the U.S. dollar against the Euro, our functional currency. If the U.S. dollar were 10% stronger against the Euro, our net assets balance would increase by approximately €0.03 million as of December 31, 2013.

In addition, we are exposed to foreign currency risks to the extent that we engage in transactions, such as investments, programming costs and accounts payable, denominated in currencies other than our functional currency. With respect to these items, changes in the exchange rate will result in unrealized or realized foreign currency transaction gains or losses, as applicable upon settlement of the transactions.

Given the volatility of exchange rates, there is no assurance that we will be able to effectively manage currency transaction and/or conversion risks. We have not entered into derivative instruments to offset the impact of foreign exchange fluctuations. Fluctuations in foreign currency exchange rates could have a material adverse effect on our results of operations and financial condition.

We are exposed to changes in interest rates primarily as a result of our borrowings. Our primary exposure to variable rate debt is through the Euribor (Euro Interbank Offered Rate). Due to the current rate of the Euribor, the outstanding amounts of the principal due on our borrowings and the scheduling of repayment of such borrowings, we have determined the risk of interest rate movement to be low. We monitor fluctuations in the interest rate market and will consider entering into interest cap agreements if we later determine that such risk has increased.

Critical Accounting Policies and Estimates

Our consolidated financial statements have been prepared in accordance with accounting principles generally accepted in the United States. The preparation of our consolidated financial statements requires management to make estimates, judgments and assumptions that affect the reported amounts of assets, liabilities, revenues, expenses and related disclosures. We base our estimates and judgments on historical experience and on various other assumptions that we believe to be reasonable under the circumstances. Actual results could differ from those estimates.

We believe the following policies to be critical to the understanding our financial condition and operating results because they require us to make estimates, assumptions and judgments about matters that are inherently uncertain.

Revenue Recognition

Our primary source of revenue is from the sale of products through our named patient and expanded access programs and from collaborative arrangements. We recognize revenue when all of the following criteria are met: persuasive evidence of an arrangement exists, delivery has occurred or services have been rendered, our price to the customer is fixed or determinable and collectability is reasonably assured. Revenues from product sales are recognized upon delivery, when title and risk of loss have passed to the customer. Provisions for returns and other adjustments related to sales are provided during the same period in which the related sales are recorded on the basis of historical rates of return. Historically, our returns have been insignificant. Revenues are recorded net of applicable allowance for contractual adjustments entered into with customers.

Collaborative arrangements generally contemplate that our technology or intellectual property will be utilized to commercialize or produce certain pharmaceutical products and that we will receive certain revenues pursuant to these agreements. Collaborative arrangements with multiple deliverables are divided into separate units of accounting if certain criteria are met, including whether the delivered element has stand-alone value to the customer and whether there is objective and reliable evidence of the fair value of the undelivered items. The consideration received from these arrangements is allocated among the separate units based on their respective fair value, and the applicable revenue recognition criteria are applied to each separate unit. Revenue associated with substantive at-risk milestones is recognized based upon the achievement of the milestones as defined in the respective agreements. We defer, and recognize as revenue, non-refundable payments received in advance that are related to the future performance over the life of the related research project. We recognize reimbursements to fund research and development efforts as such qualified expenditures are made. Finally, royalty revenues are recognized when earned after the applicable sales are made.

Inventories

Inventories consist of raw materials, work in progress and finished APIs and defibrotide distributed through our named patient and expanded access programs. We state inventories at the lower of cost or net realizable value, determining cost on an average cost basis. We periodically review inventories and reduce items to their estimated net realizable values as they become outdated or obsolete. We estimate reserves for excess and obsolete inventories based on inventory levels on hand and current and forecast product demand. Our reserve level and, as a result, our overall profitability, is subject to our ability to reasonably forecast future sales levels versus quantities on hand and existing purchase commitments. Forecasting demand and resource

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planning are based, in part, on assumptions that we must make regarding expected market changes, overall demand, pricing incentives and raw material availability, among other variables. Significant changes in these estimates could indicate that inventory levels are excessive, which would require us to reduce inventories to their estimated net realizable values.

In the highly regulated industry in which we operate, raw materials, work in progress and finished goods inventories have expiration dates that must be factored into our judgments as to the recoverability of inventory cost. Additionally, if our estimate of a product's demand and pricing is such that we may not fully recover the cost of inventory, we must consider that in our judgment as well. We also review our inventory and the manufacturing process for quality assurance and quality control issues to determine if a write-down is necessary. In the context of reflecting inventory at the lower of cost or market, we record an inventory reserve as soon as a need for such a reduction in net realizable value is determined.

Prior to commencing the sale of defibrotide through the named patient and expanded access programs, we expensed all costs associated with the production of defibrotide as research and development expenses. Subsequent to signing the agreements associated with our named patient and expanded access programs, we began to capitalize the costs of manufacturing defibrotide as inventory, including costs to convert existing raw materials to active pharmaceutical ingredients and costs to package and label previously manufactured inventory, which had already been expensed as research and development expenses. Until we sell the inventory for which a portion of the costs was previously expensed, the carrying value of our inventory and our cost of sales will reflect only incremental costs incurred subsequent to the signing of the agreements associated with our named patient and expanded access programs.

We expense costs relating to the production of clinical products which are not expected to be sold through our named patient and expanded access programs as research and development expenses in the period incurred.

Research and Development Expenses

Our research and development expenses consist primarily of salaries and benefits of our direct employees, employee stock-based compensation expenses, facility costs, overhead costs, clinical trial costs and related trial product manufacturing costs, contracted services and subcontractor costs. Clinical trial costs include costs associated with clinical research organizations, or CROs. The billings we receive from CROs for services rendered may not be received for several months following the services. Development timelines and costs are difficult to estimate and may vary significantly for each product candidate and from quarter to quarter. We accrue the estimated costs of the CROs' related services based on our estimate of management fees, site management and monitoring costs and data management costs. Differences between estimated and actual trial costs have not been material to date, and any changes have been made when they become known. At December 31, 2013, we had €5.10 million of future payables under outstanding contracts. Most of these contracts are on a cost plus or actual cost basis.

Stock-Based Compensation

Employee stock-based compensation is estimated on the date of grant, based on the fair value of the employee stock award. Employee stock-based compensation is recognized ratably over the requisite service period, which is generally the vesting period, in a manner similar to other forms of compensation paid to employees. The fair values of all option grants were estimated on the grant date using a binomial valuation model. The binomial model considers characteristics of fair value option pricing that are not available under the Black-Scholes model. Similar to the Black-Scholes model, the binomial model takes into account variables such as volatility, dividend yield rate, and risk free interest rate. However, unlike the Black-Scholes model, the binomial model also considers the contractual term of the option, the probability that the option will be exercised prior to the end of its contractual life, the probability of termination or retirement of the option holder in computing the value of the option, and the exchange rate between the Euro and the U.S. dollar. For these reasons, we believe that the binomial model provides a fair value that is more representative of actual experience and future expected experience than the value calculated using the Black-Scholes model.

The option-pricing model requires the use of certain subjective assumptions or estimates regarding the expected volatility of the market price of our stock, the expected term of the award and the expected forfeiture rate. In estimating the expected term of an award, we consider the vesting period of the award, our historical experience with

employee stock option exercise and the expected volatility, and use relevant peer group data as a comparative measure.

As of February 17, 2014, all outstanding option awards had been exercised and all ordinary shares issued upon such exercise had been subsequently tendered pursuant to the tender offer.

We review our assumptions periodically and we may change the assumptions we use to value share-based awards granted in future periods. Such changes may lead to a significant change in the expenses we recognize in connection with share-based payments.

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In using the option pricing model that we have selected, changes in the underlying assumptions have the following effect on the resulting fair value output:

An increase to the:	Results in a fair value estimate that is:
Price of the underlying share	Higher
Exercise price of option	Lower
Expected volatility of stock	Higher
Risk-free interest rate	Higher
Expected term of option	Higher

In our current valuation, we consider the volatility factor to be an important factor in determining the fair value of the options granted. For options granted in 2013, we have used an 88.32% factor based on what we believe is a representative sample of similar biopharmaceutical companies. However, this sample is not perfect, as it omits, for example, Italian companies, due to the fact that there are a limited number of companies publicly traded in the U.S. market. Significant changes to these estimates could have a material impact on the results of our operations.

Valuation Allowance

As of December 31, 2012 and 2013, we had net operating loss, or NOL, carryforwards of approximately €53.48 million and €47.39 million, respectively.

Management has determined, at this time, that it is more likely than not that the domestic deferred tax assets, which are comprised principally of NOL and research and experimentation credit carryforwards, will be recoverable and the related valuation allowance is no longer needed based on an assessment of the relative impact of all positive and negative evidence that existed at December 31, 2013, including an evaluation of cumulative income in recent years, future sources of taxable income, exclusive of reversing temporary differences, and risks and uncertainties related to our business.

Recent Accounting Pronouncements

Reference should be made to Note 2 of our consolidated financial statements, “Summary of Significant Accounting Policies to our Consolidated Financial Statements,” for a discussion of new accounting standards.

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Results of Operations

The following tables set forth our results of operations for the periods presented (in thousands, except share and per share data):

	Year Ended December 31,		
	2011	2012	2013
Revenues:			
API product sales	€4,848	€4,856	€6,172
NPP product sales	16,886	22,774	33,653
Total product sales	21,734	27,630	39,825
Other revenues	123	152	490
Other revenues from related party	2,026	1,257	2,602
Total revenues	23,883	29,039	42,917
Operating costs and expenses:			
Cost of goods sold	6,035	5,778	6,055
Research and development	5,533	10,531	15,672
Selling, general and administrative	7,727	10,829	12,883
Charges from related parties	222	186	189
Depreciation and amortization	870	1,003	1,031
Total costs and expenses	20,387	28,327	35,830
Operating income	3,496	712	7,087
Foreign currency exchange gain/(loss), net	46	(67)	55
Interest income/(expense), net	(21)	155	237
Income before income tax provision/(benefit)	3,521	800	7,379
Income tax provision/(benefit)	811	26	(17,222)
Net income	€2,710	€774	€24,601
Net income per share:			
Basic	€0.18	€0.05	€1.61
Diluted	€0.18	€0.05	€1.48
Weighted-average shares used to compute net income per share:			
Basic	14,964,021	15,014,411	15,261,799
Diluted	15,340,859	15,639,890	16,602,743

Revenues

The following table presents product sales, other revenues and total revenues for the years ended December 31, 2011, 2012 and 2013 (amounts in thousands):

	For The Year Ended December 31,		
	2011	2012	2013
Product sales:			
API - Urokinase	€1,612	€1,404	€1,531
API - Sulglicotide	3,236	3,154	4,641
API - Other	—	298	—
named patient/expanded access program sales	16,886	22,774	33,653
Total product sales	21,734	27,630	39,825
Other revenues	2,149	1,409	3,092
Total revenue	€23,883	€29,039	€42,917

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Year Ended December 31, 2013 Compared to Year Ended December 31, 2012

Product sales

Total product sales, which include sales of defibrotide and APIs, were €39.83 million for 2013 compared to €27.63 million for 2012, an increase of €12.20 million or 44%. The variance was primarily due to the increase in 2013 in sales of defibrotide through the named patient programs, attributable to a price increase which came into effect in the second quarter, and an increase in sales volumes of one of our APIs. In addition, with effect from the second quarter of 2012, the level of service fees, associated with the named patient program managed by one of our European partners, decreased.

Revenues from the distribution of defibrotide through the named patient and expanded access programs amounted to €33.65 million for the year ended December 31, 2013, compared to €22.77 million for the year ended December 31, 2012, an increase of €10.88 million or 48%. For the years ended December 31, 2013 and 2012, named patient and expanded access program sales were net of €0.52 million and €1.55 million in service fees, respectively.

API sales were €6.17 million for 2013 compared to €4.86 million for 2012. Sulglicotide API sales were €4.64 million for 2013 compared to €3.15 million for 2012, accounting for 75% and 65% of our total API sales, respectively. The variance is primarily due to an increase in volume versus the prior year. Urokinase sales were €1.53 million for 2013 compared to €1.40 million for 2012. The variance is attributable to an increase in volume versus the prior year. In 2012, we realized €0.30 million in sales of the API heparin. We did not record any revenues from sales of the API heparin in 2013.

Other revenues

Other revenues were €3.09 million for 2013 compared to €1.41 million for 2012, an increase of €1.68 million. The variance was mainly due to an increase in activities, such as clinical trials, that were reimbursed by Sigma-Tau, which were classified as other revenues from a related party and amounted to €2.56 million and €1.26 million for the years ended December 31, 2013 and 2012, respectively.

Cost of goods sold

Our cost of goods sold was €6.06 million in 2013 compared to €5.78 million in 2012. Overall, the cost of goods sold as a percentage of product sales was 15% in 2013 compared to 21% in 2012 mainly due to a different composition of product mix, with proportionately increased sales of defibrotide, which has a higher margin compared to sales of our other APIs. Also contributing to the variance was an increase in price and decrease in service fees for defibrotide distributed on a named patient basis and we also earned a higher price on some API sales in 2013 following a contract renegotiation.

Research and development expenses

We incurred research and development expenses of €15.67 million in 2013 compared to €10.53 million for 2012, an increase of €5.14 million or 49%. Research and development expenses were primarily for the development of defibrotide to treat and prevent VOD. The increase from the comparable period in 2012 was primarily due to the engagement of CROs and outside scientific, regulatory and quality consultants, travel and conference expenses and scientific advisory board meetings necessary to assist the Company in addressing issues related to our NDA and support the Company through the EMA’s regulatory review process.

Selling, general and administrative expenses

Our selling, general and administrative expenses were €12.88 million for 2013 compared to €10.83 million for 2012, an increase of €2.05 million or 19%. The increase in selling, general and administrative expenses was primarily due to an increase in salary and benefit related expenses of €0.45 million, driven primarily by increased retention and performance bonus expense, and €1.0 million of legal and consultant expense related to the due diligence process in connection with the tender offer agreement entered into with Jazz and Jazz Italy. There was also an increase in travel, administrative and tax consultant expenses in 2013.

Income tax provision/(benefit)

We recognised an income tax benefit of €17.22 million for 2013 compared to an income tax provision of €0.03 million for 2012, a variance of €17.25 million. The variance is attributable to the release of a domestic deferred tax assets valuation allowance of €17.86 million and higher taxable income in 2013 compared to 2012.

Net income

Our net income was €24.60 million in 2013 compared to €0.77 million in 2012. The increase was primarily due to the release of a domestic deferred tax valuation allowance of €17.86 million, an increase in sales of defibrotide through the named patient programs, a slight increase in API sales and other income and revenues under our agreements with Sigma-Tau, partially offset by an increase in selling, general and administrative expenses and research and development expenses.

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Year Ended December 31, 2012 Compared to Year Ended December 31, 2011

Product sales

Total product sales, which included sales of defibrotide and APIs, were €27.63 million for 2012 compared to €21.73 million for 2011, an increase of €5.90 million or 27%. The increase was primarily due to a higher volume of defibrotide distributed through the named patient and expanded access programs, which can be partially attributed to the partnerships entered into in 2012, an increased awareness of defibrotide and a decrease in service fees associated with a named patient program managed by one of our European partners. Revenues from the distribution of defibrotide through the named patient and expanded access programs amounted to €22.77 million for the year ended December 31, 2012, compared to €16.89 million for the year ended December 31, 2011, recording an increase of €5.88 million or 35%. For the years ended December 31, 2012 and 2011, named patient and expanded access program sales were net of €1.55 million and €3.17 million in service fees, respectively.

API sales were €4.86 million for 2012 compared to €4.85 million for 2011. Sulglicotide API sales were €3.15 million for 2012 compared to €3.24 million for 2011, accounting for 65% and 67% of our total API sales, respectively. The variance in sulglicotide sales was partially due to a decrease in volume which had a negative impact of €0.3 million, which was partially offset by €0.2 million due to an increase in the price of sulglicotide. Urokinase sales were €1.40 million for 2012 compared to €1.61 million for 2011. The variance is attributable to a decrease in volume versus the prior year. In 2012, we realized €0.30 million in sales of the API heparin, which we did not have in prior year.

Other revenues

Other revenues were €1.41 million for 2012 compared to €2.15 million for 2011, a decrease of €0.74 million or 34%. Other revenues for the prior year included a ratable recognition of the upfront payment of €5.11 million (US$7.00 million) made by Sigma-Tau in 2010 in connection with the amendment of a license and supply agreement with us to include the indication of prevention of VOD in the Americas, which amounted to €1.70 million (US$ 2.33 million). The decrease was partially offset by an increase in activities, such as clinical trials, that were reimbursed by Sigma-Tau, which amounted to €1.26 million and €0.32 million for the years ended December 31, 2012 and 2011, respectively.

Cost of goods sold

Our cost of goods sold was €5.78 million in 2012 compared to €6.04 million in 2011. Overall, the cost of goods sold as a percentage of product sales was 21% in 2012 compared to 28% in 2011, mainly due to a different composition of product mix with proportionately increased sales of defibrotide which has a higher margin compared to sales of our other APIs. Also contributing to the variance was a net release of an inventory reserve in the amount of €0.46 million, a slight increase in the margin of sulglicotide due to the renegotiation of the sales price and a slight decrease in labor costs due to temporary lay-offs of certain employees in connection with a temporary shut-down of our manufacturing activities.

Research and development expenses.

We incurred research and development expenses of €10.53 million in 2012 compared to €5.53 million for 2011, an increase of €5.00 million or 90%. 2011 research and development expenses include severance, employee termination benefits and other exit costs associated with a change in management in the amount of €0.43 million. Research and development expenses were primarily for the development of defibrotide to treat and prevent VOD. The increase from the comparable period in 2011 was primarily due to the engagement of CROs and outside scientific, regulatory and quality consultants, travel and conference expenses and scientific advisory board meetings necessary to assist the Company in addressing issues related to our NDA and support us through the EMA’s regulatory review process.

Selling, general and administrative expenses

Our selling, general and administrative expenses were €10.83 million for 2012 compared to €7.73 million for 2011, an increase of €3.10 million or 40%. The increase was due to higher legal and tax consultant expenses, personnel and recruiting expenses in connection with an increase in our headcount, corporate governance expenses and stock-based compensation costs. Contributing to the variance were also costs incurred in connection with the establishment of our subsidiary’s European commercial team, which primarily occurred in the second half of 2011. Therefore, sales and marketing expenses for the twelve-month period ended December 31, 2012 account for expenses related to new

appointments for our commercial team that we did not have during the prior-year period.

Income tax expense

Income tax expense was €0.03 million for 2012 compared to €0.81 million for 2011, a decrease of €0.78 million. The decrease is mainly attributable to a decrease in profitability and lower taxable income compared to the prior year. In addition, we released previously accrued income taxes in the amount of €0.02 million, recorded a tax credit of €0.07 million in connection with the enactment of a new tax reform, and accrued Italian corporate taxes of €0.11 million and Swiss corporate taxes of €0.01 million.

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Net income

Our net income was €0.77 million in 2012 compared to €2.71 million in 2011. The difference was primarily due to an increase in research and development expenses, an increase in sales and marketing expenses associated with the establishment of a commercial team, which did not exist until the second half of 2011, an increase in general and administrative expenses, a decrease in other income and revenues from a related party offset by an increase in the volume of defibrotide sold through the named patient and expanded access programs, an increase in gross margin and a decrease in income tax expenses.

LIQUIDITY AND CAPITAL RESOURCES

The following table shows a summary of our cash and cash equivalents and our cash flows for the periods indicated (in thousands):

	As of December 31,	
	2012	2013
Cash and cash equivalents	€12,485	€22,038

	Years Ended December 31,		
	2011	2012	2013
Net cash provided by operating activities	€2,401	€3,422	€11,510
Net cash used in investing activities	(393)	(611)	(4,540)
Net cash provided by/(used in) financing activities	(801)	(249)	2,641
Effect of exchange rates on cash and cash equivalents	41	(67)	(58)
Cash and cash equivalent, at beginning of period	8,742	9,990	12,485
Cash and cash equivalents, at end of period	9,990	12,485	22,038

We require cash to fund our operating activities and service our debt. We have been cash flow positive in 2013, primarily due to revenue and cash flow generated from our named patient programs for defibrotide. Our excess funds are currently invested in short-term investments with a maturity date of three to six months. Based on our historical needs and on current estimates, our current cash position and expected cash flow from our revenues will be sufficient to fund our operations for the next twelve months. However, if we are unable to successfully commercialize defibrotide, unable to generate sufficient revenue and cash flow through our expanded access and named patient programs, or if we increase expenditures above our current expectations, we may need to obtain additional funding either through arrangements with Jazz or its other subsidiaries, or through debt financings or collaborative agreements with unrelated parties.

Operating Activities

In 2013, our primary sources of funding were our operating activities. We closed the year with a net income of €24.60 million after recording a deferred tax asset of €17.86 million. Included in net income in 2013 were certain non-cash income and costs, primarily, the release of a valuation allowance on domestic deferred tax assets of €17.86 million, a provision for income taxes of €0.61 million, depreciation and amortization expense of €1.47 million, stock-based compensation expense of €1.82 million and inventory write-off costs of €0.23 million. Contributing to net income in 2013 were a release of a previously accrued inventory reserve of €0.19 million and gain on fixed assets disposal of €0.31 million. In addition, we had a net change in other operating assets and liabilities of €1.14 million, including an increase in accounts receivable of €4.55 million driven by our increased revenues in 2013, an increase in inventories of €0.50 million, an increase in accounts payable, other accrued expenses and income tax payables of €6.66 million and a release of previously accrued deferred revenues of €0.15 million.

In 2013, as compared to 2012, we recorded an increase in research and development expenses necessary to assist us in addressing issues related to our NDA and supporting us through the EMA regulatory review process, along with an increase in selling, general and administrative expenses, primarily due to an increase in salary and benefit related expenses, and legal and consultant expenses related to the due diligence process in connection with the tender offer agreement entered into with Jazz and Jazz Italy.

In 2012, our primary sources of funding were our operating activities and working capital. We closed the year with net income of €0.77 million. Included in net income in 2012 were certain non-cash charges, primarily depreciation and amortization costs of €1.46 million, stock-based compensation expenses of €1.92 million and a release of a previously accrued inventory reserve of €0.63 million related to inventory sold or destroyed. In addition to non-cash charges, we also had a net change in other operating assets and liabilities of €0.30 million, including a decrease in accounts receivable of €0.98 million, a decrease in inventory of €1.41 million, a decrease in accounts payable, other accrued expenses and income tax payables of €2.01 and a release of previously accrued deferred revenues of €0.33 million. In 2012, as compared to 2011, we recorded an increase in research and development expenses necessary to assist us in addressing issues related to our NDA and supporting us

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through the EMA regulatory review process, along with an increase in sales and marketing expenses in connection with building out our European commercial team, which was established in November 2011.

In 2011, our primary sources of funding were our operating activities and we closed the year with a net income of €2.71 million. Included in net income in 2011 were certain non-cash charges, such as depreciation and amortization costs in the amount of €1.32 million, stock-based compensation expenses of €1.67 million, a provision for income taxes of €0.58 million, and a write-off of inventory of €0.34 million. In addition to non-cash charges, we also had a net change in other operating assets and liabilities of €4.15 million, including an increase in accounts receivable of €1.28 million, an increase in inventory of €0.92 million, a decrease in accounts payable and other accrued expenses of €0.65 million, a decrease in termination indemnities of €0.13 million and a decrease in deferred revenues of €1.21 million, primarily due to the recognition of €1.70 million of previously deferred revenue related to our license and supply agreement with Sigma-Tau. In 2011, we formed Gentium GmbH, a wholly-owned subsidiary organized under the laws of Switzerland, as the headquarters for our commercial operations. We also appointed our commercial leadership team in major European countries.

Investing Activities

In 2013, we had capital expenditures of €0.85 million, which were principally due to investments made in our manufacturing facilities located in Villa Guardia, Como, Italy and proceeds of €0.31 million from the sale of the Italian marketing authorization and trademarks for Genkinase to Stada S.p.A. and manufacturing equipment. As of December 31, 2013 we had €4.00 million invested in short-term deposits with a maturity date of April 30, 2014.

In 2012, we had capital expenditures of €0.61 million, which were principally allocated to leasehold improvements for our corporate offices located in Villa Guardia, Como, Italy.

In 2011, we had capital expenditures of €0.72 million, which were principally allocated to furniture and leasehold improvements for our corporate offices located in Villa Guardia, Como, Italy. Such investment activities were partially financed through the sale of marketable securities for €0.26 million and partially reimbursed by the landlord.

Financing Activities

In 2013, we used approximately €0.41 million to reimburse a portion of our long term debt. Proceeds from the exercise of stock options amounted to €2.45 million and proceeds from new loans entered into in order to finance investments made in our manufacturing facilities were €0.60 million.

In 2012, we used approximately €0.53 million to reimburse a portion of our long term debt and capital lease obligations. Proceeds from the exercise of stock options amounted to €0.28 million.

In 2011, we used approximately €0.88 million to reimburse a portion of our long term debt and capital lease obligations. Proceeds from the exercise of stock options amounted to €0.07 million.

At December 31, 2013, we had an aggregate of €1.74 million in debt outstanding, €22.04 million in cash and cash equivalents and €4.00 million in short-term deposits maturing April 30, 2014. Additional information on the maturity, repayment obligations and interest rate structure with respect to this debt, and our material commitments for capital expenditures is provided below under “Contractual Obligations and Commitments.”

We expect to devote substantial resources toward the continuation of our research and development efforts and related regulatory expenses, the expansion of our licensing and collaboration efforts, the launch and marketing of Defitelio in Europe and obtaining marketing approval in other countries. Our funding requirements will depend on numerous factors including:

- whether we are able to successfully commercialize and sell Defitelio in Europe for the treatment of severe VOD;
- whether we will be able to obtain pricing and reimbursement for Defitelio in the countries in Europe in line with our estimates;
- the timing of, and the costs involved in, obtaining regulatory approvals in the United States and other countries outside the United States and the EU, if any;
- the scope and results of any future clinical trials to support the use of defibrotide in its currently approved indications and any potential future indications; and
- the cost of manufacturing activities.

Factors affecting our external sources of cash include:

- our ability to successfully commercially launch Defitelio for the treatment of severe VOD in Europe and obtain sustained acceptance by patients, physicians and payors; and
- our ability to obtain FDA regulatory marketing approval for defibrotide.

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We have accumulated a deficit of approximately €67.47 million since our inception. We have been cash flow positive since 2010, primarily due to revenue generated from our expanded access and named patient programs. We expect that existing cash and cash equivalents together with the anticipated cash flow from product sales will be sufficient to support our current operations for at least the next twelve months. However, if we are unable to successfully commercialize defibrotide, unable to generate sufficient revenue and cash flow through our expanded access and named patient programs, or if we increase expenditures above our current expectations, we may need to obtain additional funding either through arrangements with Jazz or its other subsidiaries, or through debt financings or collaborative agreements with unrelated parties.

2014 Outlook

On December 19, 2013, we entered into a definitive tender offer agreement with Jazz and Jazz Italy, pursuant to which Jazz Italy made an offer to purchase all our outstanding ordinary shares and ADSs at a purchase price of $57.00 per ordinary share and per ADS (without duplication for ordinary shares underlying ADSs). On February 21, 2014, Jazz Italy completed its tender offer, having acquired approximately 98 percent of our combined ordinary shares and ADSs. Jazz is now our indirect majority shareholder. We filed a Notice of Voluntary Delisting with The NASDAQ Stock Market on March 5, 2014, and trading in our ADSs on NASDAQ was suspended on March 7, 2014. We filed a Form 25 with the SEC on March 17, 2014 to terminate registration of our ordinary shares and ADSs under Section 12(b) of the Exchange Act and intend to file a Form 15 with the SEC promptly after this annual report is filed with the SEC to terminate registration of our ordinary shares and ADSs under Section 12(g) of the Exchange Act and to suspend our duty to file reports under Sections 13(a) and 15(d) of the Exchange Act. We expect that this will be the final report we will be required to file under Sections 13(a) and 15(d) of the Exchange Act.

We and the ADS depositary entered into an amendment dated March 21, 2014 to the deposit agreement dated as of June 15, 2005, or the deposit agreement, which amendment will be effective on April 13, 2014. Upon effectiveness, the amendment to the deposit agreement will reduce from one year to 60 days the period that must elapse between the date of termination of the deposit agreement and the date on which the ADS depositary may sell the ordinary shares of the Company it then holds and provides for the ADS depositary to accept any offer by a subsidiary of Jazz to purchase those shares at a price of no less than $57.00 per ordinary share, unless the ADS depositary has received what it considers to be a superior bona fide offer from another party. The ADS depositary has given notice such that the deposit agreement will terminate at 5:00 pm (Eastern time) on April 13, 2014. While the date or dates on which the ADS depositary will sell the remaining shares have not been determined, the sale will not occur before June 13, 2014. Jazz Italy has indicated that it intends to offer to purchase ordinary shares from the ADS depositary at a per share price equal to $57.00.

Consistent with our long-term manufacturing strategy, and in order to secure an additional source to manufacture commercial supply of Defitelio, we have initiated work with Fresenius Kabi, a global health care company specialized in lifesaving medicines and technologies for infusion, to conduct a technology transfer of our manufacturing process for the finished form of Defitelio to their manufacturing site in Graz, Austria. Subject to a successful technology transfer, including manufacture of process validation batches, and receipt of all necessary regulatory approvals, we intend to qualify Fresenius Kabi as a second source of Defitelio.

Our wholly-owned subsidiary, Gentium GmbH, together with other subsidiaries of Jazz, commenced the launch of Defitelio in Europe in March 2014, with an initial launch in Germany and Austria. We expect to launch in additional European countries on a rolling basis during 2014 and 2015 and are engaged in pricing and reimbursement submissions as applicable in preparation for planned launches in those countries. We intend eventually to promote Defitelio in all European markets where it has marketing authorization. In addition, we expect to continue to give patients access to defibrotide in countries where it is not commercially available through our expanded access program in the United States and through our named patient program throughout the rest of the world.

We are currently assessing what we believe would be the optimal path for potential approval of defibrotide in the United States, which may include filing a new application with existing clinical data or generating additional clinical data before a new application is ready for submission and FDA review. We are also assessing what other indications, if any, we may pursue for defibrotide, such as acute GvHD.

We have entered into license and/or supply and distribution agreements with specialized regional partners to distribute defibrotide, including on a named patient basis, in the following territories: the Asian Pacific, the Middle East and North Africa, other countries in Europe, the Nordics and Baltics, Turkey, Israel and the Palestinian Authority. We intend to continue to expand our presence into new regional territories through license and/or supply and distribution agreements with additional specialized regional partners. We expect revenue associated with the distribution of defibrotide on a named patient basis to increase as more patients throughout the world gain access to defibrotide. We do not expect a material change to our API business. We plan to incur capital expenditures moving production of urokinase from leased laboratory and manufacturing space to our existing premises and replacing obsolete equipment in

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connection with the relocation. We will need to obtain regulatory approval before we begin to produce urokinase with the new equipment at the new location.

Our ability to achieve our goals and the expected results described above and to implement our strategy is subject to a number of risks and uncertainties, including those discussed in Part I, Item 3 of this annual report in the section entitled "Risk Factors." These risks include, but are not limited to, the uncertainty as to whether Defitelio will become a successful commercial product and our ability to generate projected revenue through our commercial sales in the EU and our continued named patient and expanded access programs, as well as risks and uncertainties common in the biotechnology industry, such as continued government pricing and reimbursement pressures, dependence on corporate partners and key personnel, protection of proprietary technology, compliance with governmental regulations and approval requirements, and potential changes in the health care industry.

RESEARCH AND DEVELOPMENT

We engage multiple third parties, such as CROs and consultants, to assist us in the development of defibrotide. We expense research and development costs as they are incurred.

Research and Development Expenses

Our research and development expenses consist primarily of salaries and benefits of our direct employees, employee stock-based compensation expenses, facility costs, overhead costs, clinical trial costs and related trial product manufacturing costs, contracted services and subcontractor costs. Clinical trial costs include costs associated with CROs. The billings we receive from CROs for services rendered may not be received for several months following the services. Development timelines and costs are difficult to estimate and may vary significantly for each product candidate and from quarter to quarter. We accrue the estimated costs of the CROs' related services based on our estimate of management fees, site management and monitoring costs and data management costs. Differences between estimated and actual trial costs have not been material to date, and any changes have been made when they became known. During the years ended December 31, 2011, 2012 and 2013, we had three major categories of research projects: defibrotide to treat VOD, defibrotide to prevent VOD and assorted other projects. The table below presents our research and development expenses by project (in thousands):

	For The Year Ended December 31,		
	2011	2012	2013
Defibrotide to treat VOD	€3,156	€5,841	€8,241
Defibrotide to prevent VOD	1,583	4,336	188
Other	794	354	7,243
Total	€5,533	€10,531	€15,672

We have completed two clinical trials of defibrotide, a Phase III trial for the treatment of severe VOD with multiple organ failure, conducted in the United States, Canada and Israel, and a Phase II/III pediatric trial conducted in Europe for the prevention of VOD. We also have an ongoing study of defibrotide for the treatment of VOD through our IND protocol. We expect to collect additional data from patients receiving defibrotide through our expanded access and named patient programs.

In order to address issues raised by the FDA, we voluntarily withdrew from consideration an NDA we submitted to the FDA in 2011 seeking approval in the United States for defibrotide for the treatment of VOD. We are currently assessing what we believe would be the optimal path for potential approval of defibrotide in the United States, which may include filing a new application with existing clinical data or generating additional clinical data before a new application is ready for submission and FDA review.

We cannot know when, if ever, defibrotide will be approved in the United States or in any other country or under what circumstances, and what, if any, additional clinical or other development activities will be required in order to potentially obtain such regulatory approval and the cost associated with such required activities if any. If we fail to obtain approval for defibrotide in other countries or for new indications, our anticipated revenue from defibrotide and our growth prospects would be negatively affected. A further discussion of the risks and uncertainties associated with

the development of defibrotide are set forth in Part I, Item 3 of this annual report under the heading “Risk Factors” including the risk factor under the heading “Conducting clinical trials is costly and time-consuming, and the outcomes are uncertain. A failure to prove safety and efficacy in clinical trials would require us to discontinue development, which could materially and adversely affect our business, financial condition, results of operations and growth prospects.”

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Patents

Our commercial success depends in part on obtaining and maintaining patent protection and/or trade secret protection of defibrotide and its use and the methods used to manufacture it, as well as successfully defending these patents against third party challenges, and successfully protecting our trade secrets. Our ability to protect defibrotide from unauthorized making, using, selling, offering to sell or importation by third parties depends on the extent to which we have rights under valid and enforceable patents, or have trade secrets that cover these activities.

The unique process of deriving defibrotide from porcine DNA is extensive and uses both chemical and biological processes that rely on complex characterization methods. We have a portfolio of U.S. and non-U.S. patents and patent applications relating to various compositions of defibrotide, methods of use and methods of characterization, which will expire at various times between April 2017 and June 2032. We intend to continue to seek patent protection for our inventions.

Changes in patent laws could increase the uncertainties and costs surrounding the enforcement or defense of our issued patents. Any patent may be challenged, invalidated, held unenforceable or circumvented. In addition, our patents may not prevent other companies from developing similar or therapeutically equivalent products. Failure to successfully defend a patent challenge could materially and adversely affect our business. We also cannot ensure that others will not be issued patents that may prevent the sale of defibrotide or require licensing and the payment of significant fees or royalties. We may be unable to avoid infringement of third party patents and may have to obtain a license, defend an infringement action, or challenge the validity of the patents in court. If we do not obtain a license under necessary patents, are found liable for infringement, or are not able to have such patents declared invalid, we may be liable for significant money damages or be precluded from the manufacture, use or sale of defibrotide in a manner requiring such licenses.

Trade secrets are difficult to protect. Protracted and costly litigation could be necessary to enforce and determine the scope of our proprietary rights. Courts in certain jurisdictions are sometimes less willing to protect trade secrets. Failure to obtain or maintain patent and trade secret protection, for any reason, could have a material adverse effect on our business.

OFF-BALANCE SHEET ARRANGEMENTS

Not applicable.

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TABULAR DISCLOSURE OF CONTRACTUAL OBLIGATIONS

Contractual Obligations and Commitments

Contractual obligations represent future cash commitments and liabilities under agreements with third parties, and exclude contingent liabilities for which we cannot reasonably predict future payment. The following chart represents our total contractual obligations, at December 31, 2013, aggregated by type (in thousands):

	Total	1 Year	2 Years	3 Years	4 Years	5 Years	Thereafter
Long-term debt obligations:							
Mortgage loans	€1,080	€240	€240	€240	€240	€120	€—
Finance loans	643	97	76	80	84	89	217
Equipment loans	13	13	—	—	—	—	—
	1,736	350	316	320	324	209	217
Operating leases	930	375	185	185	185	—	—
Research and development programs	5,096	4,224	481	364	27	—	—
Selling, general and administrative programs	1,836	1,529	183	74	45	5	—
Manufacturing programs	106	106	—	—	—	—	—
	7,968	6,234	849	623	257	5	—
Total	€9,704	€6,584	€1,165	€943	€581	€214	€217

On June 14, 2006, we obtained a loan in the amount of €2.80 million from Banca Nazionale Del Lavoro S.p.A. The loan is secured by a mortgage on certain of our land and buildings and bears interest at the six-month Euribor rate plus 1.00%. Originally, the principal was repayable in fourteen installments, every six months, from December 27, 2007 until final maturity in 2014, and interest was payable every six months from June 27, 2006. In December 2009 and in June 2011, Banca Nazionale Del Lavoro S.p.A. agreed to defer payment of the loan principal for 48 months, extending the original term of the loan to 2018. At December 31, 2013, the principal amount outstanding under this loan was €1.08 million.

On December 20, 2006, we obtained three loans from Banca Intesa S.p.A. (now Banca Intesasanpaolo S.p.A.).

The first of these loans had an original principal amount of €0.23 million for a term of 60 months, maturing on December 31, 2011. Principal and interest are due in quarterly installments beginning on March 31, 2007. The loan bears interest at the three-month Euribor rate plus 1%. In December 2009 and June 2011, Banca Intesasanpaolo S.p.A. agreed to defer payment of the loan principal for 30 months, extending the term of the loan to 2014. At December 31, 2013, the amount outstanding under this loan was €0.01 million.

The second loan had an original principal amount of €0.50 million for a term of 60 months, maturing on December 31, 2011. Principal and interest are due in monthly installments beginning on January 31, 2007. The loan bears interest at the three-month Euribor rate plus 1%. In December 2009 and June 2011, Banca Intesasanpaolo S.p.A. agreed to defer payment of the loan principal for 30 months, extending the term of the loan to 2014. At December 31, 2013, the amount outstanding under this loan was €0.03 million.

The third loan had an original principal amount of €0.22 million for a term of 57 months (after a technical pre-amortization period from December 20, 2006 to March 15, 2007), maturing on December 15, 2011. Under the terms of the related loan agreement, the proceeds were required to be used within six months of disbursement for investments in the innovation of products and/or production processes or to buy manufacturing equipment. Principal and interest payments are due in quarterly installments beginning on June 15, 2007. The loan bears interest at the three-month Euribor rate plus 0.8%. In December 2009 and June 2011, Banca Intesasanpaolo S.p.A. agreed to defer payment of the loan principal for 30 months, extending the term of the loan to 2014. At December 31, 2013, the amount outstanding under this loan was €0.01 million.

On November 11, 2013, we obtained a loan in the amount of €0.60 million from Banca Popolare di Sondrio for the acquisition and installation of manufacturing equipment. The loan bears interest at the three-month Euribor rate plus 1.25%. Principal and interest payments are due in quarterly installments beginning on April 30, 2014. At December 31, 2013, the principal amount outstanding under this loan was €0.60 million.

Our commitments for clinical research consist of fixed price contracts with third-party research organizations for clinical trials for the development of defibrotide and related consulting services for advice regarding FDA issues.

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The table above does not reflect approximately €39 million in transaction-related expenses we incurred as a result of the closing of the tender offer, including €18 million in investment banker fees and €11 million in payments to selected employees. In addition, under certain circumstances, selected employees of Gentium S.p.A. and its subsidiary Gentium GmbH are entitled to a change of control bonus of approximately €5 million, which may become due in the third and fourth quarters of 2014.

ITEM 6. DIRECTORS, SENIOR MANAGEMENT AND EMPLOYEES

DIRECTORS AND SENIOR MANAGEMENT

Set forth below are the names, ages, positions and a brief account of the business experience of each of our executive officers, significant employees and directors as of March 17, 2014.

Name	Age	Position
Fintan Keegan	54	Chairman and Chief Executive Officer
Salvatore Calabrese	44	Senior Vice President, Finance, Chief Financial Officer and Chief Operating Officer
Adrian Haigh	54	Senior Vice President, Commercial Operations and Chief Operating Officer of Gentium GmbH
Dr. Giorgio Mosconi, M.D., Ph.D.	52	Senior Vice President and Scientific Director
Joyce Victoria Bigio (1)	59	Director
Suzanne Sawochka Hooper	48	Director
Iain McGill	41	Director
Elmar Schnee (2)	55	Director

(1) Member of the audit committee (chairperson).
(2) Member of the audit committee

Fintan Keegan has served as our Chairman of the board of directors and our Chief Executive Officer since January 2014. Mr. Keegan was appointed Executive Vice President, Technical Operations of Jazz Pharmaceuticals plc in July 2012 and served as its Senior Vice President of Technical Operations from January 2012 until July 2012. Prior to January 2012, he was Senior Vice President and Chief Technical Officer of Azur Pharma plc since 2006, where he was responsible for quality, regulatory, compliance, supply chain and development. Prior to his work with Azur Pharma, Mr. Keegan most recently served as Vice President of Quality and Regulatory for Elan Corporation, plc. He also held various positions with Wyeth Pharmaceuticals, Inc., Merck & Co., Inc. and at a clinical contract research organization. Mr. Keegan holds a B.Sc and a H. Dip in Pharmaceutical Manufacturing Technology from Trinity College Dublin and a M.Sc from the School of Chemistry, University of Bristol, in the United Kingdom.

Salvatore Calabrese has served as our Chief Operating Officer since October 2013, Chief Financial Officer since December 2010, Senior Vice President of Finance since February 2010, and our Vice President of Finance since February 2005. From December 2003 until February 2005, he was an Accounting and Finance Manager for Novuspharma, S.p.A., a development stage biopharmaceutical company focused on the discovery and development of cancer drugs and a subsidiary of Cell Therapeutics, Inc., a public reporting company, which then merged into Cell Therapeutics, Inc. From September 1996 until November 2003, Mr. Calabrese was employed by PricewaterhouseCoopers as an accountant and was a Manager in Assurance Business Advisory Services at the time of his departure. From October 2000 to June 2003, Mr. Calabrese worked in the Boston, MA office of PricewaterhouseCoopers. He earned a Bachelors' Degree in Economics at the University of Messina and a Masters' Degree in Accounting, Audit and Financial Control at the University of Pavia. He is also a chartered accountant in the Republic of Italy.

Adrian Haigh has served as our Senior Vice President, Commercial Operations since March 2011 and was additionally appointed Chief Operating Officer of Gentium GmbH, our wholly-owned subsidiary, in June 2013. Prior to joining Gentium, Mr. Haigh was Regional Vice President, Commercial Operations at Biogen Idec Inc., where he

managed several of Biogen's subsidiaries and its global distributor business. From 2002 to 2007, he held leadership positions at Amgen Inc., including General Manager of Portugal and Scandinavia and Head of its International Oncology Franchise. Prior to that, Mr. Haigh held management positions at SmithKline Beecham, Schering-Plough, Organon and Novo-Nordisk. Mr. Haigh received a Bachelor of Arts with Honours in Economic History from Huddersfield Polytechnic, West Yorkshire, England and a Diploma in Marketing from the Institute of Marketing at Maidenhead, United Kingdom.

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Giorgio Mosconi, M.D., Ph.D., has served as our Senior Vice President and Scientific Director since December 2013 and was previously our Vice President, Medical Affairs since June 2013. Dr. Mosconi was President and Chief Business Officer of Formula Pharmaceuticals, Inc., which he co-founded in 2009. Prior to this, he served as Senior Vice President, Business Development, at Vicuron Pharmaceuticals, Inc. (acquired by Pfizer in 2005), the result of a merger between Biosearch Italia and Versicor Inc. Previously, Dr. Mosconi served as Executive Vice President, Medical Affairs and Business Development at Biosearch Italia and as European Executive Director for R&D Anti-infectives and Immunology at Bristol-Myers-Squibb. Dr. Mosconi began his career at Marion Merrell Dow (now Sanofi-Aventis) serving as Italian Medical Leader, Anti-infectives, and then transitioning to European Therapeutic Leader. Over the years, he has been honored with prestigious awards such as the BMS President Award 1999 and in 2010 was named "Italian protagonist in the world" by the Ministry of Foreign Affairs and in 2013 “Cavaliere della Repubblica Italiana.” Dr. Mosconi holds an M.D. from the Medical School University of Milan and a Ph.D. in ENT, head and neck surgery from the University of Milan. He holds a B.A. in chemistry from Instituto Tecnico Industriale Statale at Bergamo, Italy.

Joyce Victoria Bigio has served as a director since May 2013. Ms. Bigio’s experience includes audit, management, governance, restructuring, reporting and consulting in a wide range of industry sectors. Ms. Bigio is the Managing Partner of International Accounting Solutions (IAS), an accounting services practice founded by Ms. Bigio in 2002, which specializes in financial reporting, accounting, bookkeeping, internal controls and strategic information system planning. Since April 2012, Ms. Bigio has served as a member of the board of directors of Fiat S.p.A. She also currently serves on the board of directors of Simmel Difesa S.p.A., the Italian subsidiary of Chemring Plc, Europa Donna – Umberto Veronesi Foundation, and Faraone Business Advisory. From 1998 to 2002, Ms. Bigio served as CFO of Sotheby’s Italy. From 1995 to 1998, she was the general manager of American International Bakeries, a start-up company in the Italian bakery market. From 1989 to 1995, Ms. Bigio worked at Waste Management Group in London, first as head of European reporting, then in its European merger and acquisition department. From 1986 to 1989, she served as controller of Euromobiliare, an investment bank. Previously, Ms. Bigio spent 10 years at Arthur Andersen in the audit divisions in Washington, D.C. and Milan. Ms. Bigio holds a degree in Economics and Commerce from the University of Virginia, with a major in Accounting.

Suzanne Sawochka Hooper has served as a director since January 2014. Ms. Hooper was appointed Executive Vice President and General Counsel of Jazz Pharmaceuticals plc as of March 2012. From 1999 through early 2012, she was a partner in the law firm Cooley LLP. Ms. Hooper served for several years as a member of Cooley’s Management Committee and as Vice Chair of the firm’s Business Department. While at Cooley, Ms. Hooper practiced corporate and securities law, primarily with companies and investors in the life sciences industry. Ms. Hooper received a J.D. from the University of California, Berkeley, Boalt Hall School of Law and a B.A. in Political Science from the University of California, Santa Barbara. Ms. Hooper is a member of the State Bar of California.

Iain McGill has served as a director since January 2014. Mr. McGill joined Jazz Pharmaceuticals plc in June 2012. Immediately prior to joining Jazz Pharmaceuticals plc, Mr. McGill spent three years serving as Chief Commercial Officer at EUSA Pharma (Europe) Ltd. Mr. McGill began his pharmaceutical career in sales, and over the past 20 years has held positions of increasing responsibility in sales management, market research, marketing, business development and general management at Syntex, Roche, Novartis and Wyeth. He has lived and worked in the United Kingdom, Switzerland, Canada and the United States. Mr. McGill has a BSc in Biochemistry from the University of London.

Elmar Schnee has served as a director since May 2012. Mr. Schnee has more than 20 years of experience in the international pharmaceutical industry, with specific expertise in strategic planning, business development and marketing acquired through various positions held at Merck, Fisons Pharmaceutical PLC, Sanofi-Synthélabo and UCB Pharma. Since 2011, Mr. Schnee has served as Executive Chairman of Cardiorentis Ltd., a biopharmaceutical company specializing in the development and commercialization of therapies for the treatment of acute heart failure (AHF). Previously, Mr. Schnee spent several years in the global pharmaceutical and chemical group at Merck KGaA. He joined Merck in 2003 as Managing Director of Merck Santé s.a.s. in Lyon, France. In January 2004, Mr. Schnee assumed responsibility for global operations of the ethical pharmaceuticals division of Merck KGaA, and in

November 2005, Mr. Schnee was appointed as Deputy Member of the Executive Board responsible for the pharmaceuticals business sector. In 2006, he was appointed as Regular Member of the Executive Board and General Partner of Merck KGaA, with responsibility for pharmaceutical products oversight. Mr. Schnee holds a degree in Marketing Management from the Swiss Institute of Business Administration in Zurich.

From January 2013 until our acquisition by a subsidiary of Jazz in January 2014, our board of directors consisted of seven members. On January 1, 2013, our board of directors consisted of Ms. Gigliola Bertoglio, Mr. Marco Brughera, Dr. Glenn Cooper, Dr. Laura Ferro, Dr. Khalid Islam, Dr. Bobby Sandage, Jr., and Mr. Schnee. Dr. Cooper resigned from the board on March 27, 2013. The remaining six directors and Ms. Bigio were elected as members of the board at our 2013 annual ordinary shareholders' meeting held on May 10, 2013 for a term ending with the approval of our financial statements as of December 31, 2013. On January 23, 2014, in connection with our acquisition by a subsidiary of Jazz, Ms. Bertoglio, Dr. Ferro and Dr. Islam resigned from our board of directors and Jazz's designees, Ms. Hooper, Mr. Keegan and Mr. McGill, were appointed to our board of directors. On January 24, 2014, Mr. Brughera and Dr. Sandage resigned from our board of directors. Ms. Bigio and Mr. Schnee have never been employed by us or any of our subsidiaries and are independent directors. Ms.

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Hooper, Mr. Keegan and Mr. McGill are employed by subsidiaries of Jazz. It is expected that Ms. Bigio and Mr. Schnee will resign from our board of directors at the time our shareholders approve our financial statements as of December 31, 2013 prepared in accordance with Italian GAAP.

COMPENSATION

Compensation of Directors and Executive Officers

For the years ended December 31, 2013 and 2012, compensation to our executive officers and directors then in office, excluding amounts due to termination of employment agreements, was €1.61 million and €1.15 million, respectively. Disclosure of compensation for our directors and executive officers is not required on an individual basis unless individual disclosure is otherwise made publicly by us. The following disclosure about individual compensation arrangements was disclosed by us in connection with the tender offer by a subsidiary of Jazz, and so is provided here. Dr. Khalid Islam was our Chairman and Chief Executive Officer until his resignation on January 23, 2014. Mr. Salvatore Calabrese, Dr. Giorgio Mosconi and Mr. Adrian Haigh were our other executive officers in 2013. Each of these executive officers had employment agreements with us. Their employment agreements address their duties and responsibilities and specify the amounts payable to them in connection with certain termination or change in control events. 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, if he resigns following a Change in Control (as defined in the employment agreement), or, in the case of Mr. Calabrese and Mr. Haigh, if either 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 options to purchase our ordinary shares and, in the case of Dr. Islam and Mr. Calabrese, reimbursement of any legal expenses incurred in connection with enforcing payment of amounts due.

Concurrently with our execution of the tender offer agreement with Jazz and Jazz Italy in December 2013, Dr. Islam entered into a transition, amendment and release agreement with his employer, our wholly-owned subsidiary, Gentium GmbH, pursuant to which, among other things, (i) he agreed to resign at the closing of our acquisition by Jazz, and (ii) he would only be eligible to receive certain change in control severance entitlements specified under his employment agreement if he successfully completed his consultancy agreement, as determined by Gentium GmbH in its discretion, and signed a waiver and release in substantially the form provided by Gentium GmbH. Dr. Islam also entered into a consultancy agreement with us whereby he agreed to work as a consultant for us for the six months following the closing of the acquisition. Dr. Islam is currently serving as a consultant to us.

We also entered into a retention and amendment agreement with each of Mr. Calabrese, Dr. Mosconi and Mr. Haigh, whereby, among other things, each executive officer agreed not to resign and to continue working for us 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 would not be entitled to receive certain change in control severance entitlements (and such retention bonus) if his employment was terminated by us during this period for Cause (as defined in the respective employment agreements), or in the case of Mr. Haigh and Mr. Calabrese, he resigned without Good Reason (as defined in the respective employment agreements), prior to the completion of the Retention Period (as defined in the retention and amendment agreements).

On September 27, 2013, our board of directors resolved to adopt a transaction bonus for selected senior managers of our 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 our company, subject to certain terms and conditions to be subsequently defined by our nominating, corporate governance and compensation committee. On September 27, 2013, our nominating, corporate governance and compensation committee 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 our company. Of the transaction bonus, 70% was allocated to Dr. Islam and the remaining 30% was to be allocated to selected senior managers of our company and Gentium GmbH expressly identified by our board of directors following the

recommendation of Dr. Islam. As the premium of the tender offer price over $22.83 exceeded 45%, the applicable transaction bonus is an aggregate amount of 1.5% of the transaction value, or approximately $15.2 million. Dr. Islam proposed to our board of directors on December 23, 2013 that the unallocated 30% should be allocated as follows: 10% of the transaction bonus to Mr. Calabrese, 10% to Mr. Haigh, 5% to Dr. Mosconi, and the remainder to a few other employees. Our board of directors approved such allocation. The transaction bonus and proposed allocations were also approved by a special board committee at meetings held on December 18, 2013 and December 23, 2013. The transaction bonus was paid in January 2014.

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Under Italian law, our shareholders determine director compensation relating to basic board service, such as annual fees for serving on the board and fees for attending board meetings. Historically, our compensation committee (merged into the nominating, corporate governance and compensation committee in May 2013) recommended director compensation to our shareholders and our board of directors. Our shareholders approved the following director compensation for the term started on May 10, 2013, the date of our 2013 annual ordinary shareholders' meeting, and ending with the approval of our financial statements as of December 31, 2013:

- an annual cash retainer of $45,000 for each director; and
- a stock option for 11,500 ordinary shares.

Additionally, the chairperson of our board of directors received an additional annual retainer of $25,000 and the chairs of the following committees of our board of directors receive the following additional annual retainers: (i) audit committee, $25,000; (ii) nominating, corporate governance and compensation committee, $15,000; and (iii) scientific oversight committee, $15,000. The non-chair members of each committee also received an additional annual retainer of $5,000. In addition, directors received reimbursement of their reasonable out-of-pocket expenses in connection with attendance at our board and committee meetings.

During the year ended December 31, 2013, we granted options to purchase an aggregate of 424,000 ordinary shares to our executive officers and directors at exercise prices ranging from $8.04 to $19.33. As of December 31, 2013, our directors and executive officers held options to purchase an aggregate of 1,625,108 ordinary shares. All outstanding and unvested stock option awards became fully vested and exercisable on January 23, 2014 following the expiration of the initial tender offer period. As of February 17, 2014, all outstanding option awards held by our directors and executive officers prior to January 23, 2014 have been exercised and all ordinary shares issued upon such exercise have been subsequently tendered pursuant to the tender offer, as extended by a subsequent offering period.

All of our stock-based compensation plans have been terminated and no options remain outstanding.

Pension and retirement plans

We do not have any pension or retirement plans, other than a 401(k) plan for one U.S. employee which was terminated on January 21, 2014.

BOARD PRACTICES

Board Composition

Our board of directors currently consists of five members: Ms. Bigio, Ms. Hooper, Mr. Keegan, Mr. McGill and Mr. Schnee. All of our directors' terms will expire on the date of our ordinary shareholders' meeting to be called to approve our financial statements for the year ending December 31, 2014.

We do not have any agreements with any of our directors that provide for benefits upon termination of employment, although under Italian law, if directors are removed by the vote of shareholders at an ordinary shareholders' meeting prior to the end of their term, without cause, they may have a claim for damages against us. These damages may include, but are not limited to, compensation that would otherwise have been paid to the director for the remainder of his or her term and damage to his or her reputation.

Board Committees

As of March 17, 2014, our board of directors has one standing committee, the audit committee. Prior to the closing of our acquisition by Jazz, there were two additional committees: a nominating, corporate governance and compensation committee and a scientific oversight committee. Both of the additional committees ceased all activities at the closing of our acquisition by Jazz and were dissolved by our board of directors on March 26, 2014.

Our audit committee currently consists of Ms. Bigio (chairperson) and Mr. Schnee. Each is an independent director. Ms. Bigio is our audit committee financial expert. Before January 23, 2014, our audit committee included Ms. Bertoglio who served as the chairperson, in addition to Ms. Bigio and Mr. Schnee, and Ms. Bertoglio was our audit committee financial expert. Previously in 2013, Dr. Cooper also served on the audit committee until his resignation from our board of directors on March 27, 2013.

The audit committee is a standing committee of, and operates under a written charter adopted by, our board of directors. According to its charter, the audit committee's primary role is to:

- establish procedures for the receipt, retention and treatment of complaints we receive regarding accounting, internal accounting controls or auditing matters and the confidential, anonymous submission by our employees of concerns regarding questionable accounting or auditing matters;

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- engage independent counsel and other advisors, as it deems necessary to carry out its duties, and to determine the compensation of such counsel and advisors, as well as ordinary administrative expenses of the committee; and
- approve related party transactions.

Our audit committee directly oversees our independent accountants and the resolution of any disagreements between management and the independent accountants. As discussed below, under Italian law, our board of statutory auditors also oversees our independent accountants with respect to our Italian GAAP financial statements. Under Italian law, our shareholders must appoint, terminate and determine the compensation for our independent accountants, although our audit committee can and does make recommendations on such matters to our board of directors, which in turn makes recommendations to our shareholders.

Board of Statutory Auditors

Under Italian law, in addition to electing our board of directors, our shareholders also elect a board of statutory auditors. The statutory auditors are elected for a term of three years, may be reelected for successive terms and may be removed only for cause and with the approval of a competent court. Each member of the board of statutory auditors must provide certain evidence that he or she is qualified to act in that capacity under Italian law, and that he or she meets certain professional standards. The board of statutory auditors is required to verify that we comply with applicable law and our articles of association and bylaws, respect the principles of correct administration and maintain an adequate organizational structure, internal controls and administrative and accounting system, and also oversees our independent accountants with respect to our Italian GAAP financial statements.

The following table sets forth the name and position of each of the three members of our board of statutory auditors and the alternate statutory auditors. The board of statutory auditors was elected at our shareholders' meeting held on February 28, 2014 for a term that ends the date of our ordinary shareholders' meeting that shall be called to approve our financial statements for the year ending December 31, 2016.

Name	Position
Mia Pasini	Chairman
Luca La Pietra	Member
Maurizio Pavia	Member
Alberto Demarchi	Alternate
Giovanni Lurani Cernuschi	Alternate

The following table sets forth the name and position of each of the three former members of our board of statutory auditors and the former alternate statutory auditor, who resigned from their respective offices as an active or an alternative member of the board of statutory auditors, effective on February 28, 2014. These former members of our board of statutory auditors were originally elected at a shareholders' meeting held on May 9, 2012.

Name	Position
Giorgio Iacobone	Chairman
Carlo Ciardiello	Member
Augusto Belloni	Member
Putignano Oronzo	Alternate
Domenico Ferrari	Alternate

In 2011, the board of statutory auditors met six times and attended eight board of directors' meetings and one shareholders' meeting. In 2012, the board of statutory auditors met five times and attended 6 board of directors meetings and 1 shareholders' meeting. In 2013, the board of statutory auditors met six times and attended 11 board of directors' meetings and one shareholders' meeting. In 2013, we accrued €64,000 as compensation for the service of members of our board of statutory auditors.

Indemnification of Directors and Executive Officers and Limitation of Liability

We have entered into indemnification agreements with certain of our directors and executive officers, which may, in some cases, provide indemnification that is broader in scope than the specific indemnification provisions of Italian law.

At present, other than the shareholder litigation matter disclosed in Part 1, Item 4 of this annual report in the section entitled “Business Overview—Legal Proceedings,” there is no pending litigation or proceeding involving any of our directors,

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officers, employees, or agents in which indemnification by us will be required or permitted, nor are we aware of any threatened litigation or proceeding that may result in a claim for such indemnification.

We historically maintained directors' and officers' liability insurance, which covered liabilities arising under the Securities Act of 1933, as amended, or the Securities Act. In connection with our acquisition by Jazz, we purchased a five-year "tail" prepaid policy on our directors' and officers' liability insurance in effect prior to the acquisition by Jazz Italy to cover our directors and officers in office before the acquisition. In addition, our current directors and officers are covered by directors' and officers' liability insurance maintained by Jazz.

EMPLOYEES

The table below shows the number, activity and geographic location of our permanent employees as of December 31, 2011, 2012 and 2013. As of December 31, 2013, Gentium S.p.A. had 76 employees, one of whom was based in the United States and the remaining were based in Italy, while Gentium GmbH had 13 employees.

	As of December 31,		
	2011	2012	2013
Administration, accounting, finance, business development	15	21	20
R&D, clinical, regulatory	14	17	15
Sales and marketing	9	7	8
Production, quality assurance control	30	32	33
Total	68	77	76

Italian law imposes certain confidentiality obligations on our employees and provides that we are entitled to either ownership of, or a right of option on, any intellectual property created by our employees while under our employ, although we must compensate our employees for such intellectual property creation. Our employees in Italy are also subject to national collective bargaining agreements. National agreements are negotiated collectively between the national associations of companies within a given industry and the respective national unions. National agreements provide a basic framework of working conditions, including pay, security and other provisions. With the exception of our executive officers in Italy, all of our employees are subject to a collective bargaining agreement that expires on December 31, 2015. Our executive officers in Italy were subject to a collective bargaining agreement that expired on December 31, 2013. Our work force is unionized, and we believe that we maintain satisfactory relations with our employees.

Under Italian law, employees who leave employment for any reason, including termination for cause or resignation, are entitled to a severance payment based on salary and years of service. Our liability for these termination indemnities at December 31, 2013 was €0.29 million. In accordance with Italian law, we make social security and national healthcare contributions to the Italian Government on behalf of our employees, which provides pension and healthcare insurance benefits.

SHARE OWNERSHIP

To our knowledge, none of our directors and officers owned any of our ordinary shares at March 17, 2014. See Part I, Item 7 of this annual report in the section entitled "Major Shareholders." No options or other rights to purchase our shares are outstanding.

ITEM 7. MAJOR SHAREHOLDERS AND RELATED PARTY TRANSACTIONS

MAJOR SHAREHOLDERS

At March 17, 2014, we had 17,741,189 ordinary shares outstanding and Jazz was the indirect beneficial owner of 17,427,624 of our ordinary shares, which represented approximately 98.2% of our ordinary shares outstanding as of that date. Jazz holds beneficial ownership of those ordinary shares through a number of wholly-owned subsidiaries, including Jazz Italy, which acquired beneficial ownership of all of those ordinary shares pursuant to a tender offer conducted through an initial offering period that expired on January 22, 2014 and a subsequent offering period that expired on February 20, 2014. Jazz controls us by virtue of Jazz Italy's ownership of those shares.

Other than our ADS depositary, The Bank of New York Mellon, as of March 17, 2014, there were no record holders of our ordinary shares located in the United States.

Jazz Italy does not have different voting rights than other holders of our ordinary shares.

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RELATED PARTY TRANSACTIONS

Except as described below, since January 1, 2013, we have not entered into or proposed to enter into any transaction or loan with any of our affiliates, directors, executive officers (other than employment agreements), holders of 10% or more of our ordinary shares, immediate family members of such persons, or any enterprise over which any such person is able to exercise a significant influence.

Arrangements in connection with the Tender Offer

Certain of our executive officers and directors may be deemed to have interests in the transactions consummated under the tender offer agreement that are different from or in addition to those of the holders of our company's ordinary shares and ADSs generally. These interests may have created potential conflicts of interest. Our board of directors was aware of these interests and considered them, among other matters, in approving the tender offer agreement and the transactions contemplated thereby. Moreover, our board of directors formed a special committee of independent directors constituted by Mr. Schnee (chairperson), Ms. Bigio and Dr. Sandage to review agreements and arrangements entered into or to be entered into with our directors and employees in connection with or in the context of the tender offer, and compensation proposed to be paid to employees as a result of the tender offer, and the special committee approved such agreements, arrangements and compensation. Consummation of the tender offer constituted a change in control for the purposes of determining the entitlements due to executive officers and directors relating to certain benefits, as described in Part I, Item 6 of this annual report in the section entitled “Compensation—Compensation of Executive Officers.”

In connection with our entry into the tender offer agreement with Jazz and Jazz Italy in December 2013, each of the following executive officers, Dr. Islam, Mr. Calabrese, Dr. Mosconi and Mr. Haigh, each of the then members of our board of directors (other than Mr. Brughera), and F3F S.r.l., the beneficial owner of approximately 16% of our then outstanding ordinary shares and ADSs, entered into support agreements with Jazz, pursuant to which they each agreed, among other things, to tender all of their shares in the tender offer, exercise any options to acquire ordinary shares held by them and tender the underlying ordinary shares in the tender offer, vote their shares to support the tender offer by Jazz, and not solicit or attempt to solicit any current or former employees of our company for a period of 18 months after January 23, 2014.

Control by Dr. Ferro’s Family

Dr. Ferro, who is one of our former directors, together with members of her family, may be deemed to control F3F S.r.l. (formerly known as FinSirton S.p.A.). As a result, Dr. Ferro and her family may be deemed to have had indirect control of approximately 16% of our then outstanding ordinary shares and ADSs in December 2013 when F3F S.r.l. entered into the support agreement with Jazz. F3F S.r.l. tendered all of its shares to Jazz on or before January 20, 2014.

Agreements with Various Entities

On January 1, 2012, we entered into a commercial lease with F3F S.r.l. The area leased is approximately 4,800 square meters in size and is used for offices, manufacturing, laboratories and storage facilities. The lease provides for an annual fee of €185,000 for the initial six-year term, which may be adjusted annually based on the cost of living index, and, in the event we exercise our six-year renewal option, €215,000 on an annual basis, subject to cost of living adjustments.

Sigma-Tau, together with its subsidiaries, may be deemed to have had control of approximately 17% of our then outstanding ordinary shares and ADSs in 2013. Pursuant to our license and supply agreement with Sigma-Tau (as assignee of Sigma-Tau Industrie Farmaceutiche Riunite S.p.A.) dated December 7, 2001, as amended, we have licensed the rights to Sigma-Tau to commercialize defibrotide for the treatment and prevention of VOD in North America, Central America and South America, subject to receipt of marketing authorization, if any, in the applicable territory. Pursuant to the license and supply agreement, between 2001 and 2010, we received milestone and other payments in the amount of $11.35 million and we are entitled to additional payment of $6 million following regulatory approval from the FDA to market defibrotide in the United States, a further $2 million payment following the transfer of the approved NDA to Sigma-Tau, royalty payments equal to 7% of Sigma-Tau’s net sales of defibrotide and a supply price equal to the greater of 31% of net sales or €50 (approximately $68) per unit of defibrotide finished

product. In addition, in connection with the license and supply agreement, we also entered into a cost sharing agreement with Sigma-Tau dated October 12, 2007, under which agreement, as amended, Sigma-Tau agreed to reimburse us 50% of certain costs associated with the development of defibrotide. We also agreed that $1.0 million in costs reimbursed by Sigma-Tau will be deductible from royalty payments that we are entitled to receive in the future under the license and supply agreement. In 2013, we received €1.40 million ($1.80 million) as reimbursement of research and development expenses and in 2014, Sigma-Tau agreed to reimburse us an estimated total of approximately $4.77 million as incurred.

Indemnification Agreements

We have entered into indemnification agreements with certain of our directors and officers, which may require us to indemnify against liabilities that arise by reason of the status of such directors and officers or service as directors or officers and

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may also require us to advance expenses incurred by our directors and officers in connection with any proceeding against them. However, we will not indemnify directors or officers with respect to liabilities arising from willful misconduct of a culpable nature.

INTERESTS OF EXPERTS AND COUNSEL
Not applicable.

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ITEM 8. FINANCIAL INFORMATION

CONSOLIDATED STATEMENTS

Please refer to Part III, Item 18 of this annual report entitled "Consolidated Financial Statements."

OTHER FINANCIAL INFORMATION

Legal Proceedings

As of the date of this report, we are involved in the following legal proceeding:

Shareholder Litigation Matter: In January 2014, we became aware of a purported class action lawsuit filed in the Southern District of New York in connection with the tender offer conducted by Jazz Italy to acquire our ordinary shares and ADSs. The lawsuit, captioned Xavion Jyles, Individually and on Behalf of All Others Similarly Situated v. Gentium S.P.A. et al., names us, each of our directors, Jazz and Jazz Italy as defendants. The lawsuit alleges, among other things, that our directors breached their fiduciary duties to our shareholders in connection with the tender offer agreement that we entered into with the Jazz entities valuing our ordinary shares and ADSs at $57.00 per share, and that the Jazz entities violated Sections 14(e) and 20(a) of the Exchange Act, by allegedly overseeing our preparation of an allegedly false and misleading Section 14D-9 Solicitation/Recommendation Statement. The lawsuit seeks, among other relief, class action status, rescission, and unspecified costs, attorneys' fees and other expenses. We cannot predict the timing or outcome of this matter.

From time to time we are involved in legal proceedings arising in the ordinary course of business. We believe there is no other litigation pending that could have, individually or in the aggregate, a material adverse effect on our results of operations or financial condition.

Dividend Policy

We have never declared or paid any cash dividends on our ordinary shares. We currently intend to retain all available funds to support our operations and to finance the growth and development of our business and therefore do not intend to pay any dividends in the foreseeable future.

SIGNIFICANT CHANGES

Please refer to Note 17 entitled "Subsequent Event" in the Notes to Consolidated Financial Statements of the Gentium S.p.A. Consolidated Financial Statements included in this annual report in response to Item 18, which Note 17 is hereby incorporated herein by reference.

ITEM 9. THE OFFER AND LISTING

OFFER AND LISTING DETAILS

Our ADSs were traded on NASDAQ from May 16, 2006 through March 7, 2014. We filed a Notice of Voluntary Delisting with NASDAQ on March 5, 2014, and trading in our ADSs on NASDAQ was suspended on March 7, 2014. Each ADS represents one ordinary share. Our ADSs are currently traded over-the-counter in the United States, and such market is subject to lack of liquidity.

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The following table sets forth the historical high and low closing prices per ADS reported by NASDAQ for each of the periods indicated.

	Price Range of ADSs	
	High	Low
2009	$3.87	$0.33
2010	$7.20	$1.32
2011	$12.13	$5.65
2012		
First Quarter	$9.20	$5.51
Second Quarter	$9.75	$8.76
Third Quarter	$11.16	$9.13
Fourth Quarter	$12.35	$9.92
Full Year	$12.35	$5.51
2013		
First Quarter	$12.59	$7.74
Second Quarter	$8.90	$7.37
Third Quarter	$27.13	$7.76
Month Ended		
October 31, 2013	$42.22	$26.11
November 30, 2013	$54.22	$40.51
December 31, 2013	$57.90	$50.82
Fourth Quarter	$57.90	$26.11
Full Year	$57.90	$7.37
2014		
Month Ended		
January 31, 2014	$57.09	$56.80
February 28, 2014	$56.98	$56.11
March 31, 2014 (through March 7, 2014)	$57.22	$55.04
First Quarter (through March 7, 2014)	$57.22	$55.04

The closing price of the ADSs on NASDAQ on March 7, 2014 was $55.04.

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Source: Nasdaq Stock Market

PLAN OF DISTRIBUTION
Not applicable.

MARKETS
Neither our ordinary shares nor our ADSs are currently listed or traded on any stock exchange or regulated market.

SELLING SHAREHOLDERS
Not applicable.

DILUTION
Not applicable.

EXPENSES OF THE ISSUE
Not applicable.

ITEM 10. ADDITIONAL INFORMATION

SHARE CAPITAL
Not applicable.

MEMORANDUM AND ARTICLES OF ASSOCIATION
The following is a summary of certain information concerning our ordinary shares, our bylaws (Statuto) and the Italian law provisions applicable to companies whose shares are not listed in a regulated market in the EU, as in effect at the date of this annual report. The summary contains all the information that we consider to be material regarding the shares but does not purport to be complete and is qualified in its entirety by reference to our bylaws and Italian law.
Under Italian law, most of the procedures regulating our company, including certain rights of shareholders, are contained in our bylaws as opposed to our articles of association. Amendments to our bylaws require approval at an extraordinary meeting of shareholders, as described below.
In January 2003, the Italian government approved a wide-ranging reform of the corporate law provisions of the Italian Civil Code, which came into force on January 1, 2004. On September 30, 2004, our shareholders approved a number of amendments to our bylaws, which were dictated or made possible by the 2003 corporate law reform. Our bylaws were further amended on April 28, 2005, November 29, 2005, April 28, 2006, April 27, 2007, June 30, 2009, April 30, 2010, and May 9, 2011. The following summary takes into account the 2003 corporate law reform and the consequent amendments to our bylaws.
General
As of March 26, 2014, our issued and outstanding share capital consisted of 17,741,189 ordinary shares, without a par value. The Euro currency was adopted in Italy on January 1, 2002. The redenomination of the ordinary shares from Italian Lira to Euro was approved by our shareholders on December 27, 2000. All the issued and outstanding shares are fully paid, non-assessable and in registered form.
We are registered with the Companies' Registry of Como. Our registered offices are located in Piazza XX Settembre n. 2, Comune di Villa Guardia, frazione Civello, Como, Italy, registration number 02098100130.
Our corporate purpose is the manufacturing, on behalf of our company and third parties, and marketing in both Italy and other countries, of pharmaceutical preparations, pharmaceutical products, raw materials for pharmaceutical and para-pharmaceutical use and in general all and any products sold by pharmacies or for hospital use, excluding, in all cases, the retail sale in Italy of pharmaceutical preparations and products, medical articles and clinical apparatuses in general and organic and inorganic products that may be used in agrotechnical and/or zootechnical fields. We may also prepare and organize for our own account, or on behalf of third parties, the documentation required for obtaining authorizations for marketing pharmaceutical products in compliance with the regulations in force in the countries of destination and be the holders of those authorizations. We may grant and/or transfer licenses to Italian and foreign enterprises or corporate bodies or acquire licenses for ourselves or third parties. For each product contemplated by our corporate purposes, we may carry out research programs in general and in particular technological, chemical, pharmacotoxicological and clinical research programs in the hospital and pharmaceuticals field. We are generally authorized to engage in any commercial transactions necessary or useful to achieve our corporate purpose, with the exclusion of investment services and other financial or professional activities reserved by Italian law for authorized entities.

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Authorization of shares

Our shareholders may authorize the issuance of additional shares at any time at an extraordinary shareholders' meeting. However, the newly issued shares may not be purchased before all the outstanding shares (i.e., the shares already subscribed) are entirely paid for. On September 30, 2004, following a recommendation by our board of directors, our shareholders approved a capital increase to allow for the issuance of:

- up to 1,560,000 ordinary shares available for grant under our plans;
- up to 1,335,000 ordinary shares upon the conversion of our Series A senior convertible promissory notes;
- up to 881,100 ordinary shares upon the exercise of the warrants; and
- 4,554,000 ordinary shares, including the shares underlying the ADSs in our initial public offering (including ordinary shares underlying the underwriters' purchase option and the over-allotment option).

The authorization for the issuance of ordinary shares authorized at this meeting expired on September 30, 2009, except that the authorization of the issuance of the 1,560,000 shares available for grant under our share option plans would have been valid until September 30, 2019, except that all our outstanding and unvested stock option awards became fully vested and exercisable on January 23, 2014 and were exercised as of February 17, 2014, and subsequently all of our share-based compensation plans were terminated and no options remain outstanding. In addition, 1,353,297 of these ordinary shares were authorized for issuance in connection with our issuance of the Series A notes and related warrants, but were not actually issued, and so became unauthorized and unissuable under Italian law.

On November 29, 2005, after a recommendation by our board of directors, our shareholders approved a capital increase of 713,518 ordinary shares to be reserved for issuance upon exercise of the warrants we issued to the participants in our October 2005 private placement and the placement agent for that private placement.

On April 28, 2006, following a recommendation by our board of directors, our shareholders approved an amendment to our bylaws, which granted certain powers to the board of directors for a five-year period, pursuant to articles 2443 and 2420-ter of the Italian Civil Code, including the power to:

- increase the capital of our company in cash, up to €90 million of par value, in one or more transactions, and to reserve all or part of such amount for the exercise of warrants issued by means of the same resolution of our board of directors providing for the relevant capital increase;
- issue convertible bonds (including subordinated) and increase the capital of our company, in one or more transactions, up to €10 million of par value, through the issuance of ordinary shares reserved for the conversion of such convertible bonds, and to reserve all or part of such convertible bonds for issuance upon the exercise of warrants issued by means of the same resolution of our board of directors providing for issuance of the convertible bonds; and
- in each case, exclude or limit the option right of our shareholders in favor of "strategic investors" (as defined by our bylaws) if our board of directors determines that exclusion or limitation to be in the interest of our company.

Such delegation of powers was renewed by our shareholders at the meeting held on May 9, 2011.

On May 31, 2006, pursuant to the board powers granted by the shareholders at the meeting of April 28, 2006, our board of directors resolved upon a capital increase of 466,446 ordinary shares, to be reserved for issuance upon exercise of warrants. On December 15, 2006, pursuant to the powers granted by the shareholders at the meeting dated April 28, 2006, our board of directors resolved upon a capital increase of 151,200 ordinary shares to be reserved for issuance upon exercise of warrants.

On February 6, 2007, pursuant to the powers granted by the shareholders at the meeting dated April 28, 2006, our board of directors resolved upon a capital increase of 2,354,000 ordinary shares to be subscribed within March 9, 2007, by "strategic investors."

On April 27, 2007, following a recommendation by our board of directors, our shareholders approved a capital increase relating to 1,000,000 ordinary shares to be reserved for issuance pursuant to exercise of options available for grant under our 2007 Stock Option Plan.

On June 30, 2009, our shareholders resolved to (i) remove the par value of our ordinary shares, including the par value of the ordinary shares previously issued by the company, and (ii) grant the board of directors the power to increase the capital in cash up to an amount equal to €100 million on a separable basis, in one or more transactions, for a rights

offering, through the issuance of up to a maximum of 120,000,000 shares, without par value, with the faculty to reserve all or part of such amount to the exercise of warrants issued by means of the same resolution of the board of directors approving the relevant capital increase, and with the faculty to reserve one quarter of any such capital increase to employees as equity incentive under our equity incentive plans in effect from time to time.

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On April 30, 2010, our shareholders resolved to update the text of article 6 of our bylaws as a consequence of the completion of certain capital increases and the expiration of the term for the subscription of certain other capital increases.

On May 9, 2011, our shareholders resolved to increase our capital in cash, by a maximum amount of €2.2 million, on a separable basis, with the exclusion of the preemptive right of the shareholders, for the issuance of options to purchase a maximum of 2,200,000 shares, without a par value, in favor of the Company's employees, directors and consultants.

In addition, on May 9, 2011, following a recommendation by our board of directors, our shareholders approved an amendment to our bylaws which granted certain powers to the board of directors for a five-year period, pursuant to articles 2443 and 2420-ter of the Italian Civil Code, including the powers to:

- increase the capital of our company in cash, up to €90 million of par value, in one or more transactions, and to reserve all or part of such amount for the exercise of warrants issued by means of the same resolution of our board of directors providing for the relevant capital increase;
- issue convertible bonds (including subordinated) and increase the capital of our company, in one or more transactions, up to €10 million of par value, through the issuance of ordinary shares reserved for the conversion of such convertible bonds, and to reserve all or part of such convertible bonds for issuance upon the exercise of warrants issued by means of the same resolution of our board of directors providing for issuance of the convertible bonds; and
- in each case, exclude or limit the option right of our shareholders in favor of "strategic investors" (as defined by our bylaws) if our board of directors determines that exclusion or limitation to be in the interest of our company.

Form and transfer of shares

Our ordinary shares are not represented by share certificates; rather, they are registered in book-entry form. All of our ordinary shares are issued through Monte Titoli, an Italian clearinghouse and depositary, and held through various participants, primarily financial institutions, on Monte Titoli's system. Transfers in our ordinary shares are processed on Monte Titoli's system. We update our shareholder book (libro soci) that we keep at our corporate offices for Italian law purposes from time to time, with the names of the record shareholders based on information provided to us by Monte Titoli participants.

This shareholder book is the controlling register of our record shareholders for Italian law purposes, including the purposes of establishing the record shareholders for shareholder meetings and declaring dividends and stock splits or a combination of the two. A shareholders' name must be entered in this shareholder book in order for the shareholder to establish its rights against us.

There are no limitations on the right to own or vote our ordinary shares, which applies to non-Italian residents and foreign residents. However, owners of our ordinary shares must establish an account with a Monte Titoli participant. There are no provisions in our articles of association or bylaws that would have the effect of delaying, deferring or preventing a change of control of our company and that would operate only with respect to a merger, acquisition or corporate restructuring involving our company. There are no provisions in our bylaws governing the ownership threshold above, which shareholder ownership must be disclosed. There are no provisions discriminating against any existing or prospective holder of our ordinary shares as a result of such shareholder owning a substantial number of our shares. There are no sinking fund provisions or provisions providing for liability for further capital calls by our company.

Dividend rights

Payment of any annual dividend must be proposed by our board of directors to the shareholders and approved by our shareholders at the annual ordinary shareholders' meeting. Before dividends may be paid out of our unconsolidated net income in any year, we must allocate an amount equal to 5% of the Italian GAAP net income to our legal reserve until such reserve is at least equal to 20% of our capital. If a loss in our capital occurs, we may not pay dividends until the capital is reconstituted or reduced by the amount of such losses. We may pay dividends out of available retained earnings from prior years, provided that after such payment, we will have a legal reserve at least equal to the legally required minimum of 20% of the capital. We may not approve or pay dividends until this minimum (i.e., 20% of the

capital) is met. If the minimum is met, the board of directors could propose the issuance of a dividend to the shareholders and the shareholders' resolution might approve that issuance. The shareholders' resolution will specify the manner and the date for dividend payment. Any dividends which shareholders do not collect within five years of the date on which they become payable will be forfeited by those shareholders and come back to us. The board of directors may not approve interim dividends at times between our annual ordinary shareholders' meetings.

Board of directors

Pursuant to our bylaws, our board of directors must consist of no less than three and no more than eleven individuals. Our board of directors is elected at an ordinary shareholders' meeting and the term of board membership is one year. Our directors, who may but are not required to be shareholders, may be reelected. Directors do not stand for reelection at staggered intervals.

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Cumulative voting rights are not permitted or required. There are no provisions in our articles of association or bylaws regarding retirement or non-retirement of our directors under an age limit requirement.

Our board of directors has complete power over our ordinary and extraordinary administration and, in particular, may perform all acts it deems advisable for the achievement of our corporate purposes, except for the actions reserved, by applicable law or the bylaws, to a vote of the shareholders at an ordinary or extraordinary shareholders' meeting. See also, Part I, Item 10 of this annual report in the section entitled "Memorandum and Articles of Association—Meetings of Shareholders."

If we cannot repay our creditors, and a court determines that our directors did not perform their duties regarding the preservation of our assets, the court may find our directors liable to our creditors.

Our board of directors may also appoint one or more senior managers (direttori generali) who report directly to the board. These senior managers may be employees, and the board may delegate certain powers to senior managers that the board has not already delegated to managing directors or an executive committee, subject to the limitations discussed below.

Under Italian law, our board of directors may not delegate certain responsibilities, including the preparation and approval of draft financial statements, the approval of merger and de-merger plans to be presented to shareholders' meetings, increases in the amount of our share capital or the issuance of convertible debentures (if any such power has been delegated to our board of directors by our shareholders at an extraordinary shareholders' meeting) and the fulfillment of the formalities required when our capital is required to be reduced as a result of accumulated losses that affect our stated capital by more than one third. See also, Part I, Item 10 of this annual report in the section entitled "Memorandum and Articles of Association—Meetings of Shareholders."

Meetings of our board of directors are called at least three days in advance or, in case of urgency, at least one day in advance. Statutory auditors are normally required to attend our board meetings, but if a meeting has been duly called, the board can validly take action at the meeting even if the board of statutory auditors does not attend. If the meeting has not been duly called, the meeting is nevertheless validly constituted if all of the directors in office and all of the statutory auditors are in attendance. The chairman may call meetings on his own initiative and meetings must be called upon the request of two directors.

Meetings of our board of directors may be held in person, or by audio-conference or video-conference, in any member state of the EU or in the United States. The quorum for meetings of our board of directors is the attendance of the majority of the directors in office. Resolutions are adopted by the vote of the majority of the directors in attendance at a meeting at which a quorum is present.

Under Italian law, directors having any interest in a proposed transaction must disclose their interest to the board and to the statutory auditors, even if such interest is not in conflict with our interest in the same transaction. The interested director is not required to abstain from voting on the resolution approving the transaction, but the resolution must state explicitly the reasons for, and the benefit to us of, the approved transaction. If these provisions are not complied with, or if the transaction would not have been approved without the vote of the interested director, the resolution may be challenged by a director or by our board of statutory auditors if the approved transaction may be prejudicial to us. A managing director, a member of the executive committee or any senior manager having any interest in a proposed transaction that he or she has authority to approve must solicit prior board approval of such transaction. The interested director or senior manager may be held liable for damages to us resulting from a resolution adopted in breach of the above rules. Finally, directors may be held liable for damages to us if they illicitly profit from insider information or corporate opportunities.

Under Italian law, directors may be removed from office at any time by the vote of shareholders at an ordinary shareholders' meeting although, if removed in circumstances where there was no just cause, such directors may have a claim for damages against us. These damages may include, but are not limited to, compensation that would otherwise have been paid to the director for the remainder of his or her term and damage to his or her reputation. Directors may resign at any time by written notice to our board of directors and to the chairman of our board of statutory auditors.

Our board of directors must appoint substitute directors to fill vacancies arising from removals or resignations, subject to the approval of the board of statutory auditors, to serve until the next ordinary shareholders' meeting. If, at any time,

more than half of the members of our board of directors resign or otherwise cease to be directors, the board of directors in its entirety ceases to be in office and our board of statutory auditors must promptly call an ordinary shareholders' meeting to appoint new directors.

Historically, our compensation committee (merged into the nominating, corporate governance and compensation committee in May 2013) recommends the compensation of our directors to our board of directors, which in turn makes recommendations to our shareholders. Under Italian law, our shareholders determine the compensation of our directors relating to basic board service, such as annual fees for serving on the board and/or fees for attending board meetings. Our board of directors, after consultation with our board of statutory auditors, may determine the remuneration of directors that serve on the various board committees and/or perform management or other special services for us, such as managing directors. Our directors are entitled to reimbursement for expenses incurred in connection with their service as directors, such as expenses

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incurred in travel to attend board meetings. Our articles of association and bylaws do not contain any provisions with respect to borrowing powers exercisable by our directors.

Effective January 1, 2004, an Italian share corporation may adopt one of three different models of corporate governance structure. The three models are:

- a board of directors and a board of statutory auditors, which is the historical model that all companies had prior to January 1, 2004;
- a one-tier model with a single board of directors, including an audit committee composed of independent non-executive directors; or
- a two-tier model, including a management board, which is entrusted with management responsibilities, and a supervisory board which is entrusted mainly with control and supervisory responsibilities and, among other functions, appoints and removes the members of the management board and approves our annual financial statements.

Replacing the historical model with the new one-tier model or two-tier model requires approval at an extraordinary shareholders' meeting. The amended bylaws approved by our shareholders on September 30, 2004 do not provide for a change in our governance structure. As a result, we continue to have a board of directors and a board of statutory auditors.

Statutory auditors

Under Italian law, at least one effective statutory auditor and one alternate statutory auditor of a company shall be chosen among those registered with the Register of Auditors established with the Ministry of Justice. The other statutory auditors shall be chosen among those registered with any register established by decree of the Ministry of Justice or among University professors in economic and law matters, if they are not registered with the Register of Auditors. The following persons may not be appointed as statutory auditors:

- one who is legally incapacitated, bankrupt, or disqualified from holding public or an executive office under Italian law;
- a spouse, parent or relative-in-law of someone who is a director of the company, a director of a company that controls the company, or a director of a company that is under common control with the company; and
- one whose independence may be jeopardized due to an employment or consultant relationship or any other economic relationship with the company, a company that controls the company, or a company that is under common control with the company.

In addition to electing our board of directors, our shareholders elect the board of statutory auditors (Collegio Sindacale) from individuals qualified to act in such capacity under Italian law. At our ordinary shareholders' meetings, the statutory auditors are elected for a term of three fiscal years, they may be re-elected for successive terms and may be removed only for cause and with the approval of a competent court. Each member of our board of statutory auditors must provide evidence that such individual is qualified to act in such capacity under Italian law and meets certain professional standards.

Our bylaws currently provide that the board of statutory auditors shall consist of three effective statutory auditors and two alternate statutory auditors (who will automatically replace a statutory auditor who resigns or is otherwise unable to serve).

Our board of statutory auditors is required, among other things, to verify that we:

- comply with applicable laws and our bylaws;
- respect principles of good governance; and
- maintain adequate organizational structure, internal controls and administrative and accounting systems.

Our board of statutory auditors is required to meet at least once every ninety days. In addition, our statutory auditors are supposed to attend meetings of our board of directors and meetings of our shareholders. In case a statutory auditor, without just cause, does not attend the shareholders' meetings or does not attend two consecutive meetings of the board of directors during the same fiscal year, such statutory auditor shall cease from his/her office. If the statutory auditors do not attend two consecutive meetings of the board of directors or shareholders, they may be terminated for cause by the shareholders. Our statutory auditors may decide to call a meeting of our shareholders, ask for information about our management from our directors, carry out inspections and verifications at our offices and exchange information

with our external auditors. Any shareholder may submit a complaint to our board of statutory auditors regarding facts that the shareholder believes should be subject to scrutiny by our board of statutory auditors, which must take any complaint into account in its report to the shareholders' meeting. If shareholders collectively representing 5% of our share capital submit such a complaint, our board of statutory auditors must promptly undertake an investigation and present its findings and any recommendations to a shareholders' meeting (which must be convened immediately if the complaint appears to have a reasonable basis and there is an

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urgent need to take action). Our board of statutory auditors may report serious breaches of directors' duties to a competent court. The court may take such actions as it feels appropriate, including inspecting our company's operations, removing directors, appointing temporary administrators to manage our company and any other actions that the court feels is necessary to preserve the value of our company for our creditors and shareholders.

External auditor

Italian law requires us to appoint an external auditor or a firm of external auditors ("revisore legale dei conti"), each of them qualified to act in such capacity under Italian law, that shall verify during the fiscal year that our accounting records are correctly kept and accurately reflect our activities, and that our consolidated financial statements correspond to the accounting records and the verifications conducted by the external auditors and comply with applicable rules. The external auditor or the firm of external auditors expresses its opinion on the consolidated financial statements in a report that may be reviewed by the shareholders at our offices prior to the annual shareholders' meeting. The report remains on file at our offices and may be reviewed after the annual shareholders' meeting as well; it is also published for review by the general public.

The external auditor or the firm of external auditors is appointed for a three-year term by the vote of our shareholders at an ordinary shareholders' meeting. At the ordinary shareholders' meeting, the shareholders may ask questions of the board of statutory auditors about its view of the auditors prior to voting on whether to appoint the auditors. Once appointed, the shareholders may remove the auditors only for cause and with the approval of the board of statutory auditors and of a competent court.

Meetings of shareholders

Shareholders are entitled to attend and vote at ordinary and extraordinary shareholders' meetings. Votes may be cast personally or by proxy. Shareholders' meetings may be called by our board of directors (or, in certain cases, by the board of statutory auditors) and must be called if requested by holders of at least 10% of the issued shares. Shareholders are not entitled to request that a meeting of shareholders be convened to vote on issues which as a matter of law shall be resolved upon the basis of a proposal, plan or report by our board of directors. If the shareholders' meeting is not called despite the shareholders' request and such refusal is unjustified, a competent court may call the meeting.

We may hold meetings of shareholders at our registered office in Villa Guardia, or elsewhere within Italy, the EU or the United States following publication of notice of the meeting in the "Gazzetta Ufficiale della Repubblica Italiana" or in the newspaper "Il Sole 24 Ore" at least 15 days before the date fixed for the meeting. Our bylaws provide that we must mail written notice of meetings to our shareholders at least 10 days before the date fixed for the meeting. The notice of a shareholders' meeting must specify two meeting dates for an ordinary or extraordinary shareholders' meeting (first and second "calls"). The notice of the shareholders' meeting also specifies the dates for further calls. The notice must contain a list of the items to be dealt with and state the day, hour and place for the meeting for both the first and second calls. However, if the above procedures are not complied with, the shareholders' meeting will still be deemed validly held if all outstanding shares are represented, all other holders having the right to vote are present and a majority of the board of directors and the board of statutory auditors are in attendance.

We must convene an ordinary shareholders' meeting at least once a year within 120 days following the end of the fiscal year. Our annual Italian GAAP financial statements must be approved by a vote of our shareholders at this annual ordinary shareholders' meeting. We may delay holding the shareholders' meeting up to 180 days following the end of the fiscal year if we are required to prepare consolidated financial statements or if particular circumstances concerning our structure or our purposes so require. At ordinary shareholders' meetings, our shareholders also appoint the external auditors, approve any distribution of dividends that have been proposed by our board of directors, elect our board of directors and statutory auditors, determine their remuneration and vote on any business matter for which resolution or authorization is entrusted to the shareholders by law.

We may call extraordinary shareholders' meetings to vote upon split-ups, dissolutions, appointment of receivers and similar extraordinary actions. We may also call extraordinary shareholders' meetings to vote upon proposed amendments to our bylaws, issuance of convertible debentures, mergers and de-mergers and capital increases and reductions, if the actions may not be authorized by the board of directors. The board of directors has the authority to

transfer our registered office within Italy, authorize, on a non-exclusive basis, amendments to our bylaws that are required by law, authorize mergers by absorption to our subsidiaries in which we hold all or at least 90% of the issued share capital, authorize reductions of our share capital in case of withdrawal of a shareholder and indicate who among the directors is our legal representative. If the shareholders authorize the issuance of shares or other securities at an extraordinary meeting, they may delegate the power to make specific issuances to the board of directors.

Once our shareholders have authorized the issuance of securities, the securities that have been subscribed must be fully paid for before the shareholders may authorize the issuance of additional securities, unless the shareholders meet and vote to cancel those authorized but unsubscribed securities.

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The quorum for an ordinary meeting of our shareholders on the first call is at least 50% of the outstanding ordinary shares, while on second call there is no quorum requirement. In either case, resolutions are adopted by the majority of ordinary shares in attendance or represented at the meeting. The quorum for an extraordinary shareholders' meeting is more than half of the outstanding ordinary shares on the first call and more than one-third of the outstanding shares on second call. Resolutions are adopted by the majority of the outstanding ordinary shares on first call and at least two-thirds of the holders of shares in attendance or represented at the meeting on second call. In addition, certain matters (such as, for example, a change in our purpose, the transfer of our registered office outside Italy or our liquidation prior to the date set forth in our bylaws) must be adopted by shareholders representing more than one-third of the outstanding ordinary shares (not just the ordinary shares in attendance or represented at the meeting).
Shareholders are entitled to one vote per ordinary share. Neither Italian law nor our bylaws limit the right of non-resident or foreign owners to hold or vote their shares. Shareholders do not need to "lodge" their share certificates (if any) or any communication from their broker in order to take part in the meeting.
Shareholders may appoint attorneys-in-fact by delivering in writing the proxies to represent them in an ordinary or extraordinary shareholders' meeting. Our directors, auditors and employees may not be proxies. Italian law provides that no proxy may represent more than 20 shareholders prior to the company "making recourse to the risk capital market." Italian scholars are undecided as to whether listing shares on an exchange outside of the EU constitutes "making recourse to the risk capital market for the purpose of the application of the Italian Civil Code." If we are deemed to make recourse to the risk capital market by means of listing ADSs representing our ordinary shares on NASDAQ, no proxy may represent more than 50 shareholders if the capital is equal to €5 million or less, and no proxy may represent more than 100 shareholders if the capital is more than €5 million but less than or equal to €25 million. If the capital is more than €25 million, no proxy may represent more than 200 shareholders.
Preemptive rights
Pursuant to Italian law, holders of outstanding ordinary shares and convertible debentures are entitled to subscribe for issuance of ordinary shares or convertible debentures in proportion to their holdings at the time that the shareholders authorize the capital increase for those issuances, unless those issuances are for non-cash considerations. The preemptive rights may be excluded or limited by resolution of the shareholders' adopted by the affirmative vote of holders of more than 50% of the ordinary shares at an extraordinary meeting of shareholders, or by a board of directors if the bylaws delegate such power to the board of directors (including the power to exclude or limit the preemptive right), and provided that such exclusion or limitation is in the interest of the company.
F3F S.r.l. (formerly known as FinSirton S.p.A.) waived its preemptive right in connection with the authorization of our private placement of the Series A notes and warrants, the issuance of options under our share option plans and the issuance of 4,554,000 additional ordinary shares, which includes the shares underlying the ADSs offered in our initial public offering and the shares issued in our October 2005 private placement. Our shareholders waived their preemptive rights in connection with the authorization of 713,518 ordinary shares to be reserved for issuance upon exercise of the warrants we issued to the participants in our October 2005 private placement and the placement agent for that private placement.
Our board of directors excluded the shareholders' preemptive rights in connection with the authorization of 1,943,525 ordinary shares and 466,446 ordinary shares to be reserved for issuance of the warrants we issued to the participants in our June 2006 private placement. Our board of directors also excluded the shareholders' preemptive rights in connection with the authorization of 2,354,000 ordinary shares we issued to the participants in our February 2007 private placement. Our shareholders waived their preemptive rights in connection with the authorization of 1,000,000 ordinary shares to be reserved for issuance upon exercise of options available for grant under one of our share option plans.
Preference shares; other securities
Italian law permits us to issue preference shares with limited voting rights, other classes of equity securities with different economic and voting rights, "participation certificates" with limited economic and voting rights, as well as "tracking shares," if our bylaws permit such issuance. Our bylaws currently allow us to issue these securities. We may also issue convertible and non-convertible debt securities. In order to issue convertible debt securities, our board of

directors would need to recommend to our shareholders that they approve the issuance of particular securities in connection with a capital increase, and the shareholders would need to vote to approve such an issuance and capital increase at an extraordinary meeting. The board of directors would also need to recommend, and the shareholders would need to approve by vote at the extraordinary meeting, specific terms of the securities. The shareholders may vote at the extraordinary shareholders' meeting to delegate authority to the board of directors to issue those securities from time to time, but not for more than five years from the date of the extraordinary shareholders' meeting.

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Debt-equity ratio

Italian law provides that we may not issue debt securities for an amount exceeding twice the value of our capital, our legal reserve and any other disposable reserves appearing on our latest Italian balance sheet approved by our shareholders. The legal reserve is a reserve to which we allocate 5% of our Italian GAAP net income each year until it equals at least 20% of our capital. One of the other reserves that we maintain on our balance sheet is a "share premium reserve", meaning amounts paid for our ordinary shares in excess of the amount of such ordinary shares that is allocated to the capital. Until our outstanding debt securities are repaid in full, we may not voluntarily reduce our capital or distribute our reserves (such as by declaring dividends) in the event the aggregate of the capital and reserves, following such reduction of capital and/or distribution of reserves, is less than half of the outstanding amount of the debt securities. If our equity is reduced by losses or otherwise such that the amount of the outstanding debt securities is more than twice the amount of our equity, we cannot distribute profits to our shareholders until the ratio between the amount of our debt securities and our capital and reserves is restored. Moreover, some legal scholars are of the opinion that in such a case the ratio must be restored by a recapitalization of our company. If our equity is reduced, we could recapitalize by means of issuing new shares or having our current shareholders contribute additional capital to our company, although there can be no assurance that we would be able to find purchasers for new shares or that any of our current shareholders would be willing to contribute additional capital. These laws regarding the ratio of debt securities to capital and reserves do not apply to issuances of debt securities to professional investors (as defined by Italian law). However, in such a case, should the professional investors transfer such debt securities to third parties not qualified as professional investors, the former remain liable to us for the payment of such securities.

Reduction of equity by losses

Italian law requires us to reduce our shareholders' equity in certain situations. Our shareholders' equity has three main components: capital, legal reserves and other shareholders' equity (such as any share premium and any retained earnings). We first apply our losses from operations against our shareholders' equity other than legal reserves and capital. If additional losses remain, or if we have no shareholders' equity other than legal reserves and capital, and the additional losses are more than one-third of the amount of our legal reserves and capital, our board of directors must call a shareholders' meeting as soon as possible. The shareholders should take appropriate measures, which may include, inter alia, either reducing the legal reserves and capital by the amount of the remaining losses, or carrying the losses forward for up to one year. If the shareholders vote to elect to carry the losses forward up to one year, and the losses are still more than one-third of the amount of the capital at the end of the year, then we must reduce our capital by the amount of the losses. However, as an S.p.A., we must maintain capital of at least €120,000. If the amount of the losses would reduce our capital to less than €120,000, then:

- we would need to increase our capital, which we could do by issuing new shares or having our shareholders contribute additional capital to our company, although there can be no assurance that we would be able to find purchasers for new shares or that any of our current shareholders would be willing to contribute additional capital;
- our shareholders would need to convert our company to an "S.r.l.", which has a lower capital requirement of €10,000; or
- if neither of these options were taken, our shareholders or, if they do not so resolve, a court of competent jurisdiction, could appoint a liquidator, not necessarily an Italian citizen, to liquidate our company.

Segregation of assets and proceeds

Pursuant to Italian law, our board of directors may resolve to segregate our assets into one or more separate pools. Such pools of assets may have an aggregate value not exceeding 10% of the net worth of the company. Each pool of assets must be used exclusively for the carrying out of a specific business and may not be attached by our general creditors. Similarly, creditors with respect to such specific business may only attach those assets that are included in the corresponding pool. Tort creditors, on the other hand, may always attach any of our assets. Our board of directors may authorize us to issue securities carrying economic and administrative rights relating to a pool. In addition, financing agreements relating to the funding of a specific business may provide that the proceeds of such business be used exclusively to repay the financing. Such proceeds may be attached only by the financing party and such financing party would have no recourse against other assets of ours.

We have no present intention to enter into any such transaction and no such transaction is currently in effect.

Liquidation rights

Pursuant to Italian law and subject to the satisfaction of the claims of all creditors, our shareholders are entitled to a distribution in liquidation that is equal to an amount resulting from the division of the positive liquidation balance by the number of shares (to the extent available out of our net assets). Preferred shareholders and holders of "participating certificates" typically do not participate in the distribution of assets of a dissolved corporation beyond their established contractual preferences. Once the rights of preferred shareholders and holders of participating certificates and the claims of all creditors have been fully satisfied, holders of ordinary shares are entitled to the distribution of any remaining assets.

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Purchase of shares by us

We are permitted to purchase our outstanding shares, subject to certain conditions and limitations provided for by Italian law. We may only purchase the shares out of profits available for dividends or out of distributable reserves, in each case as appearing on the latest shareholder-approved consolidated financial statements. Further, we may only repurchase fully paid-in shares. Such purchases must be authorized by our shareholders by vote at an ordinary shareholders' meeting and the authorization may be issued for a period not exceed the term of eighteen months.

A corresponding reserve equal to the purchase price of such shares must be created in the balance sheet, and such reserve is not available for distribution, unless such shares are sold or canceled. Shares purchased and held by us may be resold only pursuant to a resolution of our shareholders adopted at an ordinary shareholders' meeting. The voting rights attaching to the shares held by us or our subsidiaries cannot be exercised, but the shares can be counted for quorum purposes in shareholders' meetings. Dividends and other rights, including preemptive rights, attaching to such shares will accrue to the benefit of other shareholders.

Notification of the acquisition of shares

In accordance with Italian antitrust laws, the Italian Antitrust Authority is required to prohibit the acquisition of control in a company which would thereby create or strengthen a dominant position in the domestic market or a significant part thereof and which would result in the elimination or substantial reduction, on a lasting basis, of competition, provided that certain turnover thresholds are exceeded. However, if the turnover of the acquiring party and the company to be acquired exceed certain other monetary thresholds, the antitrust review of the acquisition falls within the exclusive jurisdiction of the European Commission.

Minority shareholders' rights; withdrawal rights

Shareholders' resolutions which are not adopted in conformity with applicable law or our bylaws may be challenged (with certain limitations and exceptions) within ninety days by absent, dissenting or abstaining shareholders representing individually or in the aggregate at least 5% of our share capital (as well as by our board of directors or our board of statutory auditors). Shareholders not reaching this threshold or shareholders not entitled to vote at our meetings may only claim damages arising from the resolution.

Dissenting or absent shareholders may withdraw from the company as a result of shareholders' resolutions approving, among others things, material modifications of our purpose or of the voting rights of our ordinary shares, our transformation from a share corporation into a different legal entity or the transfer of our registered seat outside Italy. In such a case, our other shareholders would have a preemptive right to purchase the shares of the withdrawing shareholder. Should no shareholder exercise that preemptive right, the shares must be offered to third parties or, in the absence of any third party wishing to buy them, they will be purchased by us by using the available reserves. In the event that no reserve is available, our capital must be reduced accordingly. Any repurchase of such shares by us must be on terms authorized by our board of directors, upon consultation with our board of statutory auditors and our external auditor, having regard to our net asset value, our prospective earnings and the market value of our ordinary shares, if any. Under Italian law, we may set forth different criteria in our bylaws for the consideration to be paid to withdrawing shareholders. We have not done so as of the date of this annual report.

Each shareholder may bring to the attention of the board of statutory auditors facts or acts which such shareholder deems wrongful. If such shareholders represent more than 5% of our share capital, our board of statutory auditors must investigate without delay and report its findings and recommendations at our shareholders' meeting. Shareholders representing more than 10% of our share capital have the right to report to the competent court serious breaches of the duties of the directors which may be prejudicial to us or to our subsidiaries. In addition, shareholders representing at least 20% of our share capital may commence derivative suits before the competent court against our directors, statutory auditors and general managers. We may waive or settle the suit unless shareholders holding at least 20% of the shares vote against such waiver or settlement. We will reimburse the legal costs of such action in the event that the claim of such shareholder is successful and the court does not award such costs against the relevant directors, statutory auditors or general managers.

Liability for mismanagement of subsidiaries

Pursuant to Italian law, if we, acting in our own interest or the interest of third parties, mismanage a company that we control, we are liable to that company's shareholders and creditors for ensuing damages. That liability is excluded if the ensuing damage is fully eliminated, including through subsequent transactions, or the damage is effectively offset by the global benefits to the company from the continued exercise of such direction and coordination powers. We are presumed to have control over, among other companies, any subsidiary whose financial statements are consolidated into ours. Since we currently have no subsidiaries in Italy, this law does not apply to us at this time.

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LIMITATION OF LIABILITY AND INDEMNIFICATION MATTERS

Insofar as indemnification for liabilities arising under the Securities Act may be permitted to directors, officers or persons controlling our company under the foregoing provisions, we have been informed that in the opinion of the SEC such indemnification is against public policy as expressed in the Securities Act and is therefore unenforceable.

DIFFERENCES IN CORPORATE LAW

The laws applicable to Italian joint stock companies and their shareholders differ from laws applicable to United States corporations and their shareholders. Set forth below is a summary of certain differences between the provisions of Italian law applicable to us and our shareholders and the law applicable to a Delaware corporation and its stockholders. This summary is not intended to be a complete discussion of the respective rights and it is qualified in its entirety by reference to Delaware law and Italian law.

Mergers and other extraordinary corporate transactions

Under Delaware law, a merger or consolidation requires the approval of a majority of the votes cast by the holders of shares entitled to vote in person or by proxy and, if any class or series is entitled to vote thereon as a class, the affirmative vote of a majority of the shares within each class or series entitled to vote as a class in person or by proxy, unless the certificate of incorporation requires a greater vote. The sale, lease, exchange or other disposition of all, or substantially all, the property and assets, of a Delaware corporation requires a majority vote unless the certificate of incorporation requires a greater vote. Under Delaware law, the dissolution of a corporation requires a majority vote unless the certificate of incorporation requires a greater vote.

Under Italian law, a merger requires the approval of more than half of the share capital at an extraordinary shareholders' meeting. Our bylaws authorize the board of directors to approve mergers of wholly-owned subsidiaries and subsidiaries of which we own at least 90%, except that, if so requested by shareholders representing at least 5% of our corporate capital, the relevant decisions shall be adopted at an extraordinary shareholders' meeting.

Amendments to charter documents

Under Delaware law, charter documents consist of a certificate of incorporation and bylaws. An amendment to the certificate of incorporation ordinarily requires a majority vote (unless the certificate of incorporation requires a greater vote). If a class or series is separately entitled to vote on an amendment, then its majority vote (unless the certificate of incorporation requires a greater vote), separately calculated, is necessary to approve the amendment. In addition, under Delaware law, the holders of outstanding shares of a class or series are entitled to vote as a class on a proposed amendment, whether or not entitled to vote thereon by the provisions of a company's certificate of incorporation, if the amendment would have certain effects identified in Delaware law. In such a case, an amendment must be approved by a majority of the voting power of the class (unless the certificate of incorporation requires a greater vote).

Under Delaware law, the stockholders entitled to vote have the power to adopt, amend or repeal bylaws. A corporation may also confer, in its certificate of incorporation, that power upon the board of directors.

Under Italian law, the charter documents consist of articles of association and bylaws. An amendment to these documents requires the approval (i) on first call, of more than one-half of the outstanding shares; and (ii) on second call, of more than two-thirds of the share capital represented at the meeting if the holders of more than one-third of the outstanding shares are represented at the meeting.

Certain extraordinary actions, such as a change in purpose, an advanced liquidation or an issuance of preferred shares, among others, require the approval of more than one-third of the outstanding shares for both first and second call.

Naming of companies

Under Delaware law, the legal name of a company must include a corporate identifier or name ending, such as "association", "company", "corporation", "club", "foundation", "fund", "incorporated," "institute", "society", "union", "syndic (or an abbreviation of any of the foregoing, with or without punctuation), or any word (or abbreviation, with or without punctuation) of like import in foreign countries or jurisdictions (provided that such word or abbreviation is written in roman characters or letters).

Under Italian law, the legal name of a corporation incorporated as a joint stock company must end in "S.p.A." or "Società per Azioni."

Capital
Delaware law permits companies to be incorporated with par value shares or no par value shares. If a Delaware company issues par value shares and receives an amount in excess of the par value, the directors may attribute a portion of the excess as

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"capital." If a Delaware company issues no par value shares, the directors may attribute a portion of the amount paid as "capital."

Italian law permits companies to be incorporated with par value shares or no par value shares. If an Italian company issues shares with par value and receives an amount in excess of the par value, the par value is attributed as "capital" and the excess is attributed to a "premium reserve," which is part of shareholders' equity.

Franchise tax

Delaware levies a franchise tax based on authorized capital. Italian law has no such tax.

Liability of shareholders

The liability of stockholders of a Delaware company is limited to the amount paid by the stockholders for their shares. The liability of shareholders of an Italian company is also limited to the amount paid by the shareholders for their shares.

Quorum of shareholders

Under Delaware law, no action may be taken at a meeting of the stockholders, with respect to any matter, unless a quorum is present. A quorum is present if the holders of a majority of the shares entitled to vote are represented at the meeting in person or by proxy, unless the certificate of incorporation provides for a greater percentage. Where a separate vote by a class or series or classes or series is required, a quorum shall be present at a meeting of stockholders if the holders of a majority of the shares entitled to vote are represented at the meeting in person or by proxy, unless the certificate of incorporation provides for a greater percentage.

Under Italian law, a quorum must be present at an ordinary meeting of shareholders on first call, and shall exist if the holders of at least 50% of the outstanding ordinary shares are represented at the meeting in person or by proxy. There is no quorum requirement on second call. A quorum must be present at an extraordinary meeting of shareholders on first call and second call. A quorum is present on first call if the holders of more than one-half of the share capital are represented at the meeting in person or by proxy, and on second call if the holders of more than one-third of the outstanding shares are represented at the meeting in person or by proxy.

Actions without a meeting-shareholders

Under Delaware law, stockholders may take an action without a meeting upon written consent, signed by the stockholders holding the minimum number of votes that would be necessary to take such action at a meeting, unless the certificate of incorporation provides otherwise.

Under Italian law, shareholders of a joint stock company may not act without a meeting.

Special/extraordinary meetings

Under Delaware law, special meetings of stockholders may be called by the board of directors or by such person or persons as may be authorized by the certificate of incorporation or the bylaws.

Under Italian law, an extraordinary shareholders' meeting may be called by the board of directors and must be called if requested by holders of at least 10% of the issued shares. Shareholders are not entitled to request that a meeting of shareholders be convened to vote on issues which as a matter of law shall be resolved upon the basis of a proposal, plan or report by the board of directors. If a request by the shareholders for an extraordinary meeting, or even an ordinary meeting, is refused by the board of directors, and such refusal is unjustified, the meeting may be called by a competent court.

Director qualifications

Under Delaware law, a director is not required to be a resident of Delaware or a stockholder of the corporation unless the certificate of incorporation or bylaws so require. The certificate of incorporation or bylaws may prescribe director qualifications.

Under Italian law, the only directorship requirement is that the individual has not been deemed "legally incompetent" to serve as a director under Italian law. "Legal incompetence" is determined by a competent court and may be declared by reason of lack of mental capacity, physical incapability, emotional instability, bankruptcy, certain criminal convictions or drug or alcohol addiction.

Election of directors

Under Delaware law, stockholders are not entitled to elect directors through cumulative voting, unless the certificate of incorporation provides otherwise. Absent a provision to the contrary, the directors of a corporation are elected by a plurality of the votes cast by the holders of shares entitled to vote in person or by proxy at a meeting of stockholders at which a quorum is present.

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Under Italian law, shareholders are not entitled to elect directors through cumulative voting. The directors of a corporation are elected by a majority of the votes cast by the shareholders entitled to vote in person or by proxy at an ordinary meeting of shareholders at which the relevant quorum is met.

Actions without a meeting - directors

Under Delaware law, any action required or permitted to be taken at any meeting of the board of directors may be taken without a meeting if all members of the board consent to the action in writing or by electronic transmission, and the writing or electronic transmission is filed with the minutes of proceedings of the board, unless otherwise restricted by the certificate of incorporation or bylaws.

Under Italian law, directors of a joint stock company may not act without a meeting.

Removal of directors

Under Delaware law, one or more directors of a corporation may be removed by the stockholders for cause or, unless the certificate of incorporation provides otherwise, without cause, upon the affirmative vote of the majority of votes cast by the holders of shares entitled to vote thereon, subject to certain exceptions.

Under Italian law, a director may be removed from office at any time by the vote of shareholders at an ordinary shareholders' meeting although, if the removal of a director was without just cause, such director may have a claim for damages against the company. These damages may include, but are not limited to, compensation that would otherwise have been paid to the director for the remainder of his or her term and damage to his or her reputation. Subject to the approval of the board of statutory auditors, the board of directors may appoint substitute directors to fill any vacancies caused by removal, who will serve until the next ordinary shareholders' meeting. If, at any time, more than half of the members of the board of directors are removed or otherwise cease to be directors, the board of directors will, in its entirety, cease to be in office, and the board of statutory auditors must promptly call an ordinary shareholders' meeting to appoint new directors.

Location of directors meetings

Delaware law provides that the board may hold its meetings outside of the State of Delaware, unless otherwise restricted by the certificate of incorporation or bylaws. Under Italian law and our bylaws, meetings of our board of directors may be held in person, or by audio-conference or video-conference, in any member state of the EU or in the United States.

Limitation of liability and indemnification

Delaware law requires that directors and members of any committee designated by the board of directors perform their duties in good faith and with the degree of diligence, care and skill that an ordinary prudent person would exercise under similar circumstances. Delaware law permits a corporation to impose limitations on director liability. Italian law requires directors and members of any committee designated by the board of directors to perform their duties in good faith and with that degree of diligence that is required by the nature of their office and under their specific level of competence. If a company cannot repay its creditors, and a court determines that the directors did not adequately perform their duties relating to the preservation of the company's assets, the court may find the directors liable to the company's creditors.

Dividends

Delaware law provides that the board of directors of a corporation may authorize and the corporation may make distributions subject to any restrictions in its certificate of incorporation. However, Delaware law provides that distributions may not be made if, after making the distribution, the corporation would not be able to pay its debts as they become due in the usual course of its business or the total assets would be less than total liabilities.

Under Italian law, payment of any annual dividend must be proposed by the board of directors to the shareholders and is subject to the approval of the shareholders at the annual ordinary shareholders' meeting. Before dividends may be paid out of profits in any year, a company must allocate an amount equal to 5% of the net profit to its legal reserve until such reserve is at least equal to 20% of its capital. If capital is reduced as a result of accumulated losses, a company may not pay dividends until the capital is reconstituted or reduced by the amount of such losses. A company may distribute reserves derived from available earnings retained from prior years, provided that after such payment, the company has a legal reserve at least equal to the legally required minimum of 20% of the capital. A company may

not approve or pay dividends, without the allocation of an amount equal to 5% of the net profit to its legal reserve, until this minimum (i.e., 20% of the capital) is met. If the minimum is met, the board of directors could propose the issuance of a dividend and the shareholders' resolution might approve that issuance. The shareholders' resolution will specify the manner and the date of dividend payment. Any dividends which shareholders do not collect within five years of the date on which they become payable will be forfeited by those shareholders and the money will come back to the company. The board of directors may not approve interim dividends at times between annual ordinary shareholders' meetings.

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Return of capital

Delaware law provides that corporations may return capital by dividend, redemption or repurchase of shares subject to certain solvency tests. Stockholder approval is not required for these transactions so long as the corporation meets the solvency tests.

Under Italian law, a company is permitted to purchase its outstanding shares, subject to certain conditions and limitations. A company may only purchase the shares out of profits available for dividends or out of distributable reserves, in each case as appearing on the latest shareholder-approved financial statements. Further, a company may only repurchase fully paid-in shares. Such purchases must be authorized by shareholder vote at an ordinary shareholders' meeting and the authorization may be issued for a period not to exceed eighteen (18) months. A corresponding reserve equal to the purchase price of such shares must be created in the balance sheet, and such reserve is not available for distribution, unless such shares are sold or canceled. Shares purchased and held by a company may be resold only pursuant to a resolution of its shareholders adopted at an ordinary shareholders' meeting. The voting rights attaching to the shares held by a company or its subsidiaries cannot be exercised, but the shares can be counted for quorum purposes in shareholders' meetings. Dividends and other rights, including preemptive rights, attaching to such shares, will accrue to the benefit of other shareholders.

Officers

Under Delaware law, a corporation is required to have at least two officers vested with the authority sign stock certificates and instruments to be filed with the Secretary of State. The corporation has complete freedom to designate any name to its executive positions and to allocate managerial power to its executives as it wishes. Any number of offices may be held by the same person, unless otherwise provided in the certificate of incorporation or the bylaws. Officers may be selected by any person or body and in any way specified in the bylaws or in a resolution of the governing body.

Under Italian law, there are no requirements with respect to the number, title or election of officers.

Share certificates

Under Delaware law, the shares of a corporation shall be represented by certificates, provided that the board of directors may resolve that some or all of any or all classes or series of its stock shall be uncertified stock. However, existing stockholders and future stockholders may, if they desire, obtain a stock certificate signed in the name of the corporation by the chairman or vice-chairman of the board of directors or the president or vice-president, and by the treasurer or an assistant treasurer, or the secretary or an assistant secretary of such corporation. The terms governing preferred stock, if any, must be expressed "in clear language" in the certificate of incorporation (or by a separate resolution authorized by the charter).

Under Italian law, the shares of a corporation may be issued in either registered or certificated form. Our bylaws provide that our ordinary shares are not certificated. Rather, they are held through various participants, primarily institutions, on Monte Titoli's system and registered in book-entry form on our shareholders' register.

Preemptive rights

Under Delaware law, stockholders do not possess preemptive rights with respect to the issuance of additional securities by the corporation, unless the certificate of incorporation provides otherwise.

Under Italian law, shareholders and holders of convertible debentures are entitled to subscribe for issuance of ordinary shares or convertible debentures in proportion to their holdings at the time of authorization of the capital increase for those issuances, except in the case of contributions in kind or if such exclusion or limitation is in the interest of the company.

Liquidation rights generally

Under Delaware law, stockholders are entitled to share ratably in the distribution of assets upon the dissolution of a corporation. Asset distribution to preferred stockholders upon corporate dissolution is typically limited to established contractual preferences. Once the rights of preferred stockholders have been fully satisfied, holders of common stock are entitled to any remaining assets.

Under Italian law, and subject to the satisfaction of the claims of all creditors, upon liquidation a company's shareholders are entitled to a distribution that is equal to an amount resulting from the division of the positive

liquidation balance by the number of shares or shareholders (to the extent available out of the company's net assets). Asset distribution to preferred shareholders and holders of "participating certificates", if any, upon corporate dissolution is regulated by the rules provided by the bylaws. Once the rights of preferred shareholders and holders of participating certificates have been fully satisfied, holders of ordinary shares are entitled to any remaining assets. As of the date of this annual report, we have not issued any "participating certificates" or preferred shares.

Shareholder derivative suits

Under Delaware law, a derivative suit may be brought only if the plaintiff was a record or beneficial owner of shares on the date of the transaction that gave rise to the suit, and the initial pleading in the suit states that the ownership requirement is

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satisfied, and also states with particularity, plaintiff's efforts to first obtain the action desired from the board of directors, or the reasons for not making such efforts. The court may require the plaintiff to give security for the expenses incurred or expected to be incurred by the defendants. The court may also require the plaintiff to pay expenses to the defendants if the court finds, upon a final judgment in favor of the defendants, that the suit was brought without reasonable cause.

Under Italian law, a shareholder's name must be entered in the shareholders' register in order to establish his shareholder rights against us. Shareholders may bring to the attention of the board of statutory auditors facts or acts which such shareholder deems wrongful. If such shareholders represent more than 5% of a company's share capital, its board of statutory auditors must investigate without delay and report its findings and recommendations at a shareholders' meeting. Shareholders representing more than 10% of a company's share capital have the right to report, to a competent court, serious breaches by directors of their duties as directors, which may be prejudicial to the company or its subsidiaries. In addition, shareholders representing at least 20% of a company's share capital may commence a derivative suit before a competent court against its directors, statutory auditors and general managers. A company may waive or settle the suit if it is approved by a shareholders' resolution and unless shareholders holding at least 20% of the shares vote against such resolution. A company will reimburse the legal costs of such action in the event that the claim of such shareholders is successful and the court does not award such costs against the relevant directors, statutory auditors or general managers.

Dissenters' rights

Any stockholder of a Delaware corporation has the right to dissent from any plan of merger or consolidation to which the corporation is a party, except that, unless the certificate of incorporation provides otherwise, a stockholder shall not have the right to dissent from any plan of merger or consolidation, with respect to shares of a class or series that are listed on a national securities exchange or held of record, by not less than 2,000 holders on the record date fixed to determine the stockholders entitled to vote upon the plan of merger or consolidation. A dissenting stockholder has a right to appraisal of its shares upon the satisfaction of certain conditions.

Under Italian law, shareholders' resolutions which are not adopted in conformity with applicable law or a company's bylaws may be challenged (with certain limitations and exceptions) within ninety days of such resolution by any absent, dissenting or abstaining shareholders representing in the aggregate at least 5% of the company's share capital (or by its board of directors or board of statutory auditors). Shareholders who do not meet the threshold or are not otherwise entitled to vote may only claim damages arising from the resolution.

Dissenting or absent shareholders may withdraw from the company as a result of shareholders' resolutions approving, among others things, material modifications to the company's purpose, the voting rights of the ordinary shares, the conversion from a share corporation into a different legal entity or a transfer of the registered office outside of Italy. Under any such circumstances, remaining shareholders would have preemptive rights to purchase the shares of the withdrawing shareholders. Should no shareholder exercise its preemptive right, the shares must be offered to third parties. If no third party desires to purchase the shares, the company can purchase them with its available reserves, if any. In the event that there are no reserves available, the company must reduce its capital accordingly. According to Italian law, the value of the shares of the withdrawing shareholders shall be determined by the board of directors, after consultation with the board of statutory auditors and the external auditors, taking into consideration the company's net asset value, prospective earnings and the market value of the ordinary shares, if any. Under Italian law, a company may include provisions in its bylaws governing the payment of consideration to shareholders in the event of withdrawal, provided that the provision of the bylaws does not limit the rights granted to the withdrawing shareholder. We have not done so as of the date of this annual report.

Interested shareholder transactions

Delaware corporations are subject to the State of Delaware's "business combination" statute. In general, that statute prohibits a publicly-traded corporation from engaging in various "business combination" transactions with any "interested stockholder" for a period of three years after the time that the stockholder became an interested stockholder, unless the business combination is approved by the board prior to the time the stockholder became an interested stockholder, the interested stockholder acquired 85% or more of the outstanding shares in a transaction in which it

became an interested stockholder, or the business combination is approved by the board and by holders of two-thirds of the shares, excluding any shares held by the interested stockholder. A “business combination” includes a merger, assets sale or other transaction resulting in a financial benefit to a stockholder. An “interested stockholder” is a person who, together with affiliates and associates, owns 15% or more of a corporation’s voting stock.

Under Italian law, a director having any interest in a proposed transaction must disclose his or her interest to the board of directors and to the board of statutory auditors, even if such interest does not conflict with the company’s interest in the transaction. The interested director is not required to abstain from voting on the resolution approving the transaction, but the resolution must explicitly state the reasons for the approved transaction and the benefit of the transaction to the company. If these provisions are not complied with, or if the transaction would not have been approved without the vote of the interested director, the resolution may be challenged by any director or by the board of statutory auditors on grounds that the approved

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transaction would be prejudicial to the company. A managing director ("amministratore delegato") of a company having any interest in a proposed transaction that he or she has authority to approve must solicit prior board approval before consenting to such transaction. The interested director may be held liable for damages to the company resulting from the breach of the above rules. Finally, a director may be held liable for illicitly profiting from insider information or a corporate opportunity.

Inspection of books and records

Under Delaware law, upon the written request of any stockholder, the corporation shall mail to such stockholder its balance sheet as of the end of the preceding fiscal year, and its profits, losses and surplus statements for such fiscal year. Inspection rights are extended to any person who beneficially owns stock through either a voting trustee or a nominee who holds the stock of record on behalf of such person. If the stockholder is not a holder of record, such person must state under oath the person's status as a stockholder and produce documentary evidence of beneficial ownership. Any stockholder is entitled to examine the relevant books and records of a corporation for any proper purpose, namely, a purpose reasonably related to such person's interest as a stockholder, upon written demand stating the purpose thereof.

Under Italian law, shareholders may review the report of the board of directors on the management of the company and the report of its statutory auditors and the report of its accounting firm on the company's financial statements during the fifteen days prior to the ordinary shareholders' meeting to approve those financial statements. The reports, together with the relevant financial statements, remain on file at the company's offices and may be reviewed after the annual shareholders' meeting. Our reports are also filed with the Companies' Registry of Como and made publicly available. Moreover, any shareholder is entitled to examine the shareholders' register and the ledger of the minutes of the shareholders' meeting, at any time.

Registered office

Delaware law requires that a corporation have a "registered office" in Delaware. Italian law requires that a corporation have a registered office in Italy.

Issuance of shares

Under Delaware law, directors have the authority to issue shares of common stock. If the certificate of incorporation so provides, the directors may also designate the terms of preferred stock and issue shares of preferred stock.

Under Italian law, the issuance of any shares, ordinary or otherwise, requires an amendment to the company's bylaws to increase its share capital, which must be recommended to the shareholders by the board of directors and approved by a vote of the shareholders at an extraordinary meeting of the shareholders. Once the shareholders' meeting has authorized the issuance of securities and the same have been subscribed, an amount equal to not less than 25% of the nominal value of the shares subscribed and the whole amount of the share premium, if any, shall be paid to have the shares issued to the relevant subscribing shareholder. The board would also need to recommend, and the shareholders would need to approve by vote at the extraordinary meeting, specific terms of the securities. Alternatively, shareholders at an the extraordinary shareholders' meeting can delegate the power to increase share capital to the board of directors, but the board's right to exercise such power, if delegated, will expire after five years. If the board does not approve a capital increase by the end of those five years, the board and shareholders would need to meet again to re-delegate this authority. With respect to shareholders' resolutions approving capital increases, Italian law provides that any interested person may challenge the resolution as void for a period of 180 days following the filing of the shareholders' resolution with the Register of Companies; except that, if the challenge is brought on the grounds that a required shareholders' meeting was not called to approve the capital increase, the challenge may be brought until 90 days following the shareholders' meeting to approve the financial statements reflecting such capital increase. Finally, once shareholders authorize a capital increase, all those authorized shares that have been subscribed must be paid-up entirely before the shareholders may authorize a new capital increase.

Debt-equity ratio

Under Delaware law, there are no restrictions on the amount of debt securities that a corporation may issue.

Under Italian law, a company may issue debt securities in an amount not to exceed twice the sum of its capital, its legal reserve and any other disposable reserves appearing on its latest Italian balance sheet approved by its

shareholders. The legal reserve is a reserve to which a company allocates 5% of its net income each year until it equals at least 20% of its capital. One of the other reserves that a company maintains on its balance sheet is a “share premium reserve”, which reflects the amount paid for its ordinary shares in excess of the amount of such ordinary shares allocated to its capital. In case of issuance of debt securities, a company may not voluntarily reduce its capital or distribute its reserves (such as by declaring dividends) if the aggregate of the capital and reserves, following such reduction of capital and/or distribution of reserves, is less than one-half of the outstanding amount of the debt securities. If a company’s equity is reduced by losses or otherwise, such that the amount of the outstanding debt securities is more than twice the amount of its equity, the company cannot distribute profits to its shareholders until the ratio between the amount of its debt securities and its capital and reserves is restored. Some legal scholars are of the opinion that the ratio must be restored by a recapitalization of the company. If our equity is reduced, we could recapitalize by means of issuing new shares by or having our current shareholders contribute additional capital to our company,

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although there can be no assurance that we would be able to find purchasers for new shares or that any of our current shareholders would be willing to contribute additional capital. These laws regarding the ratio of debt securities to capital and reserves do not apply: (i) to the issuance of debt securities to professional investors (as defined by Italian law), however, professional investors who transfer such debt securities to third parties not qualified as professional investors would remain liable to the transferee for the payment of such securities; (ii) to the issuance of debt securities listed on a regulated market; (iii) to convertible or exchangeable debt securities, whether or not listed on a regulated market; and (iv) to debt securities secured by mortgages over real estate owned by the company up to two-thirds of their value.

Reduction of equity by losses

Under Delaware law, stockholder equity in a corporation is reduced by losses and may become negative.

Italian law requires a company to reduce its share capital in certain situations. Shareholders' equity has three main components: share capital, legal reserves and other shareholders' equity (such as any premium paid for the shares over the par value and any retained earnings). Losses from operations are applied against, first, legal reserves and, if the reserves are not sufficient, the losses will affect the corporate capital. If share capital is reduced by more than one-third as a result of losses, the board of directors must call a shareholders' meeting as soon as possible. The shareholders should take appropriate measures, which may include, inter alia, reducing the share capital by the amount of the remaining losses, or carrying the losses forward up to one year. If the shareholders vote to carry the losses forward up to one year, and the losses are still more than one-third of the amount of the share capital at the end of that year, then the company must reduce its capital by the amount of the losses suffered. An S.p.A. must maintain capital of at least €120,000. If the amount of the losses would reduce an S.p.A.'s capital to less than €120,000, then:

- the S.p.A. would need to increase its capital, which it could do by issuing new shares or having its shareholders contribute additional capital to the company;
- the shareholders would need to convert the S.p.A. to an "S.r.l.", a private limited liability company, which has a lower capital requirement of €10,000; or
- if neither of these options is pursued, the shareholders or, if they do not so resolve, a court of competent jurisdiction, could appoint a liquidator, who need not be an Italian citizen, to liquidate the company.

MATERIAL CONTRACTS

The contracts described below have been in existence during the last two years and, as of the date of this annual report, contain provisions under which we have an obligation or right that is or may be material to us. This discussion is not complete and should be read in conjunction with the agreements described below, each of which has been filed with the SEC as an exhibit to this annual report.

In connection with our license and supply agreement with Sigma-Tau, we entered into a cost sharing agreement with Sigma-Tau dated October 12, 2007. Under the cost sharing agreement, as amended most recently in January 2014, Sigma-Tau agreed to reimburse us 50% of certain costs associated with the development of defibrotide and we agreed that $1.0 million in costs reimbursed by Sigma-Tau will be deductible from royalty payments that we are entitled to receive in the future under the license and supply agreement. In 2013, we received €1.40 million ($1.80 million) as reimbursement of research and development expenses. In 2014, Sigma-Tau agreed to reimburse us an estimated total of approximately $4.77 million as incurred.

We entered into a manufacturing services agreement with Patheon effective September 15, 2013, pursuant to which Patheon will process the defibrotide compound into finished form at its Italian manufacturing plant and supply the finished product to us. The initial term of the agreement ends on December 31, 2018, subject to automatic extension for additional two-year terms unless either party provides notice to the other of its intent to terminate the agreement at least 18 months before the end of the then-current term. Either party has the right to terminate the agreement in the event of the other party's uncured material breach or insolvency.

On October 24, 2013, we amended our clinical research agreement with US Oncology Clinical Development, or US Oncology, dated September 29, 2009 to extend the agreement for an indefinite term. US Oncology serves as a clinical research organization to help administer certain aspects of our expanded access program and associated cost recovery program. We can generally terminate the agreement at any time upon 30 days advance written notice to US Oncology

and, in certain circumstances, we may terminate the agreement immediately upon written notice. US Oncology may terminate the agreement with 90 days written notice in the event of our uncured material breach.

On December 19, 2013, we entered into a definitive tender offer agreement with Jazz and Jazz Italy, a wholly-owned subsidiary of Jazz, pursuant to which Jazz Italy made an offer to purchase all our outstanding ordinary shares and ADSs, each representing one ordinary share, at a purchase price of $57.00 per ordinary share and per ADS (without duplication for ordinary

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shares underlying ADSs). As of February 21, 2014, Jazz Italy completed its tender offer, having acquired approximately 98 percent of our combined ordinary shares and ADSs. Jazz is now our indirect majority shareholder. We and the ADS depositary entered into an amendment dated March 21, 2014 to the deposit agreement dated as of June 15, 2005, or the deposit agreement, which amendment will be effective on April 13, 2014. Upon effectiveness, the amendment to the deposit agreement will reduce from one year to 60 days the period that must elapse between the date of termination of the deposit agreement and the date on which the ADS depositary may sell the ordinary shares of the Company it then holds and provides for the ADS depositary to accept any offer by a subsidiary of Jazz to purchase those shares at a price of no less than $57.00 per ordinary share, unless the ADS depositary has received what it considers to be a superior bona fide offer from another party. The ADS depositary has given notice such that the deposit agreement will terminate at 5:00 pm (Eastern time) on April 13, 2014. While the date or dates on which the ADS depositary will sell the remaining shares have not been determined, the sale will not occur before June 13, 2014. Jazz Italy has indicated that it intends to offer to purchase ordinary shares from the ADS depositary at a per share price equal to $57.00.

We are party to agreements and other arrangements with Dr. Islam, Mr. Calabrese, Dr. Mosconi and Mr. Haigh that are described in Part I, Item 6 of this annual report in the section entitled “Compensation—Compensation of Executive Officers,” which description is incorporated herein by reference.

EXCHANGE CONTROLS

No exchange control consent is required in Italy for the transfer of dividends or other distributions with respect to shares of an Italian company or proceeds from the sale thereof to persons outside of Italy.

TAXATION

Tax Consequences Applicable to United States Holders

This section contains a description of the principal United States federal and Italian tax consequences of the ownership and disposition of ADSs or ordinary shares by a U.S. holder, as defined below. This summary does not purport to be a comprehensive description of all of the tax considerations that may be relevant to the ownership and disposition of ADSs representing our ordinary shares and each potential purchaser is therefore urged to consult an independent tax advisor.

In particular, this summary deals only with U.S. holders who hold ADSs as capital assets and does not address the tax treatment of a U.S. holder:

- who have held ADSs representing 10% or more of our voting shares (either directly or through attribution);
- who holds ADSs in connection with a permanent establishment or fixed base of business located in Italy;
- who holds ADSs in the ordinary course or as an integral part of the holder’s trade or business or as part of a hedging, straddle, integrated or conversion transaction;
- who is subject to special treatment under the U.S. income tax laws (such as securities dealers, brokers, traders that elect to market, insurance companies, banks, tax-exempt organizations, partnerships and other pass-through entities);
- whose functional currency is not the U.S. dollar; or
- who is a resident of Italy for purposes of Italian domestic law or the Income Tax Treaty between the United States and Italy, or the Income Tax Convention, or acts through an Italian permanent establishment or fixed base to which the ADSs are connected.

In addition, the following discussion does not address any aspect of state, local or non-U.S. tax laws (other than certain Italian tax laws) or any alternative minimum tax consequences.

The summary is based upon tax laws of the United States and the Republic of Italy and on the provisions of the Income Tax Convention in each case as in effect on the date hereof, all of which are subject to change (possibly with retroactive effect). We will not update this summary to reflect changes in laws and if such a change occurs, this summary could become inaccurate. For purposes of these laws and Income Tax Conventions, beneficial owners of ADSs should be treated as the beneficial owners of the ordinary shares represented by the ADSs. Prospective purchasers of the ADSs are advised to consult an independent tax advisor as to the tax consequences of the ownership

and disposition of the ADSs including, in particular, state and local tax consequences.

For purposes of this section, a U.S. holder means:

- an individual citizen or resident of the United States;

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- a corporation (or other entity taxable as a corporation for U.S. federal income tax purposes) organized in or under the laws of the United States or any political subdivision thereof;
- an estate, the income of which is includible in gross income for U.S. federal income tax purposes regardless of its source;
- a trust, if a U.S. court is able to exercise primary jurisdiction over the administration of the trust and one or more U.S. persons have the authority to control all substantial decisions of the trust; and
- any other person subject to U.S. federal income taxation on a net income basis in respect of income attributable to its ownership of the ADSs.
- A U.S. owner means a U.S. holder that is considered a resident of the United States for purposes of the Income Tax Convention and who is not subject to an anti-treaty shopping provision.

Italian Taxation of United States Holders

General. Under Italian law, financial instruments issued by an Italian company are subject to the same tax regime as are shares, provided that their remuneration is entirely represented by participation in the economic results of the issuer. Pursuant to Article 10(3) of the Income Tax Convention, the tax regime of dividends applies to income from corporate rights of an Italian company, which is subject to the same taxation treatment as income from shares under the laws of Italy. One interpretation of these laws is that a beneficial owner of an ADS should be subject to the same tax regime as a beneficial owner of a share for purposes of both Italian law and the Income Tax Convention. However, no official interpretation has been issued by the Italian tax authorities on this subject matter to date.

Income Tax Withholding on Dividends. We do not anticipate making any distributions on our ordinary shares in the foreseeable future. However, if we were to make distributions on our ordinary shares, we would generally be required under Italian law, except as otherwise discussed below, to apply a 20% final withholding tax on payments made to holders of ADSs who are not residents of Italy for tax purposes. Under Italian law, United States owners can claim a refund of up to one-fourth of the Italian withholding tax withheld on dividends (effectively reducing the rate of withholding to 15%) upon presenting evidence to the Italian tax authorities that income taxes have been fully paid on the dividends in the country of residence of the United States owners in an amount at least equal to the total refund claimed. United States holders should consult an independent tax advisor concerning the availability of this refund, which has traditionally become payable only after extensive delays.

Under the Income Tax Convention, dividends paid to United States owners will be subject to Italian withholding tax at a reduced rate of 15% for individuals not engaged in entrepreneurial activities. However, the amount that we will initially make available to the depositary for payment to United States owners will reflect withholding at the 20% rate. United States owners who comply with the certification procedures described below may claim a refund of the difference between the 20% rate and the 15% rate (referred to herein as a “treaty refund”). The certification procedure will require the United States owner to:

- obtain from the United States Internal Revenue Service, or IRS, (generally, by filing Form 8802) a form of certification required by the Italian tax authorities with respect to each dividend payment (Form 6166, printed on U.S. Department of Treasury stationary), unless a previously filed certification is effective with respect to the payment,
- produce a statement whereby the United States owner represents that it is a United States owner that does not maintain a permanent establishment in Italy, and
- set forth certain other required information.

The time for processing requests for certification by the IRS can be lengthy. Accordingly, United States owners should begin the process of obtaining a certification from the IRS as soon as possible after receiving instructions from the depositary.

The depositary’s instructions will specify certain deadlines for delivering the documentation required to obtain a treaty refund, including the certification that the United States owners must obtain from the IRS. In the case of ADSs held by United States owners through a broker or other financial intermediary, the required documentation should be delivered to such financial intermediary for transmission to the depositary. In all other cases, United States owners should deliver the required documentation directly to the depositary. We have agreed with the depositary that if the required documentation is received by the depositary on or within 30 days after the dividend payment date and, in our

reasonable judgment, such documentation satisfies the requirements for a refund of Italian withholding taxes under the Income Tax Convention then in effect between the United States and Italy, we will (within 45 days after that period) pay an amount equal to the treaty refund to the depositary for the benefit of the United States owners entitled thereto. If the depositary does not receive a United States owner's required documentation within 30 days after the dividend payment date, the United States owner may, for a short grace period (specified in the depositary's instructions), continue to

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claim an amount equal to the treaty refund by delivering the required documentation (either through the United States owner's financial intermediary or directly, as the case may be) to the depositary. However, after this grace period, the treaty refund must be claimed directly from the Italian tax authorities rather than through the depositary. United States owners seeking refunds from the Italian tax authorities have encountered expensive and extensive delays.

Income Tax on Capital Gains. Under Italian law, capital gains realized by a person who is not a resident of Italy (meaning that such person does not have a permanent establishment or fixed base in Italy to which the ADSs are connected) on the disposal of a "qualified" shareholding, contribute to determine the overall taxable income for income tax purposes. Ministerial Decree April 2, 2008 - issued pursuant to Article 1, paragraph 38 of the Law December 24, 2007 (Budget Law 2008) - sets out that 49.72% (it was 40% until 2008) of the capital gains would contribute to determine the overall taxable income. This rate applies to capital gains realized from January 1, 2009. The 40% previously in effect still applies to capital gains realized in connection with disposal deeds executed before January 1, 2009. Losses can be offset against taxable gains for a corresponding amount and, if in excess, can be carried forward up to four years. A "qualified" shareholding is defined as ordinary shares and/or rights (including ADSs) that represent more than 20% of share capital voting in the ordinary shareholders' meeting or 25% of the company's total share capital. A "disposal" of a qualified shareholding occurs if, in any 12-month period following the date when a shareholding meets one of the thresholds illustrated above, a shareholder disposes of shares or ADSs that, individually or in the aggregate, constitute a "qualified" shareholding. Generally, Italian capital gain tax, levied at a rate of 20%, is imposed on gains realized upon the transfer or sale of "non-qualified" shareholdings whether held within or outside Italy. A "non-qualified" shareholding is defined as an interest in ordinary shares and/or rights (including ADSs) which does not reach the thresholds described above for a qualified shareholding.

Furthermore, save for any applicable anti-avoidance provision, pursuant to the Income Tax Convention, a United States owner will not be subject to Italian capital gain tax or to Italian individual or corporate income tax unless such United States owner has a permanent establishment or fixed base in Italy to which the owner's ADSs is effectively connected. To this end, United States owners selling ADSs and claiming benefits under the Income Tax Convention may be required to produce appropriate documentation establishing that the above-mentioned conditions have been met.

Estate and Gift Tax. Inheritance and gift taxes, which were abolished in 2001, have been re-introduced in the Italian system by Law Decree No. 262 of October 3, 2006 (converted into law, with amendments, by Law Decree No. 286 of November 24, 2006), as amended. Such taxes will apply to the overall net value of the relevant assets, at the following rates, depending on the relationship between the testate (or donor) and the beneficiary (or donee): (a) 4%, if the beneficiary (or donee) is the spouse or a direct ascendant or descendant (such rate only applying on the net asset value exceeding, for each person, €1 million); (b) 6%, if the beneficiary (or donee) is a brother or sister (such rate only applying on the net asset value exceeding, for each person, €100 thousand); (c) 6% if the beneficiary (or donee) is another relative within the fourth degree or a direct relative-in-law as well as an indirect relative-in-law within the third degree; and (d) 8% if the beneficiary is a person other than those mentioned under (a), (b) and (c), above. If the beneficiary has a serious disability recognized under applicable law, inheritance and gift taxes will apply to its portion of the net asset value exceeding €1.5 million.

Transfer tax. In connection with the Italian stamp duty tax on the transfer of shares and ADSs, according to article 37 of Law No. 248 of December 31, 2007, converted with amendments into Law No. 31 of February 28, 2008, the stamp duty has been abolished with regard to contracts having as their object the transfer of shares. In certain cases the relevant transfer acts would be subject to the registration tax at a flat amount equal to €168.

New Stamp Duty. A stamp duty has been introduced under article 19 of Law Decree No. 201 of December 6, 2011, converted into Law No. 214 of December 22, 2011, to be imposed on communications (issued by banks and financial intermediaries) to clients relating to securities, even where the deposit of such securities is not mandatory (although certain entities are excluded). The amount of the stamp duty is based on the market value of the securities or, in the absence of a market value, on the nominal amount or the amount payable on redemption. The following rates apply:

- 0.1% for 2012; and
- 0.15% for 2013,

subject to a minimum amount of €34.20 and, for the year 2012, a maximum amount of €1,200. The communication is deemed to be sent to clients at least once a year, even where there is no obligation to issue any such communication.

United States Taxation of United States Holders

Taxation of Distributions Made on ADSs. As previously indicated, we do not anticipate making any distributions on our ordinary shares in the foreseeable future. However, if we were to make distributions with respect to our ordinary shares, the amount of any such distribution (including the amount of any Italian taxes withheld therefrom) would generally be includible in the gross income of a U.S. holder of an ADS (on the date of receipt by the depositary) as foreign source dividend income to the extent that such distributions are paid out of our current or accumulated earnings and profits, as determined for United States federal income tax purposes. If the amount of any distribution paid on our ordinary shares exceeds our current and accumulated

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earnings and profits, that excess will first reduce a holder's basis in its ADSs and, to the extent the distribution is in excess of the holder's basis, the excess amount will be treated as a capital gain. Dividends paid to U.S. holders that are corporations will not be eligible for the dividends-received deduction (which is generally applicable only to dividends paid by U.S. corporations).

The U.S. dollar amount of dividends received by individuals with respect to our shares or ADSs will be subject to taxation at a maximum rate of 20% subject to certain limited exceptions. Dividends received from a "qualified foreign corporation" generally qualify for the reduced rate. In this regard, a foreign corporation that is not a passive foreign investment company (PFIC) in the year that the dividends are paid or in the preceding taxable year will generally constitute a qualified foreign corporation with respect to any dividends paid by it on its stock if such corporation is eligible for benefits of a comprehensive income tax treaty with the United States which includes an exchange of information program. Because we qualify for benefits under the Income Tax Convention and because the Income Tax Convention is a comprehensive income tax treaty that includes an exchange of information program, we should constitute a qualified foreign corporation and dividends paid on our ordinary shares and received by U.S. holders of ADSs that are individuals should qualify for the reduced rate, so long as we are not a PFIC in the year the dividends are paid or in the preceding taxable year. While we do not believe that we qualify as a PFIC at present, no assurances can be provided that we will not constitute a PFIC in any year during which we make a distribution on our ordinary shares (or in the taxable year preceding the year of distribution).

The amount of any cash distribution received in Euro with respect to the ADSs will equal the U.S. dollar value of the distribution, including the amount of any Italian taxes withheld therefrom, determined on the basis of the spot exchange rate in effect on the date that the distribution is received by the depositary (regardless of whether or not the distribution is in fact converted into U.S. dollars), and a U.S. holder will have a tax basis in the Euro equal to that same value. Upon a subsequent sale or other disposition of the Euro, any gain or loss recognized by the U.S. holder will be ordinary income or loss for U.S. federal income tax purposes.

Subject to general foreign tax credit limitations, a U.S. holder may elect to credit any Italian income taxes withheld on dividends paid with respect to the ADSs against the holder's U.S. federal income tax liability (provided, inter alia, that the U.S. holder satisfies certain holding period requirements with respect to the ADSs). Amounts withheld in excess of the applicable rate under the Income Tax Convention in effect between the United States and Italy in respect of a U.S. holder who qualifies for the benefits of the convention will not be eligible for this credit, but the U.S. holder may claim a refund for this excess from the Italian tax authorities. See Part I, Item 10 of this annual report in the section entitled "Taxation—Italian Taxation of U.S. Holders—Income Tax Withholding on Dividends." As an alternative to claiming a foreign tax credit, a U.S. holder may claim a deduction for any Italian income taxes withheld, but only with respect to a year from which the U.S. holder elects to do so with respect to all of its foreign income taxes. There are complex rules that limit the amount of foreign income taxes that may be credited against a U.S. holder's federal income tax liability, and U.S. holders are strongly urged to consult an independent tax adviser as to the applicability of these limitations.

Sales or other Disposition of the ADSs. Subject to the discussion set forth below regarding PFICs, a U.S. holder will recognize capital gain or loss for U.S. federal income tax purposes on the sale or other disposition of the ADSs equal to the difference between the amounts realized on the disposition and the holder's basis in the ADSs. Such gain or loss will be long-term capital gain or loss if the U.S. holder has owned the ADSs for more than one year at the time of the sale or other disposition. Net long-term capital gains are subject to tax at a maximum capital gain tax rate of 20%.

Special Rules Applicable to PFICs. Special federal income tax rules apply to U.S. holders who own stock in a PFIC. In this regard, a foreign corporation is generally considered a PFIC for any taxable year in which 75% or more of its gross income is passive income or in which 50% or more of the average value of its assets are considered "passive assets" (generally assets that generate passive income or assets held for the production of passive income, including cash). We believe that we do not qualify as a PFIC at present and we do not anticipate that we will become a PFIC in the future.

However, if we were to be classified as a PFIC, a U.S. holder would generally be subject to a special tax at ordinary income tax rates on so-called "excess distributions"-which include certain distributions received on the ADSs and gain

recognized on any sale or other disposition of the ADSs. To compensate for any tax deferral, the amount of income tax on these excess distributions will be increased by an interest charge, calculated as if the excess distributions were earned ratably over the period during which the U.S. holder held the ADSs. In addition, the tax on excess distributions treated as earned in prior years will be subject to tax at the maximum ordinary income rate applicable in the year in which such income is deemed to have been earned. The harsh consequences of these rules may be avoided if the U.S. holder properly elects to include such holder's pro rata share of our ordinary earnings in its ordinary income each year and to include such holder's pro rata share of our net capital gain, whether or not distributed, in its long-term capital gain income each year. However, we do not intend to provide U.S. holders with the information that they would need in order to make this election. Alternatively, a holder of ADSs may avoid the tax consequences detailed above by making a mark-to-market election, but only if the ADSs are "regularly traded" for purposes of Section 1296 of the Code. No assurances can be made that the ADSs will be regularly traded and, in any event, a U.S. holder should consult an independent tax advisor before making any election under Section 1296 of the Code.

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In addition, if we were to be classified as a PFIC, U.S. holders would not qualify for the benefit of the reduced U.S. federal tax rate applicable to certain dividends received by individuals, as described above in "United States Taxation of U.S. Holders-Taxation of Distributions Made on the ADSs."

Medicare Tax. In general, a U.S. holder that is an individual or estate, or a trust that does not fall into a special class of trusts that is exempt from such tax, is subject to a 3.8% tax on the lesser of (1) the U.S. holder's "net investment income" for the relevant taxable year and (2) the excess of the U.S. holder's modified adjusted gross income for the taxable year over a certain threshold (which in the case of individuals will be between $125,000 and $250,000, depending on the individual's circumstances). A U.S. holder's net investment income will include its gross dividend income and its net gains from the disposition of ADSs, unless such dividends or net gains are derived in the ordinary course of the conduct of a trade or business (other than a trade or business that consists of certain passive or trading activities). If you are a U.S. holder that is an individual, estate or trust, you are encouraged to consult your tax advisors regarding the applicability of the Medicare tax to your income and gains in respect of your investment in ADSs.

Information Reporting and Backup Withholding. U.S. holders may be required to file certain U.S. information reporting returns with the IRS with respect to an investment in ADSs, including, among others, IRS Form 8938 (Statement of Specified Foreign Financial Assets). Substantial penalties may be imposed upon a U.S. holder that fails to comply with the required information reporting.

Dividends on and proceeds from the sale or other disposition of ADSs may be reported to the IRS unless the U.S. holder establishes a basis for exemption. Backup withholding may apply to amounts subject to reporting if (1) the holder fails to provide an accurate taxpayer identification number or otherwise establish a basis for exemption, or (2) is described in certain other categories of persons.

Any amounts withheld under the backup withholding rules generally will be allowed as a refund or a credit against a U.S. holder's U.S. federal income tax liability if the required information is furnished by the U.S. holder on a timely basis to the IRS.

THE DISCUSSION ABOVE IS A GENERAL SUMMARY. IT DOES NOT COVER ALL TAX MATTERS THAT MAY BE OF IMPORTANCE TO A U.S. HOLDER. EACH U.S. HOLDER IS URGED TO CONSULT ITS OWN TAX ADVISOR ABOUT THE TAX CONSEQUENCES TO IT OF AN INVESTMENT IN ADSs IN LIGHT OF THE INVESTOR'S OWN CIRCUMSTANCES.

DIVIDENDS AND PAYING AGENTS

Not applicable.

STATEMENTS BY EXPERTS

Not applicable.

DOCUMENTS ON DISPLAY

We filed a Notice of Voluntary Delisting with The NASDAQ Stock Market on March 5, 2014 and trading in our ADSs on NASDAQ was suspended on March 7, 2014. We filed a Form 25 with the SEC on March 17, 2014, to terminate registration of our ordinary shares and ADSs under Section 12(b) of the Exchange Act and intend to file a Form 15 with the SEC promptly after this annual report is filed with the SEC to terminate registration of our ordinary shares and ADSs under Section 12(g) of the Exchange Act and to suspend our duty to file reports under Sections 13(a) and 15(d) of the Exchange Act. We expect that this will be the final report we will be required to file under Sections 13(a) and 15(d) of the Exchange Act.

We were previously subject to the periodic reporting and other informational requirements of the Exchange Act applicable to a foreign private issuer. Under the Exchange Act, we have been required to file annual reports on Form 20-F within four months of our fiscal year end and to submit other reports and information under cover of Form 6-K with the SEC. Copies of the registration statements, their accompanying exhibits, as well as such reports and other information previously filed by us may be inspected without charge and may be obtained at prescribed rates at the SEC's Public Reference Room located at 450 Fifth Street, N.W., Room 1200, Washington, D.C. 20549. You

may obtain information regarding the Washington, D.C. Public Reference Room by calling the SEC at 1-800-SEC-0330 or by contacting the SEC at its website at www.sec.gov.

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As a foreign private issuer, we were exempt under the Exchange Act from, among other things, the rules prescribing the furnishing and content of proxy statements, and our executive officers, directors and principal shareholders have been exempt from the reporting and short-swing profit recovery provisions contained in Section 16 of the Exchange Act. In addition, we were not required under the Exchange Act to file periodic reports and consolidated financial statements with the SEC as frequently or as promptly as United States companies whose securities are registered under the Exchange Act.

SUBSIDIARY INFORMATION
Not applicable.

ITEM 11. QUANTITATIVE AND QUALITATIVE DISCLOSURES ABOUT MARKET RISK
Market risk represents the risk of loss arising from adverse changes in market rates and foreign exchange rates. The carrying amounts of cash and cash equivalents, accounts receivable and other receivables, and the interest rate on our debt with floating rates represents our principal exposure to credit risk in relation to our financial assets.
As of December 31, 2013, substantially all of our cash and cash equivalents were held in accounts at financial institutions located in the Republic of Italy, Switzerland and in the United States, which we believe are of acceptable credit quality. The goals of our investment policy are liquidity and capital preservation. To achieve this objective, we invest our cash in liquid instruments that meet high credit quality standards and generally have a maturity of less than three months from the date of purchase. We are exposed to exchange rate risk with respect to certain of our cash balances, accounts receivable and accounts payable that are denominated in the U.S. dollar. As of December 31, 2013, we held a cash balance of $3.99 million, accounts receivable of $2.95 million and accounts payable of $7.28 million that were denominated in U.S. dollars. As the net positions of our unhedged foreign currency transactions fluctuate, our earnings might be negatively affected. As of December 31, 2013, our foreign currency transactions were minimal and changes to the exchange rate between the U.S. dollar and Euro would have an immaterial effect on our earnings. If the U.S. dollar were 10% stronger against the Euro, our net assets balance would increase by approximately €0.03 million as of December 31, 2013.
As of December 31, 2013, we had floating debts in the principal amount of €1.74 million. Our exposure includes changes in interest rates, as borrowing under our debts bears interest at floating rates based on Euribor plus an applicable margin. The rate is currently variable based on Euribor interest rates, subject to certain minimums, that range from 1.09% to 1.49%. Each 100 basis point increase in interest rates will cause interest payments in 2014 to increase by approximately €0.13 million. Substantially all of our current revenue generating transactions and substantially all of our assets and liabilities are denominated in the Euro. In the future, we expect to transact business in U.S. dollars and other currencies. The value of the Euro against the U.S. dollar and other currencies may fluctuate and is affected by, among other things, changes in political and economic conditions. Any change in the value of the Euro relative to other currencies in which we transact business in the future could materially and adversely affect our cash flow, revenues and financial condition. To the extent that we hold assets denominated in U.S. dollars, any appreciation of the Euro against the U.S. dollar could result in a charge to our operating results and a reduction in the value of our U.S. dollar denominated assets upon re-measurement.

ITEM 12. DESCRIPTION OF SECURITIES OTHER THAN EQUITY SECURITIES

DEBT SECURITIES
Not applicable.

WARRANTS AND RIGHTS
Not applicable.

OTHER SECURITIES

Not applicable.

AMERICAN DEPOSITARY SHARES

The Bank of New York Mellon serves as the depositary for our ADS program and collects fees for depositing shares or surrendering ADSs. The Bank of New York Mellon is headquartered at One Wall Street, New York, New York, 10286.

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A deposit agreement among us, the ADS depositary and the ADS holders sets out the ADS holder rights as well as the rights and obligations of the depositary. New York law governs the deposit agreement and the ADSs. A copy of the deposit agreement is incorporated by reference as an exhibit to this annual report.

Each ADS represents one ordinary share. Holders of ADSs will not be able to independently exercise voting rights attaching to the ordinary shares evidenced by the ADSs. Holders of ADSs will only have the right to instruct the ADS depositary, as the holders' representative, to exercise these voting rights. The ADS depositary will mail to all ADS record holders a notice containing a summary of all information included in any notice of a shareholders' meeting received by the ADS depositary, and will solicit proxies from ADS holders for instructions on how to vote its ordinary shares at our shareholder meetings.

Holders of ADSs may not be able to participate in rights offerings and may experience dilution of their holdings as a result. We may from time to time offer to our existing shareholders the right to purchase our securities. Under our deposit agreement for the ADSs, the ADS depositary will not offer those rights to ADS holders unless both the rights and the underlying securities to be distributed to ADS holders are either registered under the Securities Act or exempt from registration under the Securities Act with respect to all holders of ADSs. We are under no obligation to file a registration statement with respect to any such rights or underlying securities or to endeavor to cause such a registration statement to be declared effective. In addition, we may not be able to take advantage of any exemptions from registration under the Securities Act. Accordingly, holders of our ADSs may be unable to participate in our rights offerings and may experience dilution in their holdings as a result.

ADSs are transferable on the books of the ADS depositary. However, the ADS depositary may close its transfer books at any time or, from time to time, when it deems expedient, in connection with the performance of its duties. In addition, the ADS depositary may refuse to deliver, transfer or register transfers of ADSs generally when our books or the books of the ADS depositary are closed, or if we or the ADS depositary deem it advisable to do so under any requirement of law, any government or governmental body, any provision of the deposit agreement, or for any other reason.

Fees and Expenses

Pursuant to the terms of the deposit agreement, the holders of ADSs are required to pay the following fees:

Persons depositing or withdrawing shares must pay:	For:
$5.00 (or less) per 100 ADSs (or portion of 100 ADSs)	Issuance of ADSs, including issuances resulting from a distribution of shares or rights; or Cancellation of ADSs for the purpose of withdrawal, including if the deposit agreement terminates Any cash distribution to ADS holders
$0.02 (or less) per ADS	For depositary services accrued on the last day of each calendar year to the extent no fee was charged for any cash distribution
A fee equivalent to the fee that would be payable if securities distributed to you had been shares and the shares had been deposited for issuance of ADSs	Distribution of securities distributed to holders of deposited securities which are distributed by the depositary to ADS holders
Registration or transfer fees	Transfer and registration of shares on our share register to or from the name of the depositary or its agent when you deposit or withdraw shares
Expenses of the ADS depositary	Cable, telex and facsimile transmissions (when expressly provided in the deposit agreement), or Converting foreign currency to U.S. dollars As necessary

Taxes and other governmental charges the depositary or the custodian have to pay on any ADS or share underlying an ADS, for example, stock transfer taxes, stamp duty or withholding taxes	
Any charges incurred by the ADS depositary or its agents for servicing the deposited securities	As necessary

The deposit agreement, including the fees listed above, may be amended from time to time by agreement between the ADS depositary and us, and without consent from holders of the ADSs. In addition, both we and the ADS depositary have the ability to terminate the deposit agreement upon proper notice given to the other party.

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We and the ADS depositary entered into an amendment dated March 21, 2014 to the deposit agreement dated as of June 15, 2005, or the deposit agreement, which amendment will be effective on April 13, 2014. Upon effectiveness, the amendment to the deposit agreement will reduce from one year to 60 days the period that must elapse between the date of termination of the deposit agreement and the date on which the ADS depositary may sell the ordinary shares of the Company it then holds and provides for the ADS depositary to accept any offer by a subsidiary of Jazz to purchase those shares at a price of no less than $57.00 per ordinary share, unless the ADS depositary has received what it considers to be a superior bona fide offer from another party. The ADS depositary has given notice such that the deposit agreement will terminate at 5:00 pm (Eastern time) on April 13, 2014. While the date or dates on which the ADS depositary will sell the remaining shares have not been determined, the sale will not occur before June 13, 2014. Jazz Italy has indicated that it intends to offer to purchase ordinary shares from the ADS depositary at a per share price equal to $57.00.

PART II

ITEM 13. DEFAULTS, DIVIDEND ARREARAGES AND DELINQUENCIES
None.

ITEM 14. MATERIAL MODIFICATIONS TO THE RIGHTS OF SECURITY HOLDERS AND USE OF PROCEEDS
None.

ITEM 15. CONTROLS AND PROCEDURES

Disclosure Controls and Procedures
We maintain disclosure controls and procedures that are designed to ensure that information required to be disclosed in our reports filed under the Securities Exchange Act of 1934, as amended, or Exchange Act, is recorded, processed, summarized and reported within the time periods specified in the Securities and Exchange Commission's, or SEC's, rules and forms and that such information is accumulated and communicated to our management to allow for timely decisions regarding required disclosure. In designing and evaluating the disclosure controls and procedures, management recognizes that any controls, no matter how well designed and operated, can provide only reasonable assurance of achieving the desired control objectives, and management is required to apply its judgment in evaluating the cost-benefit relationship of possible controls and procedures.
As required by Rule 13a-15(b) of the Exchange Act, an evaluation of the effectiveness of the design and operation of our disclosure controls and procedures (as defined in Rule 13a-15(e) and Rule 15d-15(e) under the Exchange Act) as of the end of the period covered by this annual report was carried out under the supervision and with the participation of our management, including our Chief Executive Officer and Chief Financial Officer. Based on that evaluation, our Chief Executive Officer and Chief Financial Officer concluded that our disclosure controls and procedures are effective to provide reasonable assurance that information required to be disclosed in the reports we file and submit under the Exchange Act is recorded, processed, summarized and reported within the time periods specified in the SEC's applicable rules and forms and that it is accumulated and communicated to our management, including the Chief Executive Officer and Chief Financial Officer, as appropriate to allow timely decisions regarding required disclosure.
Management's Annual Report on Internal Control over Financial Reporting
Our management is responsible for establishing and maintaining adequate internal control over financial reporting. Our internal control system was designed to provide reasonable assurance to our management and board of directors regarding the preparation and fair presentation of published consolidated financial statements. All internal control systems, no matter how well designed, have inherent limitations. Therefore, even those systems determined to be effective can provide only reasonable assurance with respect to consolidated financial statement preparation and

presentation.

Our management assessed the effectiveness of our internal control over financial reporting as of December 31, 2013. In making this assessment, it used the criteria set forth by the Committee of Sponsoring Organizations of the Treadway Commission (COSO) in Internal Control-Integrated Framework (1992). Based on our assessment, management has concluded that, as of December 31, 2013, our internal control over financial reporting is effective. The effectiveness of internal controls over Financial Reporting as of December 31, 2013 has been audited by Reconta Ernst & Young S.p.A., an independent registered public accounting firm, in their report on our internal control over financial reporting, which follows below.

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Attestation Report of Registered Public Accounting Firm
To the Board of Directors and Shareholders of Gentium S.p.A.
We have audited Gentium S.p.A.'s internal control over financial reporting as of December 31, 2013, based on criteria established in Internal Control-Integrated Framework issued by the Committee of Sponsoring Organizations of the Treadway Commission (1992 framework)(the COSO criteria). Gentium's S.p.A. management is responsible for maintaining effective internal control over financial reporting, and for its assessment of the effectiveness of internal control over financial reporting included in the accompanying Management's annual report on internal control over financial reporting. Our responsibility is to express an opinion on the company's internal control over financial reporting based on our audit.
We conducted our audit in accordance with the standards of the Public Company Accounting Oversight Board (United States). Those standards require that we plan and perform the audit to obtain reasonable assurance about whether effective internal control over financial reporting was maintained in all material respects. Our audit included obtaining an understanding of internal control over financial reporting, assessing the risk that a material weakness exists, testing and evaluating the design and operating effectiveness of internal control based on the assessed risk, and performing such other procedures as we considered necessary in the circumstances. We believe that our audit provides a reasonable basis for our opinion.
A company's internal control over financial reporting is a process designed to provide reasonable assurance regarding the reliability of financial reporting and the preparation of financial statements for external purposes in accordance with generally accepted accounting principles. A company's internal control over financial reporting includes those policies and procedures that (1) pertain to the maintenance of records that, in reasonable detail, accurately and fairly reflect the transactions and dispositions of the assets of the company; (2) provide reasonable assurance that transactions are recorded as necessary to permit preparation of financial statements in accordance with generally accepted accounting principles, and that receipts and expenditures of the company are being made only in accordance with authorizations of management and directors of the company; and (3) provide reasonable assurance regarding prevention or timely detection of unauthorized acquisition, use or disposition of the company's assets that could have a material effect on the financial statements.
Because of its inherent limitations, internal control over financial reporting may not prevent or detect misstatements. Also, projections of any evaluation of effectiveness to future periods are subject to the risk that controls may become inadequate because of changes in conditions, or that the degree of compliance with the policies or procedures may deteriorate.
In our opinion, Gentium S.p.A. maintained, in all material respects, effective internal control over financial reporting as of December 31, 2013, based on the COSO criteria.
We also have audited, in accordance with the standards of the Public Company Accounting Oversight Board (United States), the 2013 consolidated financial statements of Gentium S.p.A. and our report dated March 31, 2014 expressed an unqualified opinion thereon.

/s/ Reconta Ernst & Young S.p.A.
Milan, Italy
March 31, 2014

Changes in Internal Control over Financial Reporting

There has not been any change in our internal control over financial reporting identified in the evaluation required by Rule 13a-15 or Rule 15d-15 of the Exchange Act that occurred during the period covered by this annual report that has materially affected, or is reasonably likely to materially affect, our internal control over financial reporting.

ITEM 16A. AUDIT COMMITTEE FINANCIAL EXPERT

We have both a board of statutory auditors and an audit committee. Our board of directors has determined that Ms. Bigio, a member of our audit committee and an independent director, qualifies as an “audit committee financial expert” within the meaning of this Item 16A.

ITEM 16B. CODE OF ETHICS

We have adopted a code of ethics, as defined in Item 16B of Form 20-F under the Exchange Act, that is applicable to, among others, our Chief Executive Officer and Chief Financial Officer. Copies of this code of ethics are available upon request by writing to us at the address on the cover page of this annual report. We have also posted the code of ethics on our website at www.gentium.it. Material appearing on our website is not incorporated by reference into this annual report. If we amend the

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provisions of our code of ethics, or if we grant any waiver of such provisions, we will disclose such amendment or waiver on our website at the same address.

ITEM 16C. PRINCIPAL ACCOUNTANT FEES AND SERVICES

The following table sets forth the fees contractually agreed to with our independent auditors, Reconta Ernst & Young S.p.A., for the years ended December 31, 2012 and 2013 (in thousands):

	Year ended December 31,	
	2012	2013
Audit fees	€180	€180
Audit-related fees	—	—
Tax fees	—	—
All other fees	—	—
Total	€180	€180

In the above table, in accordance with the SEC's definitions and rules, "audit fees" are fees for professional services in respect of the audit of a company's consolidated financial statements, and for services that are normally provided by the accountant in connection with statutory and regulatory filings or engagements. Reconta Ernst & Young S.p.A. did not provide any tax compliance services or advice on specific changes in tax regulations for the years ended December 31, 2012 and 2013.

To help ensure the independence of our independent registered public accounting firm, the audit committee is required to pre-approve all audit and non-audit services to be performed for us by our independent registered public accounting firm. All audit and permitted non-audit services, including the fees and terms thereof, to be performed by our independent registered public accounting firm must be approved in advance by the audit committee and submitted to shareholders for final approval.

ITEM 16D. EXEMPTION FROM THE LISTING STANDARDS FOR AUDIT COMMITTEES

Under Italian law, our shareholders, not the audit committee, must be the party that appoints, terminates and determines the compensation for our independent accountants, although our audit committee does make recommendations on such matters to our board of directors, which in turn makes recommendations to our shareholders. As a result, our audit committee is not able to perform all of the duties required by Rule 10A-3 of the Exchange Act. Our audit committee has established procedures for the receipt, retention and treatment of complaints regarding accounting, internal accounting controls and auditing matters, and has authority to engage independent counsel and other advisors and to determine the compensation of such advisors, as well as its ordinary administrative expenses, and together with the board of statutory auditors, oversees our independent accountants (including resolution of disagreements between management and the independent accountants regarding financial reporting). Rule 10A-3 provides that foreign private issuers with a board of statutory auditors established in accordance with local law or listing requirements and which meets specified requirements with regard to independence and responsibilities (including the performance of most of the specific tasks assigned to audit committees by the rule, to the extent prohibited by local law) are exempt from the audit committee requirements established by the rule. Our board of directors has determined that, because of the existence and nature of our board of statutory auditors, together with the performance of other duties under Rule 10A-3 by our shareholders and the performance of the remaining duties by our audit committee, we either satisfy Rule 10A-3 or qualify for an exemption from the audit committee requirements of Rule 10A-3, as provided in Rule 10A-3.

ITEM 16E. PURCHASES OF EQUITY SECURITIES BY THE ISSUER AND AFFILIATED PURCHASERS

Not applicable.

ITEM 16F. CHANGE IN CERTIFYING ACCOUNTANT

Not applicable.

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ITEM 16G. CORPORATE GOVERNANCE

Not applicable.

ITEM 16F. MINE SAFETY DISCLOSURE

Not applicable.

PART III

ITEM 17. CONSOLIDATED FINANCIAL STATEMENTS
Our audited financial statements have been prepared in accordance with Item 18 of Form 20-F.

ITEM 18. CONSOLIDATED FINANCIAL STATEMENTS
Our audited financial statements are included in this annual report beginning on page F-1.

Item 19. Exhibits

Exhibit	Description
Charter Documents	
1.1	Articles of Association of Gentium S.p.A., formerly known as Pharma Research S.r.l., dated November 11, 1993, incorporated by reference to Exhibit 3(i) to the Registration Statement on Form F-1, Registration No. 333-122233, previously filed with the SEC on January 24, 2005.
1.2	Amended and Restated Bylaws of Gentium S.p.A. dated May 9, 2011, incorporated by reference to Exhibit 1.2 to the Annual Report on Form 20-F previously filed with the SEC on March 30, 2012.
American Depositary Share Documents	
2.1	Form of Deposit Agreement among Gentium S.p.A., The Bank of New York and the owners and beneficial owners from time to time of American Depositary Receipts (including as an exhibit the form of American Depositary Receipt), incorporated by reference to Exhibit 4.6 to Amendment No. 5 to the Registration Statement on Form F-1, Registration No. 333-122233, previously filed with the SEC on June 9, 2005.
2.2	Form of American Depositary Receipt (see Exhibit 2.1).
2.3	Amendment, dated as of March 21, 2014, to the Deposit Agreement dated as of June 15, 2005 among Gentium S.p.A., The Bank of New York Mellon and the owners and beneficial owners from to time of American Depositary Receipts.
Loan Agreements	
4.1	Loan Agreement between Banca Nazionale del Lavoro S.p.A. and Gentium S.p.A. dated June 14, 2006 incorporated by reference to Exhibit 10.7.3 to the Registration Statement on Form F-3, Registration No. 333-135622, previously filed with the SEC on July 6, 2006.
4.2	Loan Agreement for €230,000 with Banca Intesa S.p.A., dated December 20, 2006, incorporated by reference to Exhibit 2 to the report on Form 6-K, previously filed with the SEC on February 2, 2007.
4.3	Loan Agreement for €500,000 with Banca Intesa S.p.A., dated December 20, 2006, incorporated by reference to Exhibit 3 to the report on Form 6-K, previously filed with the SEC on February 2, 2007.
4.4	Loan Agreement for €225,000 with Banca Intesa S.p.A., dated December 20, 2006, incorporated by reference to Exhibit 4 to the report on Form 6-K, previously filed with the SEC on February 2, 2007.
4.5	Loan Agreement, dated June 30, 2006, between San Paolo IMI S.p.A. and Gentium S.p.A., incorporated by reference to Exhibit 4.43 to the Annual Report on Form 20-F for the year ended December 31, 2006, previously filed with the SEC on April 30, 2007.
4.6	

	Loan Agreement (English translation), dated November 11, 2013, between Banca Popolare Di Sondrio and Gentium S.p.A.
License and Distribution Agreements	
4.7.1	License and Supply Agreement by and between Gentium S.p.A. 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.

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4.7.2	Letter Agreement, dated October 12, 2007, between Gentium S.p.A. 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.
4.7.3*	Amendment to License and Supply Agreement and Letter Agreement, dated December 7, 2001 and October 12, 2007, respectively, effective January 7, 2010, between Gentium S.p.A. 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.
4.7.4**	Amendment No. 2 to Letter Agreement dated October 12, 2007, as amended, dated May 11, 2012, between Gentium S.p.A. and Sigma-Tau Pharmaceuticals, Inc.
4.8	Distribution Agreement, dated December 28, 2006, between Gentium S.p.A. and Crinos S.p.A., incorporated by reference to Exhibit 6 to the report on Form 6-K, previously filed with the SEC on January 3, 2007.
4.9*	Technical Transfer Services Agreement, dated February 2, 2009, between Gentium S.p.A. and Patheon Italia S.p.A, incorporated by reference to Exhibit 4.21 to the Annual Report on Form 20-F for the year ended December 31, 2008, previously filed with the SEC on March 31, 2009.
4.10.1*	Master Contract Clinical Research Agreement, dated September 29, 2009, between US Oncology Clinical Development and Gentium S.p.A., incorporated by reference to Exhibit 2 to the report on Form 6-K, previously filed with the SEC on December 1, 2009.
4.10.2**	Amendment, dated October 24, 2013, to Master Contract Clinical Research Agreement dated December 29, 2009 between US Oncology Clinical Development and Gentium S.p.A.
4.11**	Manufacturing Services Agreement, effective September 15, 2013, between Patheon UK Limited and Gentium S.p.A.
Leases	
4.12	Commercial Lease Contract between Gentium S.p.A. and Sirton Pharmaceuticals S.p.A. dated January 1, 2005, incorporated by reference to Exhibit 10.33 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.
4.13	Commercial Lease Contract between Gentium S.p.A. and FinSirton S.p.A. (now F3F S.r.l.) dated January 1, 2012, incorporated by reference to Exhibit 4.21 to the Annual Report on Form 20-F previously filed with the SEC on March 30, 2012.
Miscellaneous	
4.14	Form of indemnification agreement between Gentium S.p.A. and certain of its officers and directors, 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.
4.15	Tender Offer Agreement, dated December 19, 2013, by and among Jazz Pharmaceuticals plc, Jazz Pharmaceuticals Italy S.r.l. and Gentium S.p.A., incorporated by reference to Exhibit 2.1 to the Form 8-K/A filed by Jazz Pharmaceuticals plc with the SEC on December 20, 2013.
4.16	Employment agreement by and between Gentium S.p.A. 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, incorporated by reference to Exhibit 99.(e)(4) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.17	Employment agreement by and between Gentium S.p.A. and Salvatore Calabrese, dated September 7, 2007, incorporated by reference to Exhibit 99.(e)(5) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.18	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, incorporated by reference to Exhibit 99.(e)(6) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.19	

	Employment agreement by and between Gentium S.p.A. 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, incorporated by reference to Exhibit 99.(e)(7) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.20	Transition, Amendment and Release Agreement, dated December 19, 2013, by and between Gentium GmbH and Dr. Khalid Islam, incorporated by reference to Exhibit 99.(e)(8) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.21	Consultancy Agreement, dated December 19, 2013, by and between Gentium S.p.A. and Dr. Khalid Islam, incorporated by reference to Exhibit 99.(e)(9) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.22	Retention and Amendment Agreement, dated December 19, 2013, by and between Gentium S.p.A. and Salvatore Calabrese, incorporated by reference to Exhibit 99.(e)(10) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.

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4.23	Retention and Amendment Agreement, dated December 19, 2013, by and between Gentium GmbH and Giorgio Mosconi, incorporated by reference to Exhibit 99.(e)(11) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.24	Retention and Amendment Agreement, dated December 19, 2013, by and between Gentium GmbH and Adrian Haigh, incorporated by reference to Exhibit 99.(e)(12) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
Subsidiaries	
8.1	List of Subsidiaries.
Certifications and Consents	
12.1	Chief Executive Officer Certification pursuant to Section 302 of the Sarbanes-Oxley Act of 2002.
12.2	Chief Financial Officer Certification pursuant to Section 302 of the Sarbanes-Oxley Act of 2002.
13.1	Chief Executive Officer Certification pursuant to Section 906 of the Sarbanes-Oxley Act of 2002.
13.2	Chief Financial Officer Certification pursuant to Section 906 of the Sarbanes-Oxley Act of 2002.
XBRL Information	
101.INS	XBRL Instance Document.
101.SCH	XBRL Taxonomy Extension Schema Document.
101.CAL	XBRL Taxonomy Extension Calculation Linkbase Document.
101.DEF	XBRL Taxonomy Extension Definition Linkbase Document.
101.LAB	XBRL Taxonomy Extension Label Linkbase Document.
101.PRE	XBRL Taxonomy Extension Presentation Linkbase Document.

* Confidential treatment has been granted for portions of this exhibit. Omitted portions have been filed separately with the SEC.

** Confidential treatment has been requested for portions of this exhibit. Omitted portions have been filed separately with the SEC.

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GENTIUM S.p.A.
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REPORT OF INDEPENDENT REGISTERED PUBLIC ACCOUNTING FIRM
To the Board of Directors and Shareholders of Gentium S.p.A.

We have audited the accompanying consolidated balance sheets of Gentium S.p.A. (the "Company") as of December 31, 2013 and 2012, and the related consolidated statements of income, shareholders' equity, and cash flows for each of the three years in the period ended December 31, 2013. These financial statements are the responsibility of the Company's management. Our responsibility is to express an opinion on these financial statements based on our audits.

We conducted our audits in accordance with the standards of the Public Company Accounting Oversight Board (United States). Those standards require that we plan and perform the audit to obtain reasonable assurance about whether the financial statements are free of material misstatement. An audit includes examining, on a test basis, evidence supporting the amounts and disclosures in the financial statements. An audit also includes assessing the accounting principles used and significant estimates made by management, as well as evaluating the overall financial statement presentation. We believe that our audits provide a reasonable basis for our opinion.

In our opinion, the financial statements referred to above present fairly, in all material respects, the consolidated financial position of Gentium S.p.A. at December 31, 2013 and 2012 and the consolidated results of its operations and its cash flows for each of the three years in the period ended December 31, 2013, in conformity with U.S. generally accepted accounting principles.

We also have audited, in accordance with the standards of the Public Company Accounting Oversight Board (United States), Gentium S.p.A.'s internal control over financial reporting as of December 31, 2013, based on criteria established in Internal Control-Integrated Framework (1992 framework) issued by the Committee of Sponsoring Organizations of the Treadway Commission and our report dated March 31, 2014 expressed an unqualified opinion thereon.

/s/ Reconta Ernst & Young S.p.A.

Milan, Italy
March 31, 2014

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GENTIUM S.p.A.
CONSOLIDATED BALANCE SHEETS
(In thousands, except share amounts)

	As of December 31,	
	2012	2013
ASSETS		
Current assets:		
Cash and cash equivalents	€12,485	€22,038
Short-term deposit	—	4,000
Accounts receivable, net of allowance of nil as of December 31, 2012 and 2013	4,870	8,006
Accounts receivable from related parties, net of allowance of €765 as of December 31, 2012 and 2013	216	1,541
Inventories, net of allowance of €332 and €374 as of December 31, 2012 and 2013	1,990	2,448
Prepaid expenses and other current assets	1,428	1,768
Deferred tax assets	—	2,539
Total current assets	20,989	42,340
Property, manufacturing facility and equipment, net	7,449	7,581
Deferred tax assets, non-current	—	15,324
Intangible and other non-current assets	200	226
Total assets	€28,638	€65,471
LIABILITIES AND SHAREHOLDERS' EQUITY		
Current liabilities:		
Accounts payable	€4,453	€9,939
Accounts payable to related parties	5	5
Accrued expenses and other current liabilities	1,728	3,684
Income taxes payable	11	370
Deferred revenues	163	11
Current maturities of long-term debt	409	350
Total current liabilities	6,769	14,359
Long-term debt, net of current maturities	1,135	1,386
Deferred tax liabilities	—	28
Termination indemnities	384	292
Other long-term liabilities	—	190
Total liabilities	8,288	16,255
Commitments and contingencies (Note 15)		
Shareholders' equity:		
Share capital (19,656,317 shares authorized as of December 31, 2012 and 2013; 15,038,483 and 15,555,131 shares issued and outstanding at December 31, 2012 and 2013, each of no par value)	112,421	116,686
Accumulated deficit	(92,071)	(67,470)
Total shareholders' equity	20,350	49,216
Total liabilities and shareholders' equity	€28,638	€65,471

The accompanying notes are an integral part of these consolidated financial statements.

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GENTIUM S.p.A.
CONSOLIDATED STATEMENTS OF INCOME
(In thousands, except share and per share amounts)

	For the Year Ended December 31,		
	2011	2012	2013
Revenues:			
API product sales	€4,848	€4,856	€6,172
NPP product sales	16,886	22,774	33,653
Total product sales	21,734	27,630	39,825
Other revenues	123	152	490
Other revenues from related party	2,026	1,257	2,602
Total revenues	23,883	29,039	42,917
Operating costs and expenses:			
Cost of goods sold	6,035	5,778	6,055
Research and development	5,533	10,531	15,672
Selling, general and administrative	7,727	10,829	12,883
Charges from related parties	222	186	189
Depreciation and amortization	870	1,003	1,031
Total costs and expenses	20,387	28,327	35,830
Operating income	3,496	712	7,087
Foreign currency exchange gain/(loss), net	46	(67)	55
Interest income/(expense), net	(21)	155	237
Income before income tax provision/(benefit)	3,521	800	7,379
Income tax provision/(benefit)	811	26	(17,222)
Net income	€2,710	€774	€24,601
Net income per share:			
Basic	€0.18	€0.05	€1.61
Diluted	€0.18	€0.05	€1.48
Weighted-average shares used to compute net income per share:			
Basic	14,964,021	15,014,411	15,261,799
Diluted	15,340,859	15,639,890	16,602,743

The accompanying notes are an integral part of these consolidated financial statements.

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GENTIUM S.p.A.
CONSOLIDATED STATEMENTS OF SHAREHOLDERS' EQUITY
(In thousands)

	Shares	Amount	Accumulated Deficit	Total Shareholders' Equity
Balance at December 31, 2010	14,956	€108,485	€(95,555)	€12,930
Stock-based compensation	—	1,666	—	1,666
Issuance of common stock upon exercise of stock options	13	77	—	77
Net income	—	—	2,710	2,710
Balance at December 31, 2011	14,969	€110,228	€(92,845)	€17,383
Stock-based compensation	—	1,916	—	1,916
Issuance of common stock upon exercise of stock options	69	277	—	277
Net income	—	—	774	774
Balance at December 31, 2012	15,038	€112,421	€(92,071)	€20,350
Stock-based compensation	—	1,816	—	1,816
Issuance of common stock upon exercise of stock options	517	2,449	—	2,449
Net income	—	—	24,601	24,601
Balance at December 31, 2013	15,555	€116,686	€(67,470)	€49,216

The accompanying notes are an integral part of these consolidated financial statements.

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GENTIUM S.p.A.
CONSOLIDATED STATEMENTS OF CASH FLOWS
(In thousands)

	For the Year Ended December 31,		
	2011	2012	2013
Operating activities			
Net income	€2,710	€774	€24,601
Adjustments to reconcile net income to net cash provided by operating activities:			
Write-down of inventory	337	173	228
Unrealized foreign exchange loss/(gain)	(48)	36	(37)
Release of inventory reserve (net)	—	(630)	(186)
Depreciation and amortization	1,320	1,456	1,474
Stock-based compensation	1,666	1,916	1,816
Gain on fixed asset disposal	(14)	—	(306)
Release of allowance for doubtful accounts	—	(27)	—
Provision for income taxes	580	117	613
Release of income tax provision	—	(91)	—
Deferred tax assets	—	—	(17,863)
Deferred tax liabilities	—	—	28
Changes in operating assets and liabilities:			
Accounts receivable	(1,281)	976	(4,549)
Inventories	(919)	1,413	(498)
Prepaid expenses and other current and non-current assets	42	(358)	(223)
Accounts payable, accrued expenses and income tax payables	(648)	(2,010)	6,656
Termination indemnities	(134)	8	(92)
Deferred revenue	(1,210)	(331)	(152)
Net cash provided by operating activities	2,401	3,422	11,510
Investing activities			
Capital expenditures	(718)	(611)	(850)
Proceeds from sale of marketing authorization, trademark and equipment	62	—	310
Short-term deposit	—	—	(4,000)
Sales of marketable securities	263	—	—
Net cash used in investing activities	(393)	(611)	(4,540)
Financing activities			
Proceeds from stock option exercises	77	277	2,449
Repayments of long-term debt	(808)	(505)	(408)
Proceeds from long-term debt	—	—	600
Principal payment of capital lease obligation	(70)	(21)	—
Net cash provided by/(used in) financing activities	(801)	(249)	2,641
Net increase in cash and cash equivalents	1,207	2,562	9,611
Effect of exchange rates on cash and cash equivalents	41	(67)	(58)
Cash and cash equivalents, at beginning of period	8,742	9,990	12,485
Cash and cash equivalents, at end of period	€9,990	€12,485	€22,038
Supplemental disclosure of cash flow information:			
Cash paid for interest	€66	€44	€20
Cash paid for income taxes	€375	€1,011	€11

The accompanying notes are an integral part of these consolidated financial statements.

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GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS

1. BUSINESS AND BASIS OF PRESENTATION

Business

We are a biopharmaceutical company primarily focused on the development of defibrotide, a sodium salt of a complex mixture of single- and double-stranded oligodeoxyribonucleotides derived from porcine DNA. Our development of defibrotide has been directed to the treatment and prevention of hepatic veno-occlusive disease, or VOD, a potentially life-threatening complication of hematopoietic stem cell transplantation, or HSCT. Stem cell transplantation is a frequently used treatment modality for hematologic cancers and other conditions in both adults and children. Certain high-dose conditioning regimens used as part of HSCT can damage the lining cells of hepatic vessels which is thought to lead to the development of VOD, a blockage of the small vessels in the liver, that leads to liver failure and can result in significant dysfunction in other organs such as the kidneys and lungs. The condition is also referred to as “sinusoidal obstruction syndrome.” Severe VOD is the most extreme form of VOD and is associated with multi-organ failure and high rates of morbidity and mortality.

On December 19, 2013, we entered into a definitive tender offer agreement with Jazz Pharmaceuticals Public Limited Company, or Jazz, and Jazz Pharmaceuticals Italy S.p.A., or Jazz Italy, a wholly-owned subsidiary of Jazz, pursuant to which Jazz Italy made an offer to purchase all our outstanding ordinary shares and American Depositary Shares, or ADSs, each representing one ordinary share, at a purchase price of $57.00 per ordinary share and per ADS (without duplication for ordinary shares underlying ADSs). On February 21, 2014, Jazz Italy completed its tender offer, having acquired approximately 98 percent of our combined ordinary shares and ADSs. Jazz is now our indirect majority shareholder. We filed a Notice of Voluntary Delisting with The NASDAQ Stock Market on March 5, 2014, and trading in our ADSs on The NASDAQ Global Market, or NASDAQ, was suspended on March 7, 2014. We filed a Form 25 with the Securities and Exchange Commission, or the SEC, on March 17, 2014 to terminate registration of our ordinary shares and ADSs under Section 12(b) of the Securities Exchange Act of 1934, as amended, or the Exchange Act, and intend to file a Form 15 with the SEC promptly after this annual report is filed with the SEC to terminate registration of our ordinary shares and ADSs under Section 12(g) of the Exchange Act and to suspend our duty to file reports under Sections 13(a) and 15(d) of the Exchange Act. We expect that this will be the final report we will be required to file under Sections 13(a) and 15(d) of the Exchange Act.

We and the ADS depositary entered into an amendment dated March 21, 2014 to the deposit agreement dated as of June 15, 2005, or the deposit agreement, which amendment will be effective on April 13, 2014. Upon effectiveness, the amendment to the deposit agreement will reduce from one year to 60 days the period that must elapse between the date of termination of the deposit agreement and the date on which the ADS depositary may sell the ordinary shares of the Company it then holds and provides for the ADS depositary to accept any offer by a subsidiary of Jazz to purchase those shares at a price of no less than $57.00 per ordinary share, unless the ADS depositary has received what it considers to be a superior bona fide offer from another party. The ADS depositary has given notice such that the deposit agreement will terminate at 5:00 pm (Eastern time) on April 13, 2014. While the date or dates on which the ADS depositary will sell the remaining shares have not been determined, the sale will not occur before June 13, 2014. Jazz Italy has indicated that it intends to offer to purchase ordinary shares from the ADS depositary at a per share price equal to $57.00.

In October 2013, the European Commission, or EC, granted marketing authorization under exceptional circumstances for our defibrotide product, Defitelio® (defibrotide), for the treatment of severe VOD in adults and children undergoing HSCT therapy. Our wholly-owned subsidiary, Gentium GmbH, together with other subsidiaries of Jazz, commenced the launch of Defitelio in Europe in March 2014, starting with Germany and Austria. We expect to launch in additional European countries on a rolling basis during 2014 and 2015 and are engaged in pricing and reimbursement submissions as applicable in preparation for planned launches in those countries. We intend eventually to promote Defitelio in all European markets where it has marketing authorization. To our knowledge, there are currently no approved treatments for VOD in the United States. Defibrotide is being distributed to patients diagnosed

with VOD in the United States through an expanded access program pursuant to a treatment investigational new drug, or IND, protocol, which we call our expanded access program. In addition, we expect to continue to give patients access to defibrotide in other countries where it is not commercially available on a named patient basis, which we refer to refer to as our named patient program.

Unless otherwise indicated or the context otherwise requires, references to “Gentium,” “the Company,” “we,” “us,” and “our” refer to Gentium S.p.A. and its consolidated subsidiary, Gentium GmbH.

GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

Basis of Presentation
The accompanying consolidated financial statements have been prepared in accordance with U.S. generally accepted accounting principles, or GAAP. These consolidated financial statements are denominated in the currency of the European Monetary Union (the Euro or €).
The accompanying consolidated financial statements have been prepared on the assumption that we will continue as a going concern, which contemplates the realization of assets and the satisfaction of liabilities in the normal course of business for the twelve-month period following the date of the balance sheet. Through December 31, 2013, the Company had accumulated losses of €67.47 million. Since 2010, we have been cash flow positive, primarily due to revenue generated from our expanded access and named patient programs. However, if we are unable to successfully commercialize defibrotide, unable to generate sufficient revenue and cash flow through our expanded access and named patient programs, or if we increase expenditures above our current expectations, we may need to obtain additional funding either through arrangements with Jazz or its other subsidiaries, or through debt financings or collaborative agreements with unrelated parties, which may not be available to us on favorable terms, if at all.

2. SUMMARY OF SIGNIFICANT ACCOUNTING POLICIES
Consolidation
Our consolidated financial statements reflect the financial position of Gentium S.p.A. and its wholly-owned subsidiary, Gentium GmbH. All intercompany balances and transactions have been eliminated upon consolidation.
Use of Estimates
The preparation of our consolidated financial statements in accordance with U.S. GAAP requires management to make judgments, estimates and assumptions that may affect the reported amounts of assets and liabilities, and related disclosure of contingent assets and liabilities at the date of the consolidated financial statements and the reported amounts of revenues and expenses during the reporting period. We base our estimates on historical experience and on various other assumptions that are believed to be reasonable under the circumstances. Actual results could differ materially from those estimates.
Segment Information
The Company is managed and operated as one business. The entire business is managed by a single management team that reports to the chief executive officer. The Company does not operate separate lines of business or separate business entities with respect to any of its products. The Company's chief operating decision maker reviews the profits and losses and manages the operations of the Company on an aggregate basis. Accordingly, we have determined that we operate in one business segment, which is the biopharmaceutical industry.
Cash and Cash Equivalents
Cash and cash equivalents include highly liquid investments, readily convertible to cash, that mature within three months or less from date of purchase.
Concentration of Credit Risk and Other Risks and Uncertainties
Financial instruments that potentially subject the Company to concentrations of credit risk consist principally of cash and cash equivalents and trade receivables. The Company limits its investments to short-term low risk instruments. The Company is exposed to credit risk with respect to its trade accounts receivable from sales of defibrotide through its named patient and expanded access programs, which are typically unsecured. As of December 31, 2013, our top two customers accounted for approximately 58% and 14% of our accounts receivable, respectively. As of December 31, 2012, our top two customers accounted for approximately 53% and 11% of our accounts receivable, respectively. We are exposed to risks associated with foreign currency transactions in which we use U.S. dollars to make contract payments denominated in Euros and vice versa. As the net positions of our unhedged foreign currency transactions fluctuate, our earnings might be affected. We currently do not utilize forward exchange contracts or any type of hedging instruments to hedge foreign exchange risk, as we believe our overall exposure is relatively limited. For the year ended December 31, 2013, our top three customers accounted for 53%, 12% and 7% of our product sales,

respectively. For the year ended December 31, 2012, our top three customers accounted for 47%, 11% and 10% of our product sales, respectively. For the year ended December 31, 2011, our top three customers accounted for 56%, 14% and 12% of our product sales, respectively.

GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

The Company is subject to a number of risks common in the biotechnology industry including, but not limited to, the uncertainty as to whether Defitelio will become a successful commercial product, our ability to generate projected revenue through our named patient and expanded access programs, our dependence on corporate partners and key personnel, protection of proprietary technology, compliance with U.S. Food and Drug Administration, or FDA, and other governmental regulations and approval requirements, our ability to obtain financing, if necessary, and potential changes in the health care industry.

Accounts Receivable

Accounts receivable are primarily comprised of amounts due from our wholesale distributors and pharmaceutical companies. Account receivables are recorded net of allowances for distributors' fees where we are not invoiced directly and doubtful accounts. Estimates for distributors' fees are based on contractual terms. Estimates for our allowance for doubtful accounts are determined based on existing contractual payment terms, historical payment patterns of our customers and individual customer circumstances. Amounts determined to be uncollectible are charged or written-off against the reserve.

Inventories

Inventories consist of raw materials, work in process, finished active pharmaceutical ingredients and defibrotide distributed through the named patient and expanded access programs. Inventories are stated at the lower of cost or market value, with cost being determined on an average cost basis, which approximates the first-in, first-out method. Prior to commencing the sale of defibrotide through the named patient and expanded access programs, we expensed all costs associated with the production of defibrotide as research and development expenses. Since signing the agreements associated with the named patient and expanded access programs, we have capitalized the subsequent costs of manufacturing defibrotide as inventory, including costs to convert the existing defibrotide active pharmaceutical ingredients to vials and costs to package and label previously manufactured inventory which had previously been expensed as research and development expenses. Until we sell the inventory, the carrying values of our inventories and our cost of goods sold will reflect only incremental costs incurred subsequent to the signing of these agreements.

The Company periodically reviews its inventories and items that are considered outdated or obsolete which are reduced to their estimated net realizable values. The Company estimates reserves for excess and obsolete inventories based on inventory levels on hand and current and forecast product demand. If an estimate of future product demand suggests that inventory levels will become obsolete, then inventories are reduced to their estimated net realizable values. We also review our inventory for quality assurance and quality control issues identified in the manufacturing process and determine whether a write-down is necessary.

We expense costs relating to the production of clinical products, which are not expected to be sold through the named patient and expanded access programs, as research and development expenses in the period in which they are incurred.

Property, Manufacturing Facility and Equipment

Property, manufacturing facility and equipment are carried at cost, subject to review for impairment whenever events or changes in circumstances indicate that the carrying amount of the asset may not be recoverable. The cost of normal, recurring, or periodic repairs and maintenance activities related to property, manufacturing facility and equipment are expensed as incurred, and the cost for major expenditures for additions and improvements are capitalized if they extend the useful life or capacity of the asset.

The cost of our property, manufacturing facility and equipment also includes a proportionate share of the Company's financing costs. The amount of interest cost to be capitalized for qualifying assets is that portion of the interest cost incurred during the assets' acquisition period that could have been avoided if expenditures for the assets had not been made. Capitalized interest expense is amortized over the same life as the underlying constructed asset.

We depreciate or amortize the cost of our property, manufacturing facility and equipment using the straight-line method over the estimated useful lives of the respective assets, which are summarized as follows:

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GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

Asset Category	Useful Lives
Land	Not depreciated
Buildings	20 years
Plant and Machinery	10 to 15 years
Industrial Equipment	10 years
Furniture and Fixtures	5 to 10 years
Leasehold Improvements	Lesser of the useful life or the term of the respective lease
Internally Developed Software	15 years

When we dispose of property, manufacturing facility and equipment, we remove the associated cost and accumulated depreciation from the related accounts on our consolidated balance sheet and include any resulting gain or loss in our consolidated statements of income.

Computer Software

We capitalize costs of computer software obtained for internal use. Such costs are included in property, manufacturing facility and equipment and are amortized over the estimated useful life of the software.

Intangibles

Intangible assets are stated at cost and amortized on a straight-line basis over the expected useful life of such assets, which is estimated to be five to ten years for licenses and trademarks.

Impairment of Long-lived Assets, including Intangibles

The Company’s long-lived assets consist primarily of property, manufacturing facility and equipment. The Company evaluates its ability to recover the carrying values of long-lived assets used in its business, considering changes in the business environment or other facts and circumstances that suggest the value of such assets may be impaired. If this evaluation indicates the carrying value will not be recoverable, based on the undiscounted expected future cash flows estimated to be generated by these assets, the Company reduces the carrying amount to the estimated fair value.

Revenue Recognition

We recognize revenues when all of the following criteria are met: persuasive evidence of an arrangement exists, delivery has occurred or services have been rendered, our price to the customer is fixed or determinable, and collectability is reasonably assured. Revenues from product sales are recognized upon delivery, when title and risk of loss have passed to the customer. Product revenues are recorded net of applicable reserves for discounts, distributor fees and allowances.

Items deducted from total product sales:

- Distributor fees: We have entered into agreements with distributors to manage defibrotide as an investigational drug on a named patient and expanded access basis. We recognize a fee to distributors based on a contractually determined fixed percentage of sales. These fees are accrued at the time of the sale and offset against product sales and are typically paid within 60 days after the issuance of a sales report. When billed directly to the Company, distributor fees are recorded as accounts payable or accrued expenses on our consolidated balance sheets.
- Cash discounts: We may offer a price discount to a customer if a minimum order quantity is reached in a calendar year. We establish a reserve based on estimates of the amounts earned or to be claimed on the related sales, which is classified under accrued expenses and other current liabilities on our balance sheets, and as a reduction of product sales.
- Product returns: We do not provide our customers with a general right of product return, although we do permit returns if the product is damaged or defective when received by the customer.

Collaborative arrangements with multiple deliverables are divided into separate units of accounting if certain criteria are met, including whether the delivered element has stand-alone value to the customer and whether there is objective and reliable evidence of the fair value of the undelivered item. The consideration received from these arrangements is

allocated among the separate units based on their respective selling prices, and the applicable revenue recognition criteria are applied to each

separate unit. Revenues from collaborative arrangements generally include manufacturing fee arrangements if the research and development efforts ever reach the commercialization phase.

Revenue from non-refundable upfront license fees and milestone payments is recognized as performance occurs and our obligations are completed. In accordance with the specific terms of the Company's obligations under these arrangements, revenue is recognized as the obligation is fulfilled or ratably over the development or manufacturing period. Revenue associated with substantive at-risk milestones is recognized based upon the achievement of the milestones as defined in the respective agreements. Revenue from the reimbursement of research costs under collaborative arrangements is recognized as the related research and development costs are incurred, as provided under the terms of these arrangements.

Advance payments received in excess of amounts earned are recorded on the balance sheets as deferred revenue until earned.

Costs incurred by the Company for shipping and handling are included in cost of goods sold.

Research and Development

Research and development expenditures are charged to operations as incurred. Research and development expenses consist of costs incurred for proprietary and collaborative research and development, including activities such as product registration and investigator-sponsored trials. Research and development expenses include salaries, benefits and other personnel-related costs, clinical trial and related clinical manufacturing expenses, fees paid to clinical research organizations, or CROs, contract and other outside service fees, employee stock-based compensation expenses and allocated facility and overhead costs. Payments we make for research and development services prior to the services being rendered are recorded as prepaid assets on our consolidated balance sheets and are expensed as the services are provided.

Clinical Trial Accruals

The Company accounts for the costs of clinical studies conducted by CROs based on the estimated costs and contractual progress over the life of the individual study. These costs can be a significant component of research and development expenses.

Income Taxes

The Company uses the liability method of accounting for income taxes. Deferred tax assets and liabilities are recognized for the estimated future tax consequences of temporary differences between the financial statements carrying amounts and the respective tax bases. Deferred tax assets and liabilities are measured using enacted tax rates expected to apply to taxable income in the year in which the temporary differences are expected to be recovered or settled. We evaluate the realizability of our deferred tax assets and establish a valuation allowance when it is more likely than not that all or a portion of deferred tax assets will not be realized. We account for uncertain tax positions using a "more-likely-than-not" threshold for recognizing and resolving uncertain tax positions. We evaluate uncertain tax positions and consider various factors, including, but not limited to, changes in tax law, the measurement of tax positions taken or expected to be taken in tax returns, the effective settlement of matters subject to audit, new audit activity and changes in facts or circumstances related to a tax position.

Foreign Currency Transactions

The functional currency of the Company's foreign subsidiary is the Euro and, therefore, there are no translation adjustments in the consolidated financial statements. However, net realized and unrealized gains and losses resulting from foreign currency transactions that are denominated in a currency other than the Company's functional currency, the Euro, are included in the statements of income.

Stock-Based Compensation

The Company recognizes stock-based compensation at fair value. Compensation expense for awards that are ultimately expected to vest is recognized as an expense on a straight-line basis over the requisite service period of the equity compensation award, which is generally the vesting period.

The fair value of the stock options is estimated on the date of grant using a binomial valuation model. The binomial model considers characteristics of fair value option pricing that are not available under the Black-Scholes model. Similar to the Black-Scholes model, the binomial model takes into account variables such as volatility, dividend yield rate, and risk free interest rate. The binomial model also considers the contractual term of the option, the probability that the option will be exercised prior to the end of its contractual life, and the probability of termination or retirement of the option holder in

GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

computing the value of the option and the exchange rate between the Euro and the U.S. dollar. For these reasons, the Company believes that the binomial model provides a fair value that is more representative of actual experience and future expected experience than the value calculated using the Black-Scholes model.

Fair Value of Financial Instruments

The carrying amounts of cash and cash equivalents, short-term deposits, accounts receivable, prepaid expenses, other current assets, accounts payable and accrued expenses approximate fair values due to the short-term maturities of these instruments.

Earnings per Share

Basic earnings per share is based upon the weighted-average number of common shares outstanding and excludes the effect of dilutive common stock issuable from stock options. In computing diluted earnings per share, only potential common shares that are dilutive, or those that reduce earnings per share, are included. The issuance of common stock from stock options is not assumed if the result is anti-dilutive.

Recent Accounting Pronouncements

In July 2013, the Financial Accounting Standards Board, or the FASB, issued Accounting Standards Update, or ASU, No. 2013-11, “Presentation of an Unrecognized Tax Benefit When a Net Operating Loss Carryforward, a Similar Tax Loss, or a Tax Credit Carryforward Exists”, or ASU No. 2013-11, which concludes that, under certain circumstances, unrecognized tax benefits should be presented in the financial statements as a reduction to a deferred tax asset for a net operating loss carryforward, a similar tax loss, or a tax credit carryforward. ASU No. 2013-11 will be effective for us beginning January 1, 2014. We do not anticipate that the adoption of this standard will have a material impact on our financial position.

In March 2013, the FASB issued ASU No. 2013-05, “Parent’s Accounting for the Cumulative Translation Adjustment upon De-recognition of Certain Subsidiaries or Groups of Assets within a Foreign Entity or of an Investment in a Foreign Entity”, or ASU No. 2013-05. The objective of ASU No. 2013-05 is to resolve the diversity in practice regarding the release into net income of the cumulative translation adjustment upon de-recognition of a subsidiary or group of assets within a foreign entity. ASU No. 2013-05 will be effective for us beginning January 1, 2014. We do not anticipate that the adoption of this standard will have an impact on our results of operations or financial position.

3. RELATED PARTIES

Historically, the Company had significant relationships with two privately owned Italian companies: F3F S.r.l. (formerly known as FinSirton S.p.A.) and its wholly-owned subsidiary, Sirton Pharmaceuticals S.p.A. (now Vifarma S.p.A.). F3F S.r.l., the parent company of several businesses, was one of the Company’s largest shareholders at December 31, 2013, with approximately 16% ownership at that date, and was originally the Company’s sole shareholder. The Company’s former Chief Executive Officer and Chairperson, Dr. Laura Ferro may be deemed to control F3F S.r.l. In addition, Dr. Ferro previously served as a member of Sirton’s (now Vifarma’s) board of directors. Sirton (now Vifarma) was put into liquidation and, on June 28, 2010, was admitted by the Court of Como to a composition with creditors’ proceedings ("concordato preventivo"). The composition with creditors was approved on February 3, 2011. At that time, Sirton’s assets were acquired by a third party, as approved by the Court of Como. A liquidator has been appointed to manage the liquidation process and the distribution of proceeds received from the sale of Sirton’s assets to Sirton’s creditors. Although the distribution allocation has not yet been finalized, we understand that the liquidator may propose to satisfy the amounts due to secured creditors in full, with a payout distribution of 18.26% to all unsecured creditors. Our net exposure to Sirton at the date of the admission to the composition with creditors was €0.85 million. If the preliminary indication from the liquidator is confirmed, we may collect 18.26% or €0.16 million of the receivables outstanding on the date of the admission to the composition with creditors. In 2012, we received a partial payment of €0.09 million. In the prior year, due to the uncertainty of the final distribution to creditors from the sales of Sirton’s assets, we established an allowance for doubtful accounts of €0.85 million, which represents our exposure against Sirton. In 2012, in connection with the partial payment received, we released €0.09 million of the

allowance. As of December 31, 2013, we still maintain an allowance of €0.77 million which represents our exposure against Sirton (now Vifarma).

The Company had a lease agreement with Sirton (now Vifarma) that expired on December 31, 2010, but was renewed for an additional six-year term. In connection with Sirton's (now Vifarma's) liquidation proceeding, the lease agreement with Sirton (now Vifarma), along with the premises to which such lease pertains, were transferred to an unrelated third party that has also acquired the rights to Sirton's name and assets.

GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

On January 1, 2012, we entered into a new commercial lease with F3F S.r.l. The area leased is approximately 4,800 square meters in size and is used for offices, manufacturing, laboratories and storage facilities. The lease provides for an annual fee of €0.19 million for the initial six-year term, which may be adjusted annually based on the cost of living index, and, in the event we exercise our six-year renewal option, €0.22 million on an annual basis, subject to cost of living adjustments.

Expenses under these operating leases for the years ended December 31, 2011, 2012 and 2013 amounted to €0.21 million, €0.20 million and €0.20 million, respectively. See Note 15 for the commitments under these leases.

For the years ended December 31, 2011, 2012 and 2013, the Company had the following transactions with F3F S.r.l. and Sirton (now Vifarma) (in thousands):

	For the Year Ended December 31,		
	2011	2012	2013
Charges from related parties	€222	€186	€189
Total	€222	€186	€189

In 2012 and 2013, transactions with the new Sirton Pharmaceuticals S.p.A. were not classified as transactions with a related party since the new Sirton Pharmaceuticals S.p.A. is no longer a related party of the Company given the change of ownership.

The Company is a party to a license and supply agreement with Sigma-Tau Pharmaceuticals, Inc., or Sigma-Tau, pursuant to which the Company has licensed to Sigma-Tau the rights to commercialize defibrotide for the treatment and prevention of VOD in North America, Central America and South America, subject to receipt of marketing authorization, if any, in the applicable territory. Sigma-Tau is an affiliate of Sigma-Tau Finanziaria S.p.A. Dr. Marco Brughera, who holds various senior-level positions within the Sigma-Tau Group, served as a member of our board of directors until January 24, 2014. See Note 4 for further discussion of our relationship with Sigma-Tau.

In connection with the license and supply agreement, the Company also entered into a cost sharing agreement with Sigma-Tau dated October 12, 2007. Under the cost sharing agreement, as amended, Sigma-Tau agreed to reimburse the Company 50% of certain costs associated with the development of defibrotide. Pursuant to the terms of this agreement, between 2007 and 2013, the Company received $11.00 million in reimbursement of research and development expenses. Furthermore, the Company agreed that $1.00 million in costs reimbursed by Sigma-Tau will be deductible from royalty payments that the Company is entitled to receive in the future under the license and supply agreement. In 2013, the Company received €1.40 million ($1.80 million) as reimbursement of research and development expenses and in 2014, Sigma-Tau agreed to reimburse the Company an estimated total of approximately $4.77 million as incurred.

The balance of any reimbursements that we are entitled to receive from Sigma-Tau was classified as accounts receivable from related parties and other revenues from related parties in the accompanying consolidated financial statements. As of December 31, 2012 and 2013, the Company had the following balances with F3F S.r.l., Sirton (now Vifarma) and Sigma-Tau (in thousands):

	As of December 31,	
	2012	2013
Accounts Receivable – Sirton (now Vifarma)	€765	€765
Accounts Receivable – Sigma Tau	216	1,541
Allowance for doubtful accounts	(765)	(765)
Accounts Receivable, net	€216	€1,541
Accounts Payable Sirton (now Vifarma)	€5	€5

The accounting policies applied in transactions with our affiliates are consistent with those policies applied in transactions with independent third parties and all related party agreements are negotiated on an arm’s length basis.

4. COLLABORATIVE ARRANGEMENTS

The Company is a party to a license and supply agreement with Sigma-Tau (as assignee of Sigma-Tau Industrie Farmaceutiche Riunite S.p.A.) dated December 7, 2001. Under the agreement, Sigma-Tau obtained exclusive rights to distribute, market and sell defibrotide to treat VOD in the United States. This license expires 8 years after product launch. In 2005, the Company expanded Sigma-Tau's current license territory to North America, Central America and South America,

GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

subject to receipt of marketing authorization, if any, in the applicable territory. In January 2010, the Company amended its existing license and supply agreement to encompass a license to Sigma-Tau for the intravenous formulation of defibrotide for the prevention of VOD in the Americas and to transfer the NDA post-approval in the United States. Pursuant to the license and supply agreement, as amended, between 2001 and 2010, the Company received milestone and other payments in the amount of $11.35 million and is entitled to an additional payment of $6.00 million following regulatory approval from the FDA to market defibrotide in the United States and a further $2.00 million following the transfer of the approved NDA to Sigma-Tau.

The license and supply agreement also envisages that the Company will produce and supply defibrotide to Sigma-Tau for marketing and distribution in the United States if and when the drug is approved by the FDA. The Company will be entitled to royalty payments equal to 7% of Sigma-Tau's net sales of defibrotide and a supply price equal to the greater of 31% of net sales of defibrotide or €50 (approximately $68) per unit of finished product.

In addition, on October 12, 2007, the Company entered into a cost sharing agreement with Sigma-Tau to address the need for additional funding in accordance with the original license and supply agreement. Sigma-Tau agreed to reimburse 50% of certain costs associated with the development of defibrotide. Between 2007 and 2013, the Company received $11.00 million in reimbursement of research and development expenses. In addition, the Company agreed that $1.00 million in costs reimbursed by Sigma-Tau will be deductible from royalty payments that the Company will be entitled to receive in the future under the license and supply agreement. In 2013 we received €1.40 million ($1.80 million) as reimbursement of research and development expenses, and in 2014, Sigma-Tau agreed to reimburse the Company an estimated total of approximately $4.77 million as incurred. We recognize the reimbursement of research and development expenses as revenue when we incur the costs subject to reimbursement.

Under the license and supply agreement, if, during the drug development stage, the Company realizes that the activities required to bring the product to completion will necessitate a material increase in expenditures, the parties will discuss the increased costs and possible revisions to the terms of the agreement; if the parties are unable to mutually agree on such revisions, either party can terminate the agreement. If the Company or Sigma-Tau terminates the agreement for that reason, or if the Company unilaterally discontinues the development of defibrotide to treat VOD (after written notice to Sigma-Tau), and the Company resumes the development of defibrotide within 36 months of the termination, substantially availing itself of the stages previously completed, either independently or with a third party, the Company will be required to promptly reimburse Sigma-Tau for the amounts received previously for development expenses.

The following table outlines the nature and amount of other revenue recognized in the accompanying consolidated statements of income (in thousands):

	For the Year Ended December 31,		
	2011	2012	2013
Research and development cost reimbursement	€323	€1,257	€2,563
Upfront payments recognized ratably	1,703	—	—
Other revenues from F3F S.r.l.	—	—	39
Total	€2,026	€1,257	€2,602

5. INVENTORIES

Inventories consisted of following (in thousands):

	As of December 31,	
	2012	2013
Raw materials	€332	€521
Work in process	236	764
Finished goods	1,422	1,163
Total	€1,990	€2,448

At December 31, 2012 and 2013, the reserves for obsolescence were €0.33 million and €0.37 million, respectively. The increase from 2012 is mainly due to the utilization of a reserve for €0.19 million in connection with the destruction of inventories that were written off in the prior year together with the establishment of a reserve of €0.23 million for APIs which did not pass our quality standards.

GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

Prior to signing the named patient and expanded access program agreements, all costs associated with the production of defibrotide were expensed as research and development expenses. As of December 31, 2012 and 2013, inventory included €0.74 million and €0.49 million, respectively, for defibrotide commercial batches classified as finished goods, which are expected to be sold through the named patient and expanded access programs.

6. PREPAID EXPENSES AND OTHER CURRENT ASSETS

Prepaid expenses and other current assets consisted of the following (in thousands):

	As of December 31,	
	2012	2013
VAT receivables	€174	€833
Tax receivables	645	69
Other prepaid expenses and current assets	609	866
Total prepaid expenses and current assets	€1,428	€1,768

Value added tax, or VAT, receivables represent the tax on the value of consumption. VAT has no effect on the Company's operating results, as payments and receipts are allowed to be netted against each other in periodic filings with the tax authorities. The VAT payment system is a "custodial" relationship. VAT liabilities are generated when the Company invoices selected customers, including the VAT amount, and VAT receivables are created when the Company purchases goods and services subject to VAT. The increase in VAT receivables is due to the following: i) utilization of €0.14 million to offset the payment of an equivalent amount of social charges and withholding taxes, ii) the reimbursement of €0.04 million, and iii) an increase in VAT receivables of €0.83 million, of which €0.37 million may be claimed back to offset an equivalent amount of social security charges and withholding taxes and €0.46 million relates to VAT receivables from German tax authorities on intercompany purchases.

Tax receivables relate to tax advance payments or withholding tax. We recorded tax receivables of €0.65 million and €0.07 million for the years ended December 31, 2012 and 2013, respectively. The decrease is attributable to: (i) the utilization of €0.38 million to offset the payment of an equivalent amount of social charges and other corporate taxes, (ii) the allocation of a tax credit of €0.24 million to offset estimated 2013 tax liabilities, and (iii) an increase of €0.04 million in tax credits matured in withholding interest which can be used to offset future taxable income.

Other prepaid expenses and current assets primarily relate to advances to vendors and prepaid premiums to insurance companies.

7. PROPERTY, MANUFACTURING FACILITY AND EQUIPMENT

Property, manufacturing facility and equipment are recorded at historical cost, net of accumulated depreciation.

Property, manufacturing facility and equipment consisted of the following (in thousands):

	As of December 31,					
	2012			2013		
	Cost	Accumulated Depreciation	Net book value	Cost	Accumulated Depreciation	Net book value
Land and building	€2,686	€1,559	€1,127	€2,851	€1,644	€1,207
Plant and machinery	15,553	11,352	4,201	16,143	12,329	3,814
Industrial equipment	1,765	1,248	517	1,798	1,367	431
Furniture and fixtures	883	571	312	937	628	309
Leasehold improvements	1,310	553	757	1,321	725	596
Internally developed software	750	300	450	765	351	414
Construction in progress	85	—	85	810	—	810
	€23,032	€15,583	€7,449	€24,625	€17,044	€7,581

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GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

As of December 31, 2012 and 2013, property, manufacturing facility and equipment included €0.46 million attributed to laboratory instruments acquired under capital lease arrangements. The related accumulated depreciation at December 31, 2012 and 2013 was €0.28 million and €0.32 million, respectively.

8. FAIR VALUE MEASUREMENT

Fair values determined on the basis of Level 1 inputs utilize quoted prices (unadjusted) in active markets for identical assets or liabilities. The Company's Level 1 assets consist of cash and cash equivalents and short term deposits. Fair values determined on the basis of Level 2 inputs utilize observable quoted prices for similar assets and liabilities in active markets and observable quoted prices for identical or similar assets in markets that are not very active. Fair values determined on the basis of Level 3 inputs utilize unobservable inputs and include valuations of assets or liabilities for which there is little, if any, market activity. Level 3 assets or liabilities include those for which fair value measurements are determined using pricing models, discounted cash flow methodologies or similar valuation techniques, as well as significant management judgment or estimation.

The table below presents information on assets measured at fair value on a recurring basis at December 31, 2012 and 2013, and includes the valuation techniques the Company utilizes to determine such fair value (in thousands):

		Fair Value Measurements at December 31, 2013 using		
	Total Carrying Value at December 31, 2013	Quoted prices in active markets (Level 1)	Significant other observable inputs (Level 2)	Significant unobservable inputs (Level 3)
Cash and cash equivalents	€ 22,038	€22,038	€—	€—
Short-term deposit	4,000	4,000		
Total	€ 26,038	€26,038	€—	€—

		Fair Value Measurements at December 31, 2012 using		
	Total Carrying Value at December 31, 2012	Quoted prices in active markets (Level 1)	Significant other observable inputs (Level 2)	Significant unobservable inputs (Level 3)
Cash and cash equivalents	€12,485	€12,485	€—	€—
Total	€12,485	€12,485	€—	€—

9. ACCRUED EXPENSES AND OTHER CURRENT LIABILITIES

Accrued expenses and other current liabilities consisted of the following (in thousands):

	As of December 31,	
	2012	2013
Accrued compensation and employee benefits	€1,176	€1,284
VAT payables	—	1,052
Due to social security	326	715
Withholding tax due	162	534
Other payables	64	99
Total	€1,728	€3,684

Accrued compensation and employee benefits include bonuses, salaries, vacation and deferred compensation due to employees, directors and management. VAT payables refer to amounts due to the German tax authorities on intercompany sales made by Gentium S.p.A.

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GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

10. CREDIT FACILITIES AND LONG-TERM DEBT

Long term debt, net of current maturities consisted of the following (in thousands):

	As of December 31,	
	2012	2013
Mortgage loan bearing interest at the Euribor 6 month rate plus 1.0% due June 2018 (1.32% and 1.39% at December 31, 2012 and 2013, respectively)	€1,320	€1,080
Equipment loan bearing interest at the Euribor 3 months rate plus 1.20% due June 2013 (1.39% and 1.49% at December 31, 2012 and 2013, respectively)	62	—
Financing loan bearing interest at the Euribor 1 months rate plus 1.00% due June 2014 (1.11% and 1.22% at December 31, 2012 and 2013, respectively)	84	29
Equipment loan bearing interest at the Euribor 3 months rate plus 1.00% due June 2014 (1.19% and 1.29% at December 31, 2012 and 2013, respectively)	39	14
Financing loan bearing interest at the Euribor 3 months rate plus 0.80% due June 2014 (0.99% and 1.09% at December 31, 2012 and 2013, respectively)	39	13
Financing loan bearing interest at the Euribor 3 months rate plus 1.25% due January 2021 (5.75% at December 31, 2013)	—	600
	1,544	1,736
Less current maturities	(409)	(350)
Total	€1,135	€1,386

On November 11, 2013, we obtained a loan in the amount of €0.60 million from Banca Popolare di Sondrio for the acquisition and installation of manufacturing equipment, bearing interest at the three-month Euribor rate plus 1.25%. Principal and interest are due in quarterly installments beginning on April 30, 2014. At December 31, 2013, the principal amount outstanding under this loan was €0.60 million.

The maturities of long-term debt outstanding as of December 31, 2013 were as follows (in thousands):

Year ending December 31,	Scheduled Long-term Debt Maturities
2015	€316
2016	320
2017	324
2018	209
Thereafter	217
Total	€1,386

11. INCOME TAXES

The components of income before the income tax provision/(benefit) were as follows (in thousands):

	For the Year Ended December 31,		
	2011	2012	2013
Domestic	€4,655	€3,969	€7,855
Foreign	(1,134)	(3,169)	(476)
Total	€3,521	€800	€7,379

GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

The following table sets forth the details of the income tax provision/(benefit) (in thousands):

	For the Year Ended December 31,		
	2011	2012	2013
Current			
Domestic	€677	€15	€613
Foreign	134	11	—
Total current income tax	811	26	613
Deferred			
Domestic income tax (benefit)	—	—	(17,863)
Domestic income tax provision	—	—	28
Foreign	—	—	—
Total deferred income tax (benefit)	—	—	(17,835)
Total income tax provision/(benefit)	€811	€26	€(17,222)

Current income tax provision represents amounts due to Italian tax authorities in payment of the Italian Regional Tax on Productive Activities, or IRAP, and the Italian corporate tax, or IRES.

A reconciliation of income taxes computed at the Italian statutory income tax rate to our effective income tax rate was as follows (in thousands):

	For the Year Ended December 31,		
	2011	2012	2013
Statutory income tax rate	27.5 %	27.5 %	27.5 %
Income tax provision at statutory rate	€968	€220	€2,029
Movement in valuation allowance	(1,252)	(1,205)	(20,664)
Effect on Swiss tax rate	140	383	521
Italian regional tax - IRAP	380	107	345
Swiss minimum tax	134	11	—
Stock options	458	527	499
Permanent differences from tax calculation	(17)	(23)	48
True-up previous years in deferred tax asset calculation	—	6	—
Total income tax provision/(benefit)	€811	€26	€(17,222)
Effective income tax rate	23.0 %	3.3 %	(233.4)%

GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

Significant components of our net deferred tax assets/(liabilities) were as follows (in thousands):

	As of December 31,	
	2012	2013
Deferred tax assets:		
Net operating losses	€14,183	€12,620
Capitalization of R&D costs	5,320	3,801
Property, manufacturing facility and equipment	344	261
Write down of intangible assets	1,086	1,501
Allowance on doubtful accounts	15	—
Inventory write-off	192	117
Other	22	61
Total deferred tax assets	21,162	18,361
Valuation allowance	(21,162)	(498)
Net deferred tax assets	—	17,863
Deferred tax liabilities	—	(28)
Net deferred tax assets	€—	€17,835

The gross domestic NOLs amounted to approximately €49.17 million and €44.07 million as of December 31, 2012 and 2013, respectively. Under Italian tax law NOLs cannot be carried back. Tax losses can be carried forward indefinitely; however such tax losses can only be used to offset a maximum of 80% of taxable income for each tax year. The Company's only foreign subsidiary, Gentium GmbH, was incorporated in Switzerland in 2011. This entity generated losses for the years ended December 31, 2011 and 2012 and taxable income for 2013 against which we utilized existing NOLs. The gross foreign NOLs amounted to approximately €4.30 million and €3.32 million as of December 31, 2012 and 2013, respectively. According to Swiss tax law, these NOLs can be carried forward for seven years and will begin to expire in 2018.

Valuation allowances require an assessment of both positive and negative evidence when determining whether it is more likely than not that deferred tax assets are recoverable. Such assessment is made on a jurisdiction by jurisdiction basis. The Company's assessment included an evaluation of cumulative income in recent years, future sources of taxable income, exclusive of reversing temporary differences, and risks and uncertainties related to our business. As of December 31, 2013 the Company has determined that it is more likely than not that the domestic deferred tax assets will be recoverable and the related valuation allowance is no longer needed. The Company has determined that it is more likely than not that the Company will not realize the benefits of the foreign deferred tax assets. Accordingly, the Company continues to maintain a full valuation allowance on the Company's foreign deferred tax assets until sufficient positive evidence exists to support reversal.

As of December 31, 2012 and 2013, the Company had no uncertain tax positions and, therefore, had no accrued interest or penalties related to such uncertain tax positions. There are no changes expected to occur in the next 12 months with respect to the status of the Company's uncertain tax positions.

The fiscal years 2008 through 2013 in respect of Gentium S.p.A are still subject to income tax examination. The fiscal years 2011 through 2013 in respect of Gentium GmbH are still subject to income tax examination.

12. SHAREHOLDERS' EQUITY

The Company had 15,038,483 and 15,555,131 ordinary shares, each of no par value, issued and outstanding as of December 31, 2012 and 2013, respectively. As of December 31, 2013, the total number of authorized shares was 19,656,317.

Authorized capital consisted of the following:

	As of December 31,	
	2012	2013

Issued and outstanding	15,038,483	15,555,131
Reserved for stock option plans	4,617,834	4,101,186
Total	19,656,317	19,656,317

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NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

On April 28, 2006, our shareholders approved an amendment to our bylaws, which granted certain powers to our board of directors, pursuant to the provisions of Articles 2443 and 2420, part 3 of the Italian Civil Code, including the power to increase the capital of the Company in cash, up to €90.00 million of par value, in one or more transactions, and to issue convertible bonds (including subordinated) and increase the capital of the Company, in one or more transactions, up to €10.00 million of par value, through the issuance of ordinary shares reserved for the conversion of such convertible bonds, and in both cases also with the faculty to issue warrants by means of the same resolution of the board of directors providing for the relevant capital increase and in each case, exclude or limit the option right of the shareholders if the board of directors determines that exclusion or limitation to be in the interest of the Company. Such delegation of powers expired after five years. On May 9, 2011, our shareholders renewed this resolution for additional five years starting from the date of the resolution of the 2011 Extraordinary Shareholders' meeting approving the amendment. As of December 31, 2013, our board of directors has approved the issuance of 4,549,435 ordinary shares in connection with this resolution by our shareholders.

On June 30, 2009, our shareholders approved an amendment to our bylaws, which granted certain powers to our board of directors, pursuant to article 2443 of the Italian Civil Code, including the power to increase the capital of our company in cash, up to an amount equal to €100.00 million, on a separable basis, in one or more transactions, for the purpose of a rights offering with the faculty to reserve all or part of such amount for the exercise of warrants issued by means of the same resolution of our board of directors providing for the relevant capital increase and with the faculty to reserve 1/4 of any such capital increase to employees under the Company's equity incentive plans in effect from time to time, and the power to cancel the par value of the ordinary shares of the Company, which was completed on June 30, 2009. As of December 31, 2013, our board of directors has not approved the issuance of any shares pursuant to this resolution by our shareholders.

13. EQUITY INCENTIVE PLANS

The Company currently has two option plans in place: an Amended and Restated 2004 Equity Incentive Plan, which includes an Amended and Restated 2004 Italy Stock Award Sub-Plan, and a 2007 Stock Option Plan (collectively, the "Plans").

Amended and Restated 2004 Equity Incentive Plan

Certain of the Company's employees and directors participate in the Amended and Restated 2004 Equity Incentive Plan and Italy Stock Award Sub-Plan. These plans were initially adopted on September 30, 2004 and amended on April 27, 2007. The plans provide for the issue of incentives awards for up 1,500,000 ordinary shares to employees, consultants, directors, and non-employee directors. Awards may be in the form of either incentive or non-qualified. Our compensation committee determines the price of share options granted under the incentive plan, with the provision that the exercise price for an incentive share option cannot be less than 100% of the fair market value of our ordinary shares on the date of grant. The term of share options granted under the incentive plan generally may not exceed ten years, although the shareholders' authorization for a capital increase relating to the ordinary shares issuable upon exercise of such options expires on September 30, 2019. As of December 31, 2013, there were 806,356 shares underlying outstanding options and 507,739 shares available for future grants under this plan. Shares subject to options that have expired or have otherwise terminated without being exercised in full become available again for issuance under the plan.

Options granted under the incentive plan vest at the rate determined by our compensation committee. Typically, options granted under the incentive plan to officers and employees vest over three years, with one-third of the shares covered by the option vesting on the first anniversary of the grant date and the remainder vesting monthly over the next two years.

2004 Italy Stock Award Sub-Plan

Our Amended and Restated 2004 Italy Stock Award Sub-Plan is a part of our Amended and Restated 2004 Equity Incentive Plan and provides for the grant of share options and the issuance of share grants to certain of our employees

who reside in the Republic of Italy and who are liable for income tax in the Republic of Italy. Generally, the exercise price for a share option under the Italy sub-plan cannot be lower than the average of the closing price of our ordinary shares as listed on NASDAQ over the 30 days preceding the date of grant.

Amended 2007 Stock Option Plan

On April 27, 2007, the Company's shareholders approved the 2007 Stock Option Plan providing for options that may be granted to the Company's directors, employees and consultants to purchase up to 3,200,000 ordinary shares. As of December 31, 2013, there were 1,385,849 ordinary shares underlying outstanding options and 1,401,242 shares available for future grants under this plan. Shares subject to options that have expired or have otherwise terminated without being exercised in full become available again for issuance under the plan.

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NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

The 2007 Stock Option Plan is administered by our board of directors or a committee appointed by our board of directors. The board or the committee determines recipients and types of options to be granted, including the number of shares subject to an option, the vesting schedule of options, the exercisability of options and, subject to applicable restrictions, other terms of the options. The board of directors has delegated responsibility for administration of the 2007 Stock Option Plan to the compensation committee.

The term of share options granted under the 2007 Stock Option Plan generally may not exceed the earlier of ten years or March 26, 2022. Our compensation committee determines the price of share options granted under the 2007 Stock Option Plan, subject to certain limitations.

Options granted under the 2007 Stock Option Plan vest at the rate determined by our compensation committee. Typically, options granted to employees under the 2007 Stock Option Plan vest over three years, with one-third of the shares covered by the option vesting on the first anniversary of the grant date and the remainder vesting monthly over the next two years.

The board of directors may amend the 2007 Stock Option Plan at any time. Amendments will be submitted for shareholder approval to the extent required under applicable laws, rules and regulations. The 2007 Stock Option Plan will terminate on March 26, 2022 unless earlier terminated by the board of directors or a committee appointed by the board of directors.

The following table lists the balances available under the Plans at December 31, 2013.

	Amended and Restated Nonstatutory Plan and Agreement	Amended and Restated 2004 Equity Incentive Plan	2007 Stock Option Plan
Number of shares authorized	60,000	1,500,000	3,200,000
Number of option granted since inception	60,000	2,568,400	2,176,578
Number of options exercised	(60,000)	(245,905)	(412,909)
Number of options canceled/expired	—	(1,516,139)	(377,820)
Number of shares available for future grant	—	507,739	1,401,242

Stock-based compensation expenses are measured at the grant date on the basis of fair value of the award ultimately expected to vest and recognized as expenses over the service period, which is generally the vesting period. The Company recorded non-cash compensation as follows (in thousands):

	For the Year Ended December 31,		
	2011	2012	2013
Cost of goods sold	€39	€46	€48
Research and development	157	120	201
Selling, general and administrative	1,470	1,750	1,567
Total stock-based compensation	€1,666	€1,916	€1,816

The weighted-average grant date fair market values of options granted to officers, employees and directors in the years ended December 31, 2011, December 31, 2012 and December 31, 2013 were $5.81, $5.64 and $5.28. The valuation of options granted was based on the following weighted-average assumptions:

	For the Year Ended December 31,		
	2011	2012	2013
Risk free interest rate	3.19%	2.10%	1.80%
Expected dividend yield	—%	—%	—%
Expected stock price volatility	92.82%	93.55%	88.32%
Expected term (in years)	5.44	5.71	6.88

The fair value of the stock options is estimated on the date of grant using a binomial valuation model. The binomial model accounts for volatility in the price of the Company's stock, the risk-free interest rate, the estimated life of the option, the closing market price of the Company's stock and the exercise price of the stock. Some of these inputs are highly subjective assumptions which can vary over time. In order to determine the expected volatility, the Company analyzed available information, including past experience of a group of stocks in the industry having similar traits. The risk-free rate for the

GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

expected term of the option is based on the U.S. Treasury yield curve in effect at the time of grant. The Company assumed that no dividends would be paid during the expected term of the options.

All of the Company's stock options vest ratably through continued employment over the vesting period. Once vested, options become exercisable immediately. Stock-based compensation expenses recognized in the statements of income are based on awards ultimately expected to vest, reduced for estimated forfeitures. Based on historical data, the pre-vesting forfeiture percentage was estimated to be approximately zero. If pre-vesting forfeitures occur in the future, the Company will record the effect of such forfeitures as they occur.

The Company expects to incur significant non-cash compensation expenses for option grants in the future. As of December 31, 2013, compensation costs not yet recognized totaled €1.37 million. All outstanding and unvested stock options became fully vested on consummation of the tender offer.

A summary of the Company's stock option activity based on the exchange rate in effect at the grant date is as follows:

	Shares Available for Grant	Shares Subject to Outstanding Options	Weighted-Average Exercise Price		Weighted-Average Remaining Contractual Term (Years)
Options outstanding at December 31, 2010	476,359	2,023,641	€4.83	$6.70	7.35
Available under revised stock option plan	2,200,000	—	—	—	
Granted	(657,300)	657,300	€5.81	$8.54	
Exercised	(12,833)	(12,833)	€6.65	$7.97	
Cancellations	482,865	(470,033)	€7.57	$9.66	
Options outstanding at December 31, 2011	2,489,091	2,198,075	€4.53	$6.61	4.28
Granted	(161,500)	161,500	€6.90	$9.02	
Exercised	(69,333)	(69,333)	€4.48	$5.30	
Cancellations	98,358	(29,025)	€4.77	$6.50	
Options outstanding at December 31, 2012	2,356,616	2,261,217	€4.70	$6.82	4.32
Granted	(629,000)	629,000	€7.13	$9.09	
Exercised	(516,648)	(516,648)	€4.38	$5.86	
Cancellations	698,013	(181,364)	€7.04	$8.79	
Options outstanding at December 31, 2013	1,908,981	2,192,205	€5.28	$7.63	4.17
Exercisable at December 31, 2013		1,686,261	€5.26	$7.25	4.17

GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

Cash received for exercised stock options amounted to €0.08 million, €0.28 million and €2.45 million in the years ended December 31, 2011, 2012 and 2013, respectively. The intrinsic values of options exercised in 2011, 2012 and 2013 were $0.01 million, $0.32 million and $9.00 million, respectively. The total fair values of options vested during 2011, 2012 and 2013 were $3.50 million, $7.47 million and $7.37 million, respectively.

	Options Outstanding			Options Exercisable	
Range of Exercise Prices	Number of Options Outstanding	Weighted-Average Years Remaining on Contractual Life	Weighted-Average Exercise Price	Number of Options Exercisable	Weighted-Average Exercise Price
$4.57 - $5.00	746,500	5.35 - 6.33	$4.57 - $5.00	746,500	$4.57 - $5.00
$5.49 - $6.00	105,000	6.92 - 7.87	$5.49 - $6.00	96,250	$5.49 - $6.00
$6.06 - $7.08	70,884	7.86 - 1.82	$6.06 - $7.08	44,342	$6.06 - $7.08
$8.04 - $8.47	127,257	8.36 - 8.23	$8.04 - $8.47	84,830	$8.04 - $8.47
$8.77 - $8.85	183,594	8.55 7.57	$8.77 - $8.85	63,136	$8.77 $8.85
$8.88 - $9.00	420,000	8.15 - 1.50	$8.88 - $9.00	173,472	$8.88 - $9.00
$9.20 - $9.26	301,862	8.16 - 7.16	$9.20 - $9.26	277,012	$9.20 - $9.26
$9.30 - $9.91	126,000	8.36 - 7.35	$9.30 - $9.91	126,000	$9.30 - $9.91
$13.98 - $16.52	47,108	4.00 3.85	$13.98 - $16.52	47,108	$13.98 - $16.52
$18.95 - $19.33	64,000	3.23 - 8.69	$18.95 - $19.33	27,611	$18.95 - $19.33
	2,192,205			1,686,261	

At December 31, 2013 the aggregate intrinsic value of the outstanding options and exercisable options were $108.21 million and $84.06 million, respectively.

14. EARNINGS PER SHARE

The computation of basic earnings per share, or EPS, is based upon the weighted-average of our ordinary shares outstanding. The computation of diluted EPS is based upon the weighted-average of our ordinary shares and the dilutive potential of ordinary shares outstanding. Dilutive potential of ordinary shares outstanding refers to the impact of ordinary equivalent shares resulting from the assumed exercise of stock options under the treasury stock method. The computation for basic and diluted EPS was as follows (in thousands, except share and per share data):

	For the Year Ended December 31		
	2011	2012	2013
Income (numerator):			
Net income for basic and diluted EPS	€2,710	€774	24,601
Shares (denominator):			
Weighted-average shares for basic EPS	14,964,021	15,014,411	15,261,799
Effect of dilutive securities	376,838	625,479	1,340,944
Weighted-average shares for diluted EPS	15,340,859	15,639,890	16,602,743
Basic EPS	€0.18	€0.05	€1.61
Diluted EPS	€0.18	€0.05	€1.48

For the years ended December 31, 2011, 2012 and 2013, there were employee stock options, calculated on a weighted-average basis, to purchase 644,146, 236,296 and nil shares, respectively, of our common stock, with exercise prices greater than the average market prices of our common stock for these periods, which are not included

in the computation of diluted EPS as their impact would have been anti-dilutive.

GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

15. COMMITMENTS AND CONTINGENCIES

Future non-cancelable minimum lease payments under operating leases as of December 31, 2013 are (in thousands):

Year ending December 31,	Lease Payments
2014	€375
2015	185
2016	185
2017	185
2018	—
Total	€930

As of December 31, 2013, in addition, we had €7.04 million in future payables under outstanding contracts of which €5.86 million is due within one year. Most of these contracts are on a cost plus or actual cost basis.

See Note 17 entitled “Subsequent Event” for a summary of a shareholder litigation matter. From time to time we are involved in legal proceedings arising in the ordinary course of business. We believe there is no other litigation pending that could have, individually or in the aggregate, a material adverse effect on our results of operations or financial condition.

16. INFORMATION REGARDING GEOGRAPHICAL AREA AND MAJOR CUSTOMERS

For the years ended December 31, 2011, 2012 and 2013, total product sales by geographic territory and customer were as follows (amounts in thousands):

	For the Year Ended December 31,								
	2011			2012			2013		
UK	€12,155	56	%	€13,290	48	%	€21,453	54	%
Korea	3,402	16		3,455	13		4,951	12	
Spain	1,137	5		2,054	7		2,955	7	
Turkey	917	4		2,671	10		2,850	7	
United States	2,622	12		2,226	8		2,645	7	
Australia	840	4		1,344	5		1,922	5	
Sweden	—	—		997	4		1,747	5	
Italy	599	3		769	3		382	1	
Israel	62	—		152	1		76	—	
Other	—	—		672	1		844	2	
Total	€21,734	100	%	€27,630	100	%	€39,825	100	%

GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

	For the Year Ended December 31,								
	2011			2012			2013		
Customer A	€12,099	56	%	€13,062	47	%	€21,196	53	%
Customer B	3,112	14		3,154	11		4,640	12	
Customer C	917	4		2,671	10		2,850	7	
Customer D	2,622	12		2,226	8		2,645	7	
Customer E	840	4		1,343	5		1,922	5	
Customer F	—	—		1,132	4		1,841	5	
Customer G	—	—		997	4		1,747	4	
Customer H	1,137	5		923	3		1,114	3	
Customer I	—	—		637	2		617	2	
Customer J	599	3		471	2		382	1	
Other Customers	408	2		1,014	4		871	1	
Total	€21,734	100	%	€27,630	100	%	€39,825	100	%

17. SUBSEQUENT EVENT

On December 19, 2013, we entered into a definitive tender offer agreement with Jazz and Jazz Italy, pursuant to which Jazz Italy made an offer to purchase all our outstanding ordinary shares and ADSs, each representing one ordinary share, at a purchase price of $57.00 per ordinary share and per ADS (without duplication for ordinary shares underlying ADSs). On February 21, 2014, Jazz Italy completed its tender offer, having acquired approximately 98 percent of our combined ordinary shares and ADSs. Jazz is now our indirect majority shareholder. We filed a Notice of Voluntary Delisting with The NASDAQ Stock Market on March 5, 2014, and trading in our ADSs on NASDAQ was suspended on March 7, 2014. See Note 1 entitled “Business and Basis of Presentation” for related discussion.

As a result of the closing of the tender offer, we incurred approximately €39 million in transaction-related expenses, including €18 million in investment banker fees and €11 million in payments to selected employees. In addition, under certain circumstances, selected employees of Gentium S.p.A. and its subsidiary Gentium GmbH are entitled to a change of control bonus of approximately €5 million, which may become due in the third and fourth quarters of 2014. As of February 17, 2014, all outstanding option awards had been exercised and all ordinary shares issued upon such exercise had been subsequently tendered pursuant to the tender offer. Proceeds from exercise of stock options amount to approximately €13.00 million. In addition, all of our stock-based compensation plans and our 401(k) saving plan have been terminated.

GENTIUM S.p.A.
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

In January 2014, we became aware of a purported class action lawsuit filed in the Southern District of New York in connection with the tender offer. The lawsuit, captioned Xavion Jyles, Individually and on Behalf of All Others Similarly Situated v. Gentium S.P.A. et al., names us, each of our directors, Jazz and Jazz Italy as defendants. The lawsuit alleges, among other things, that our directors breached their fiduciary duties to our shareholders in connection with the tender offer agreement that we entered into with the Jazz entities valuing our ordinary shares and ADSs at $57.00 per share, and that the Jazz entities violated Sections 14(e) and 20(a) of the Exchange Act, by allegedly overseeing our preparation of an allegedly false and misleading Section 14D-9 Solicitation/Recommendation Statement. The lawsuit seeks, among other relief, class action status, rescission, and unspecified costs, attorneys' fees and other expenses. We cannot predict the timing or outcome of this matter.

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SIGNATURES

The registrant hereby certifies that it meets all of the requirements for filing on Form 20-F and that it has duly caused and authorized the undersigned to sign this annual report on its behalf.

Date: March 31, 2014

GENTIUM S.P.A.
(Registrant)
/s/ Fintan Keegan
Fintan Keegan
Chairman and Chief Executive Officer

/s/ Salvatore Calabrese
Salvatore Calabrese
Senior Vice President, Finance, Chief Financial Officer and Chief Operating Officer

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INDEX OF EXHIBITS

Exhibit	Description
Charter Documents	
1.1	Articles of Association of Gentium S.p.A., formerly known as Pharma Research S.r.l., dated November 11, 1993, incorporated by reference to Exhibit 3(i) to the Registration Statement on Form F-1, Registration No. 333-122233, previously filed with the SEC on January 24, 2005.
1.2	Amended and Restated Bylaws of Gentium S.p.A. dated May 9, 2011, incorporated by reference to Exhibit 1.2 to the Annual Report on Form 20-F previously filed with the SEC on March 30, 2012.
American Depositary Share Documents	
2.1	Form of Deposit Agreement among Gentium S.p.A., The Bank of New York and the owners and beneficial owners from time to time of American Depositary Receipts (including as an exhibit the form of American Depositary Receipt), incorporated by reference to Exhibit 4.6 to Amendment No. 5 to the Registration Statement on Form F-1, Registration No. 333-122233, previously filed with the SEC on June 9, 2005.
2.2	Form of American Depositary Receipt (see Exhibit 2.1).
2.3	Amendment, dated as of March 21, 2014, to the Deposit Agreement dated as of June 15, 2005 among Gentium S.p.A., The Bank of New York Mellon and the owners and beneficial owners from to time of American Depositary Receipts.
Loan Agreements	
4.1	Loan Agreement between Banca Nazionale del Lavoro S.p.A. and Gentium S.p.A. dated June 14, 2006 incorporated by reference to Exhibit 10.7.3 to the Registration Statement on Form F-3, Registration No. 333-135622, previously filed with the SEC on July 6, 2006.
4.2	Loan Agreement for €230,000 with Banca Intesa S.p.A., dated December 20, 2006, incorporated by reference to Exhibit 2 to the report on Form 6-K, previously filed with the SEC on February 2, 2007.
4.3	Loan Agreement for €500,000 with Banca Intesa S.p.A., dated December 20, 2006, incorporated by reference to Exhibit 3 to the report on Form 6-K, previously filed with the SEC on February 2, 2007.
4.4	Loan Agreement for €225,000 with Banca Intesa S.p.A., dated December 20, 2006, incorporated by reference to Exhibit 4 to the report on Form 6-K, previously filed with the SEC on February 2, 2007.
4.5	Loan Agreement, dated June 30, 2006, between San Paolo IMI S.p.A. and Gentium S.p.A., incorporated by reference to Exhibit 4.43 to the Annual Report on Form 20-F for the year ended December 31, 2006, previously filed with the SEC on April 30, 2007.
4.6	Loan Agreement (English translation), dated November 11, 2013, between Banca Popolare Di Sondrio and Gentium S.p.A.
License and Distribution Agreements	
4.7.1	License and Supply Agreement by and between Gentium S.p.A. 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.
4.7.2	Letter Agreement, dated October 12, 2007, between Gentium S.p.A. 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.
4.7.3*	Amendment to License and Supply Agreement and Letter Agreement, dated December 7, 2001 and October 12, 2007, respectively, effective January 7, 2010, between Gentium S.p.A. 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.
4.7.4**	Amendment No. 2 to Letter Agreement dated October 12, 2007, as amended, dated May 11, 2012, between Gentium S.p.A. and Sigma-Tau Pharmaceuticals, Inc.

4.8	Distribution Agreement, dated December 28, 2006, between Gentium S.p.A. and Crinos S.p.A., incorporated by reference to Exhibit 6 to the report on Form 6-K, previously filed with the SEC on January 3, 2007.
4.9*	Technical Transfer Services Agreement, dated February 2, 2009, between Gentium S.p.A. and Patheon Italia S.p.A, incorporated by reference to Exhibit 4.21 to the Annual Report on Form 20-F for the year ended December 31, 2008, previously filed with the SEC on March 31, 2009.
4.10.1*	Master Contract Clinical Research Agreement, dated September 29, 2009, between US Oncology Clinical Development and Gentium S.p.A., incorporated by reference to Exhibit 2 to the report on Form 6-K, previously filed with the SEC on December 1, 2009.
4.10.2**	Amendment, dated October 24, 2013, to Master Contract Clinical Research Agreement dated December 29, 2009 between US Oncology Clinical Development and Gentium S.p.A.

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4.11**	Manufacturing Services Agreement, effective September 15, 2013, between Patheon UK Limited and Gentium S.p.A.
Leases	
4.12	Commercial Lease Contract between Gentium S.p.A. and Sirton Pharmaceuticals S.p.A. dated January 1, 2005, incorporated by reference to Exhibit 10.33 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.
4.13	Commercial Lease Contract between Gentium S.p.A. and FinSirton S.p.A. (now F3F S.r.l.) dated January 1, 2012, incorporated by reference to Exhibit 4.21 to the Annual Report on Form 20-F previously filed with the SEC on March 30, 2012.
Miscellaneous	
4.14	Form of indemnification agreement between Gentium S.p.A. and certain of its officers and directors, 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.
4.15	Tender Offer Agreement, dated December 19, 2013, by and among Jazz Pharmaceuticals plc, Jazz Pharmaceuticals Italy S.r.l. and Gentium S.p.A., incorporated by reference to Exhibit 2.1 to the Form 8-K/A filed by Jazz Pharmaceuticals plc with the SEC on December 20, 2013.
4.16	Employment agreement by and between Gentium S.p.A. 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, incorporated by reference to Exhibit 99.(e)(4) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.17	Employment agreement by and between Gentium S.p.A. and Salvatore Calabrese, dated September 7, 2007, incorporated by reference to Exhibit 99.(e)(5) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.18	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, incorporated by reference to Exhibit 99.(e)(6) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.19	Employment agreement by and between Gentium S.p.A. 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, incorporated by reference to Exhibit 99.(e)(7) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.20	Transition, Amendment and Release Agreement, dated December 19, 2013, by and between Gentium GmbH and Dr. Khalid Islam, incorporated by reference to Exhibit 99.(e)(8) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.21	Consultancy Agreement, dated December 19, 2013, by and between Gentium S.p.A. and Dr. Khalid Islam, incorporated by reference to Exhibit 99.(e)(9) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.22	Retention and Amendment Agreement, dated December 19, 2013, by and between Gentium S.p.A. and Salvatore Calabrese, incorporated by reference to Exhibit 99.(e)(10) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.23	Retention and Amendment Agreement, dated December 19, 2013, by and between Gentium GmbH and Giorgio Mosconi, incorporated by reference to Exhibit 99.(e)(11) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
4.24	Retention and Amendment Agreement, dated December 19, 2013, by and between Gentium GmbH and Adrian Haigh, incorporated by reference to Exhibit 99.(e)(12) to the Solicitation/Recommendation Statement on Schedule 14D-9 filed with the SEC on December 24, 2013.
Subsidiaries	
8.1	List of Subsidiaries.

Certifications and Consents

12.1	Chief Executive Officer Certification pursuant to Section 302 of the Sarbanes-Oxley Act of 2002.
12.2	Chief Financial Officer Certification pursuant to Section 302 of the Sarbanes-Oxley Act of 2002.
13.1	Chief Executive Officer Certification pursuant to Section 906 of the Sarbanes-Oxley Act of 2002.
13.2	Chief Financial Officer Certification pursuant to Section 906 of the Sarbanes-Oxley Act of 2002.

XBRL Information

101.INS	XBRL Instance Document.
101.SCH	XBRL Taxonomy Extension Schema Document.
101.CAL	XBRL Taxonomy Extension Calculation Linkbase Document.

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101.DEF	XBRL Taxonomy Extension Definition Linkbase Document.
101.LAB	XBRL Taxonomy Extension Label Linkbase Document.
101.PRE	XBRL Taxonomy Extension Presentation Linkbase Document.

* Confidential treatment has been granted for portions of this exhibit. Omitted portions have been filed separately with the SEC.

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