

BIOMARIN PHARMACEUTICAL INC

FORM 10-Q (Quarterly Report)

Filed 05/02/14 for the Period Ending 03/31/14

Address	105 DIGITAL DRIVE NOVATO, CA 94949
Telephone	4155066700
CIK	0001048477
Symbol	BMRN
SIC Code	2834 - Pharmaceutical Preparations
Industry	Biotechnology & Drugs
Sector	Healthcare
Fiscal Year	12/31

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

Form 10-Q

(Mark One)

QUARTERLY REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934

For the quarterly period ended March 31, 2014

Or

TRANSITION REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934

For the transition period from _____ to _____.

Commission File Number: 000-26727

BioMarin Pharmaceutical Inc.

(Exact name of registrant as specified in its charter)

Delaware
(State or other jurisdiction of
incorporation or organization)

770 Lindero Street, San Rafael, California
(Address of principal executive offices)

68-0397820
(I.R.S. Employer
Identification No.)

94901
(Zip Code)

(415) 506-6700
(Registrant's telephone number including area code)

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 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 No

Indicate by check mark whether the registrant is a large accelerated filer, an accelerated filer, a non-accelerated filer, or a smaller reporting company. See the definitions of "large accelerated filer," "accelerated filer" and "smaller reporting company" in Rule 12b-2 of the Exchange Act.

Large accelerated filer

Accelerated filer

Non-accelerated filer (Do not check if a smaller reporting company)

Smaller reporting company

Indicate by check mark whether the registrant is a shell company (as defined in Rule 12b-2 of the Exchange Act.) Yes No

Applicable only to issuers involved in bankruptcy proceedings during the preceding five years:

Indicate by check mark whether the registrant has filed all documents and reports required to be filed by Sections 12, 13 or 15(d) of the Securities Exchange Act of 1934 subsequent to the distribution of securities under a plan confirmed by a court. Yes No

Applicable only to corporate issuers:

Indicate the number of shares outstanding of each of the issuer's classes of common stock, as of the latest practicable date: 145,926,798 shares of common stock, par value \$0.001, outstanding as of April 18, 2014.

Table of Contents

BIOMARIN PHARMACEUTICAL INC.

TABLE OF CONTENTS

	<u>Page</u>
PART I. FINANCIAL INFORMATION	3
Item 1. Financial Statements	3
Condensed Consolidated Balance Sheets as of March 31, 2014 (Unaudited) and December 31, 2013	3
Condensed Consolidated Statements of Comprehensive Loss (Unaudited) for the three months ended March 31, 2014 and 2013	4
Condensed Consolidated Statements of Cash Flows (Unaudited) for the three months ended March 31, 2014 and 2013	5
Notes to Condensed Consolidated Financial Statements (Unaudited)	6
Item 2. Management's Discussion and Analysis of Financial Condition and Results of Operations	22
Item 3. Quantitative and Qualitative Disclosures about Market Risk	35
Item 4. Controls and Procedures	35
PART II. OTHER INFORMATION	35
Item 1. Legal Proceedings	35
Item 1A. Risk Factors	35
Item 2. Unregistered Sales of Equity Securities and Use of Proceeds	53
Item 3. Defaults Upon Senior Securities	53
Item 4. Mine Safety Disclosures	53
Item 5. Other Information	53
Item 6. Exhibits	54
SIGNATURE	55

BioMarin[®], Naglazyme[®] and Kuvan[®] are registered trademarks of BioMarin Pharmaceutical Inc., or its affiliates. Aldurazyme[®] is a registered trademark of BioMarin/Genzyme LLC. VIMIZIM[™] and Firdapse[™] are trademarks of BioMarin Pharmaceutical Inc., or its affiliates.

Table of Contents

BIOMARIN PHARMACEUTICAL INC.
CONDENSED CONSOLIDATED BALANCE SHEETS
March 31, 2014 and December 31, 2013
(In thousands of U.S. dollars, except per share amounts)

	<u>March 31,</u> <u>2014</u>	<u>December 31,</u> <u>2013</u> ⁽¹⁾
	(unaudited)	
ASSETS		
Current assets:		
Cash and cash equivalents	\$ 639,778	\$ 568,781
Short-term investments	247,703	215,942
Accounts receivable, net (allowance for doubtful accounts: \$551 and \$529, respectively)	110,462	117,822
Inventory	176,893	162,605
Current deferred tax assets	30,561	30,561
Other current assets	41,013	41,707
Total current assets	<u>1,246,410</u>	<u>1,137,418</u>
Noncurrent assets:		
Investment in BioMarin/Genzyme LLC	478	816
Long-term investments	251,450	267,700
Property, plant and equipment, net	437,066	319,316
Intangible assets, net	165,397	163,147
Goodwill	54,258	54,258
Long-term deferred tax assets	144,124	145,234
Other assets	41,545	156,171
Total assets	<u>\$ 2,340,728</u>	<u>\$ 2,244,060</u>
LIABILITIES AND STOCKHOLDERS' EQUITY		
Current liabilities:		
Accounts payable and accrued liabilities	\$ 156,038	\$ 183,271
Total current liabilities	156,038	183,271
Noncurrent liabilities:		
Long-term convertible debt	661,419	655,566
Long-term contingent acquisition consideration payable	38,430	30,790
Other long-term liabilities	24,720	33,392
Total liabilities	<u>880,607</u>	<u>903,019</u>
Stockholders' equity:		
Common stock, \$0.001 par value: 250,000,000 shares authorized at March 31, 2014 and December 31, 2013; 145,738,396 and 143,463,668 shares issued and outstanding at March 31, 2014 and December 31, 2013, respectively.	146	144
Additional paid-in capital	2,213,347	2,059,101
Company common stock held by Nonqualified Deferred Compensation Plan	(6,731)	(7,421)
Accumulated other comprehensive income	7,275	5,018
Accumulated deficit	(753,916)	(715,801)
Total stockholders' equity	<u>1,460,121</u>	<u>1,341,041</u>
Total liabilities and stockholders' equity	<u>\$ 2,340,728</u>	<u>\$ 2,244,060</u>

(1) December 31, 2013 balances were derived from the audited Consolidated Financial Statements included in the Company's Annual Report on Form 10-K for the year ended December 31, 2013, filed with the Securities and Exchange Commission (the SEC) on February 26, 2014.

The accompanying notes are an integral part of these Condensed Consolidated Financial Statements.

BIOMARIN PHARMACEUTICAL INC.

CONDENSED CONSOLIDATED STATEMENTS OF COMPREHENSIVE LOSS
 Three Months Ended March 31, 2014 and 2013
 (In thousands of U.S. dollars, except per share amounts)
 (Unaudited)

	2014	2013
REVENUES:		
Net product revenues	\$ 149,004	\$ 127,344
Collaborative agreement revenues	415	135
Royalty, license and other revenues	2,133	449
Total revenues	<u>151,552</u>	<u>127,928</u>
OPERATING EXPENSES:		
Cost of sales (excludes amortization of certain acquired intangible assets)	22,816	20,500
Research and development	86,166	83,743
Selling, general and administrative	60,069	51,050
Intangible asset amortization and contingent consideration	8,957	5,556
Total operating expenses	<u>178,008</u>	<u>160,849</u>
LOSS FROM OPERATIONS	(26,456)	(32,921)
Equity in the loss of BioMarin/Genzyme LLC	(338)	(401)
Interest income	1,123	718
Interest expense	(9,106)	(1,725)
Debt conversion expense	0	(10,420)
Other income	153	228
LOSS BEFORE INCOME TAXES	(34,624)	(44,521)
Provision for (benefit from) income taxes	3,491	(4,711)
NET LOSS	<u>\$ (38,115)</u>	<u>\$ (39,810)</u>
NET LOSS PER SHARE, BASIC	<u>\$ (0.26)</u>	<u>\$ (0.31)</u>
NET LOSS PER SHARE, DILUTED	<u>\$ (0.27)</u>	<u>\$ (0.31)</u>
Weighted average common shares outstanding, basic	<u>143,983</u>	<u>127,969</u>
Weighted average common shares outstanding, diluted	<u>144,157</u>	<u>127,969</u>
COMPREHENSIVE LOSS	<u>\$ (35,858)</u>	<u>\$ (38,453)</u>

The accompanying notes are an integral part of these Condensed Consolidated Financial Statements.

BIOMARIN PHARMACEUTICAL INC.

CONDENSED CONSOLIDATED STATEMENTS OF CASH FLOWS
Three Months Ended March 31, 2014 and 2013
(In thousands of U.S. dollars)
(Unaudited)

	<u>2014</u>	<u>2013</u>
CASH FLOWS FROM OPERATING ACTIVITIES:		
Net loss	\$ (38,115)	\$ (39,810)
Adjustments to reconcile net loss to net cash used in operating activities:		
Depreciation and amortization	12,023	11,348
Non-cash interest expense	6,698	216
Accretion of discount on investments	1,900	1,456
Equity in the loss of BioMarin/Genzyme LLC	338	401
Stock-based compensation	17,267	11,508
Gain on termination of lease	(8,858)	0
Deferred income taxes	(179)	(43)
Excess tax benefit from stock option exercises	(278)	(128)
Unrealized foreign exchange (gain) loss on forward contracts	1,323	(364)
Non-cash changes in the fair value of contingent acquisition consideration payable	8,151	4,751
Debt conversion expense	0	10,420
Changes in operating assets and liabilities:		
Accounts receivable, net	7,360	(11,279)
Inventory	(14,288)	(7,127)
Other current assets	(927)	(8,404)
Other assets	(955)	(1,016)
Accounts payable and accrued liabilities	(18,921)	(6,732)
Other long-term liabilities	587	2,774
Net cash used in operating activities	<u>(26,874)</u>	<u>(32,029)</u>
CASH FLOWS FROM INVESTING ACTIVITIES:		
Purchases of property, plant and equipment	(24,177)	(11,826)
Maturities and sales of investments	69,391	82,531
Purchase of available-for-sale investments	(84,306)	(68,770)
Business acquisitions, net of cash acquired	0	(9,875)
Net cash used in investing activities	<u>(39,092)</u>	<u>(7,940)</u>
CASH FLOWS FROM FINANCING ACTIVITIES:		
Proceeds from exercises of stock options	19,712	24,580
Taxes paid related to net share settlement of equity awards	(473)	(225)
Proceeds from public offering of common stock, net	117,463	0
Excess tax benefit from stock option exercises	278	128
Payments for debt conversion	0	(10,420)
Payment on maturity of 2013 convertible note	0	(98)
Repayment of capital lease obligations	(17)	(143)
Net cash provided by financing activities	<u>136,963</u>	<u>13,822</u>
NET INCREASE (DECREASE) IN CASH AND CASH EQUIVALENTS	70,997	(26,147)
Cash and cash equivalents:		
Beginning of period	\$ 568,781	\$ 180,527
End of period	<u>\$ 639,778</u>	<u>\$ 154,380</u>
SUPPLEMENTAL CASH FLOW DISCLOSURES:		
Cash paid for interest, net of interest capitalized into fixed assets	\$ 1	\$ 1,998
Cash paid for income taxes	381	646
Stock-based compensation capitalized into inventory	2,053	993
Depreciation capitalized into inventory	2,924	2,607
SUPPLEMENTAL CASH FLOW DISCLOSURES FROM INVESTING AND FINANCING ACTIVITIES:		
Decrease in accounts payable and accrued liabilities related to fixed assets	\$ (9,171)	\$ (6,407)
Conversion of convertible debt	0	238,277
Deferred offering costs reclassified into additional paid-in-capital as a result of conversion of convertible debt	0	2,315
Release of escrow balance for purchase of San Rafael Corporate Center	116,500	0

The accompanying notes are an integral part of these Condensed Consolidated Financial Statements.

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

(1) NATURE OF OPERATIONS AND BUSINESS RISKS

BioMarin Pharmaceutical Inc. (the Company or BioMarin), a Delaware corporation, develops and commercializes innovative biopharmaceuticals for serious diseases and medical conditions. BioMarin selects product candidates for diseases and conditions that represent a significant unmet medical need, have well-understood biology and provide an opportunity to be first-to-market or offer a significant benefit over existing products. The Company's product portfolio is comprised of five approved products and multiple investigational product candidates. The Company's approved products are VIMIZIM (elosulfase alpha), Naglazyme (galsulfase), Kuvan (sapropterin dihydrochloride), Aldurazyme (laronidase) and Firdapse (amifampridine phosphate).

Through March 31, 2014, the Company had accumulated losses of approximately \$753.9 million. If the Company elects to increase its spending on development programs significantly above current long-term plans or enters into potential licenses and other acquisitions of complementary technologies, products or companies, the Company may need additional capital. The Company expects to continue to finance future cash needs that exceed its operating activities primarily through its current cash, cash equivalents, short-term and long-term investments, and to the extent necessary, through proceeds from equity or debt financings, loans and collaborative agreements with corporate partners.

The Company is subject to a number of risks, including: the financial performance of VIMIZIM, Naglazyme, Kuvan, Aldurazyme and Firdapse; the potential need for additional financings; the Company's ability to successfully commercialize its product candidates, if approved; the uncertainty of the Company's research and development efforts resulting in future successful commercial products; the Company's ability to successfully obtain regulatory approval for new products; significant competition from larger organizations; reliance on the proprietary technology of others; dependence on key personnel; uncertain patent protection; dependence on corporate partners and collaborators; and possible restrictions on reimbursement from governmental agencies and healthcare organizations, as well as other changes in the health care industry.

(2) BASIS OF PRESENTATION

The accompanying Condensed Consolidated Financial Statements have been prepared pursuant to the rules and regulations of the SEC for Quarterly Reports on Form 10-Q and do not include all of the information and note disclosures required by U.S. generally accepted accounting principles (U.S. GAAP) for complete financial statements. The Condensed Consolidated Financial Statements should therefore be read in conjunction with the Consolidated Financial Statements and Notes thereto for the fiscal year ended December 31, 2013 included in the Company's Annual Report on Form 10-K.

The accompanying Condensed Consolidated Financial Statements have been prepared in accordance with U.S. GAAP, which requires management to make estimates and assumptions that affect amounts reported in the Condensed Consolidated Financial Statements and accompanying disclosures. Although these estimates are based on management's best knowledge of current events and actions that the Company may undertake in the future, actual results may be different from those estimates. The Condensed Consolidated Financial Statements reflect all adjustments of a normal, recurring nature that are, in the opinion of management, necessary for a fair presentation of results for these interim periods. The results of operations for the three months ended March 31, 2014 are not necessarily indicative of the results that may be expected for the fiscal year ending December 31, 2014.

The Company has evaluated events and transactions subsequent to the balance sheet date. Based on this evaluation, the Company is not aware of any events or transactions that occurred subsequent to the balance sheet date but prior to filing this Quarterly Report on Form 10-Q that would require recognition or disclosure in the Condensed Consolidated Financial Statements.

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

(3) SIGNIFICANT ACCOUNTING POLICIES

There have been no material changes to the Company’s significant accounting policies during the three months ended March 31, 2014, as compared to the significant accounting policies disclosed in Note 3 of the Consolidated Financial Statements in the Company’s Annual Report on Form 10-K for the year ended December 31, 2013.

Reclassifications

Certain items in the Company’s prior year Condensed Consolidated Financial Statements have been reclassified to conform to the current presentation.

(4) RECENT ACCOUNTING PRONOUNCEMENTS

There have been no new accounting pronouncements or changes to accounting pronouncements during the three months ended March 31, 2014, as compared to the recent accounting pronouncements described in the Company’s Annual Report on Form 10-K for the year-ended December 31, 2013, that are of significance or potential significance to the Company.

(5) ACQUISITION OF SAN RAFAEL CORPORATE CENTER

On March 10, 2014, the Company completed the acquisition of the real estate commonly known as the San Rafael Corporate Center (SRCC), located in San Rafael, California. SRCC is a multi-building, commercial property where, prior to the transaction, the Company was leasing a certain portion of the space for its headquarters and related operating activities. The purpose of this acquisition is to allow for future expansion of the Company’s corporate headquarters to accommodate anticipated headcount growth. The acquisition of SRCC has been accounted for as a business combination because the building and the in-place leases met the definition of a business in Accounting Standards Codification 805 (ASC 805), *Business Combinations*. The purchase price for SRCC was \$116.5 million. The fair value of the consideration paid was \$116.5 million, all of which was paid in cash, which was held in escrow as of December 31, 2013.

The following table summarizes the estimated fair values of assets acquired as of the date of acquisition:

	Estimated Fair Value	Estimated Useful Lives
Building and improvements	\$ 94,414	50 years
Land	14,565	
Land improvements	3,616	10 years
Intangible assets	3,905	Remaining lease term
Total identifiable net assets	\$ 116,500	

The fair values assigned to tangible and identifiable intangible assets acquired assumed are based on management’s estimates and assumptions based on the information that was available as of the date of the acquisition. The Company believes that the information provides a reasonable basis for estimating the fair values of assets acquired.

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

The following table sets forth the fair value of the components of the identifiable intangible assets acquired by asset class:

Above market leases	\$ 351
In-place leases	3,554
Total intangible assets subject to amortization	<u>\$ 3,905</u>

The value of any in-place leases are estimated to be equal to the property owners' avoidance of costs necessary to release the property for a lease term equal to the remaining primary in-place lease term and the value of investment grade tenancy, which is derived by estimating, based on a review of the market, the cost to be borne by a property owner to replicate a market lease for the remaining in-place term. These costs consist of: (i) rent lost during downtime (e.g., assumed periods of vacancy), (ii) estimated expenses that would be incurred by the property owner during periods of vacancy, (iii) rent concessions (e.g., free rent), (iv) leasing commissions and (v) tenant improvement allowances. The Company determined these values using management's estimates along with third-party appraisals. The Company will amortize the capitalized value of in-place lease intangible assets to expense over the remaining initial term of each lease. The Company will amortize the capitalized value of above market leases to expense over the remaining lives of the underlying leases.

The amount of third-party tenant revenue (included in the line item Royalty, License and Other Revenues) and net income from third-party tenants included in the Company's Condensed Consolidated Statements of Comprehensive Loss from the acquisition date of March 10, 2014, through the period ended March 31, 2014, were \$0.4 million and \$0.3 million, respectively.

SRCC's results of operations prior to the acquisition were insignificant to the Company's Condensed Consolidated Financial Statements.

Included in Selling, General and Administrative (SG&A) expenses are transaction costs incurred in connection with the acquisition of \$0.2 million during the three months ended March 31, 2014. In connection with the purchase of SRCC, the Company recognized a gain of \$8.8 million in the three months ended March 31, 2014, due to the early termination of the Company's lease and the realization of the remaining balance in deferred rent and the reversal of the related asset retirement obligation upon acquisition of the SRCC. \$2.7 million and \$6.1 million of the gain were included in SG&A and Research and Development (R&D) expenses, respectively. The allocation of the gain to SG&A and R&D is consistent with the Company's allocation practices for facility costs for this previously leased space.

(6) STOCKHOLDERS' EQUITY

In March 2014, the Company sold 1.5 million shares of its common stock at a price of \$78.45 per share in an underwritten public offering pursuant to an effective registration statement previously filed with the SEC. The Company received net proceeds of approximately \$117.5 million from this public offering.

(7) NET LOSS PER COMMON SHARE

Potentially issuable shares of common stock include shares issuable upon the exercise of outstanding employee stock option awards, common stock issuable under the Company's Amended and Restated Employee Stock Purchase Plan (the ESPP), unvested restricted stock, common stock held by the Company's Nonqualified Deferred Compensation Plan and contingent issuances of common stock related to convertible debt.

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

The following table sets forth the computation of basic and diluted earnings per common share:

	Three Months Ended March 31,	
	2014	2013
Numerator:		
Net loss, basic	\$ (38,115)	\$ (39,810)
Gain on Company common stock issued to the Nonqualified Deferred Compensation Plan	(374)	0
Net loss, diluted	<u>\$ (38,489)</u>	<u>\$ (39,810)</u>
Denominator (in thousands of common shares):		
Basic weighted-average shares outstanding	143,983	127,969
Effect of dilutive securities:		
Common stock issued to the Nonqualified Deferred Compensation Plan	174	0
Fully diluted weighted-average shares	<u>144,157</u>	<u>127,969</u>
Basic loss per common share	<u>\$ (0.26)</u>	<u>\$ (0.31)</u>
Diluted loss per common share	<u>\$ (0.27)</u>	<u>\$ (0.31)</u>

In addition to the equity instruments included in the table above, the table below presents potential shares of common stock that were excluded from the computation as they were anti-dilutive using the treasury stock method (in thousands of common shares):

	Three Months Ended March 31,	
	2014	2013
Options to purchase common stock	12,444	12,884
Common stock issuable under the 2017 Notes	3,047	5,396
Common stock issuable under the 2018 Notes and the 2020 Notes	7,966	0
Unvested restricted stock units	1,326	1,375
Potentially issuable common stock for ESPP purchases	209	325
Common stock held by the Nonqualified Deferred Compensation Plan	0	202
Total number of potentially issuable shares	<u>24,992</u>	<u>20,182</u>

The Company accounts for the effect of the 2018 Notes and the 2020 Notes on diluted net loss per share using the treasury stock method since they may be settled in cash or shares at the Company's option. As a result, the 2018 Notes and the 2020 Notes have no effect on diluted net loss per share until the Company's stock price exceeds the conversion price of \$94.15 per share for the Notes. In the period of conversion, the Notes will have no impact on diluted net loss if the Notes are settled in cash and will have an impact on dilutive loss per share if the Notes are settled in shares upon conversion.

(8) INVESTMENTS

All investments were classified as available-for-sale at March 31, 2014 and December 31, 2013. The amortized cost, gross unrealized holding gains or losses, and fair value of the Company's available-for-sale securities by major security type at March 31, 2014 and December 31, 2013 are summarized in the tables below:

	Amortized Cost	Gross Unrealized Holding Gains	Gross Unrealized Holding Losses	Aggregate Fair Value at March 31, 2014
Certificates of deposit	\$ 53,171	\$ 5	\$ 0	\$ 53,176
Corporate debt securities	383,897	413	(379)	383,931
Commercial paper	52,946	19	0	52,965
U.S. Government agency securities	8,900	2	0	8,902
Greek government-issued bonds	52	127	0	179
Total	<u>\$ 498,966</u>	<u>\$ 566</u>	<u>\$ (379)</u>	<u>\$ 499,153</u>

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

	Amortized Cost	Gross Unrealized Holding Gains	Gross Unrealized Holding Losses	Aggregate Fair Value at December 31, 2013
Certificates of deposit	\$ 47,008	\$ 2	\$ 0	\$ 47,010
Corporate debt securities	341,519	313	(423)	341,409
Commercial paper	86,154	24	0	86,178
U.S. Government agency securities	8,900	1	0	8,901
Greek government-issued bonds	52	92	0	144
Total	<u>\$ 483,633</u>	<u>\$ 432</u>	<u>\$ (423)</u>	<u>\$ 483,642</u>

The Company has an investment in marketable equity securities which is measured using quoted prices in its respective active market that is considered a strategic investment. As of March 31, 2014, the fair value of the Company's marketable equity securities of \$15.1 million included an unrealized gain of \$12.1 million. As of December 31, 2013, the fair value of the Company's marketable equity securities of \$13.0 million includes an unrealized gain of \$10.1 million. This investment is recorded in Other Assets in the Company's Condensed Consolidated Balance Sheets.

The fair values of available-for-sale securities by contractual maturity were as follows:

	March 31, 2014	December 31, 2013
Maturing in one year or less	\$ 247,703	\$ 215,942
Maturing after one year through three years	251,450	267,700
Total	<u>\$ 499,153</u>	<u>\$ 483,642</u>

Impairment assessments are made at the individual security level each reporting period. When the fair value of an investment is less than its cost at the balance sheet date, a determination is made as to whether the impairment is other-than-temporary and, if it is other-than-temporary, an impairment loss is recognized in earnings equal to the difference between the investment's amortized cost and fair value at such date. As of March 31, 2014, some of the Company's investments were in an unrealized loss position. However, none of the underlying investments have been in a continuous loss position longer than twelve months, and no other-than-temporary impairment is deemed to have occurred.

See Note 14 to these Condensed Consolidated Financial Statements for additional discussion regarding the fair value of the Company's available-for-sale securities.

(9) INTANGIBLE ASSETS

Intangible assets consisted of the following:

	March 31, 2014	December 31, 2013
Intangible assets:		
Finite-lived intangible assets	\$ 123,147	\$ 118,242
Indefinite-lived intangible assets	74,430	74,430
Gross intangible assets:	197,577	192,672
Less: Accumulated amortization	<u>(32,180)</u>	<u>(29,525)</u>
Net carrying value	<u>\$ 165,397</u>	<u>\$ 163,147</u>

Indefinite-Lived Intangible Assets

Indefinite-lived intangible assets consist of IPR&D assets related to both early and late stage product candidates purchased in the acquisitions of Huxley Pharmaceuticals Inc. (Huxley), LEAD Therapeutics, Inc. (LEAD), ZyStor Therapeutics, Inc. (ZyStor) and Zacharon.

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

Intangible assets related to IPR&D assets are considered to be indefinite-lived until the completion or abandonment of the associated research and development efforts. During the period the assets are considered indefinite-lived, they will not be amortized but will be tested for impairment on an annual basis and between annual tests if the Company becomes aware of any events occurring or changes in circumstances that would indicate a reduction in the fair value of the IPR&D assets below their respective carrying amounts.

See Note 10 to the Consolidated Financial Statements included in the Company's Annual Report on Form 10-K for the year ended December 31, 2013, for additional information related to the Company's Intangible Assets.

(10) PROPERTY, PLANT AND EQUIPMENT

Property, plant and equipment, net consisted of the following:

	March 31,	December 31,
	<u>2014</u>	<u>2013</u>
Leasehold improvements	\$ 58,527	\$ 73,973
Building and improvements	270,441	159,125
Manufacturing and laboratory equipment	99,978	95,126
Computer hardware and software	78,729	74,948
Furniture and equipment	12,661	12,367
Land improvements	3,616	0
Land	26,145	11,608
Construction-in-progress	81,559	77,212
	<u>631,656</u>	<u>504,359</u>
Less: Accumulated depreciation	(194,590)	(185,043)
Total property, plant and equipment, net	<u>\$ 437,066</u>	<u>\$ 319,316</u>

Depreciation expense for the three months ended March 31, 2014 and 2013 was \$9.6 million and \$8.7 million, respectively, of which \$2.9 million and \$2.6 million was capitalized into inventory, respectively.

Capitalized interest related to the Company's property, plant and equipment purchases for each of the three months ended March 31, 2014 and 2013 was insignificant.

(11) SUPPLEMENTAL BALANCE SHEET INFORMATION

Inventory consisted of the following:

	March 31,	December 31,
	<u>2014</u>	<u>2013</u>
Raw materials	\$ 14,491	\$ 15,309
Work-in-process	105,663	88,417
Finished goods	56,739	58,879
Total inventory	<u>\$ 176,893</u>	<u>\$ 162,605</u>

Other Assets consisted of the following:

	March 31,	December 31,
	<u>2014</u>	<u>2013</u>
Deposits	\$ 7,239	\$ 7,196
Escrow balance for SRCC purchase	0	116,500
Deferred offering costs	14,364	15,374
Strategic investment	15,067	13,000
Other	4,875	4,101
Total other assets	<u>\$ 41,545</u>	<u>\$ 156,171</u>

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

Accounts payable and accrued liabilities consisted of the following:

	March 31,	December 31,
	<u>2014</u>	<u>2013</u>
Accounts payable	\$ 15,328	\$ 36,894
Accrued accounts payable	64,395	58,408
Accrued compensation expense	22,006	33,496
Accrued vacation expense	12,210	10,487
Current portion of contingent acquisition consideration payable	12,393	11,882
Accrued rebates payable	10,763	10,429
Accrued royalties payable	4,670	5,829
Value added taxes payable	3,504	3,603
Other accrued operating expenses	3,822	4,875
Current portion of nonqualified deferred compensation liability	905	1,363
Other	6,042	6,005
Total accounts payable and accrued liabilities	<u>\$ 156,038</u>	<u>\$ 183,271</u>

(12) CONVERTIBLE DEBT

The following table summarizes information regarding the Company's convertible debt:

	March 31,	December 31,
	<u>2014</u>	<u>2013</u>
Convertible Notes due 2020, net of unamortized discount of \$85,301 and \$87,975, respectively	\$ 289,699	\$ 287,025
Convertible Notes due 2018, net of unamortized discount of \$65,320 and \$68,500, respectively	309,680	306,500
Convertible Notes due 2017	62,040	62,041
Total long-term convertible debt, net of unamortized discount	<u>\$ 661,419</u>	<u>\$ 655,566</u>
Fair value of fixed rate convertible debt		
Convertible Notes due in 2020 ⁽¹⁾	\$ 416,618	\$ 400,879
Convertible Notes due in 2018 ⁽¹⁾	411,742	397,691
Convertible Notes due in 2017 ⁽¹⁾	207,577	213,765
Total	<u>\$ 1,035,937</u>	<u>\$ 1,012,335</u>

- (1) The fair value of the Company's fixed rate convertible debt is based on open market trades and is classified as Level 1 in the fair value hierarchy.

Interest expense on the Company's convertible debt was comprised of the following:

	Three Months Ended March, 31	
	<u>2014</u>	<u>2013</u>
Coupon interest	\$ 2,408	\$ 1,509
Amortization of issuance costs	843	216
Accretion of debt discount	5,855	0
Total interest expense on convertible debt	<u>\$ 9,106</u>	<u>\$ 1,725</u>

See Note 5 to the Consolidated Financial Statements included in the Company's Annual Report on Form 10-K for the year ended December 31, 2013, for additional information related to the Company's Convertible Debt.

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

(13) DERIVATIVE INSTRUMENTS AND HEDGING STRATEGIES*Foreign Currency Exchange Rate Exposure*

The Company uses forward foreign currency exchange contracts to hedge certain operational exposures resulting from potential changes in foreign currency exchange rates. Such exposures result from portions of the Company's forecasted revenues and operating expenses being denominated in currencies other than the U.S. dollar, primarily the Euro, the British Pound and the Brazilian Real.

The Company designates certain of these forward foreign currency exchange contracts as hedging instruments and enters into some forward foreign currency exchange contracts that are considered to be economic hedges that are not designated as hedging instruments. Whether designated or undesignated, these forward foreign currency exchange contracts protect against the reduction in value of forecasted foreign currency cash flows resulting from Naglazyme product revenues, Aldurazyme royalty revenues, operating expenses and net asset or liability positions designated in currencies other than the U.S. dollar. The fair values of forward foreign currency exchange contracts are estimated using current exchange rates and interest rates, and take into consideration the current creditworthiness of the counterparties or the Company, as applicable. Details of the specific instruments used by the Company to hedge its exposure to foreign currency exchange rate fluctuations are discussed below. See Note 14 to these Condensed Consolidated Financial Statements for additional discussion regarding the fair value of forward foreign currency exchange contracts.

At March 31, 2014, the Company had 105 forward foreign currency exchange contracts outstanding to sell a total of 141.4 million Euros with expiration dates ranging from April 2014 through February 2017. These hedges were entered into in order to protect against the fluctuations in revenue associated with Euro denominated Naglazyme and Aldurazyme sales. The Company has formally designated these forward foreign currency exchange contracts as cash flow hedges and expects them to be highly effective in offsetting fluctuations in revenues denominated in Euros related to changes in foreign currency exchange rates.

The Company also enters into forward foreign currency exchange contracts that are not designated as hedges for accounting purposes. The changes in fair value of these forward foreign currency exchange contracts are included as a part of SG&A expense in the Company's Condensed Consolidated Statements of Comprehensive Loss. At March 31, 2014, the Company had two outstanding forward foreign currency exchange contract to sell 38.6 million Euros and 2.8 million British Pounds, which were not designated as a hedge for accounting purposes and matured on April 30, 2014.

The maximum length of time over which the Company is hedging its exposure to the reduction in value of forecasted foreign currency cash flows through forward foreign currency exchange contracts is through February 2017. Over the next twelve months, the Company expects to reclassify \$1.4 million from accumulated other comprehensive income to earnings as the forecasted revenue transactions occur.

The fair value carrying amounts of the Company's derivative instruments were as follows:

	Asset Derivatives March 31, 2014		Liability Derivatives March 31, 2014	
	Balance Sheet Location	Fair Value	Balance Sheet Location	Fair Value
Derivatives designated as hedging instruments:				
Forward foreign currency exchange contracts	Other current assets	\$ 38	Accounts payable and accrued liabilities	\$ 1,329
Forward foreign currency exchange contracts	Other assets	275	Other long-term liabilities	73
Total		<u>\$ 313</u>		<u>\$ 1,402</u>
Derivatives not designated as hedging instruments:				
Forward foreign currency exchange contracts	Other current assets	\$ 0	Accounts payable and accrued liabilities	\$ 115
Total		<u>0</u>		<u>115</u>
Total value of derivative contracts		<u>\$ 313</u>		<u>\$ 1,517</u>

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

	Asset Derivatives December 31, 2013		Liability Derivatives December 31, 2013	
	Balance Sheet Location	Fair Value	Balance Sheet Location	Fair Value
Derivatives designated as hedging instruments:				
Forward foreign currency exchange contracts	Other current assets	\$ 0	Accounts payable and accrued liabilities	\$ 2,186
Forward foreign currency exchange contracts	Other assets	0	Other long-term liabilities	0
Total		<u>\$ 0</u>		<u>\$ 2,186</u>
Derivatives not designated as hedging instruments:				
Forward foreign currency exchange contracts	Other current assets	\$ 59	Accounts payable and accrued liabilities	\$ 0
Total		<u>59</u>		<u>0</u>
Total value of derivative contracts		<u>\$ 59</u>		<u>\$ 2,186</u>

The effect of the Company's derivative instruments on the Condensed Consolidated Financial Statements for the three months ended March 31, 2014 and 2013 was as follows:

	Forward Foreign Currency Exchange Contracts Three Months Ended March 31,	
	2014	2013
Derivatives Designated as Hedging Instruments:		
Net gain recognized in Other Comprehensive Income (OCI) ⁽¹⁾	\$ 1,396	\$ 679
Net gain (loss) reclassified from accumulated OCI into income ⁽²⁾	(567)	320
Net gain (loss) recognized in income ⁽³⁾	(121)	105
Derivatives Not Designated as Hedging Instruments:		
Net gain recognized in income ⁽⁴⁾	\$ 56	\$ 901

(1) Net change in the fair value of the effective portion classified as OCI.

(2) Effective portion classified as net product revenue.

(3) Ineffective portion and amount excluded from effectiveness testing classified as selling, general and administrative expense.

(4) Classified as selling, general and administrative expense.

At March 31, 2014 and December 31, 2013, accumulated other comprehensive income before taxes associated with forward foreign currency exchange contracts qualifying for hedge accounting treatment was a gain of \$1.1 million and a loss of \$2.4 million, respectively.

The Company is exposed to counterparty credit risk on all of its derivative financial instruments. The Company has established and maintains strict counterparty credit guidelines and enters into hedges only with financial institutions that are investment grade or better to minimize the Company's exposure to potential defaults. The Company does not require collateral to be pledged under these agreements.

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

(14) FAIR VALUE MEASUREMENTS

The Company measures certain financial assets and liabilities at fair value on a recurring basis, including available-for-sale fixed income securities and foreign currency derivatives. The tables below present the fair value of these financial assets and liabilities determined using the following input levels.

	Fair Value Measurements at March 31, 2014			
	Quoted Price in			
	Active Markets	Significant Other	Significant	Total
	for Identical	Observable Inputs	Unobservable	
Assets	(Level 2)	Inputs		
	(Level 1)	(Level 3)		
Assets:				
Cash and cash equivalents:				
Overnight deposits	\$ 137,884	\$ 0	\$ 0	\$ 137,884
Money market instruments	0	501,894	0	501,894
Total cash and cash equivalents	\$ 137,884	\$ 501,894	\$ 0	\$ 639,778
Available-for-sale securities:				
Short-term:				
Certificates of deposit	\$ 0	\$ 36,207	\$ 0	\$ 36,207
Corporate debt securities	0	158,531	0	158,531
Commercial paper	0	52,965	0	52,965
Long-term:				
Certificates of deposit	0	16,969	0	16,969
Corporate debt securities	0	225,400	0	225,400
U.S. Government agency securities	0	8,902	0	8,902
Greek government-issued bonds	0	179	0	179
Total available-for-sale securities	\$ 0	\$ 499,153	\$ 0	\$ 499,153
Other Current Assets:				
Nonqualified Deferred Compensation Plan assets	\$ 0	\$ 219	\$ 0	\$ 219
Forward foreign currency exchange contract assets ⁽¹⁾	0	38	0	38
Restricted investments ⁽²⁾	0	2,350	0	2,350
Total other current assets	\$ 0	\$ 2,607	\$ 0	\$ 2,607
Other Assets:				
Nonqualified Deferred Compensation Plan assets	\$ 0	\$ 4,366	\$ 0	\$ 4,366
Forward foreign currency exchange contract assets ⁽¹⁾	0	275	0	275
Strategic investment ⁽³⁾	15,067	0	0	15,067
Total other assets	\$ 15,067	\$ 4,641	\$ 0	\$ 19,708
Total assets	\$ 152,951	\$ 1,008,295	\$ 0	\$ 1,161,246
Liabilities:				
Current Liabilities:				
Nonqualified Deferred Compensation Plan liability	\$ 686	\$ 219	\$ 0	\$ 905
Forward foreign currency exchange contract liability ⁽¹⁾	0	1,444	0	1,444
Contingent acquisition consideration payable	0	0	12,393	12,393
Total current liabilities	\$ 686	\$ 1,663	\$ 12,393	\$ 14,742
Other long-term liabilities:				
Nonqualified Deferred Compensation Plan liability	\$ 11,184	\$ 4,366	\$ 0	\$ 15,550
Forward foreign currency exchange contract liability ⁽¹⁾	0	73	0	73
Contingent acquisition consideration payable	0	0	38,430	38,430
Asset retirement obligation (ARO)	0	0	3,850	3,850
Total other long-term liabilities	\$ 11,184	\$ 4,439	\$ 42,280	\$ 57,903
Total liabilities	\$ 11,870	\$ 6,102	\$ 54,673	\$ 72,645

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

	Fair Value Measurements at December 31, 2013			
	Quoted Price in			Total
	Active Markets for Identical Assets (Level 1)	Significant Other Observable Inputs (Level 2)	Significant Unobservable Inputs (Level 3)	
Assets:				
Cash and cash equivalents:				
Overnight deposits	\$ 156,228	\$ 0	\$ 0	\$ 156,228
Money market instruments	0	412,553	0	412,553
Total cash and cash equivalents	\$ 156,228	\$ 412,553	\$ 0	\$ 568,781
Available-for-sale securities:				
Short-term:				
Certificates of deposit	\$ 0	\$ 30,513	\$ 0	\$ 30,513
Corporate debt securities	0	99,251	0	99,251
Commercial paper	0	86,178	0	86,178
Long-term:				
Certificates of deposit	0	16,497	0	16,497
Corporate debt securities	0	242,158	0	242,158
U.S. Government agency securities	0	8,901	0	8,901
Greek government-issued bonds	0	144	0	144
Total available-for-sale securities	\$ 0	\$ 483,642	\$ 0	\$ 483,642
Other Current Assets:				
Nonqualified Deferred Compensation Plan assets	\$ 0	\$ 136	\$ 0	\$ 136
Forward foreign currency exchange contract assets ⁽¹⁾	0	59	0	59
Restricted investments ⁽²⁾	0	5,670	0	5,670
Total other current assets	\$ 0	\$ 5,865	\$ 0	\$ 5,865
Other Assets:				
Nonqualified Deferred Compensation Plan assets	\$ 0	\$ 3,459	\$ 0	\$ 3,459
Restricted investments ⁽²⁾	0	412	0	412
Strategic investment ⁽³⁾	13,000	0	0	13,000
Total other assets	\$ 13,000	\$ 3,871	\$ 0	\$ 16,871
Total assets	\$ 169,228	\$ 905,931	\$ 0	\$ 1,075,159
Liabilities:				
Current Liabilities:				
Nonqualified Deferred Compensation Plan liability	\$ 1,227	\$ 136	\$ 0	\$ 1,363
Forward foreign currency exchange contract liability ⁽¹⁾	0	2,186	0	2,186
Contingent acquisition consideration payable	0	0	11,882	11,882
Total current liabilities	\$ 1,227	\$ 2,322	\$ 11,882	\$ 15,431
Other long-term liabilities:				
Nonqualified Deferred Compensation Plan liability	\$ 12,345	\$ 3,459	\$ 0	\$ 15,804
Contingent acquisition consideration payable	0	0	30,790	30,790
Asset retirement obligation	0	0	4,122	4,122
Total other long-term liabilities	\$ 12,345	\$ 3,459	\$ 34,912	\$ 50,716
Total liabilities	\$ 13,572	\$ 5,781	\$ 46,794	\$ 66,147

- (1) See Note 13 to these Condensed Consolidated Financial Statements for further information regarding the derivative instruments.
- (2) The restricted investments at March 31, 2014 secure the Company's irrevocable standby letter of credit obtained in connection with certain commercial agreements. The restricted investments at December 31, 2013 secure the Company's irrevocable standby letter of credit obtained in connection with the Company's SRCC lease and certain commercial agreements.
- (3) The Company has an investment in marketable equity securities measured using quoted prices in an active market that is considered a strategic investment. See Note 8 to these Condensed Consolidated Financial Statements for additional discussion regarding the Company's strategic investment.

There were no transfers between levels during the three months ended March 31, 2014.

The Company's Level 2 securities are valued using third-party pricing sources. The pricing services utilize industry standard valuation models, including both income and market-based approaches, for which all significant inputs are observable, either directly or indirectly, to estimate fair value. These inputs include reported trades of and broker/dealer quotes on the same or similar securities, issuer credit spreads, benchmark securities, prepayment/default projections based on historical data and other observable inputs.

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

The Company validates the prices provided by its third-party pricing services by understanding the models used, obtaining market values from other pricing sources, analyzing pricing data in certain instances and confirming those securities traded in active markets. See Note 8 to these Condensed Consolidated Financial Statements for further information regarding the Company’s financial instruments.

Liabilities measured at fair value using Level 3 inputs were comprised of contingent acquisition consideration payable and asset retirement obligations.

The Company’s contingent acquisition consideration payable is estimated using a probability-based income approach utilizing an appropriate discount rate. Key assumptions used by management to estimate the fair value of contingent acquisition consideration payable include estimated probabilities, the estimated timing of when a milestone may be attained and assumed discount periods and rates. Subsequent changes in the fair value of the contingent acquisition consideration payable, resulting from management’s revision of key assumptions, will be recorded in Intangible Asset Amortization and Contingent Consideration in the Company’s Condensed Consolidated Statements of Comprehensive Loss. The probability-based income approach used by management to estimate the fair value of the contingent acquisition consideration is most sensitive to changes in the estimated probabilities.

Contingent acquisition consideration payable at December 31, 2013	\$ 42,672
Changes in the fair value of the contingent acquisition consideration payable	<u>8,151</u>
Contingent acquisition consideration payable at March 31, 2014	<u>\$ 50,823</u>

Under certain of the Company’s lease agreements, the Company is contractually obligated to return leased space to its original condition upon termination of the lease agreement. The Company records an asset retirement obligation liability and a corresponding capital asset in an amount equal to the estimated fair value of the obligation when estimable. In subsequent periods, for each such lease, the Company records Interest Expense to accrete the asset retirement obligation liability to full value and depreciates each capitalized asset retirement obligation asset, both over the term of the associated lease agreement.

Asset retirement obligations at December 31, 2013	\$ 4,122
Accretion	30
Release of SRCC ARO accruals	<u>(302)</u>
Asset retirement obligations at March 31, 2014	<u>\$ 3,850</u>

The Company acquired intangible assets as a result of various business acquisitions. The estimated fair value of these long-lived assets was measured using Level 3 inputs as of the acquisition date.

(15) STOCK-BASED COMPENSATION

The Company’s stock-based compensation plans include the 2006 Share Incentive Plan and the ESPP and the 2012 Inducement Plan, which expired in May 2013. The Company’s stock-based compensation plans are administered by the Compensation Committee of the Board of Directors, which selects persons to receive awards and determines the number of shares subject to each award and the terms, conditions, performance measures and other provisions of the award. See Note 16 to the Consolidated Financial Statements included in the Company’s Annual Report on Form 10-K for the year ended December 31, 2013, for additional information related to these stock-based compensation plans.

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

Determining the Fair Value of Stock Options and Stock Purchase Rights

The fair value of each option award is estimated on the date of grant using the Black-Scholes valuation model and the assumptions noted in the tables below. The expected life of options is based on observed historical exercise patterns. Groups of employees that have similar historical exercise patterns were considered separately for valuation purposes, but none were identified that had distinctly different exercise patterns as of March 31, 2014. The expected volatility of stock options is based upon the weighted average of the historical volatility of the Company's common stock and the implied volatility of traded options on the Company's common stock for fiscal periods in which there is sufficient trading volume in options on the Company's common stock. The risk-free interest rate is based on the implied yield on a U.S. Treasury zero-coupon issue with a remaining term equal to the expected term of the option. The dividend yield reflects that the Company has not paid any cash dividends since inception and does not intend to pay any cash dividends in the foreseeable future. The assumptions used to estimate the per share fair value of stock options granted under the 2012 Inducement Plan and the 2006 Share Incentive Plan were as follows:

	<u>Three Months Ended March 31,</u>	
	<u>2014</u>	<u>2013</u>
Expected volatility	44 – 45%	44%
Dividend yield	0.0%	0.0%
Expected life	6.9 years	6.7 years
Risk-free interest rate	2.1 – 2.3%	1.1%

During the three months ended March 31, 2014, the Company granted 107,922 options with a weighted average option value of \$37.44 per option.

The Company did not grant any new stock purchase rights under the ESPP during the three months ended March 31, 2014.

Restricted Stock Unit Awards with Service-Based Vesting Conditions

RSUs are generally subject to forfeiture if employment terminates prior to the release of vesting restrictions. The Company expenses the cost of the RSUs, which is determined to be the fair market value of the shares of common stock underlying the RSUs at the date of grant, ratably over the period during which the vesting restrictions lapse. During the three months ended March 31, 2014, the Company granted 27,880 RSUs with a weighted average fair market value of \$77.33 per share.

Restricted Stock Unit Awards with Performance and Market-Based Vesting Conditions

Pursuant to the approval of the Board of Directors, the Company granted RSU awards with performance and market-based vesting conditions during 2012 and 2011 to certain executive officers. As of March 31, 2014, these awards provide for a base award of 860,000 RSUs (the Base RSUs), with a weighted-average grant date fair value of \$34.66. The number of RSUs that could potentially vest from the Base RSUs granted is contingent upon achievement of specific performance goals and will be multiplied by the Total Shareholder Return multiplier which could range from 75% to 125% to determine the number of earned RSUs.

Stock-based compensation expense for this award will be recognized over the remaining service period beginning in the period the Company determines the strategic performance goal or goals is probable of achievement. During the fourth quarter of 2013, management concluded that regulatory approval of VIMIZIM was probable and the Company began recognizing compensation expense related to the performance based RSUs allocated to this performance goal. The Company recognized compensation expense of \$0.6 million for these awards for the three months ended March 31, 2014. For the three months ended March 31, 2013, the Company did not recognize any expense related to these awards because the Company's management had not yet determined the goals were probable of achievement.

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

Compensation expense included in the Company's Condensed Consolidated Statements of Comprehensive Loss for all stock-based compensation arrangements was as follows:

	<u>Three Months Ended March 31,</u>	
	<u>2014</u>	<u>2013</u>
Cost of sales	\$ 1,086	\$ 1,044
Research and development	7,115	5,324
Selling, general and administrative	8,103	5,197
Total stock-based compensation expense	<u>\$ 16,304</u>	<u>\$ 11,565</u>

Stock-based compensation of \$2.1 million and \$1.0 million was capitalized into inventory, for the three months ended March 31, 2014 and 2013, respectively. Capitalized stock-based compensation is recognized as cost of sales when the related product is sold.

(16) COMPREHENSIVE LOSS

The following table summarizes amounts reclassified out of Accumulated Other Comprehensive Income/(Loss) (AOCI) and their effect on the Company's Condensed Consolidated Statements of Comprehensive Loss for the three months ended March 31, 2014 and 2013.

Details about AOCI Components	<u>Amount Reclassified from AOCI</u> <u>(Gain) Loss</u> <u>Three Months Ended March, 31</u>		<u>Condensed Consolidated Statement of</u> <u>Comprehensive Loss Classification</u>
	<u>2014</u>	<u>2013</u>	
(Gains) Losses on cash flow hedges:			
Forward foreign currency exchange contracts	\$ 887	\$ (472)	Net product revenues
Forward foreign currency exchange contracts	0	(27)	Selling, general and administrative
Income tax effect of the above items	(320)	179	Provision for (benefit from) income taxes
	<u>\$ 567</u>	<u>\$ (320)</u>	Net loss

The following table summarizes changes in the accumulated balances for each component of AOCI, including current period other comprehensive income and reclassifications out of AOCI, for the three months ended March 31, 2014 and 2013.

	<u>Before</u> <u>Tax</u> <u>Amount</u>	<u>Tax (Expense)</u> <u>Benefit</u>	<u>Net-of-Tax</u> <u>Amount</u>
	AOCI balance at December 31, 2013	\$ 7,757	\$ (2,739)
Foreign currency translation adjustment	5	0	5
Unrealized gain (loss) on available-for-sale securities			
Unrealized holding gains (loss)	2,244	(821)	1,423
Less: reclassification adjustment for gain (loss) realized in net loss	0	0	0
Net unrealized holding gain (loss)	<u>2,244</u>	<u>(821)</u>	<u>1,423</u>
Net unrealized holding gain (loss) on cash flow hedges			
Unrealized holding gain (loss)	2,183	(787)	1,396
Less: reclassification adjustment for gain (loss) realized in net loss	(887)	320	(567)
Net unrealized holding gain (loss)	<u>1,296</u>	<u>(467)</u>	<u>829</u>
Other comprehensive income	<u>3,545</u>	<u>(1,288)</u>	<u>2,257</u>
AOCI balance at March 31, 2014	<u>\$11,302</u>	<u>\$ (4,027)</u>	<u>\$ 7,275</u>

	<u>Before Tax</u> <u>Amount</u>	<u>Tax (Expense)</u> <u>Benefit</u>	<u>Net-of-Tax</u> <u>Amount</u>
	AOCI balance at December 31, 2012	\$ (222)	\$ 20
Foreign currency translation adjustment	148	0	148
Unrealized gain (loss) on available-for-sale securities			
Unrealized holding gains (loss)	330	(120)	210
Less: reclassification adjustment for gain (loss) realized in net loss	0	0	0

Net unrealized holding gain (loss)	<u>330</u>	<u>(120)</u>	<u>210</u>
Net unrealized holding gain (loss) on cash flow hedges			
Unrealized holding gain (loss)	1,062	(383)	679
Less: reclassification adjustment for gain (loss) realized in net loss	<u>499</u>	<u>(179)</u>	<u>320</u>
Net unrealized holding gain (loss)	<u>1,561</u>	<u>(562)</u>	<u>999</u>
Other comprehensive income	<u>2,039</u>	<u>(682)</u>	<u>1,357</u>
AOCI balance at March 31, 2013	<u>\$ 1,817</u>	<u>\$ (662)</u>	<u>\$ 1,155</u>

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

(17) REVENUE AND CREDIT CONCENTRATIONS

Net Product Revenue— The Company considers there to be revenue concentration risks for regions where net product revenue exceeds ten percent of consolidated net product revenue. The concentration of the Company’s net product revenue within the regions below may have a material adverse effect on the Company’s revenue and results of operations if sales in the respective regions experience difficulties.

The table below summarizes net product revenue concentrations based on patient location for VIMIZIM, Naglazyme, Kuvan and Firdapse and the headquarters for Genzyme Corporation (Genzyme) for Aldurazyme. Although Genzyme sells Aldurazyme worldwide, the royalties earned by the Company on Genzyme’s net sales are included in the U.S. region, as the transactions are with Genzyme whose headquarters are located in the U.S.

Region:	Three Months Ended March 31,	
	2014	2013
United States	47%	48%
Europe	20%	21%
Latin America	16%	16%
Rest of world	17%	15%
Total net product revenue	<u>100%</u>	<u>100%</u>

The following table illustrates the percentage of the Company’s consolidated net product revenue attributed to the Company’s four largest customers.

	Three Months Ended March 31,	
	2014	2013
Customer A	16%	15%
Customer B ⁽¹⁾	12%	13%
Customer C	11%	13%
Customer D	11%	10%
Total	<u>50%</u>	<u>51%</u>

- (1) Genzyme is the Company’s sole customer for Aldurazyme and is responsible for marketing and selling Aldurazyme to third-parties. Net product revenues from Genzyme are comprised of royalties on worldwide net Aldurazyme sales and incremental product transfer revenue.

The accounts receivable balances at March 31, 2014 and December 31, 2013 were comprised of amounts due from customers for net product sales of VIMIZIM, Naglazyme, Kuvan and Firdapse and Aldurazyme product transfer and royalty revenues. On a consolidated basis, the Company’s two largest customers accounted for 40% and 16% of the March 31, 2014 accounts receivable balance, respectively, compared to December 31, 2013 when the two largest customers accounted for 45% and 15% of the accounts receivable balance, respectively. As of March 31, 2014 and December 31, 2013, accounts receivable for the Company’s largest customer balance included \$22.2 million and \$26.3 million, respectively, of unbilled accounts receivable related to net incremental Aldurazyme product transfers to Genzyme. The Company does not require collateral from its customers, but does perform periodic credit evaluations of its customers’ financial condition and requires immediate payment in certain circumstances.

The Company’s product sales to government-owned or government-funded customers in certain European countries, including Italy, Spain, Portugal, Greece and Russia, are subject to payment terms that are statutorily determined. Because these customers are government-owned or government-funded, the Company may be impacted by declines in sovereign credit ratings or sovereign defaults in these countries. A significant or further decline in sovereign credit ratings or a default in these countries may decrease the likelihood that the Company will collect accounts receivable or may increase the discount rates and the length of time until receivables are collected, which could result in a negative impact to the Company’s operating results. For the three months ended March 31, 2014, approximately 5% of the Company’s net product revenues were from these countries. Additionally, approximately 20% of the Company’s outstanding accounts receivable at March 31, 2014 related to such countries.

BIOMARIN PHARMACEUTICAL INC.

NOTES TO CONDENSED CONSOLIDATED FINANCIAL STATEMENTS – (Continued)
(In thousands of U.S. dollars, except per share amounts or as otherwise disclosed)

The following table summarizes the accounts receivable by country that were past due related to Italy, Spain, Portugal, Greece and Russia, the number of days past due and the total allowance for doubtful accounts related to each of these countries at March 31, 2014.

	Days Past Due			Total Amount Past Due	Allowance for Doubtful Accounts
	< 180 Days	180 — 360 Days	> 360 Days		
Italy	\$ 0	\$ 0	\$ 0	\$ 0	\$ 0
Spain	1,798	333	46	2,177	0
Portugal	467	0	0	467	0
Greece	0	0	365	365	365
Russia	0	0	0	0	0
Total	<u>\$ 2,265</u>	<u>\$ 333</u>	<u>\$ 411</u>	<u>\$ 3,009</u>	<u>\$ 365</u>

The Company also sells its products in other countries that face economic crises and local currency devaluation. Although the Company has historically collected receivables from customers in those countries, sustained weakness or further deterioration of the local economies and currencies may cause customers in those countries to be unable to pay for the Company's products. The Company has not historically experienced a significant level of uncollected receivables and has received continued payments from its more aged accounts. The Company believes that the allowances for doubtful accounts related to these countries is adequate based on its analysis of the specific business circumstances and expectations of collection for each of the underlying accounts in these countries.

(18) INCOME TAXES

The Company has historically computed interim period tax expense by applying its forecasted effective tax rate to year-to-date earnings. However, due to a significant amount of U.S. permanent differences relative to the amount of U.S. forecasted loss used in computing the effective tax rate, the effective tax rate is highly sensitive to minor fluctuations in forecasted income. As such, the Company has computed U.S. tax expense for the three months ended March 31, 2014 using an actual year-to-date tax calculation. Foreign tax expense was computed using a forecasted annual effective tax rate.

(19) COMMITMENTS AND CONTINGENCIES

The Company is also subject to contingent payments totaling approximately \$581.5 million as of March 31, 2014 which are due upon achievement of certain regulatory and licensing milestones if they occur before certain dates in the future. Of this amount, \$56.4 million relates to programs that are no longer being developed.

In the normal course of business, the Company enters into various firm purchase commitments primarily related to research and development and certain inventory related items. As of March 31, 2014, these commitments for the next five years were approximately \$37.4 million. These amounts primarily relate to active pharmaceutical ingredients and represent minimum purchase requirements and post marketing commitments related to the Company's approved products.

Item 2. Management’s Discussion and Analysis of Financial Condition and Results of Operations

Forward-Looking Statements

This Quarterly Report on Form 10-Q contains “forward-looking statements” as defined under securities laws. Many of these statements can be identified by the use of terminology such as “believes,” “expects,” “anticipates,” “plans,” “may,” “will,” “projects,” “continues,” “estimates,” “potential,” “opportunity” or the negative versions of these terms and other similar expressions. These forward-looking statements may be found in “*Overview*,” of this Item 2 and other sections of this Quarterly Report on Form 10-Q. Our actual results or experience could differ significantly from the forward-looking statements. Factors that could cause or contribute to these differences include those discussed in “Risk Factors,” in Part II, Item 1A of this Quarterly Report on Form 10-Q as well as information provided elsewhere in this Quarterly Report on Form 10-Q and our Annual Report on Form 10-K for the year end December 31, 2013. You should carefully consider that information before you make an investment decision.

You should not place undue reliance on these types of forward-looking statements, which speak only as of the date that they were made. These forward-looking statements are based on the beliefs and assumptions of our management based on information currently available to management and should be considered in connection with any written or oral forward-looking statements that we may issue in the future as well as other cautionary statements we have made and may make. We do not undertake any obligation to release publicly any revisions to these forward-looking statements after completion of the filing of this Quarterly Report on Form 10-Q to reflect later events or circumstances or the occurrence of unanticipated events.

The following discussion of our financial condition and results of operations should be read in conjunction with our Condensed Consolidated Financial Statements and the related Notes thereto included elsewhere in this Quarterly Report on Form 10-Q.

Overview

We develop and commercialize innovative biopharmaceuticals for serious diseases and medical conditions. We select product candidates for diseases and conditions that represent a significant unmet medical need, have well-understood biology and provide an opportunity to be first-to-market or offer a significant benefit over existing products.

Key components of our results of operations include the following (in millions):

	<u>Three Months Ended March 31,</u>	
	<u>2014</u>	<u>2013</u>
Total net product revenues	\$ 149.0	\$ 127.3
Cost of sales	22.8	20.5
Research and development expense	86.2	83.7
Selling, general and administrative expense	60.1	51.1
Intangible asset amortization and contingent consideration expense	9.0	5.6
Net loss	(38.1)	(39.8)
Stock-based compensation expense	16.3	11.6

See “*Results of Operations*” below for a discussion of the detailed components and analysis of the amounts above.

Our product portfolio is comprised of five approved products and multiple investigational product candidates. Our approved products are VIMIZIM (elosulfase alpha), Naglazyme (galsulfase), Kuvan (sapropterin dihydrochloride), Aldurazyme (laronidase) and Firdapse (amifampridine phosphate).

Management's Discussion and Analysis of Financial Condition and Results of Operations – (Continued)

VIMIZIM, a treatment mucopolysaccharidosis Type IVA or Morquio Syndrome Type A, a lysosomal storage disorder, received marketing approval in the U.S. in February 2014 and in the European Union (the EU) in April 2014. We immediately began marketing VIMIZIM in the U.S. using our existing sales force and commercial organization and completed our first commercial sale in the U.S. in February 2014. VIMIZIM net product revenues for the three months ended March 31, 2014 totaled \$0.9 million.

Naglzyme, a recombinant form of N-acetylgalactosamine 4-sulfatase indicated for patients with mucopolysaccharidosis VI (MPS VI), a debilitating life-threatening genetic disease for which no other drug treatment currently exists and which is caused by the deficiency of arylsulfatase B, received marketing approval in the U.S. in May 2005, in the EU in January 2006 and subsequently in other countries. Naglzyme net product revenues for the three months ended March 31, 2014 totaled \$80.1 million, compared to \$69.4 million for the three months ended March 31, 2013.

Kuvan was granted marketing approval for the treatment of phenylketonuria (PKU) in the U.S. in December 2007 and in the EU in December 2008. Kuvan net product revenues for the three months ended March 31, 2014 totaled \$45.2 million, compared to \$37.6 million for the three months ended March 31, 2013.

Aldurazyme, which was developed in collaboration with Genzyme Corporation (Genzyme), was approved in 2003 for marketing in the U.S. and the EU and subsequently in other countries for patients with mucopolysaccharidosis I (MPS I). Aldurazyme net product revenues for the three months ended March 31, 2014 totaled \$18.1 million, compared to \$16.7 million for the three months ended March 31, 2013.

In December 2009, the European Medicines Agency granted marketing approval for Firdapse, a proprietary form of 3-4-diaminopyridine (amifampridine phosphate), for the treatment of Lambert-Eaton Myasthenic Syndrome (LEMS). We launched this product on a country-by-country basis in the EU beginning in April 2010. Firdapse net product revenues for the three months ended March 31, 2014 totaled \$4.7 million, compared to \$3.6 million for the three months ended March 31, 2013.

We are conducting clinical trials on several investigational product candidates for the treatment of various diseases including:

- PEG PAL, an enzyme substitution therapy for the treatment of PKU;
- BMN 701, an enzyme replacement therapy for Pompe disease, a glycogen storage disorder;
- BMN 673, an orally available poly-ADP ribose polymerase inhibitor for the treatment of patients with certain cancers;
- BMN 111, a peptide therapeutic for the treatment of achondroplasia, the leading cause of dwarfism; and
- BMN 190 for the treatment of late infantile neuronal ceroid lipofuscinosis (CLN2), lysosomal storage disorder primarily affecting the brain.

We are conducting or planning to conduct preclinical development of several other product candidates for genetic and other metabolic diseases and recently announced the selection of two new drug development candidates, BMN 270 and BMN 250. BMN 270 is a Factor VIII gene therapy drug development candidate, an AAV VIII vector, for the treatment of hemophilia A. BMN 250 is a novel fusion of alpha-N-acetylglucosaminidase (NAGLU) with a peptide derived from insulin-like growth factor 2 (IGF2), for the treatment of Sanfilippo B syndrome or Mucopolysaccharidosis type IIIB (MPS IIIB).

Cost of sales includes raw materials, personnel and facility and other costs associated with manufacturing VIMIZIM, Naglzyme and Aldurazyme at our production facility in Novato, California. Cost of sales also includes third-party manufacturing costs for the production of the active ingredient in Kuvan and Firdapse and third-party production costs related to final formulation and packaging services for all products and cost of royalties payable to third-parties for all products.

Management's Discussion and Analysis of Financial Condition and Results of Operations – (Continued)

Research and development includes costs associated with the research and development of product candidates and post-marketing research commitments related to our approved products. These costs primarily include preclinical and clinical studies, personnel and raw materials costs associated with manufacturing product candidates, quality control and assurance, research and development facilities and regulatory costs.

Selling, general and administrative expense primarily includes expenses associated with the commercialization of approved products and general and administrative costs to support our operations. These expenses include: product marketing and sales operations personnel; corporate facility operating expenses; information technology expenses and depreciation; and core corporate support functions, including human resources, finance and legal, and other external corporate costs such as insurance, legal fees and other professional services.

Intangible asset amortization and contingent consideration includes amortization expense related to our finite-lived intangible assets associated with marketing rights in the EU for Firdapse, impairment losses (if any) on intangible assets and changes in the fair value of contingent acquisition consideration payable. Changes in fair value can result from changes in estimated probability adjustments, changes in estimated timing of when a milestone may be achieved, changes in assumed discount periods and rates and passage of time.

Our cash, cash equivalents, short-term investments and long-term investments totaled \$1,138.9 million as of March 31, 2014, compared to \$1,052.4 million as of December 31, 2013. We have historically financed our operations primarily through our cash flows from operating activities, the issuance of common stock and convertible debt and by relying on equipment and other commercial financing. We will be highly dependent on our net product revenue to supplement our current liquidity and fund our operations for the foreseeable future. We may in the future elect to supplement this with further debt or equity offerings or commercial borrowing. Further, depending on market conditions, our financial position and performance and other factors, we may in the future choose to use a portion of our cash or cash equivalents to repurchase our convertible debt or other securities. See “*Financial Position, Liquidity and Capital Resources*” below for a further discussion of our liquidity and capital resources.

Critical Accounting Policies and Estimates

In preparing our Condensed Consolidated Financial Statements in accordance with accounting principles generally accepted in the U.S. and pursuant to the rules and regulations promulgated by the SEC, we make assumptions, judgments and estimates that can have a significant impact on our net income/(loss) and affect the reported amounts of certain assets, liabilities, revenue and expenses, and related disclosures. We base our assumptions, judgments and estimates on historical experience and various other factors that we believe to be reasonable under the circumstances. Actual results could differ materially from these estimates under different assumptions or conditions. On a regular basis, we evaluate our assumptions, judgments and estimates. We also discuss our critical accounting policies and estimates with the Audit Committee of our Board of Directors.

We believe that the assumptions, judgments and estimates involved in the accounting for business combinations, contingent acquisition consideration payable, income taxes, long-lived assets, revenue recognition and inventory have the greatest impact on our Condensed Consolidated Financial Statements, so we consider these to be our critical accounting policies. Historically, our assumptions, judgments and estimates relative to our critical accounting policies have not differed materially from actual results.

There have been no significant changes to our critical accounting policies and estimates during three months ended March 31, 2014, as compared to the critical accounting policies and estimates disclosed in “*Management's Discussion and Analysis of Financial Condition and Results of Operations*” included in our Annual Report on Form 10-K for the year ended December 31, 2013, which was filed with the SEC on February 26, 2014.

Recent Accounting Pronouncements

See Note 4 to our accompanying Condensed Consolidated Financial Statements for a full description of recent accounting pronouncements and our expectation of their impact, if any, on our results of operations and financial condition.

Table of Contents

Management's Discussion and Analysis of Financial Condition and Results of Operations – (Continued)

Results of Operations

Net Loss

Our net loss for the three months ended March 31, 2014 was \$38.1 million, compared to a net loss of \$39.8 million for the three months ended March 31, 2013. The decrease in net loss was primarily a result of the following (in millions):

Net loss for the period ended March 31, 2013	\$ (39.8)
Increased gross profit from product sales	19.4
Absence of debt conversion expense	10.4
Increased selling, general and administrative expense	(9.0)
Decreased benefit from income taxes	(8.2)
Increased interest expense	(7.4)
Increased intangible asset amortization and contingent consideration	(3.4)
Increased research and development expense	(2.4)
Other individually insignificant fluctuations	2.3
Net loss for the period ended March 31, 2014	<u>\$ (38.1)</u>

The increase in gross profit from product sales during the three months ended March 31, 2014 as compared to the three months ended March 31, 2013 was primarily a result of additional Naglazyme patients initiating therapy globally and additional Kuvan patients initiating therapy in the U.S. The increase in research and development expense was primarily attributed to increased development expenses for our PEG PAL program and earlier stage development programs. The increase in selling, general and administrative expense was primarily due to increased sales and marketing expenses related to our commercial products and increased expenses related to the commercial launch of VIMIZIM.

See below for additional information related to the primary net loss fluctuations presented above, including details of our operating expense fluctuations.

Net Product Revenues, Cost of Sales and Gross Profit

Net product revenues were as follows (in millions):

	Three Months Ended March 31,		
	2014	2013	Change
VIMIZIM	\$ 0.9	\$ 0	\$ 0.9
Naglazyme	80.1	69.4	10.7
Kuvan	45.2	37.6	7.6
Aldurazyme	18.1	16.7	1.4
Firdapse	4.7	3.6	1.1
Total net product revenues	<u>\$ 149.0</u>	<u>\$ 127.3</u>	<u>\$ 21.7</u>

Gross profit by product was as follows (in millions):

	Three Months Ended March 31,		
	2014	2013	Change
VIMIZIM	\$ 0.8	\$ 0	\$ 0.8
Naglazyme	69.0	59.7	9.3
Kuvan	38.1	31.6	6.5
Aldurazyme	14.8	12.7	2.1
Firdapse	3.5	2.8	0.7
Total gross profit	<u>\$ 126.2</u>	<u>\$ 106.8</u>	<u>\$ 19.4</u>

Table of Contents

Management's Discussion and Analysis of Financial Condition and Results of Operations – (Continued)

Net product revenues attributed to our collaboration with Genzyme were as follows (in millions):

	Three Months Ended March 31,		
	2014	2013	Change
Aldurazyme revenue reported by Genzyme	\$ 55.9	\$ 48.4	\$ 7.5

	Three Months Ended March 31,		
	2014	2013	Change
Royalties earned from Genzyme	\$ 21.9	\$ 19.3	\$ 2.6
Incremental (previously recognized) Aldurazyme product transfer revenue	(3.8)	(2.6)	(1.2)
Total Aldurazyme net product revenues	\$ 18.1	\$ 16.7	\$ 1.4

Net product revenues for Naglazyme for the three months ended March 31, 2014 totaled \$80.1 million, of which \$70.7 million was earned from customers based outside the U.S., compared to \$69.4 million for the three months ended March 31, 2013, of which \$60.1 million was earned from customers based outside the U.S. The increase in Naglazyme net product revenues was attributed to new patients initiating therapy. The impact of foreign currency exchange rates on Naglazyme sales denominated in currencies other than the U.S. dollar was positive by \$0.1 million for the three months ended March 31, 2013. Naglazyme gross margins for each of the three months ended March 31, 2014 and 2013 were 86%. Naglazyme gross margins for the three months ended March 31, 2014 were consistent with expectations and are not expected to fluctuate significantly in the future.

Net product revenue for Kuvan for the three months ended March 31, 2014 totaled \$45.2 million, compared to \$37.6 million for the three months ended March 31, 2013. The increase in Kuvan net product revenues was attributed to new patients initiating therapy. Kuvan gross margins for each of the three months ended March 31, 2014 and 2013 were 84%. Cost of goods sold for the three months ended March 31, 2014 and 2013 reflect royalties paid to third-parties of approximately 10%. Kuvan gross margins for the three months ended March 31, 2014 were consistent with expectations and are not expected to fluctuate significantly in the future. The 4% royalties earned from Merck Serono's net sales of Kuvan for the three months ended March 31, 2014 were \$0.5 million, compared to \$0.4 million for the three months ended March 31, 2013.

Aldurazyme gross margins were 82% for the three months ended March 31, 2014, compared to 76% for the three months ended March 31, 2013. Aldurazyme gross margins reflect the profit earned on royalty revenue and net incremental product transfer revenue. Aldurazyme gross margins are expected to fluctuate depending on the mix of royalty revenue, from which we earn higher gross profit, and product transfer revenue, from which we earn lower gross profit.

Net product revenue for Firdapse for the three months ended March 31, 2014 totaled \$4.7 million, compared to \$3.6 million for the three months ended March 31, 2013. Firdapse gross margins for the three months ended March 31, 2014 were 75%, compared to 77% for the three months ended March 31, 2013. Cost of goods sold for the three months ended March 31, 2014 and 2013 reflect royalties paid to third-parties of approximately 8%. Firdapse gross margins for the three months ended March 31, 2014 decreased due to increased manufacturing costs and the depletion of manufactured product that was previously expensed as research and development expense. Firdapse gross margins for the three months ended March 31, 2014 were consistent with expectations and are not expected to fluctuate significantly in the future.

In February 2014, the FDA granted marketing approval for VIMIZIM and we began marketing the product immediately. Net product revenues for VIMIZIM for the three months ended March 31, 2014 totaled \$0.9 million and gross margins were 91%.

Total cost of sales for the three months ended March 31, 2014 was \$22.8 million, compared to \$20.5 million for the three months ended March 31, 2013. The increase in cost of sales was primarily attributed to the increase in product sales.

Table of Contents

Management's Discussion and Analysis of Financial Condition and Results of Operations – (Continued)

Research and Development

We manage our research and development expense by identifying the research and development activities we anticipate will be performed during a given period and then prioritizing efforts based on scientific data, probability of successful development, market potential, available human and capital resources and other similar considerations. We continually review our pipeline and the development status of product candidates and, as necessary, reallocate resources among the research and development portfolio that we believe will best support the future growth of our business.

Research and development expense increased to \$86.2 million for the three months ended March 31, 2014, from \$83.7 million for the three months ended March 31, 2013. The increase in research and development expense was primarily a result of the following (in millions):

Research and development expense for the period ended March 31, 2013	\$ 83.7
Gain on early lease termination	6.1
Increased PEG PAL development expenses	4.9
Increased development expenses on early development stage programs	3.0
Increased BMN 190 development expenses	1.9
Increased stock-based compensation expenses related to research and development	1.8
Increased BMN 111 development expenses	1.0
Increased BMN 673 development expenses	0.3
Decreased VIMIZIM development expenses	(4.0)
Decreased BMN 701 development expenses	(2.0)
Decreased development expenses related to mature commercial products	(1.3)
Decrease in non-allocated research and development expenses and other net changes	(9.2)
Research and development expense for the period ended March 31, 2014	<u>\$ 86.2</u>

The increase in PEG PAL and BMN 673 development expense was attributed to increased clinical trial activities related to these product candidates. The increase in development expense on early development stage programs was primarily attributed to the pre-clinical activity related to BMN 270, BMN 250 and development costs related to the programs acquired from Zacharon Pharmaceuticals, Inc. (Zacharon). The increase in stock-based compensation is primarily attributed to an increase in the number of options outstanding due to an increased number of employees and an increase in the weighted-average fair value of the equity awards granted during 2013. The increases in BMN 190 and BMN 111 development expense were attributed to increased pre-clinical activities related to these product candidates. The decrease in non-allocated research and development expense is primarily attributed to a decline in research and development personnel costs and facility costs that are not allocated to specific programs. The gain in the three months ended March 31, 2014, resulted from the early termination of our lease and the recognition of the remaining deferred rent and asset retirement liabilities upon acquisition of SRCC where our corporate headquarters are located.

During 2014, we expect our research and development spending to increase over 2013 levels due to our PEG PAL, BMN 673, BMN 701, BMN 111 and BMN 190 programs progressing, including a few of those programs progressing to more advanced phases of clinical studies. Phase 3 clinical trials for PEG PAL and BMN 673 were initiated in the second and fourth quarters of 2013, respectively, and we expect to initiate a Phase 3 trial of BMN 701 in the second quarter of 2014. We also expect increased spending on pre-clinical and clinical activities for our early development stage programs including BMN 270, programs acquired from Zacharon and BMN 250. Additionally, we expect to continue incurring significant research and development expense for the foreseeable future due to long-term clinical activities related to post-approval regulatory commitments for our approved products. We continuously evaluate the recoverability of costs associated with pre-launch manufacturing activities, and if it is determined that recoverability is highly likely and therefore future revenues are expected, the costs subsequently incurred related to pre-launch manufacturing activities may be capitalized. When regulatory approval and the likelihood of future revenues for a product candidate are less certain, the related manufacturing costs are expensed as research and development expenses.

Selling, General and Administrative

Selling, general and administrative expense increased to \$60.1 million for the three months ended March 31, 2014, from \$51.1 million for the three months ended March 31, 2013. The increase in selling, general and administrative expenses was primarily a result of the following (in millions):

Table of Contents

Management's Discussion and Analysis of Financial Condition and Results of Operations – (Continued)

Selling, general and administrative expense for the period ended March 31, 2013	\$ 51.1
Gain on early lease termination	2.7
Increased VIMIZIM commercial launch expenses	7.6
Increased stock-based compensation	2.9
Decreased sales and marketing expenses related to mature commercial products	(1.5)
Decreased foreign exchange losses on unhedged transactions	(0.1)
Net decrease in corporate support and other administrative expenses	(2.6)
Selling, general and administrative expense for the period ended March 31, 2014	<u>\$ 60.1</u>

The increase in stock-based compensation is attributed to an increase in the number of options outstanding due to an increased number of employees, an increase in the weighted-average fair value of the equity awards granted during 2013. We continue to incur sales and marketing expense for Naglazyme and Kuvan as a result of continued expansion of our international and U.S. activities, respectively. Additionally, transaction costs associated with the SRCC acquisition increased selling, general and administrative expenses by \$0.2 million for the three months ended March 31, 2014. The gain in the three months ended March 31, 2014, resulted from the early termination of our lease and the recognition of the remaining deferred rent and asset retirement liabilities upon acquisition of the SRCC where our corporate headquarters are located. We expect selling, general and administrative expenses to increase in future periods as a result of the international expansion of Naglazyme, the U.S. commercialization activities for Kuvan, commercial launch activities for VIMIZIM and the administrative support of our expanding operations.

Intangible Asset Amortization and Contingent Consideration

Intangible asset amortization and contingent consideration expense is comprised of amortization of the European marketing rights for Firdapse, changes in the fair value of contingent acquisition consideration payable to former stockholders of our acquired businesses and impairment loss (if any) on intangible assets. Changes in the fair value of contingent acquisition consideration payable result from updates to the estimated probability of achievement or assumed timing of milestones and adjustments to the discount periods and rates. Intangible asset amortization and contingent consideration expense consisted of the following (in millions):

	Three Months Ended March 31,		
	2014	2013	Change
Changes in the fair value of contingent acquisition consideration payable	\$ 8.2	\$ 4.8	\$ 3.4
Amortization of Firdapse European marketing rights	0.8	0.8	0
Total intangible asset amortization and contingent consideration	<u>\$ 9.0</u>	<u>\$ 5.6</u>	<u>\$ 3.4</u>

The changes in the fair value of the contingent acquisition consideration payable were primarily attributed to changes in the estimated probability of achieving development milestones based on the current status of the related development programs as well as changes in the discount rate utilized in the fair value calculations. During the three months ended March 31, 2014, the majority of the changes related to the development progress of BMN 701.

Equity in the Loss of BioMarin/Genzyme LLC

Equity in the loss of BioMarin/Genzyme LLC (the LLC) includes our 50% share of the joint venture's loss for the period. The LLC's operations consist primarily of certain research and development activities and the intellectual property that are managed by the joint venture, with costs shared equally by BioMarin and Genzyme.

Equity in the loss of the joint venture totaled \$0.3 million for the three months ended March 31, 2014, compared to \$0.4 million for the three months ended March 31, 2013.

Table of Contents

Management's Discussion and Analysis of Financial Condition and Results of Operations – (Continued)

Interest Income

We invest our cash, short-term and long-term investments in government and other high credit quality securities in order to limit default and market risk. Interest income totaled \$1.1 million for the three months ended March 31, 2014, compared to \$0.7 million for the three months ended March 31, 2013. The increase in interest income during the three months ended March 31, 2014, as compared to the three months ended March 31, 2013 was primarily due to higher cash and investment balances. We expect future interest income to increase due to the \$696.4 million of net proceeds from our October 2013 debt offering and our March 2014 equity offering. See Note 5 to the Consolidated Financial Statements included in our Annual Report on Form 10-K for the year ended December 31, 2013, for additional information.

Interest Expense and Debt Conversion Expense

We incur interest expense on our convertible debt and our capital leases. Interest expense consisted of the following (in millions):

	Three Months Ended March 31,		
	2014	2013	Change
Coupon interest	\$ 2.4	\$ 1.5	\$ 0.9
Amortization of issuance costs	0.8	0	0.8
Accretion of discount on convertible notes	5.9	0.2	5.7
Total interest expense	<u>\$ 9.1</u>	<u>\$ 1.7</u>	<u>\$ 7.4</u>

The increase in interest expense in the three months ended March 31, 2014 compared to the three months ended March 31, 2013 was attributed to our October 2013 debt offering. In connection with the early conversion in March 2013 of \$215.0 million in aggregate principle of the 2017 Notes, we recognized debt conversion expense of \$10.4 million. We expect future interest expense to increase due to the October 2013 debt offering and the accretion of the related debt discount. See Note 5 to the Consolidated Financial Statements included in our Annual Report on Form 10-K for the year ended December 31, 2013, for additional information regarding our Convertible Debt.

Provision for (Benefit from) Income Taxes

During the three months ended March 31, 2014 we recognized income tax expense of \$3.5 million, compared to an income tax benefit of \$4.7 million in the three months ended March 31, 2013. We have historically computed interim period tax expense by applying our forecasted effective tax rate to year-to-date earnings. However, due to a significant amount of U.S. permanent differences relative to the amount of U.S. forecasted loss used in computing the effective tax rate, the effective tax rate is highly sensitive to minor fluctuations in forecasted income. As such, we have computed U.S. tax expense for the three months ended March 31, 2014 using an actual year-to-date tax calculation. Foreign tax expense was computed using a forecasted annual effective tax rate. The income tax benefit for the three months ended March 30, 2013 had previously been calculated using an estimate of the annual effective rate for the full fiscal year.

Income tax expense for the three months ended March 31, 2014 and 2013 consisted of state, federal and foreign current tax expense, which were offset by deferred tax benefits from federal orphan drug credits and California Research and Development (R&D) credits and resulted in a net benefit for the first quarter of 2013. Income tax expense for the three months ended March 31, 2013 was also offset by federal R&D credits. The provisions for the three months ended March 31, 2014 and 2013 were further reduced by the benefit related to stock option exercises during the three months ended March 31, 2014 and 2013. Additionally, the American Taxpayer Relief Act of 2012 (the Relief Act), was enacted on January 2, 2013. The Relief Act reinstated the federal R&D credit retroactively to January 1, 2012 through December 31, 2013. In accordance with Financial Accounts Standards Board Accounting Standards Codification Topic 740, *Income Taxes*, we accounted for the effects of change in the tax law in the period that included the enactment date of the change, resulting in the recognition of a deferred tax benefit of \$1.9 million related to R&D expenses incurred during 2012 as a discrete item during the three months ended March 31, 2013, which further increased our income tax benefit for the current period provision. See Note 20 to our Consolidated Financial Statements included in our Annual Report on Form 10-K for the year ended December 31, 2013 for additional discussion of the components of our provision for (benefit from) income taxes.

Management's Discussion and Analysis of Financial Condition and Results of Operations – (Continued)

Financial Position, Liquidity and Capital Resources

We expect to fund our operations with our net product revenues from our commercial products, cash, cash equivalents, short-term and long-term investments supplemented by proceeds from equity or debt financings and loans or collaborative agreements with corporate partners, each to the extent necessary. This expectation could change depending on how much we elect to spend on our development programs, potential licenses, and acquisitions of complementary technologies, products and companies or if we elect to settle all or a portion of our debt in cash. We will be highly dependent on our net product revenue to supplement our current liquidity and fund our operations for the foreseeable future. We may in the future elect to supplement this with further debt or equity offerings or commercial borrowing, even after giving effect to our October 2013 debt offering and our March 2014 equity offering.

We consider the unrepatriated cumulative earnings of certain of our foreign subsidiaries to be invested indefinitely outside the U.S. As of March 31, 2014, \$83.7 million of our \$1,138.9 million balance of cash, cash equivalents and marketable securities was from foreign subsidiary operations and is intended to fund future foreign operations. In managing our liquidity needs in the U.S., we do not rely on the unrepatriated earnings as a source of funds and we have not provided for U.S. federal or state income taxes on these undistributed foreign earnings.

We are mindful that conditions in the current macroeconomic environment could affect our ability to achieve our goals. Some of the factors that could affect our business include: future changes to healthcare reform in the U.S., a continuation or worsening of global economic conditions, patent expirations of competitive products and the launch of generic competitors, continued government pricing pressures internationally and the potential volatility in foreign currency exchange rates. We will continue to monitor these conditions and will adjust our business processes, as appropriate, to mitigate these risks to our business.

Our financial condition as of March 31, 2014 and December 31, 2013 was as follows (in millions):

	March 31,	December 31,	
	2014	2013	Change
Cash and cash equivalents	\$ 639.8	\$ 568.8	\$ 71.0
Short-term investments	247.7	215.9	31.8
Long-term investments	251.4	267.7	(16.3)
Cash, cash equivalents and investments	<u>\$ 1,138.9</u>	<u>\$ 1,052.4</u>	<u>\$ 86.5</u>
Current assets	\$ 1,246.4	\$ 1,137.4	\$ 109.0
Current liabilities	156.0	183.3	(27.3)
Working capital	<u>\$ 1,090.4</u>	<u>\$ 954.1</u>	<u>\$ 136.3</u>
Convertible debt	\$ 661.4	\$ 655.6	\$ 5.8

Our cash flows for each of the three months ended March 31, 2014 and 2013 are summarized as follows (in millions):

	Three Months Ended March 31,		
	2014	2013	Change
Cash and cash equivalents at the beginning of the period	\$ 568.8	\$ 180.5	\$ 388.3
Net cash used in operating activities	(26.9)	(32.0)	5.1
Net cash used in investing activities	(39.1)	(7.9)	(31.2)
Net cash provided by financing activities	137.0	13.8	123.2
Cash and cash equivalents at the end of the period	\$ 639.8	\$ 154.4	\$ 485.4
Short-term and long-term investments	499.1	368.0	127.8
Cash, cash equivalents and investments	<u>\$ 1,138.9</u>	<u>\$ 522.4</u>	<u>\$ 613.2</u>

Table of Contents

Management's Discussion and Analysis of Financial Condition and Results of Operations – (Continued)

Cash, Cash Equivalents and Investments

The increase in cash, cash equivalents and investments at March 31, 2014 from December 31, 2013 was primarily attributed to the net proceeds of \$117.5 million from our March 2014 public offering of common stock and employee stock exercises, offset by increases in cash used in operating activities and purchases of property, plant and equipment.

Working Capital

Working capital increased by \$136.3 million, from \$954.1 million at December 31, 2013 to \$1,090.4 million at March 31, 2014. The increase in working capital was attributed to the following (in millions):

Working capital at December 31, 2013	\$ 954.1
Increased cash, cash equivalents and short-term investments	102.8
Decreased accounts payable and accrued liabilities	27.3
Net increase in other current operating assets	6.2
Working capital at March 31, 2014	<u>\$ 1,090.4</u>

The increase in cash, cash equivalents and short-term investments was primarily attributed to the net proceeds of \$117.5 from our March 2014 equity offering and proceeds of \$19.7 million from employee stock exercises. The net proceeds from the public offering of our common stock and employee stock exercises were partially offset by \$26.9 million of cash used in operating activities.

The net increase in other current operating assets is attributed to a \$14.3 million increase in inventory, offset by a decrease of \$7.4 million in accounts receivable. The increase in inventory was primarily attributed buildup of Naglazyme and Aldurazyme inventories during the quarter. The decrease in accounts receivable is attributed to timing of net product revenues and cash receipts from customers.

Our product sales to government-owned or government-funded customers in certain countries, including Russia and the Southern European countries of Greece, Spain, Italy and Portugal, are subject to payment terms that are imposed by government authority. Because these customers are government-owned or government-funded, we may be impacted by declines in sovereign credit ratings or sovereign defaults in these countries. A significant or further decline in sovereign credit ratings or default in the Southern European countries or Russia, may decrease the likelihood that we will collect accounts receivable or may increase the discount rates and the length of time until receivables are collected, which could result in a negative impact to our operating results. Historically we have not experienced a significant level of uncollected receivables and have received continued payments from our more aged accounts. We believe that the allowances for doubtful accounts for these countries are adequate based on our analysis of the specific business circumstances and expectations of collection for each of the underlying accounts in these countries. As of March 31, 2014, approximately 20% of our outstanding accounts receivable relate to such countries. See Note 17 to our accompanying Condensed Consolidated Financial Statements for additional discussion. We also sell our products in other countries that face economic crises and local currency devaluation. Although we have historically collected receivables from customers in those countries, sustained weakness or further deterioration of the local economies and currencies may cause our customers in those countries to be unable to pay for our products with the same negative effect on our operations.

Cash Used in Operating Activities

Cash used in operating activities for the three months ended March 31, 2014 was \$26.9 million, compared to cash used in operating activities of \$32.0 million for the three months ended March 31, 2013. The decrease in cash used in operating activities was primarily related to increased payments of \$12.2 million and \$7.2 million for accounts payable and accrued liabilities and inventory purchases, respectively, offset by an \$18.6 million decrease in accounts receivable and a \$7.5 million increase in other current assets.

Management's Discussion and Analysis of Financial Condition and Results of Operations – (Continued)***Cash Used in Investing Activities***

Net cash used in investing activities during the three months ended March 31, 2014 was \$39.1 million compared to net cash used in investing activities of \$7.9 million during the three months ended March 31, 2013. Our investing activities have consisted primarily of purchases and sales and maturities of investments and capital expenditures, such as manufacturing equipment and facility improvements. The increase in net cash used in investing activities for the three months ended March 31, 2014 was primarily comprised of a \$12.4 million decrease in capital expenditures and \$28.7 million in net purchases of investments, offset by a decrease of \$9.9 million in business acquisitions. We expect to make significant capital investments in our Shanbally, Ireland manufacturing facility during the remainder of 2014 to enable future commercial manufacturing of our products at the facility.

Cash Provided by Financing Activities

Net cash provided by financing activities for the three months ended March 31, 2014 was \$137.0 million, compared to net cash provided by financing activities of \$13.8 million for the three months ended March 31, 2013. Historically, our financing activities primarily included payments related to our contingent acquisition obligations, payments related to our convertible debt obligations and proceeds from employee stock purchases under the ESPP and employee stock option exercises. The increase in net cash provided by financing activities for the three months ended March 31, 2014 was primarily attributed to of the net proceeds from our March 2014 equity offering, offset by decreased proceeds from employee stock option exercises of \$4.9 million.

Other Information

On October 15, 2013, we completed an offering of \$750.0 million in aggregate principal amount of senior subordinated convertible notes consisting of \$375.0 million in aggregate principal the 2018 Notes and \$375.0 million in aggregate principal the 2020 Notes. The net proceeds from the offering were \$696.4 million, after deducting commissions and offering expenses and the purchase of capped calls. The 2018 Notes and the 2020 Notes were issued at face value and accrue interest at annual rates of 0.75% and 1.50%, respectively, which is payable semiannually in arrears on April 15 and October 15 of each year beginning on April 15, 2014. See Note 5 to the Consolidated Financial Statements included in our Annual Report on Form 10-K for the year ended December 31, 2013, for additional information regarding our Convertible Debt.

In April 2007, we sold approximately \$324.9 million of the 2017 Notes of which \$62.0 million remained outstanding at March 31, 2014. The debt was issued at face value and bears interest at the rate of 1.875% per annum, payable semi-annually in cash. During 2013, we entered into separate agreements with 18 of the existing holders of the 2017 Notes pursuant to which such holders converted \$262.8 million in aggregate principal of the 2017 Notes into 12.9 million shares of our common stock. In addition to issuing the requisite number of shares of common stock pursuant to the 2017 Notes, we also made varying cash payments to each of the holders, totaling an aggregate of \$14.8 million, of which \$13.0 million was recognized as Debt Conversion Expense in our Consolidated Statement of Operations for the year ended December 31, 2013. The remaining 2017 Notes are convertible, at the option of the holder, at any time prior to maturity, into shares of our common stock at a conversion price of approximately \$20.36 per share, subject to adjustment in certain circumstances. Our debt does not contain a call provision and we are unable to unilaterally redeem the debt prior to maturity in 2017. We also must repay the debt if there is a qualifying change in control or termination of trading of our common stock. If a change of control occurs, we will pay a make whole premium by increasing the conversion rate applicable to the 2017 Notes. See Note 5 to the Consolidated Financial Statements included in our Annual Report on Form 10-K for the year ended December 31, 2013, for additional information regarding our Convertible Debt.

In March 2006, we sold approximately \$172.5 million the 2013 Notes, which fully matured on March 29, 2013. The debt was issued at face value and bore interest at the rate of 2.5% per annum, payable semi-annually in cash. The debt did not contain a call provision and we were unable to unilaterally redeem the remaining debt prior to maturity in March 2013. Upon maturity of the 2013 Notes, we issued 1.4 million shares of our common stock pursuant to the terms of the 2013 Notes and paid a bond holder \$98,000 in cash for the par value at maturity. See Note 5 to the Consolidated Financial Statements included in our Annual Report on Form 10-K for the year ended December 31, 2013, for additional information regarding our Convertible Debt.

Table of Contents

Management's Discussion and Analysis of Financial Condition and Results of Operations – (Continued)

Our \$812.0 million (undiscounted) of total convertible debt as of March 31, 2014 will impact our liquidity due to the semi-annual cash interest payments and will further impact our liquidity if we elect to settle all or portions of the 2018 Notes or the 2020 Notes in cash upon conversion or if the holders of our 2017 Notes do not convert on or prior to the scheduled repayments of the debt. Further, depending on market conditions, our financial position and performance and other factors, we may in the future choose to use a portion of our cash or cash equivalents to repurchase our convertible debt or other securities.

On January 4, 2013, we acquired Zacharon, which focused on developing small molecules targeting pathways of glycan and glycolipid metabolism, for a net cash upfront payment of \$9.7 million. In connection with the acquisition, we agreed to pay the Zacharon stockholders additional consideration in future periods of up to \$134.0 million (undiscounted) in milestone payments if certain clinical, development and sales milestones are met.

In March 2014, we sold 1.5 million shares of our common stock at a price of \$78.45 per share in an underwritten public offering pursuant to an effective registration statement previously filed with the SEC. We received net proceeds of approximately \$117.5 million from this public offering.

Funding Commitments

We cannot estimate with certainty the cost to complete any of our product development programs. Additionally, except as disclosed under "Overview" above, we cannot precisely estimate the time to complete any of our product development programs or when we expect to receive net cash inflows from any of our product development programs. Please see "Risk Factors" included in Part II Item 1A of this Quarterly Report on Form 10-Q, for a discussion of the reasons we are unable to estimate such information, and in particular the following risk factors:

- *If we fail to obtain or maintain regulatory approval to commercially market and sell our drugs, or if approval is delayed, we will be unable to generate revenue from the sale of these products, our potential for generating positive cash flow will be diminished, and the capital necessary to fund our operations will be increased;*
- *If we are unable to successfully develop and maintain manufacturing processes for our drug products to produce sufficient quantities at acceptable costs, we may be unable to meet demand for our products and lose potential revenue, have reduced margins or be forced to terminate a program;*
- *If we fail to compete successfully with respect to product sales, we may be unable to generate sufficient sales to recover our expenses related to the development of a product program or to justify continued marketing of a product and our revenue could be adversely affected; and*
- *If we do not achieve our projected development goals in the timeframes we announce and expect, the commercialization of our products may be delayed and the credibility of our management may be adversely affected and, as a result, our stock price may decline.*

Our investment in our product development programs and continued development of our existing commercial products has a major impact on our operating performance. Our research and development expenses in each of the three months ended March 31, 2014 and 2013 and the period since inception of the major programs were as follows (in millions):

	Three Months Ended March 31,		Since Program Inception
	2014	2013	
VIMIZIM	\$ 18.0	\$ 22.0	\$ 311.8
Naglazyme	2.6	2.9	179.9
Kuvan	3.0	4.0	158.2
Firdapse	1.2	1.2	35.6
BMN 673	5.0	4.7	61.6
BMN 701	10.8	12.8	108.0
BMN 111	4.3	3.3	51.2
BMN 190	5.1	3.2	36.6
PEG PAL	16.0	11.1	183.7
Not allocated to specific major current projects	20.2	18.5	Not meaningful
Totals	\$ 86.2	\$ 83.7	

Table of Contents

Management's Discussion and Analysis of Financial Condition and Results of Operations – (Continued)

We may elect to increase our spending above our current long-term plans and consequently we may be unable to achieve our long-term goals. This may increase our capital requirements, including: costs associated with the commercialization of our products; additional clinical trials; investments in the manufacturing of VIMIZIM, Naglazyme, Kuvan, Aldurazyme and Firdapse; preclinical studies and clinical trials for our other product candidates; potential licenses and other acquisitions of complementary technologies, products and companies; and general corporate purposes.

Our future capital requirements will depend on many factors, including, but not limited to:

- product sales and profitability of VIMIZIM, Naglazyme, Kuvan, Aldurazyme and Firdapse;
- manufacture, supply or distribution of VIMIZIM, Naglazyme, Kuvan, Aldurazyme and Firdapse;
- progress of our product candidates through the regulatory process and our ability to successfully commercialize any such products that receive regulatory approval;
- results of clinical trials, announcements of technological innovations or new products by us or our competitors;
- government regulatory action affecting our product candidates or our competitors' drug products in both the U.S. and in non-U.S. countries;
- developments or disputes concerning patent or proprietary rights;
- general market conditions and fluctuations for the emerging growth and pharmaceutical market sectors;
- economic conditions in the U.S. or abroad;
- broad market fluctuations in the U.S., the EU or in other parts of the world;
- actual or anticipated fluctuations in our operating results; and
- changes in Company assessments or financial estimates by securities analysts.

Off-Balance Sheet Arrangements

We do not have any off-balance sheet arrangements that are currently material or reasonably likely to be material to our consolidated financial position or results of operations.

Contractual and Commercial Obligations

We have contractual and commercial obligations under our debt, operating leases and other obligations related to research and development activities, purchase commitments, licenses and sales royalties with annual minimums. Our contractual obligations for non-cancelable purchase commitments as of March 31, 2014 are presented in the table below (in millions).

	Payments Due within				Total
	Less Than 1 Year	1-3 Years	3-5 Years	More Than 5 Years	
2017 Notes and related interest	\$ 1.2	\$ 2.4	\$ 62.6	\$ 0	\$ 66.2
2018 Notes and related interest	2.8	5.6	380.6	0	389.0
2020 Notes and related interest	5.6	11.2	11.2	386.3	414.3
Operating leases	4.8	11.1	7.7	4.6	28.2
Research and development and purchase commitments	25.3	9.3	2.8	0	37.4
Total	<u>\$ 39.7</u>	<u>\$ 39.6</u>	<u>\$ 464.9</u>	<u>\$ 390.9</u>	<u>\$ 935.1</u>

We are also subject to contingent payments totaling approximately \$581.5 million as of March 31, 2014, which are due upon achievement of certain regulatory and licensing milestones if they occur before certain dates in the future. Of this amount, \$56.4 million relates to programs that are no longer being developed.

Table of Contents

Item 3. Quantitative and Qualitative Disclosures About Market Risk

Our market risks during the three months ended March 31, 2014 have not materially changed from those discussed in Part II, Item 7A of our Annual Report on Form 10-K for the year ended December 31, 2013, which was filed with the SEC on February 26, 2014.

Item 4. Controls and Procedures

(a) Controls and Procedures

An evaluation was carried out, under the supervision of and with the participation of our management, including our Chief Executive Officer and our Chief Financial Officer, regarding the effectiveness of our disclosure controls and procedures (as defined in Rules 13a-15(e) and 15d-15(e) under the Securities Exchange Act of 1934, as amended (the Exchange Act)) as of the end of the period covered by this report.

Based on the evaluation, our Chief Executive Officer and our Chief Financial Officer have concluded that our disclosure controls and procedures are effective to ensure that the information required to be disclosed by us in the reports that we file or submit under the Exchange Act is recorded, processed, summarized and reported within the time periods specified in the SEC's rules and forms.

(b) Change in Internal Controls over Financial Reporting

There were no changes in our internal control over financial reporting, as such term is defined in Rules 13a-15(f) and 15d-15(f) under the Exchange Act, during our most recently completed quarter that have materially affected or are reasonably likely to materially affect our internal control over financial reporting. We are utilizing the Committee of Sponsoring Organizations of the Treadway Commission (COSO) 1992 Framework on internal control.

PART II. OTHER INFORMATION

Item 1. Legal Proceedings.

None.

Item 1A. Risk Factors

An investment in our securities involves a high degree of risk. We operate in a dynamic and rapidly changing industry that involves numerous risks and uncertainties. The risks and uncertainties described below are not the only ones we face. Other risks and uncertainties, including those that we do not currently consider material, may impair our business. If any of the risks discussed below actually occur, our business, financial condition, operating results or cash flows could be materially adversely affected. This could cause the value of our securities to decline, and you may lose all or part of your investment.

We have marked with an asterisk (*) those risk factors below that include a substantive change from or update to the risk factors included in our Annual Report on Form 10-K, for the year ended December 31, 2013, which was filed with the SEC on February 26, 2014.

Risk Related to Our Business

*** If we fail to obtain or maintain regulatory approval to commercially market and sell our drugs, or if approval is delayed, we will be unable to generate revenue from the sale of these products, our potential for generating positive cash flow will be diminished, and the capital necessary to fund our operations will be increased.**

We must obtain and maintain regulatory approval to market and sell our drug products in the U.S. and in jurisdictions outside of the U.S. In the U.S., we must obtain FDA approval for each drug that we intend to commercialize. The FDA approval process is typically lengthy and expensive, and approval is never certain. Products distributed abroad are also subject to government regulation by international regulatory authorities. The approval process in the EU and other countries can also be lengthy and expensive and regulatory approval is also never certain. Naglazyme, Aldurazyme and Kuvan have received regulatory approval to be commercially marketed and sold in the U.S., EU and other countries. Firdapse has received regulatory approval to be commercially marketed only in the EU. VIMIZIM received regulatory approval in the U.S. on February 14, 2014 and in the EU on April 28, 2014 but has not been approved in any other jurisdiction and may never receive additional regulatory approvals for any jurisdiction.

As part of the recent reauthorization of the Prescription Drug User Fee Act, new biologics are included in a new product review program intended to enhance FDA-sponsor communications to lead to greater first-cycle approval decisions. As part of this program, applications for new biologics are subject to either a 12-month standard or 8-month priority review period that begins from the date of application submission. However, since this is a new product review program and few products have completed this new review process, the priority review period may take longer than eight months and the standard review period may take longer than 12 months. Similarly, although the EMA has an accelerated approval process, the timelines mandated by the regulations are subject to the possibility of substantial delays.

In addition, the FDA and its international equivalents have substantial discretion over the approval process for pharmaceutical products. As such, these regulatory agencies may in the end not agree that we have demonstrated the requisite level of product safety and efficacy to grant approval and may require additional data. If we fail to obtain regulatory approval for our product candidates, we will be unable to market and sell those drug products. Because of the risks and uncertainties in pharmaceutical development, our product candidates could take a significantly longer time to gain regulatory approval than we expect or may never gain approval. We also rely on independent third-party contract research organizations (CROs), to file some of our ex-U.S. and ex-EU marketing applications and important aspects of the services performed for us by the CROs are out of our direct control. If we fail to adequately manage our CROs, if the CRO elects to prioritize work on our projects below other projects or if there is any dispute or disruption in our relationship with our CROs, the filing of our applications may be delayed.

From time to time during the regulatory approval process for our products and our product candidates, we engage in discussions with the FDA and comparable international regulatory authorities regarding the regulatory requirements for our development programs. To the extent appropriate, we accommodate the requests of the regulatory authorities and, to date, we have generally been able to reach reasonable accommodations and resolutions regarding the underlying issues. However, we are often unable to determine the outcome of such deliberations until they are final. If we are unable to effectively and efficiently resolve and comply with the inquiries and requests of the FDA and other non-U.S. regulatory authorities, the approval of our product candidates may be delayed and their value may be reduced.

After any of our products receive regulatory approval, they remain subject to ongoing regulation, which can impact, among other things product labeling, manufacturing practices, adverse event reporting, storage, expiration, distribution, advertising and promotion, and record keeping. If we do not comply with the applicable regulations, the range of possible sanctions includes issuance of adverse publicity, product recalls or seizures, fines, total or partial suspensions of production and/or distribution, suspension of marketing applications, and enforcement actions, including injunctions and civil or criminal prosecution. The FDA and comparable international regulatory agencies can withdraw a product's approval under some circumstances, such as the failure to comply with regulatory requirements or unexpected safety issues. Further, the FDA often requires post-marketing testing and surveillance to monitor the effects of approved products. The FDA and comparable international regulatory agencies may condition

Table of Contents

approval of our product candidates on the completion of such post-marketing clinical studies. These post-marketing studies may suggest that a product causes undesirable side effects or may present a risk to the patient. If data we collect from post-marketing studies suggest that one of our approved products may present a risk to safety, the government authorities could withdraw our product approval, suspend production or place other marketing restrictions on our products. If regulatory sanctions are applied or if regulatory approval is delayed or withdrawn, the value of our company and our operating results will be adversely affected. Additionally, we will be unable to generate revenue from the sale of these products, our potential for generating positive cash flow will be diminished and the capital necessary to fund our operations will be increased.

If we fail to obtain or maintain orphan drug exclusivity for some of our products, our competitors may sell products to treat the same conditions and our revenues will be reduced.

As part of our business strategy, we intend to develop some drugs that may be eligible for FDA and EU orphan drug designation. Under the Orphan Drug Act, the FDA may designate a product as an orphan drug if it is intended to treat a rare disease or condition, defined as a patient population of fewer than 200,000 in the U.S. The company that first obtains FDA approval for a designated orphan drug for a given rare disease receives marketing exclusivity for use of that drug for the stated condition for a period of seven years. Orphan drug exclusive marketing rights may be lost if the FDA later determines that the request for designation was materially defective or if the manufacturer is unable to assure sufficient quantity of the drug. Similar regulations are available in the EU with a ten-year period of market exclusivity.

Because the extent and scope of patent protection for some of our drug products is limited, orphan drug designation is especially important for our products that are eligible for orphan drug designation. For eligible drugs, we plan to rely on the exclusivity period under the Orphan Drug Act to maintain a competitive position. If we do not obtain orphan drug exclusivity for our drug products that do not have broad patent protection, our competitors may then sell the same drug to treat the same condition and our revenues will be reduced.

Even though we have obtained orphan drug designation for certain of our products and product candidates and even if we obtain orphan drug designation for our future product candidates, due to the uncertainties associated with developing pharmaceutical products, we may not be the first to obtain marketing approval for any particular orphan indication. Further, even if we obtain orphan drug exclusivity for a product, that exclusivity may not effectively protect the product from competition because different drugs can be approved for the same condition. Even after an orphan drug is approved, the FDA can subsequently approve the same drug for the same condition if the FDA concludes that the later drug is safer, more effective or makes a major contribution to patient care. Orphan drug designation neither shortens the development time or regulatory review time of a drug, nor gives the drug any advantage in the regulatory review or approval process.

We may face competition from biological products approved through an abbreviated regulatory pathway.

Our Naglazyme, Aldurazyme and VIMIZIM products are regulated by the FDA as biologics under the Federal Food, Drug and Cosmetic Act (FDC Act), and the Public Health Service Act (the PHS Act). Biologics require the submission of a Biologics License Application (BLA), and approval by the FDA prior to being marketed in the U.S. Historically, a biologic product approved under a BLA was not subject to the generic drug review and approval provisions of the FDC Act. However, the Patient Protection and Affordable Care Act of 2010, as amended by the Health Care and Education Reconciliation Act of 2010 (collectively, the PPACA), created a regulatory pathway under the PHS Act for the abbreviated approval for biological products that are demonstrated to be “biosimilar” or “interchangeable” with an FDA-approved biological product. In order to meet the standard of interchangeability, a sponsor must demonstrate that the biosimilar product can be expected to produce the same clinical result as the reference product, and for a product that is administered more than once, that the risk of switching between the reference product and biosimilar product is not greater than the risk of maintaining the patient on the reference product. Such biosimilars would reference biological products approved in the U.S. The law establishes a period of 12 years of data exclusivity for reference products, which protects the data in the original BLA by prohibiting sponsors of biosimilars from gaining FDA approval based in part on reference to data in the original BLA. Our products approved under BLAs, as well as products in development that may be approved under BLAs, could be reference products for such abbreviated BLAs.

Table of Contents

To obtain regulatory approval to market our products, preclinical studies and costly and lengthy preclinical and clinical trials are required and the results of the studies and trials are highly uncertain.

As part of the regulatory approval process we must conduct, at our own expense, preclinical studies in the laboratory and clinical trials on humans for each product candidate. We expect the number of preclinical studies and clinical trials that the regulatory authorities will require will vary depending on the product candidate, the disease or condition the drug is being developed to address and regulations applicable to the particular drug. Generally, the number and size of clinical trials required for approval increase based on the expected patient population that may be treated with a drug. We may need to perform multiple preclinical studies using various doses and formulations before we can begin clinical trials, which could result in delays in our ability to market any of our product candidates. Furthermore, even if we obtain favorable results in preclinical studies, the results in humans may be significantly different. After we have conducted preclinical studies, we must demonstrate that our drug products are safe and efficacious for use in the targeted human patients in order to receive regulatory approval for commercial sale.

Adverse or inconclusive clinical results would stop us from filing for regulatory approval of our product candidates. Additional factors that can cause delay or termination of our clinical trials include:

- slow or insufficient patient enrollment;
- slow recruitment of, and completion of necessary institutional approvals at, clinical sites;
- longer treatment time required to demonstrate efficacy;
- lack of sufficient supplies of the product candidate;
- adverse medical events or side effects in treated patients;
- lack of effectiveness of the product candidate being tested; and
- regulatory requests for additional clinical trials or pre-clinical studies.

Typically, if a drug product is intended to treat a chronic disease, as is the case with some of our product candidates, safety and efficacy data must be gathered over an extended period of time, which can range from nine months to three years or more. We also rely on independent third-party CROs to perform most of our clinical studies and many important aspects of the services performed for us by the CROs are out of our direct control. If we fail to adequately manage our CROs, or if there is any dispute or disruption in our relationship with our CROs, our clinical trials may be delayed. Moreover, in our regulatory submissions, we rely on the quality and validity of the clinical work performed by third-party CROs. If any of our CROs' processes, methodologies or results were determined to be invalid or inadequate, our own clinical data and results and related regulatory approvals could adversely be impacted.

If we continue to incur operating losses for a period longer than anticipated, we may be unable to continue our operations at planned levels and be forced to reduce our operations.

Since we began operations in March 1997, we have been engaged in very substantial research and development and operated at a net loss until 2008. Although we were profitable in 2008 and 2010, we operated at a net loss in 2009, 2011 and 2012. Based upon our current plan for investments in research and development for existing and new programs, we expect to operate at a net loss for at least the next 12 months. Our future profitability depends on our marketing and selling of VIMIZIM, Naglazyme, Kuvan and Firdapse, the successful continued commercialization of Aldurazyme by Genzyme, the receipt of regulatory approval of our product candidates, our ability to successfully manufacture and market any approved drugs, either by ourselves or jointly with others, our spending on our development programs and the impact of any possible future business development transactions. The extent of our future losses and the timing of profitability are highly uncertain. If we fail to become profitable or are unable to sustain profitability on a continuing basis, then we may be unable to continue our operations at planned levels and be forced to reduce our operations.

If we fail to comply with manufacturing regulations, our financial results and financial condition will be adversely affected.

Before we can begin commercial manufacture of our products, we or our contract manufacturers, must obtain regulatory approval of our manufacturing facilities, processes and quality systems. In addition, our

Table of Contents

pharmaceutical manufacturing facilities are continuously subject to inspection by the FDA and international regulatory authorities, before and after product approval. Our manufacturing facilities in the U.S. have been approved by the FDA, the European Commission (EC), and health agencies in other countries for the manufacture of Aldurazyme and Naglazyme. In addition, our third-party manufacturers' facilities involved with the manufacture of VIMIZIM, Naglazyme, Kuvan, Aldurazyme and Firdapse have also been inspected and approved by various regulatory authorities. The manufacturing facility located in Shanbally, Cork, Ireland that we purchased in 2011 has not yet been approved by the FDA or the EMA. We intend to make a substantial investment in the build-out of the Shanbally facility in order to manufacture VIMIZIM and other products. If the facility is not ultimately approved by the FDA or the EMA, we will not be able to manufacture VIMIZIM or other products at this facility and we may not be able to meet the anticipated commercial demand for VIMIZIM which would have an adverse effect on our financial results.

Due to the complexity of the processes used to manufacture our products and product candidates, we may be unable to continue to pass or initially pass federal or international regulatory inspections in a cost effective manner. For the same reason, any potential third-party manufacturer of VIMIZIM, Naglazyme, Kuvan, Aldurazyme and Firdapse or our product candidates may be unable to comply with GMP regulations in a cost effective manner and may be unable to initially or continue to pass a federal or international regulatory inspection.

If we, or third-party manufacturers with whom we contract, are unable to comply with manufacturing regulations, we may be subject to delay of approval of our product candidates, warning or untitled letters, fines, unanticipated compliance expenses, recall or seizure of our products, total or partial suspension of production and/or enforcement actions, including injunctions, and criminal or civil prosecution. These possible sanctions would adversely affect our financial results and financial condition.

***If we fail to obtain the capital necessary to fund our operations, our financial results and financial condition will be adversely affected and we will have to delay or terminate some or all of our product development programs.**

As of March 31, 2014, we had cash, cash equivalents and short and long-term investments totaling \$1,138.9 million and long-term debt obligations of \$812.0 million (undiscounted). In October 2013, we completed an offering of senior subordinated convertible notes and received net proceeds of approximately \$696.4 million, after deducting commissions, estimated offering expenses payable by us and the purchase of the related capped calls. We will need cash to not only repay the principal amount of the Notes but also the ongoing interest due on the Notes during their term. In March 2014, we sold 1.5 million shares of our common stock at a price of \$78.45 per share in an underwritten public offering pursuant to an effective registration statement previously filed with the SEC. We received net proceeds of \$117.5 million for this public offering; however we may require additional financing to fund our future operations, including the commercialization of our approved drugs and drug product candidates currently under development, preclinical studies and clinical trials, and potential licenses and acquisitions. We may be unable to raise additional financing, if needed, due to a variety of factors, including our financial condition, the status of our product programs, and the general condition of the financial markets. If we fail to raise any necessary additional financing we may have to delay or terminate some or all of our product development programs and our financial condition and operating results will be adversely affected.

We expect to continue to spend substantial amounts of capital for our operations for the foreseeable future. The amount of capital we will need depends on many factors, including:

- our ability to successfully market and sell VIMIZIM, Naglazyme, Kuvan and Firdapse;
- Genzyme's ability to continue to successfully commercialize Aldurazyme;
- the progress and success of our preclinical studies and clinical trials (including studies and the manufacture of materials);
- the timing, number, size and scope of our preclinical studies and clinical trials;
- the time and cost necessary to obtain regulatory approvals and the costs of post-marketing studies which may be required by regulatory authorities;
- the time and cost necessary to develop commercial manufacturing processes, including quality systems, and to build or acquire manufacturing capabilities;
- the progress of research programs carried out by us;

Table of Contents

- our possible achievement of milestones identified in our purchase agreements with the former stockholders of LEAD Therapeutics, Inc., ZyStor, Huxley Pharmaceuticals, Inc., and Zacharon Pharmaceuticals Inc. that trigger related milestone payments;
- any changes made to, or new developments in, our existing collaborative, licensing and other commercial relationships or any new collaborative, licensing and other commercial relationships that we may establish; and
- whether our convertible debt is converted to common stock in the future.

Moreover, our fixed expenses such as rent, license payments, interest expense and other contractual commitments are substantial and may increase in the future. These fixed expenses may increase because we may enter into:

- additional licenses and collaborative agreements;
- additional contracts for product manufacturing; and
- additional financing facilities.

We may need to raise additional funds from equity or debt securities, loans or collaborative agreements if we are unable to satisfy our liquidity requirements. The sale of additional securities may result in additional dilution to our stockholders. Furthermore, additional financing may not be available in amounts or on terms satisfactory to us or at all. This could result in the delay, reduction or termination of our research, which could harm our business.

If we are unable to successfully develop and maintain manufacturing processes for our drug products to produce sufficient quantities at acceptable costs, we may be unable to meet demand for our products and lose potential revenue, have reduced margins or be forced to terminate a program.

Due to the complexity of manufacturing our products, we may not be able to manufacture drug products successfully with a commercially viable process or at a scale large enough to support their respective commercial markets or at acceptable margins.

The development of commercially viable manufacturing processes typically is very difficult to achieve and is often very expensive and may require extended periods of time. Changes in manufacturing processes (including manufacturing cell lines), equipment or facilities may require us to complete clinical trials to receive regulatory approval of any manufacturing improvements. Also, we may be required to demonstrate product comparability between a biological product made after a manufacturing change and the product made before implementation of the change through additional types of analytical and functional testing or may have to complete additional clinical studies. If we contract for manufacturing services with an unproven process, our contractor is subject to the same uncertainties, high standards and regulatory controls, and may therefore experience difficulty if further process development is necessary.

Even a developed manufacturing process can encounter difficulties. Problems may arise during manufacturing for a variety of reasons, including human error, mechanical breakdowns, problems with raw materials and cell banks, malfunctions of internal information technology systems, and other events that cannot always be prevented or anticipated. Many of the processes include biological systems, which add significant complexity, as compared to chemical synthesis. We expect that, from time to time, consistent with biotechnology industry expectations, certain production lots will fail to produce product that meets our quality control release acceptance criteria. To date, our historical failure rates for all of our product programs, including Naglazyme, Aldurazyme and VIMIZIM, have been within our expectations, which are based on industry norms. If the failure rate increased substantially, we could experience increased costs, lost revenue, damage to customer relations, time and expense investigating the cause and, depending upon the cause, similar losses with respect to other lots or products. If problems are not discovered before the product is released to the market, recall and product liability costs may also be incurred.

In order to produce product within our time and cost parameters, we must continue to produce product within our expected success rate and yield expectations. Because of the complexity of our manufacturing processes, it may be difficult or impossible for us to determine the cause of any particular lot failure and we must effectively take corrective action in response to any failure in a timely manner.

Table of Contents

Although we have entered into contractual relationships with third-party manufacturers to produce the active ingredient in Kuvan and Firdapse, if those manufacturers are unwilling or unable to fulfill their contractual obligations, we may be unable to meet demand for these products or sell these products at all and we may lose potential revenue. We have contracts for the production of final product for Kuvan and Firdapse. We also rely on third-parties for portions of the manufacture of Naglazyme and Aldurazyme. If those manufacturers are unwilling or unable to fulfill their contractual obligations or satisfy demand outside of or in excess of the contractual obligations, we may be unable to meet demand for these products or sell these products at all and we may lose potential revenue. Further, the availability of suitable contract manufacturing capacity at scheduled or optimum times is not certain.

In addition, our manufacturing processes subject us to a variety of federal, state and local laws and regulations governing the use, generation, manufacture, storage, handling and disposal of hazardous materials and wastes resulting from their use. We may incur significant costs in complying with these laws and regulations.

If we are unable to effectively address manufacturing issues, we may be unable to meet demand for our products and lose potential revenue, have reduced margins, or be forced to terminate a program.

Our manufacturing facility for Naglazyme, Aldurazyme and VIMIZIM is located near known earthquake fault zones, and the occurrence of an earthquake or other catastrophic disaster could cause damage to our facility and equipment, or that of our third-party manufacturers or single-source suppliers, which could materially impair our ability to manufacture Naglazyme, Aldurazyme and VIMIZIM or our third-party manufacturer's ability to manufacture Kuvan or Firdapse.

Our Galli Drive facility located in Novato, California is currently our only manufacturing facility for Naglazyme, Aldurazyme and VIMIZIM. It is located in the San Francisco Bay Area near known earthquake fault zones and is vulnerable to significant damage from earthquakes. We, the third-party manufacturers with whom we contract and our single-source suppliers of raw materials, which include many of our critical raw materials, are also vulnerable to damage from other types of disasters, including fires, floods, power loss and similar events. If any disaster were to occur, or any terrorist or criminal activity caused significant damage to our facilities or the facilities of our third-party manufacturers and suppliers, our ability to manufacture Naglazyme, Aldurazyme and VIMIZIM, or to have Kuvan or Firdapse manufactured, could be seriously, or potentially completely impaired, and our commercialization efforts and revenue could be seriously impaired. The insurance that we carry, the inventory that we maintain and our risk mitigation plans may not be adequate to cover our losses resulting from disasters or other business interruptions.

Supply interruptions may disrupt our inventory levels and the availability of our products and cause delays in obtaining regulatory approval for our product candidates, or harm our business by reducing our revenues.

Numerous factors could cause interruptions in the supply of our finished products, including:

- timing, scheduling and prioritization of production by our contract manufacturers or a breach of our agreements by our contract manufacturers;
- labor interruptions;
- changes in our sources for manufacturing;
- the timing and delivery of shipments;
- our failure to locate and obtain replacement manufacturers as needed on a timely basis; and conditions affecting the cost and availability of raw materials.

Any interruption in the supply of finished products could hinder our ability to distribute finished products to meet commercial demand.

With respect to our product candidates, production of product is necessary to perform clinical trials and successful registration batches are necessary to file for approval to commercially market and sell product candidates. Delays in obtaining clinical material or registration batches could delay regulatory approval for our product candidates.

Table of Contents

Because the target patient populations for our products are small, we must achieve significant market share and maintain high per-patient prices for our products to achieve profitability.

All of our products target diseases with small patient populations. As a result, our per-patient prices must be relatively high in order to recover our development and manufacturing costs and achieve profitability. For Naglazyme and VIMIZIM, if approved outside of the U.S., we must market worldwide to achieve significant market penetration of the product. In addition, because the number of potential patients in the disease populations are small, it is not only important to find patients who begin therapy to achieve significant market penetration of the product, but we also need to be able to maintain these patients on therapy for an extended period of time. Due to the expected costs of treatment for our products for genetic diseases, we may be unable to maintain or obtain sufficient market share at a price high enough to justify our product development efforts and manufacturing expenses.

If we fail to obtain an adequate level of coverage and reimbursement for our drug products by third-party payers, the sales of our drugs would be adversely affected or there may be no commercially viable markets for our products.

The course of treatment for patients using our products is expensive. We expect patients to need treatment for extended periods, and for some products throughout the lifetimes of the patients. We expect that most families of patients will not be capable of paying for this treatment themselves. There will be no commercially viable market for our products without coverage and reimbursement from third-party payers. Additionally, even if there is a commercially viable market, if the level of reimbursement is below our expectations, our revenue and gross margins will be adversely affected.

Third-party payers, such as government or private health care insurers, carefully review and increasingly challenge the prices charged for drugs. Reimbursement rates from private companies vary depending on the third-party payer, the insurance plan and other factors. Reimbursement systems in international markets vary significantly by country and by region, and reimbursement approvals must be obtained on a country-by-country basis.

Reimbursement in the EU and many other territories must be negotiated on a country-by-country basis and in many countries the product cannot be commercially launched until reimbursement is approved. The timing to complete the negotiation process in each country is highly uncertain, and in some countries we expect that it may exceed 12 months. Even after a price is negotiated, countries frequently request or require adjustments to the price and other concessions over time.

For our future products, we will not know what the reimbursement rates will be until we are ready to market the product and we actually negotiate the rates. If we are unable to obtain sufficiently high reimbursement rates for our products, they may not be commercially viable or our future revenues and gross margins may be adversely affected.

A significant portion of our international sales are made based on special access programs, and changes to these programs could adversely affect our product sales and revenue in these countries.

We make a significant portion of our international sales of Naglazyme through special access or “named patient” programs, which do not require full product approval. We expect to also utilize these programs for VIMIZIM. The specifics of the programs vary from country to country. Generally, special approval must be obtained for each patient. The approval normally requires an application or a lawsuit accompanied by evidence of medical need. Generally, the approvals for each patient must be renewed from time to time.

These programs are not well defined in some countries and are subject to changes in requirements and funding levels. Any change to these programs could adversely affect our ability to sell our products in those countries and delay sales. If the programs are not funded by the respective government, there could be insufficient funds to pay for all patients. Further, governments have in the past undertaken and may in the future undertake, unofficial measures to limit purchases of our products, including initially denying coverage for purchasers, delaying orders and denying or taking excessively long to approve customs clearance. Any such actions could materially delay or reduce our revenues from such countries.

Table of Contents

Without the special access programs, we would need to seek full product approval to commercially market and sell our products. This can be an expensive and time-consuming process and may subject our products to additional price controls. Because the number of patients is so small in some countries, it may not be economically feasible to seek and maintain a full product approval, and therefore the sales in such country would be permanently reduced or eliminated. For all of these reasons, if the special access programs that we are currently using are eliminated or restricted, our revenues could be adversely affected.

If we fail to compete successfully with respect to product sales, we may be unable to generate sufficient sales to recover our expenses related to the development of a product program or to justify continued marketing of a product and our revenue could be adversely affected.

Our competitors may develop, manufacture and market products that are more effective or less expensive than ours. They may also obtain regulatory approvals for their products faster than we can obtain them (including those products with orphan drug designation) or commercialize their products before we do. If we do not compete successfully, our revenue would be adversely affected, and we may be unable to generate sufficient sales to recover our expenses related to the development of a product program or to justify continued marketing of a product.

Government price controls or other changes in pricing regulation could restrict the amount that we are able to charge for our current and future products, which would adversely affect our revenue and results of operations.

We expect that coverage and reimbursement may be increasingly restricted both in the U.S. and internationally. The escalating cost of health care has led to increased pressure on the health care industry to reduce costs. Governmental and private third-party payers have proposed health care reforms and cost reductions. A number of federal and state proposals to control the cost of health care, including the cost of drug treatments, have been made in the U.S. In some international markets, the government controls the pricing, which can affect the profitability of drugs. Current government regulations and possible future legislation regarding health care may affect coverage and reimbursement for medical treatment by third-party payers, which may render our products not commercially viable or may adversely affect our future revenues and gross margins.

International operations are also generally subject to extensive price and market regulations, and there are many proposals for additional cost-containment measures, including proposals that would directly or indirectly impose additional price controls or mandatory price cuts or reduce the value of our intellectual property portfolio. As part of these cost containment measures, some countries have imposed or threatened to impose revenue caps limiting the annual volume of sales of Naglazyme. To the extent that these caps are significantly below actual demand, our future revenues and gross margins may be adversely affected.

We cannot predict the extent to which our business may be affected by these or other potential future legislative or regulatory developments. However, future price controls or other changes in pricing regulation could restrict the amount that we are able to charge for our current and future products, which would adversely affect our revenue and results of operations.

Government health care reform could increase our costs, and would adversely affect our revenue and results of operations.

Our industry is highly regulated and changes in law may adversely impact our business, operations or financial results. The PPACA is a sweeping measure intended to expand healthcare coverage within the U.S., primarily through the imposition of health insurance mandates on employers and individuals and expansion of the Medicaid program.

Several provisions of the new law, which have varying effective dates, may affect us and will likely increase certain of our costs. For example, the Medicaid rebate rate was increased and the volume of rebated drugs has been expanded to include beneficiaries in Medicaid managed care organizations. Among other things, the PPACA also expanded the 340B drug discount program (excluding orphan drugs), including the creation of new penalties for non-compliance; included a 50% discount on brand name drugs for Medicare Part D participants in the

Table of Contents

coverage gap, or “donut hole,” and imposed a new fee on certain manufacturers and importers of branded prescription drugs (excluding orphan drugs under certain conditions). The law also revised the definition of “average manufacturer price” for reporting purposes, which could increase the amount of the Medicaid drug rebates paid to states.

In addition, other legislative changes have been proposed and adopted since the PPACA was enacted. These changes include aggregate reductions in Medicare payments to providers of up to 2% per fiscal year, which went into effect on April 1, 2013. In January 2013, President Obama signed into law the American Taxpayer Relief Act of 2012, which, among other things, further reduced Medicare payments to several types of providers and increased the statute of limitations period for the government to recover overpayments to providers from three to five years. These new laws may result in additional reductions in Medicare and other healthcare funding, which could have a material adverse effect on our customers and, accordingly, our financial operations.

We anticipate that PPACA, as well as other healthcare reform measures that may be adopted in the future, may result in more rigorous coverage criteria and an additional downward pressure on the reimbursement our customers may receive for our products. Any reduction in reimbursement from Medicare and other government programs may result in a similar reduction in payments from private payors. The implementation of cost containment measures or other healthcare reforms may prevent us from being able to generate revenue, attain profitability or commercialize our products.

We face credit risks from customers outside of the U.S. that may adversely affect our results of operations.

Our product sales to government-owned or supported customers in various countries outside of the U.S. are subject to significant payment delays due to government funding and reimbursement practices. This has resulted and may continue to result in an increase in days sales outstanding due to the average length of time that we have accounts receivable outstanding. If significant changes were to occur in the reimbursement practices of these governments or if government funding becomes unavailable, we may not be able to collect on amounts due to us from these customers and our results of operations would be adversely affected.

If we are found in violation of federal or state “fraud and abuse” laws, we may be required to pay a penalty or be suspended from participation in federal or state health care programs, which may adversely affect our business, financial condition and results of operation.

We are subject to various federal and state health care “fraud and abuse” laws, including anti-kickback laws, false claims laws and laws related to ensuring compliance. The federal health care program anti-kickback statute makes it illegal for any person, including a pharmaceutical company, to knowingly and willfully offer, solicit, pay or receive any remuneration, directly or indirectly, in exchange for or to induce the referral of business, including the purchase, order or prescription of a particular drug, for which payment may be made under federal health care programs, such as Medicare and Medicaid. Under federal government regulations, certain arrangements, or safe harbors, are deemed not to violate the federal anti-kickback statute. However, the exemptions and safe harbors are drawn narrowly, and practices that involve remuneration not intended to induce prescribing, purchases or recommendations may be subject to scrutiny if they do not qualify for an exemption or safe harbor. Our practices may not in all cases meet all of the criteria for safe harbor protection from anti-kickback liability, although we seek to comply with these safe harbors. Violations of the anti-kickback statute are punishable by imprisonment, criminal fines, civil monetary penalties and exclusion from participation in federal healthcare programs.

Federal and state false claims laws prohibit any person from knowingly presenting, or causing to be presented, a false claim for payment to the federal government, or knowingly making, or causing to be made, a false statement to have a false claim paid. In addition, certain marketing practices, including off-label promotion, may also violate false claims laws. Under the Health Insurance Portability and Accountability Act of 1996, we also are prohibited from knowingly and willfully executing a scheme to defraud any health care benefit program, including private payers, or knowingly and willfully falsifying, concealing or covering up a material fact or making any materially false, fictitious or fraudulent statement in connection with the delivery of or payment for health care benefits, items or services. Sanctions under these federal and state laws may include civil monetary penalties, exclusion of a manufacturer’s products from reimbursement under government programs, criminal fines and imprisonment.

Table of Contents

Many states have adopted laws similar to the federal anti-kickback statute, some of which apply to referral of patients for health care services reimbursed by any source, not just governmental payers.

Substantial new provisions affecting compliance also have been adopted, which may require us to modify our business practices with health care practitioners. The PPACA, among other things, requires drug manufacturers to collect and report information on payments or transfers of value to physicians and teaching hospitals, as well as investment interests held by physicians and their immediate family members during the preceding calendar year. Failure to submit required information may result in civil monetary penalties. The CMS has issued a final rule that requires manufacturers to begin collecting required information on August 1, 2013 with the first reports due March 31, 2014 (and by the 90th day of each calendar year thereafter) and publication of the reported data in a searchable form on a public website beginning September 30, 2014.

In addition, there has been a recent trend of increased state regulation of payments made to physicians. Certain states mandate implementation of compliance programs, compliance with the Office of Inspector General Compliance Program Guidance for Pharmaceutical Manufacturers and the PhRMA Code on Interactions with Healthcare Professionals, and/or the tracking and reporting of gifts, compensation and other remuneration to physicians. The shifting compliance environment and the need to implement systems to comply with multiple jurisdictions with different compliance and/or reporting requirements increases the possibility that a pharmaceutical manufacturer may violate one or more of the requirements.

While we believe we have structured our business arrangements to comply with these laws, because of the breadth of these laws, the narrowness of available statutory and regulatory exceptions and the increased focus by law enforcement agencies in enforcing such laws, it is possible that some of our business activities could be subject to challenge under one or more of such laws. In addition, recent health care reform legislation has strengthened, these laws. For example, the PPACA, among other things, amends the intent requirement of the federal anti-kickback and criminal healthcare fraud statutes. A person or entity no longer needs to have actual knowledge of this statute or specific intent to violate it. Moreover, the PPACA provides that the government may assert that a claim including items or services resulting from a violation of the federal anti-kickback statute constitutes a false or fraudulent claim for purposes of the False Claims Act. If we are found in violation of one of these laws, we may be subject to criminal, civil or administrative sanctions, including debarment, suspension or exclusion from participation in federal or state health care programs any of which could adversely affect our business, financial condition and results of operation.

We conduct a significant amount of our sales and operations outside of the U.S., which subjects us to additional business risks that could adversely affect our revenue and results of operations.

A significant portion of the sales of Aldurazyme and Naglazyme and all of the sales of Firdapse are generated from countries other than the U.S. Additionally, we have operations in several European countries, Brazil, other Latin American countries, Turkey and other Asian countries. We expect that we will continue to expand our international operations in the future. International operations inherently subject us to a number of risks and uncertainties, including:

- changes in international regulatory and compliance requirements that could restrict our ability to manufacture, market and sell our products;
- political and economic instability;
- diminished protection of intellectual property in some countries outside of the U.S.;
- trade protection measures and import or export licensing requirements;
- difficulty in staffing and managing international operations;
- differing labor regulations and business practices;
- potentially negative consequences from changes in or interpretations of tax laws;
- changes in international medical reimbursement policies and programs;
- financial risks such as longer payment cycles, difficulty collecting accounts receivable and exposure to fluctuations in foreign currency exchange rates; and
- regulatory and compliance risks that relate to maintaining accurate information and control over sales and distributors' and service providers' activities that may fall within the purview of the Foreign Corrupt Practices Act.

Table of Contents

Any of these factors may, individually or as a group, have a material adverse effect on our business and results of operations.

As we continue to expand our existing international operations, we may encounter new risks. For example, as we focus on building our international sales and distribution networks in new geographic regions, we must continue to develop relationships with qualified local distributors and trading companies. If we are not successful in developing and maintaining these relationships, we may not be able to grow sales in these geographic regions. These or other similar risks could adversely affect our revenue and profitability.

If we are unable to protect our proprietary technology, we may not be able to compete as effectively.

Where appropriate, we seek patent protection for certain aspects of our technology. Patent protection may not be available for some of the products we are developing. If we must spend significant time and money protecting or enforcing our patents, designing around patents held by others or licensing, potentially for large fees, patents or other proprietary rights held by others, our business and financial prospects may be harmed.

The patent positions of biopharmaceutical products are complex and uncertain. The scope and extent of patent protection for some of our products and product candidates are particularly uncertain because key information on some of our product candidates has existed in the public domain for many years. The composition and genetic sequences of animal and/or human versions of Naglazyme, Aldurazyme and many of our product candidates have been published and are believed to be in the public domain. The chemical structure of BH4 (the active ingredient in Kuvan) and 3,4-DAP (the active ingredient in Firdapse) have also been published. Publication of this information may prevent us from obtaining or enforcing patents relating to our products and product candidates, including without limitation composition-of-matter patents, which are generally believed to offer the strongest patent protection.

We own or have licensed patents and patent applications related to VIMIZIM, Naglazyme, Kuvan, Aldurazyme and Firdapse. However, these patents and patent applications do not ensure the protection of our intellectual property for a number of reasons, including without limitation the following:

- With respect to pending patent applications, unless and until actually issued, the protective value of these applications is impossible to determine. We do not know whether our patent applications will result in issued patents.
- Competitors may interfere with our patent process in a variety of ways. Competitors may claim that they invented the claimed invention prior to us or that they filed their application for a patent on a claimed invention before we did. Competitors may also claim that we are infringing on their patents and therefore we cannot practice our technology. Competitors may also contest our patents by showing the patent examiner or a court that the invention was not original, was not novel or was obvious, for example. In litigation, a competitor could claim that our issued patents are not valid or are unenforceable for a number of reasons. If a court agrees, we would not be able to enforce that patent. We have no meaningful experience with competitors interfering with or challenging the validity or enforceability of our patents or patent applications.
- Enforcing patents is expensive and may absorb significant time of our management. Management would spend less time and resources on developing products, which could increase our operating expenses and delay product programs. We may not have the financial ability to sustain a patent infringement action, or it may not be financially reasonable to do so.
- Receipt of a patent may not provide much, if any, practical protection. For example, if we receive a patent with a narrow scope, then it will be easier for competitors to design products that do not infringe on our patent.
- The recently enacted America Invents Act, which reformed certain patent laws in the U.S., may create additional uncertainty. Among the significant changes are switching from a “first-to-invent” system to a “first-to-file” system, and the implementation of new procedures that permit competitors to challenge our patents in the U.S. Patent and Trademark Office after grant.

Table of Contents

It is also unclear whether our trade secrets are adequately protected. Our employees, consultants or contractors may unintentionally or willfully disclose trade secrets to competitors. Enforcing a claim that someone else illegally obtained and is using our trade secrets, as with patent litigation, is expensive and time consuming, requires significant resources and the outcome is unpredictable. In addition, courts outside of the U.S. are sometimes less willing to protect trade secrets. Furthermore, our competitors may independently develop equivalent knowledge, methods and know-how, in which case we would not be able to enforce our trade secret rights against such competitors.

If we are unable to protect our intellectual property, third parties could develop competing products, which could adversely affect our revenue and financial results generally.

Competitors and other third parties may have developed intellectual property that could limit our ability to market and commercialize our products and product candidates, if approved.

Similar to us, competitors continually seek intellectual property protection for their technology. Several of our development programs, such as BMN 673, BMN 701, BMN 111 and BMN 270, focus on therapeutic areas that have been the subject of extensive research and development by third parties for many years. Due to the amount of intellectual property in our field of technology, we cannot be certain that we do not infringe intellectual property rights of competitors or that we will not infringe intellectual property rights of competitors granted or created in the future. For example, if a patent holder believes our product infringes its patent, the patent holder may sue us even if we have received patent protection for our technology. If someone else claims we infringe its intellectual property, we would face a number of issues, including the following:

- Defending a lawsuit takes significant executive resources and can be very expensive.
- If a court decides that our product infringes a competitor's intellectual property, we may have to pay substantial damages.
- With respect to patents, in addition to requiring us to pay substantial damages, a court may prohibit us from making, selling, offering to sell, importing or using our product unless the patent holder licenses the patent to us. The patent holder is not required to grant us a license. If a license is available, it may not be available on commercially reasonable terms. For example, we may have to pay substantial royalties or grant cross licenses to our patents and patent applications.
- We may need to redesign our product so it does not infringe the intellectual property rights of others.
- Redesigning our product so it does not infringe the intellectual property rights of competitors may not be possible or could require substantial funds and time.

We may also support and collaborate in research conducted by government organizations, hospitals, universities or other educational institutions. These research partners may be unwilling to grant us any exclusive rights to technology or products derived from these collaborations.

If we do not obtain required licenses or rights, we could encounter delays in our product development efforts while we attempt to design around other patents or may be prohibited from making, using, importing, offering to sell or selling products requiring these licenses or rights. There is also a risk that disputes may arise as to the rights to technology or products developed in collaboration with other parties. If we are not able to resolve such disputes and obtain the licenses or rights we need, we may not be able to develop or market our products.

If our Manufacturing, Marketing and Sales Agreement with Genzyme were terminated, we could be prevented from continuing to commercialize Aldurazyme or our ability to successfully commercialize Aldurazyme would be delayed or diminished.

Either party may terminate the Manufacturing, Marketing and Sales Agreement (the MMS Agreement), between Genzyme and us related to Aldurazyme for specified reasons, including if the other party is in material breach of the MMS Agreement, has experienced a change of control, as such term is defined in the MMS Agreement, or has declared bankruptcy and also is in breach of the MMS Agreement. Although we are not currently in breach of the MMS Agreement, there is a risk that either party could breach the MMS Agreement in the future. Either party may also terminate the MMS Agreement upon one year prior written notice for any reason.

Table of Contents

If the MMS Agreement is terminated for breach, the breaching party will transfer its interest in BioMarin/Genzyme LLC (the LLC), to the non-breaching party, and the non-breaching party will pay a specified buyout amount for the breaching party's interest in Aldurazyme and in the LLC. If we are the breaching party, we would lose our rights to Aldurazyme and the related intellectual property and regulatory approvals. If the MMS Agreement is terminated without cause, the non-terminating party would have the option, exercisable for one year, to buy out the terminating party's interest in Aldurazyme and in the LLC at a specified buyout amount.

If such option is not exercised, all rights to Aldurazyme will be sold and the LLC will be dissolved. In the event of termination of the buyout option without exercise by the non-terminating party as described above, all right and title to Aldurazyme is to be sold to the highest bidder, with the proceeds to be split between Genzyme and us in accordance with our percentage interest in the LLC.

If the MMS Agreement is terminated by either party because the other party declared bankruptcy, the terminating party would be obligated to buy out the other party and would obtain all rights to Aldurazyme exclusively. If the MMS Agreement is terminated by a party because the other party experienced a change of control, the terminating party shall notify the other party, the offeree, of its intent to buy out the offeree's interest in Aldurazyme and the LLC for a stated amount set by the terminating party at its discretion. The offeree must then either accept this offer or agree to buy the terminating party's interest in Aldurazyme and the LLC on those same terms. The party who buys out the other party would then have exclusive worldwide rights to Aldurazyme. The Amended and Restated Collaboration Agreement between us and Genzyme will automatically terminate upon the effective date of the termination of the MMS Agreement and may not be terminated independently from the MMS Agreement.

If we were obligated or given the option to buy out Genzyme's interest in Aldurazyme and the LLC, and thereby gain exclusive rights to Aldurazyme, we may not have sufficient funds to do so and we may not be able to obtain the financing to do so. If we fail to buy out Genzyme's interest, we may be held in breach of the agreement and may lose any claim to the rights to Aldurazyme and the related intellectual property and regulatory approvals. We would then effectively be prohibited from developing and commercializing Aldurazyme. If this happened, not only would our product revenues decrease, but our share price would also decline.

Based on our strategic alliance with Merck Serono, unless Merck Serono "opts in" to the PEG PAL program, we will not realize any cost sharing for the development expenses, development milestones, or royalties for ex-U.S. sales.

In May 2005, we entered into an agreement with Merck Serono for the further development and commercialization of Kuvan (and any other product containing 6R-BH4) and PEG PAL for PKU. Pursuant to that agreement, we received development milestones on Kuvan and receive royalties on sales by Merck Serono. Additionally, we may be entitled to development milestones and royalties related to PEG PAL. However, Merck Serono has "opted out" of the PEG PAL development program. Unless and until it elects to opt in, it is not obligated to pay any of the milestones related to the program or to reimburse us for any of the development costs. Additionally, even though Merck Serono has opted out of the PEG PAL development program, we do not have any right to commercialize PEG PAL outside of the U.S. and Japan or to grant anyone else such rights.

Merck Serono may elect to opt in at any time. If Merck Serono opts in to the PEG PAL development program before the unblinding of the first Phase 3 trial for PEG PAL, it must pay 75% of the Phase 3 costs incurred prior to the opt-in and the \$7,000,000 Phase 3 initiation milestone. If it opts in after unblinding of the first Phase 3 trial for PEG PAL, it must pay 100% of the Phase 3 costs incurred prior to the opt-in and the \$7,000,000 Phase 3 initiation milestone. Additionally, in all cases after it opts in to the PEG PAL development program, Merck Serono would be obligated to pay one half of future development costs under the agreement and any further milestones due under the agreement. If Merck Serono does not opt in, it will not have the right to use any of the clinical or other independently developed data.

We cannot determine when or if Merck Serono will opt in to the PEG PAL development program. If Merck Serono does not opt in, we will not receive any milestones under the agreement nor will there be any sales outside of the U.S. or Japan generating revenue from royalties or otherwise.

Table of Contents

If we fail to compete successfully with respect to acquisitions, joint ventures or other collaboration opportunities, we may be limited in our ability to develop new products and to continue to expand our product pipeline.

Our competitors compete with us to attract organizations for acquisitions, joint ventures, licensing arrangements or other collaborations. To date, several of our product programs have been acquired through acquisitions, such as BMN 701 and BMN 673 and several of our product programs have been developed through licensing or collaborative arrangements, such as Naglazyme, Aldurazyme, Kuvan and Firdapse. These collaborations include licensing proprietary technology from, and other relationships with, academic research institutions. Our future success will depend, in part, on our ability to identify additional opportunities and to successfully enter into partnering or acquisition agreements for those opportunities. If our competitors successfully enter into partnering arrangements or license agreements with academic research institutions, we will then be precluded from pursuing those specific opportunities. Since each of these opportunities is unique, we may not be able to find a substitute. Several pharmaceutical and biotechnology companies have already established themselves in the field of genetic diseases. These companies have already begun many drug development programs, some of which may target diseases that we are also targeting, and have already entered into partnering and licensing arrangements with academic research institutions, reducing the pool of available opportunities.

Universities and public and private research institutions also compete with us. While these organizations primarily have educational or basic research objectives, they may develop proprietary technology and acquire patents that we may need for the development of our product candidates. We will attempt to license this proprietary technology, if available. These licenses may not be available to us on acceptable terms, if at all. If we are unable to compete successfully with respect to acquisitions, joint venture and other collaboration opportunities, we may be limited in our ability to develop new products and to continue to expand our product pipeline.

If generic manufacturers use litigation and regulatory means to obtain approval for generic versions of Kuvan, our revenue and results of operations would be adversely affected.

The Hatch Waxman Act permits the FDA to approve abbreviated new drug applications (ANDAs) for generic versions of branded drugs. We refer to this process as the “ANDA process”. The ANDA process permits competitor companies to obtain marketing approval for a drug with the same active ingredient for the same uses but does not generally require the conduct and submission of clinical efficacy studies for that product. In place of such clinical studies, an ANDA applicant usually needs only to submit data demonstrating that its product is bioequivalent to the branded product based on pharmacokinetic studies. Pursuant to the Hatch-Waxman Act, companies were able to file an ANDA application for the active ingredient in Kuvan at any time after December 2011. At present, we have not received information that any other party has filed or has conducted the bioequivalency study necessary to file an ANDA for Kuvan.

The Hatch Waxman Act requires an applicant for a drug that relies, at least in part, on our data regarding the safety and efficacy of Kuvan, to notify us of their application and potential infringement of our patents listed in the FDA’s Approved Drug Products with Therapeutic Equivalence Evaluations (Orange Book). Upon receipt of a notice alleging that our patents listed in the Orange Book are invalid or not infringed by the proposed competitor product (a paragraph iv notice), we would have 45 days to bring a patent infringement suit in federal district court against the company seeking approval for its product. The discovery, trial and appeals process in such suits can take several years. If we commence such a suit alleging infringement of one or more of our Orange Book listed patents within 45 days from receipt of the paragraph iv notice, the Hatch-Waxman Act provides a 30-month stay on the FDA’s approval of the competitor’s application. If the litigation is resolved in favor of the applicant or the challenged patent expires during the 30-month stay period, the stay is lifted and the FDA’s review of the application may be completed. Such litigation is often time-consuming, costly and may result in competition if such patent(s) are not upheld or if the competitor does not infringe such patent(s). However, generic versions of Kuvan would be prohibited until the expiration of orphan drug exclusivity in December 2014 or June 2015 if we receive pediatric exclusivity.

The filing of an ANDA application in respect to Kuvan could have an adverse impact on our stock price and litigation to enforce our patents is likely to cost a substantial amount and require significant management attention. If the patents covering Kuvan were not upheld in litigation or if the generic competitor is found to not infringe these patents, the resulting generic competition following the expiration of orphan exclusivity would have a material adverse effect on our revenue and results of operations.

Table of Contents

If we do not achieve our projected development goals in the timeframes we announce and expect, the commercialization of our products may be delayed and the credibility of our management may be adversely affected and, as a result, our stock price may decline.

For planning purposes, we estimate the timing of the accomplishment of various scientific, clinical, regulatory and other product development goals, which we sometimes refer to as milestones. These milestones may include the commencement or completion of scientific studies and clinical trials and the submission of regulatory filings. From time to time, we publicly announce the expected timing of some of these milestones. All of these milestones are based on a variety of assumptions. The actual timing of these milestones can vary dramatically compared to our estimates, in many cases for reasons beyond our control. If we do not meet these milestones as publicly announced, the commercialization of our products may be delayed and the credibility of our management may be adversely affected and, as a result, our stock price may decline.

We depend upon our key personnel and our ability to attract and retain employees.

Our future growth and success will depend in large part on our continued ability to attract, retain, manage and motivate our employees. The loss of the services of any member of our senior management or the inability to hire or retain experienced management personnel could adversely affect our ability to execute our business plan and harm our operating results.

Because of the specialized scientific and managerial nature of our business, we rely heavily on our ability to attract and retain qualified scientific, technical and managerial personnel. In particular, the loss of one or more of our senior executive officers could be detrimental to us if we do not have an adequate succession plan or if we cannot recruit suitable replacements in a timely manner. While our senior executive officers are parties to employment agreements with us, these agreements do not guarantee that they will remain employed with us in the future. In addition, in many cases, these agreements do not restrict our senior executive officers' ability to compete with us after their employment is terminated. The competition for qualified personnel in the pharmaceutical field is intense, and there is a limited pool of qualified potential employees to recruit. Due to this intense competition, we may be unable to continue to attract and retain qualified personnel necessary for the development of our business or to recruit suitable replacement personnel. If we are unsuccessful in our recruitment and retention efforts, our business may be harmed.

Our success depends on our ability to manage our growth.

Product candidates that we are currently developing or may acquire in the future may be intended for patient populations that are significantly larger than any of MPS I, MPS VI, PKU or LEMS. In order to continue development and marketing of these products, if approved, we will need to significantly expand our operations. To manage expansion effectively, we need to continue to develop and improve our research and development capabilities, manufacturing and quality capacities, sales and marketing capabilities, financial and administrative systems and standard processes for global operations. Our staff, financial resources, systems, procedures or controls may be inadequate to support our operations and may increase our exposure to regulatory and corruption risks and our management may be unable to manage successfully future market opportunities or our relationships with customers and other third-parties.

Changes in methods of treatment of disease could reduce demand for our products and adversely affect revenues.

Even if our drug products are approved, if doctors elect a course of treatment which does not include our drug products, this decision would reduce demand for our drug products and adversely affect revenues. For example, if gene therapy becomes widely used as a treatment of genetic diseases, the use of enzyme replacement therapy, such as Naglazyme and Aldurazyme in MPS diseases, could be greatly reduced. Changes in treatment method can be caused by the introduction of other companies' products or the development of new technologies or surgical procedures which may not directly compete with ours, but which have the effect of changing how doctors decide to treat a disease.

Table of Contents

If product liability lawsuits are successfully brought against us, we may incur substantial liabilities.

We are exposed to the potential product liability risks inherent in the testing, manufacturing and marketing of human pharmaceuticals. We currently maintain insurance against product liability lawsuits for the commercial sale of our products and for the clinical trials of our product candidates. Pharmaceutical companies must balance the cost of insurance with the level of coverage based on estimates of potential liability. Historically, the potential liability associated with product liability lawsuits for pharmaceutical products has been unpredictable. Although we believe that our current insurance is a reasonable estimate of our potential liability and represents a commercially reasonable balancing of the level of coverage as compared to the cost of the insurance, we may be subject to claims in connection with our clinical trials and commercial use of VIMIZIM, Naglazyme, Kuvan, Aldurazyme and Firdapse, or our clinical trials for PEG PAL, BMN 701, BMN 673, BMN 111, BMN 190 or BMN 270 for which our insurance coverage may not be adequate and we may be unable to avoid significant liability if any product liability lawsuit is brought against us. If we are the subject of a successful product liability claim that exceeds the limits of any insurance coverage we obtain, we may incur substantial charges that would adversely affect our earnings and require the commitment of capital resources that might otherwise be available for the development and commercialization of our product programs.

We rely significantly on information technology and any failure, inadequacy, interruption or security lapse of that technology, including any cybersecurity incidents, could harm our ability to operate our business effectively.

We rely significantly on our information technology and manufacturing infrastructure to effectively manage and maintain our inventory and internal reports, to manufacture and ship products to customers and to timely invoice them. Any failure, inadequacy or interruption of that infrastructure or security lapse of that technology, including cybersecurity incidents could harm our ability to operate our business effectively. Our ability to manage and maintain our inventory and internal reports, to manufacture and ship our products to customers and timely invoice them depends significantly on our enterprise resource planning, production management and other information systems. Cybersecurity attacks in particular are evolving and include, but are not limited to, malicious software, attempts to gain unauthorized access to data and other electronic security breaches that could lead to disruptions in systems, misappropriation of our confidential or otherwise protected information and corruption of data. Cybersecurity incidents resulting in the failure of our enterprise resource planning system, production management or other systems to operate effectively or to integrate with other systems, or a breach in security or other unauthorized access of these systems, may affect our ability to manage and maintain our inventory and internal reports, and result in delays in product fulfillment and reduced efficiency of our operations. A breach in security, unauthorized access resulting in misappropriation, theft, or sabotage with respect to our proprietary and confidential information, including research or clinical data, could require significant capital investments to remediate and could adversely affect our business, financial condition and results of operations.

***Our business is affected by macroeconomic conditions.**

Various macroeconomic factors could adversely affect our business and the results of our operations and financial condition, including changes in inflation, interest rates and foreign currency exchange rates and overall economic conditions and uncertainties, including those resulting from the current and future conditions in the global financial markets. For instance, if inflation or other factors were to significantly increase our business costs, it may not be feasible to pass through price increases on to our customers due to the process by which health care providers are reimbursed for our products by the government. Interest rates, the liquidity of the credit markets and the volatility of the capital markets could also affect the value of our investments and our ability to liquidate our investments in order to fund our operations. We purchase or enter into a variety of financial instruments and transactions, including investments in commercial paper, the extension of credit to corporations, institutions and governments and hedging contracts. If any of the issuers or counter parties to these instruments were to default on their obligations, it could materially reduce the value of the transaction and adversely affect our cash flows.

Table of Contents

For the three months ended March 31, 2014 approximately 5% of our net product revenues were from the countries of Italy, Spain, Portugal, Greece and Russia. Approximately 20% of our total accounts receivable as of March 31, 2014 related to such countries and we have included an allowance for doubtful accounts for certain accounts receivable from Greece. If the financial conditions of these countries continues to decline, a substantial portion of the receivables may be uncollectable, which would mean we would have to provide for additional allowances for doubtful accounts or cease selling products in these countries, either of which could adversely affect our results of operations. Additionally, if one or more of these countries were unable to purchase our products, our revenue would be adversely affected. We also sell our products in other countries that face economic crises and local currency devaluation. Although we have historically collected receivables from customers in those countries, sustained weakness or further deterioration of the local economies and currencies may cause our customers in those countries to be unable to pay for our products with the same negative effect on our operations.

Interest rates and the ability to access credit markets could also adversely affect the ability of our customers/distributors to purchase, pay for and effectively distribute our products. Similarly, these macroeconomic factors could affect the ability of our contract manufacturers, sole-source or single-source suppliers to remain in business or otherwise manufacture or supply product. Failure by any of them to remain a going concern could affect our ability to manufacture products.

Risks Related to Ownership of Our Securities

Our stock price may be volatile, and an investment in our stock could suffer a decline in value.

Our valuation and stock price since the beginning of trading after our initial public offering have had no meaningful relationship to current or historical earnings, asset values, book value or many other criteria based on conventional measures of stock value. The market price of our common stock will fluctuate due to factors including:

- product sales and profitability of VIMIZIM, Naglazyme, Kuvan, Aldurazyme and Firdapse;
- manufacture, supply or distribution of VIMIZIM, Naglazyme, Kuvan, Aldurazyme and Firdapse;
- progress of our product candidates through the regulatory process and our ability to successfully commercialize any such products that receive regulatory approval;
- results of clinical trials, announcements of technological innovations or new products by us or our competitors;
- government regulatory action affecting our product candidates or our competitors' drug products in both the U.S. and non-U.S. countries;
- developments or disputes concerning patent or proprietary rights;
- general market conditions and fluctuations for the emerging growth and pharmaceutical market sectors;
- economic conditions in the U.S. or abroad;
- broad market fluctuations in the U.S., the EU or in other parts of the world;
- actual or anticipated fluctuations in our operating results; and
- changes in company assessments or financial estimates by securities analysts.

In the past, following periods of large price declines in the public market price of a company's securities, securities class action litigation has often been initiated against that company. Litigation of this type could result in substantial costs and diversion of management's attention and resources, which would hurt our business. Any adverse determination in litigation could also subject us to significant liabilities. In addition, our stock price can be materially adversely affected by factors beyond our control, such as disruptions in global financial markets or negative trends in the biotechnology sector of the economy, even if our business is operating well.

Recent and future regulatory actions and other events may adversely affect the trading price and liquidity of our senior subordinated convertible notes.

We expect that many investors in, and potential purchasers of, the Notes will employ, or seek to employ, a convertible arbitrage strategy with respect to the Notes. Investors would typically implement such a strategy by selling short the common stock underlying the Notes and dynamically adjusting their short position while continuing to hold the Notes. Investors may also implement this type of strategy by entering into swaps on our common stock in lieu of or in addition to short selling the common stock.

Table of Contents

The SEC and other regulatory and self-regulatory authorities have implemented various rules and taken certain actions, and may in the future adopt additional rules and take other actions, that may impact those engaging in short selling activity involving equity securities (including our common stock). Such rules and actions include Rule 201 of SEC Regulation SHO, the adoption by the Financial Industry Regulatory Authority, Inc. of a “Limit Up-Limit Down” program, the imposition of market-wide circuit breakers that halt trading of securities for certain periods following specific market declines, and the implementation of certain regulatory reforms required by the Dodd-Frank Wall Street Reform and Consumer Protection Act of 2010. Any governmental or regulatory action that restricts the ability of investors in, or potential purchasers of, the Notes to effect short sales of our common stock or enter into swaps on our common stock could adversely affect the trading price and the liquidity of the Notes.

In addition, if investors and potential purchasers seeking to employ a convertible arbitrage strategy are unable to borrow or enter into swaps on our common stock, in each case on commercially reasonable terms, the trading price and liquidity of the Notes may be adversely affected.

Anti-takeover provisions in our charter documents and under Delaware law may make an acquisition of us, which may be beneficial to our stockholders, more difficult.

We are incorporated in Delaware. Certain anti-takeover provisions of Delaware law and our charter documents as currently in effect may make a change in control of our company more difficult, even if a change in control would be beneficial to the stockholders. Our anti-takeover provisions include provisions in our certificate of incorporation providing that stockholders’ meetings may only be called by our Board of Directors and provisions in our bylaws providing that the stockholders may not take action by written consent and requiring that stockholders that desire to nominate any person for election to our Board of Directors or to make any proposal with respect to business to be conducted at a meeting of our stockholders be submitted in appropriate form to our Secretary within a specified period of time in advance of any such meeting. Additionally, our Board of Directors has the authority to issue shares of preferred stock and to determine the terms of those shares of stock without any further action by our stockholders. The rights of holders of our common stock are subject to the rights of the holders of any preferred stock that may be issued. The issuance of preferred stock could make it more difficult for a third-party to acquire a majority of our outstanding voting stock. Delaware law also prohibits corporations from engaging in a business combination with any holders of 15% or more of their capital stock until the holder has held the stock for three years unless, among other possibilities, our Board of Directors approves the transaction. Our Board of Directors may use these provisions to prevent changes in the management and control of our company. Also, under applicable Delaware law, our Board of Directors may adopt additional anti-takeover measures in the future.

Item 2. Unregistered Sales of Equity Securities and Use of Proceeds.

None.

Item 3. Defaults Upon Senior Securities.

None.

Item 4. Mine Safety Disclosures

None.

Item 5. Other Information.

None.

Table of Contents

Item 6. Exhibits.

10.1*#	Amended and Restated BioMarin Pharmaceutical Inc., Non-Qualified Defined Compensation Plan, as adopted on December 1, 2005, as amended and restated on July 1, 2009 and as further amended on December 19, 2013.
31.1*	Certification of Chief Executive Officer pursuant to Rules 13a-14(a)/15d-14(a) of the Securities Exchange Act of 1934, as amended.
31.2*	Certification of Chief Financial Officer pursuant to Rules 13a-14(a)/15d-14(a) of the Securities Exchange Act of 1934, as amended.
32.1*	Certification of Chief Executive Officer and Chief Financial Officer pursuant to 18 U.S.C. Section 1350, as adopted pursuant to Section 906 of the Sarbanes-Oxley Act of 2002. This Certification accompanies this report and shall not, except to the extent required by the Sarbanes-Oxley Act of 2002, be deemed filed for purposes of §18 of the Securities Exchange Act of 1934, as amended.
101.INS*	XBRL Instance Document
101.SCH*	XBRL Taxonomy Extension Schema Document
101.CAL*	XBRL Taxonomy Extension Calculation Document
101.DEF*	XBRL Taxonomy Extension Definition Linkbase
101.LAB*	XBRL Taxonomy Extension Labels Linkbase Document
101.PRE*	XBRL Taxonomy Extension Presentation Link Document

* Filed herewith.

Management contract or compensatory Plan or arrangement

Attached as Exhibit 101 to this report are documents formatted in XBRL (Extensible Business Reporting Language): (i) Condensed Consolidated Balance Sheets as of March 31, 2014 and December 31, 2013, (ii) Condensed Consolidated Statements of Comprehensive Loss for the three months ended March 31, 2014 and 2013, (iii) Condensed Consolidated Statements of Cash Flows for the three months ended March 31, 2014 and 2013, and (iv) Notes to Condensed Consolidated Financial Statements.

Table of Contents

EXHIBIT INDEX

10.1*#	Amended and Restated BioMarin Pharmaceutical Inc., Non-Qualified Defined Compensation Plan, as adopted on December 1, 2005, as amended and restated on July 1, 2009 and as further amended on December 19, 2013.
31.1*	Certification of Chief Executive Officer pursuant to Rules 13a-14(a)/15d-14(a) of the Securities Exchange Act of 1934, as amended.
31.2*	Certification of Chief Financial Officer pursuant to Rules 13a-14(a)/15d-14(a) of the Securities Exchange Act of 1934, as amended.
32.1*	Certification of Chief Executive Officer and Chief Financial Officer pursuant to 18 U.S.C. Section 1350, as adopted pursuant to Section 906 of the Sarbanes-Oxley Act of 2002. This Certification accompanies this report and shall not, except to the extent required by the Sarbanes-Oxley Act of 2002, be deemed filed for purposes of §18 of the Securities Exchange Act of 1934, as amended.
101.INS*	XBRL Instance Document
101.SCH*	XBRL Taxonomy Extension Schema Document
101.CAL*	XBRL Taxonomy Extension Calculation Document
101.DEF*	XBRL Taxonomy Extension Definition Linkbase
101.LAB*	XBRL Taxonomy Extension Labels Linkbase Document
101.PRE*	XBRL Taxonomy Extension Presentation Link Document

* Filed herewith.

Management contract or compensatory Plan or arrangement

Attached as Exhibit 101 to this report are documents formatted in XBRL (Extensible Business Reporting Language): (i) Condensed Consolidated Balance Sheets as of March 31, 2014 and December 31, 2013, (ii) Condensed Consolidated Statements of Comprehensive Loss for the three months ended March 31, 2014 and 2013, (iii) Condensed Consolidated Statements of Cash Flows for the three months ended March 31, 2014 and 2013, and (iv) Notes to Condensed Consolidated Financial Statements.

**AMENDED AND RESTATED
BIOMARIN PHARMACEUTICAL INC.
NONQUALIFIED DEFERRED COMPENSATION PLAN**

**AMENDED AND RESTATED
BIOMARIN PHARMACEUTICAL INC.
NONQUALIFIED DEFERRED COMPENSATION PLAN**

RECITALS

This BioMarin Pharmaceutical Inc. Nonqualified Deferred Compensation Plan (“Plan”) was adopted by BioMarin Pharmaceutical Inc. effective as of December 1, 2005, was thereafter amended and restated, effective January 1, 2009, and is now being amended and restated effective December 19, 2013 in order to establish special distribution rules for Qualified Employees, as defined below. The Plan has been adopted primarily for the purpose of providing deferred compensation for a select group of management or highly compensated employees of the Company and its related entities. This Plan shall also be available to non-employee members of the Board of Directors. Accordingly, it is intended that this Plan be exempt from the requirements of Parts II, III and IV of Title I of the Employee Retirement Income Security Act of 1974, as amended (“ERISA”), pursuant to Sections 201(2), 301(a)(3) and 401(a)(1) of ERISA. This Plan is intended to be an unfunded, nonqualified deferred compensation plan. Plan participants shall have the status of unsecured creditors of BioMarin Pharmaceutical with respect to the payment of Plan benefits.

1. **DEFINITIONS** .

- 1.1 **“Account”** means the book entry account(s) established under the Plan for each Participant’s Compensation Deferrals, Discretionary Employer Contributions and any contribution credits and deemed income, gains and losses credited thereto or debited therefrom. Account balances shall be reduced by any distributions made to the Participant or the Participant’s Beneficiary(ies) therefrom and any charges that may be imposed on such Account(s) pursuant to the terms of the Plan. Separate Subaccounts may be established to which shall be credited a Participant’s Compensation Deferrals for each separate Plan Year, the Discretionary Employer Contributions, if any, and the gains and losses with respect thereto. Where Subaccounts have been established, Account shall refer to all of the Participants’ Subaccounts, collectively, as the context may require.
- 1.2 **“Affiliate”** means, with respect to any entity, all other entities with which the subject entity would be aggregated and treated as a single employer under Code Section 414(b) (controlled group of corporations) and Code Section 414(c) (a group of trades or businesses, whether or not incorporated, under common control), as applicable.
- 1.3 **“Beneficiary”** means any person or persons so designated in accordance with the provisions of **Section 7.1** .
- 1.4 **“Board”** means the Board of Directors of the Company. If one or more committees have been appointed by the Board to determine eligibility under the Plan, Discretionary Employer Contributions to be made to the Plan, or to exercise any other Company discretion with respect to such Plan, “Board” also means such committee(s).
- 1.5 **“Change Of Control”** shall mean either (i) a merger, consolidation, share exchange, business combination, issuance of securities, direct or indirect acquisition of securities, tender offer, exchange offer or other similar transaction as a result of which the persons that beneficially owned, directly or indirectly, the shares of the Company’s voting stock immediately prior to such transaction cease to beneficially own, directly or indirectly, shares of voting stock representing more than fifty percent (50%) of the total voting power of all outstanding classes of voting stock of the Company or the continuing or surviving corporation if Company is not the continuing or surviving corporation in such transaction, or (ii) a sale of all or substantially all of the assets of Company.
- 1.6 **“Code”** means the Internal Revenue Code of 1986 and the regulations thereunder, as amended from time to time.

- 1.7 **“Committee”** means the Administrative Committee composed of such individuals as may be appointed by the Board which shall function as the administrator of the Plan.
- 1.8 **“Company”** means BioMarin Pharmaceutical Inc., a Delaware company, and any successor organization thereto.
- 1.9 **“Compensation”** means, in the case of a Participant who is an employee of the Company such Participant’s regular cash salary and cash bonuses, and Restricted Stock Compensation, and in the case of a Participant who is a Director of the Company, such Participant’s annual Board retainer and Board meeting fees (including Board Committee meeting fees). Compensation shall also include any “Performance Based Compensation” as that term is defined under Section 409A of the Code and any regulations thereunder.
- 1.10 **“Compensation Deferrals”** means the percentage or dollar amount of an Eligible Employee’s Compensation which the Eligible Employee elects to defer pursuant to **Section 3.1** .
- 1.11 **“Designation Date”** means the date or dates as of which a designation of deemed investment directions by an individual pursuant to **Section 4.3**, or any change in a prior designation of deemed investment directions by an individual pursuant to **Section 4.3** , shall become effective. The Designation Dates in any Plan Year shall be determined by the Committee.
- 1.12 **“Director”** means a non-employee member of the Board of Directors.
- 1.13 **“Disability”** will be determined to exist if the Participant is, by reason of any medically determinable physical or mental impairment which can be expected to result in death or can be expected to last for a continuous period of not less than twelve (12) months, eligible to receive income replacement benefits for a period of not less than three (3) months under any disability benefit plan for covered Employees of the Employer, or, if the Participant does not participate in such plan, would have been eligible to receive such benefits had the Participant participated in such plan.
- 1.14 **“Discretionary Employer Contributions”** means the amount, if any, of contributions awarded to a Participant pursuant to **Section 3.2** .
- 1.15 **“Effective Date”** means the effective date of the Plan, which shall be December 1, 2005, or if later, the date the Plan is approved by the Board.
- 1.16 **“Election”** means the form on which a Participant (i) elects to make Compensation Deferrals pursuant to **Article 3** or (ii) elects a fixed payment date pursuant to **Article 5** , or (iii) elects the method by which his or her Account will be distributed pursuant to Article 6. The Election shall be in such form, including specifically by electronic means, as may be prescribed by the Committee.
- 1.17 **“Eligible Employee”** means, for any Plan Year (or applicable portion thereof), an employee of the Employer who is a member of the select group of management or highly compensated employees as more particularly described in **Article 2** and who has been designated by the Committee, in its sole discretion, as eligible to participate in the Plan.
- 1.18 **“Employer”** shall be defined as follows:
- (a) Except as otherwise provided in part (b) of this Section, the term “Employer” shall mean the Company and/or any of its subsidiaries or affiliates (now in existence or hereafter formed or acquired) that have been selected by the Committee to participate in the Plan and have adopted the Plan for the benefit of its Eligible Employees.

- (b) For the purpose of determining whether a Participant has experienced a Separation from Service, the term “Employer” shall mean the entity for which the Participant performs services and with respect to which the legally binding right to compensation deferred or contributed under this Plan arises, and all of its Affiliates.
- 1.19 **“Entry Date”** means the first day of any Plan Year and, as to any Eligible Employee, the date which is thirty (30) days from the date on which such Eligible Employee is first notified by the Committee of his or her eligibility to participate in the Plan. Notwithstanding the foregoing, for any individual first designated as an Eligible Employee on or before the Effective Date, his or her Entry Date shall be the Effective Date
- 1.20 **“Open Enrollment Period”** means such period as the Committee may specify which ends prior to the first day of each Plan Year, or, with respect to an Eligible Employee or Director who first becomes eligible to participate in the Plan during a Plan Year, ends within thirty (30) days of becoming an Eligible Employee or Director, provided that the newly Eligible Employee or Director was not previously eligible in another individual account deferred compensation plan that would be aggregated with this Plan pursuant to Treasury Regulation Section 1.409A-1(c) at any time during the 24-month period ending on the date he or she became eligible to participate in the Plan. If an Eligible Employee or Director first becomes eligible after the first day of the Plan Year, the Compensation Deferral shall be limited to Compensation paid for services performed after the date of the Election. For Compensation that is earned based upon a specific performance period (e.g., annual bonus or retainer), a Compensation Deferral shall be limited to Compensation paid for services performed after the Election if the Compensation Deferral applies to no more than an amount equal to the total amount of the Compensation for the performance period multiplied by the ratio of the number days remaining in the performance period after the Election over the total number of days in the performance period. Notwithstanding the foregoing, the Open Enrollment Period for deferrals of Performance Based Compensation may end no later than six (6) months prior to the end of the performance period for which services are to be rendered.
- 1.21 **“Participant”** means an Eligible Employee or Director who has elected to participate in the Plan by executing and submitting an Election to the Committee. A Participant shall also mean an Eligible Employee for whom Discretionary Employer Contributions are made, regardless of whether such Eligible Employee has executed and submitted an Election.
- 1.22 **“Performance-Based Compensation”** means any incentive bonus or other compensation amount to the extent that it is (a) variable and contingent on the satisfaction of pre-established organizational or individual performance criteria, (b) not readily ascertainable at the time the deferral election is made, and (c) based on services performed over a period of at least twelve (12) months. For this purpose, performance criteria are “pre-established” if they are established in writing no later than ninety (90) days after the related service period begins.
- 1.23 **“Plan”** means this BioMarin Pharmaceutical Inc. Nonqualified Deferred Compensation Plan, as amended from time to time.
- 1.24 **“Plan Year”** means the twelve (12) month period beginning on each January 1 and ending on the following December 31.
- 1.25 **“Qualified Employee”** means a Participant employed by the Company who has at least 5 years of service at the time of Separation from Service.
- 1.26 **“Restricted Stock Compensation,”** means any restricted stock, restricted stock unit, phantom stock or similar award granted by the Employer to a Participant under any Employer-sponsored equity compensation plan.

- 1.27 **“Retirement”** means, in the case of a Participant employed by the Company, Separation from Service on or after age 55 with 10 years or more of service, and in the case of a Director, Separation of Service as a Director.
- 1.28 **“Separation from Service”** (or **“Separates from Service”**) shall mean the termination of services provided by a Participant to his or her Employer, whether voluntarily or involuntarily, as determined by the Committee in accordance with Treasury Regulation Section 1.409A-1(h). In determining whether a Participant has experienced a Separation from Service, the following provisions shall apply:
- (a) For a Participant who provides services to the Employer as an Employee, except as otherwise provided in **Section 1.28(b)**, a Separation from Service shall occur when such Participant experiences a termination of employment with such Employer. A Participant shall be considered to have experienced a termination of employment when the facts and circumstances indicate that either (i) the Participant is not reasonably expected to perform further services for the Employer after a certain date, or (ii) that the level of bona fide services the Participant will perform for the Employer after such date (whether as an Employee or as an independent contractor) will permanently decrease to no more than 49% of the average level of bona fide services performed by such Participant (whether as an Employee or an independent contractor) over the immediately preceding 36-month period (or full period of services to the Employer if the Participant has been providing services to the Employer for less than 36 months).
 - (b) If a Participant is on military leave, sick leave, or other bona fide leave of absence, the employment relationship between the Participant and the Employer shall be treated as continuing intact, provided that the period of such leave does not exceed six months, or longer, so long as the Participant retains a right to reemployment with the Employer under an applicable statute or by contract. If the period of leave exceeds six (6) months and the Participant does not retain a right to reemployment under an applicable statute or by contract, the Participant will incur a Separation from Service as of the first day immediately following the end of such 6-month period. However, where a Participant’s leave of absence is due to his or her inability to perform the duties of his or her position or any similar position as the result of any medically determinable physical or mental impairment that can be expected to result in death or last for a continuous period of not less than six (6) months, a 29-month period of absence will be substituted for such 6-month period. In applying the provisions of this paragraph, a leave of absence shall be considered a bona fide leave of absence only if there is a reasonable expectation that the Participant will return to perform services for the Employer.
 - (c) Notwithstanding the foregoing, if a Participant who provides services to the Employer as both an Employee and a member of the Board, then to the extent permitted by Treasury Regulation Section 1.409A-1(h)(5), the services provided by such Participant as a Board member shall not be taken into account in determining whether the Participant experiences a Separation from Service as an Employee, and the services provided by such Participant as an Employee shall not be taken into account in determining whether the Participant has experienced a Separation from Service as a Board member.
 - (d) Notwithstanding the foregoing, if, in connection with the Employer’s sale of substantial assets to an unrelated buyer (within the meaning of Treasury Regulation Section 1.409A-1(h)(4)), a Participant would otherwise experience a Separation of Service, then, in accordance with Treasury Regulation Section 1.409A-1(h)(4), the asset purchase agreement may specify whether or not such Participant has experienced a Separation from Service, provided that all Participants affected by the asset sale are treated consistently under the Plan.

- 1.29 “**Trust**” means any trust, including a grantor trust within the meaning of subpart E, part I, subchapter J, chapter I, subtitle A of the Code, created by the Trust agreement, to hold Compensation Deferrals and Discretionary Employer Contributions.
- 1.30 “**Trustee**” means the trustee of the Trust described in **Article 11** .
- 1.31 “**Valuation Date**” means any business day on which the New York Stock Exchange is open, or such other date that the Committee, in its sole discretion, designates as a Valuation Date.

2. ELIGIBILITY AND PARTICIPATION.

- 2.1 Eligibility . Eligibility for participation in the Plan shall be limited to Directors and a select group of management or highly compensated employees of the Employer, who are designated by the Committee, in its sole discretion, as eligible to participate in the Plan. Eligible Individuals shall be notified as to their eligibility to participate in the Plan. Participation in the Plan is voluntary, other than for Discretionary Employer Contributions. A person shall cease to be an Eligible Person for future plan years at such a time as he or she is neither a member of a select group of management nor highly compensated employees of the Company nor a Director of the Company.
- 2.2 Commencement Of Participation . An Eligible Employee may begin participation in the Plan upon any Entry Date, subject to the execution and submission of an Election pursuant to **Article 3** . In addition, participation of an Eligible Employee who has not otherwise commenced participation in the Plan, shall commence when a Discretionary Employer Contribution is made to the Account of such Eligible Employee pursuant to the provisions of **Section 3.2**.
- 2.3 Cessation Of Participation . Active participation in the Plan shall end when a Participant’s employment terminates for any reason or at such time as a Participant is notified by the Committee pursuant to **Section 2.4** that he or she is no longer eligible to participate in the Plan. Upon Separation from Service of employment or termination of eligibility, a Participant shall remain an inactive Participant in the Plan until the vested Account of the Participant under this Plan has been paid in full.
- 2.4 Change Of Employment Category . During any period in which a Participant remains with the company, but ceases to be an Eligible Person, he or she shall not be eligible to make Compensation Deferrals, or to receive Discretionary Employer Contributions hereunder for future plan years.

3. CONTRIBUTIONS AND CREDITS .

3.1 Participant Contributions And Credits .

- (a) Time and Manner of Election . In accordance with rules established by the Committee, a Participant may elect to defer Compensation that would otherwise be paid to the Participant that is attributable to services first performed after the end of the applicable Open Enrollment Period. Notwithstanding the foregoing, the Open Enrollment Period for deferrals of Performance Based Compensation may end no later than six (6) months prior to the end of the performance period for which services are to be rendered. A Participant who is a Director may elect to defer up to 100% of his or her Compensation earned during the Plan Year. Amounts so deferred shall be considered a Participant’s “Compensation Deferrals” and shall be deducted by the Company from the Compensation of the deferring Participant and shall be credited to the Compensation Deferral Account of the deferring Participant. The Participant may, on an applicable election form provided by the Committee, elect for his or her Compensation deferrals to be paid in a lump sum within a specific calendar year, subject to such limitations as the Committee may set forth in the applicable election form. In addition, if permitted by the

Committee, a Participant may elect to receive a distribution in annual installments. The annual installments shall commence within a specific calendar year as set forth within the election form. Furthermore, in accordance with rules established by the Committee, a Participant may elect to defer Restricted Stock Compensation that would otherwise be payable to the Participant to the Participant's Account. The value of the Restricted Stock Compensation to be credited to the Participant's Account shall be equal to fair market value of the number of shares vesting to the Participant as of the date of the shares vesting.

- (b) Timing of Election. The Election must be filed with the Committee during the Open Enrollment Period for the Plan Year to which such Election applies. Elections to defer Restricted Stock Compensation must be made no later than twelve (12) months prior to the vesting of the shares and no later than thirty (30) days after the granting of the award.
- (c) Irrevocable Election. The Participant's Election with respect to his or her Compensation Deferrals is irrevocable. Unless increased, decreased or terminated during any subsequent Open Enrollment Period, an Election shall remain in effect until so changed by the Participant during such subsequent Open Enrollment Period.
- (d) Limitation on Compensation Deferrals. A Participant's Compensation Deferral Elections shall be subject to the following:
 - (i) A Participant electing to defer compensation for a given Plan Year must elect to defer a minimum of the greater of 1% of his or her Compensation or \$10,000 each Plan Year.
 - (ii) A Participant may elect to defer up to a maximum of one hundred percent (100%) of his or her Compensation and 100% of Performance Based Compensation.
 - (iii) For each Plan Year in which a Compensation Deferral is being withheld from a Participant, the Participant's Employer (s) shall withhold from that portion of the Participant's Compensation that is not being deferred, in a manner determined by the Employer(s), the Participant's share of FICA or other employment taxes on such Compensation Deferral. If necessary, the Committee may reduce the Compensation Deferral in order to comply with this Section.
- (e) No Withdrawal. Except as provided in **Section 5.2**, amounts credited to a Participant's Account may not be withdrawn by a Participant and shall be paid only in accordance with the provisions of this Plan and applicable Participant Election.
- (f) Vesting. A Participant shall at all times be 100% vested in the amounts credited to his or her Compensation Deferral Account.

3.2 Discretionary Employer Contributions And Credits.

- (a) Discretionary Employer Contributions. Apart from Compensation Deferral Contributions, the Board shall retain the right to make discretionary contributions for any Participant under this Plan at the times and in the amount(s) designated by the Employer, in its sole discretion. Amounts so credited will be considered a Participant's "Discretionary Employer Contributions."
- (b) Company Contribution Account. There shall be established and maintained a separate Company Contribution Account in the name of each Participant to which shall be credited the amount of any Company Contributions during a plan Year and any earnings thereon, and from which shall be debited the amount of any losses thereon and the amount of any distributions made to the Participant therefrom.

- (c) Vesting. Amounts credited to the Company Contribution Account shall become 100% vested after the Participant has had three full Years of Service with the Company following the Plan Year in which the Company Contribution was made, or at such other time as the Company may designate at the time the Contribution is made. Notwithstanding the preceding, if (i) the Participant dies or incurs a Disability prior to vesting or (ii) there is a Change of Control, all amounts credited to his or her Company Contribution Account shall become 100% vested. Any Participant that experiences a severance prior to full vesting shall irrevocably forfeit the portion not vested at the time of severance, and the amount so forfeited shall be returned to the Company. Any Participant that experiences a Retirement prior to full vesting shall, unless the Committee determines otherwise in its sole discretion (which shall include the discretion to fully vest amounts credited to the Company Contribution Account on Retirement), irrevocably forfeit the portion not vested at the time of Retirement, and the amount so forfeited shall be returned to the Company.
- (d) Forfeitures for Misconduct. If a Participant Separates from Service with the Employer as a result of the Participant's gross misconduct, as determined by the Committee, or if the Participant engages in unlawful business competition with the Employer, the Participant shall forfeit all amounts allocated to his or her Discretionary Employer Contribution Account(s) under this **Section 3.2** (regardless of the vesting of such amounts). Such forfeitures shall be retained by the Employer.

4. ALLOCATION OF FUNDS.

- 4.1 Allocation Of Deemed Earnings Or Losses On Accounts. Subject to such limitations as may from time to time be required by law, imposed by the Committee or the Trustee or contained elsewhere in the Plan, and subject to such operating rules and procedures as may be imposed from time to time by the Committee, prior to the date on which a direction will become effective, the Participant shall have the right to direct the Committee as to how amounts in his or her Compensation Deferral Account shall be deemed to be invested. However, with regard to Restricted Stock Compensation the amount deferred will be deemed to be invested in Company stock and will remain in Company stock until such amount is distributed in shares to the Participant pursuant to **Section 6**. The Committee may, but is not required to, invest assets held by the Company on behalf of the Participant pursuant to the deemed investment directions the Committee has properly received from the Participant, and may utilize the Trust for the same in its discretion.

As of each Valuation Date, the Participant's Account will be credited or debited to reflect the Participant's deemed investments, The Participant's Account will be credited or debited with the increase or decrease in the realizable net asset value of the designated deemed investments, as follows. As of each Valuation Date, an amount equal to the net increase or decrease in realizable net asset value (as determined by the Committee) of each deemed investment option within the Account since the preceding Valuation Date shall be allocated among all Participants' Accounts deemed to be invested in that investment option in accordance with the ratio which the portion of the Account of each Participant which is deemed to be invested within that investment option, determined as provided herein, bears to the aggregate of all amounts deemed to be invested within that investment option.

- 4.2 Accounting For Distributions. As of the date of any distribution hereunder, the distribution made hereunder to the Participant or his or her Beneficiary or Beneficiaries shall be charged to such Participant's Account. Such amounts shall be charged on a pro rata basis against the investments of the Plan in which the Participant's Account is deemed to be invested.

4.3 Deemed Investment Directions Of Participants. Subject to such limitations as may from time to time be required by law, imposed by the Employer or the Trustee or contained elsewhere in the Plan, and subject to such operating rules and procedures as may be imposed from time to time by the Employer, prior to and effective for each Designation Date, each Participant may communicate to the Employer a direction (in accordance with (a), below) as to how his or her Account should be deemed to be invested among such categories of deemed investments as may be made available by the Employer hereunder. Such direction shall designate the percentage (in any whole percent multiples) of each portion of the Participant's Account which is requested to be deemed to be invested in such categories of deemed investments, and shall be subject to the following rules:

- (a) Any initial or subsequent deemed investment direction shall be in writing, on a form supplied by and filed with the Committee, and/or, as required or permitted by the Committee, shall be by written designation and/or electronic transmission designation. A designation shall be effective as of the Designation Date next following the date the direction is received and accepted by the Committee on which it would be reasonably practicable for the Committee to effect the designation. The Participant may, if permitted by the Committee, make a deemed investment direction for his or her existing Account balance as of the Designation Date and a separate deemed investment direction for contribution credits occurring after the Designation Date.
- (b) All amounts credited to the Participant's Account shall be deemed to be invested in accordance with the then effective deemed investment direction, and as of the Designation Date with respect to any new deemed investment direction, all or a portion of the Participant's Account at that date shall be reallocated among the designated deemed investment funds according to the percentages specified in the new deemed investment direction unless and until a subsequent deemed investment direction shall be filed and become effective. An election concerning deemed investment choices shall continue indefinitely as provided in the Participant's most recent Election, or other form specified by the Committee.
- (c) If the Employer receives an initial or revised deemed investment direction which it deems to be incomplete, unclear or improper, the Participant's investment direction then in effect shall remain in effect (or, in the case of a deficiency in an initial deemed investment direction, the Participant shall be deemed to have filed no deemed investment direction) until the next Designation Date, unless the Employer provides for, and permits the application of, corrective action prior thereto.
- (d) If the Employer possesses (or is deemed to possess as provided in (c), above) at any time directions as to the deemed investment of less than all of a Participant's Account, the Participant shall be deemed to have directed that the undesignated portion of the Participant's Account be deemed to be uninvested. Or, in its discretion, the Employer may direct such undesignated portion of the Account to be deemed to be invested in a money market, fixed income or similar fund made available under the Plan as determined by the Employer.
- (e) Each reference in this Section to a Participant shall be deemed to include, where applicable, a reference to a Beneficiary.

5. ENTITLEMENT TO BENEFITS.

5.1 Fixed Payment Dates; Separation from Service. During the Open Enrollment Period of each Plan Year and on his or her Election a Participant may select a fixed payment date for the payment of amounts (or a portion of amounts) credited to his or her vested Account during the Plan Year for which the Participant Election is effective, which will be valued and payable according to the provisions of **Article 6**. Such fixed payment dates or distribution methods set forth on the Election may be postponed to later dates so long as elections to so postpone the dates or changes

the distribution methods are made by the Participant at least twelve (12) months prior to the date on which the distribution was originally scheduled to be made, the election will not take effect until at least twelve (12) months after the date on which the election is made, and the new postponed distribution date is at least five (5) years after the originally scheduled date.

A Participant who selects a fixed payment date for amounts credited to his or her Account during a Plan Year shall receive payment of such vested amounts at the earlier of such fixed payment date (as postponed, if applicable) or his or her Separation from Service.

Any fixed payment date elected by a Participant as provided above must be a date no earlier than the January 1 of the second calendar year after the calendar year for which the election is effective.

During the first Open Enrollment Period for which a Participant elects Compensation Deferrals, the Participant may specify on his or her Election whether he or she wishes to elect installment distributions in accordance with **Section 6.3** (b) for distributions on account of the Participant's Separation from Service either while a Qualified Employee or within twelve (12) months after a Change of Control that qualifies as change of control event under Treasury Regulation Section 1.409A-3(i)(5). In the absence of such a timely election such payments will be in a lump sum in accordance with **Section 6** , but a Participant may thereafter elect to receive installment distributions in accordance with **Section 6.3** (b) at any time that is at least (12) months prior to the Participant's Separation from Service if the election does not take effect until at least twelve (12) months after the date on which the election is made, and payments do not commence until at least five (5) years after the originally scheduled distribution date.

If a Participant does not make an election as provided above for any particular amounts hereunder, and the Participant Separates from Service for any reason, other than at a time when the Participant is a Qualified Employee or within twelve (12) months after a Change of Control that qualifies as "change in control event" under Treasury Regulation Section 1.409A-3(i)(5), the Participant's vested Account at the date of such Separation from Service shall be valued and payable in a single lump sum after such Separation from Service according to the provisions of Section 6.

- 5.2 **Hardship Distributions**. In the event of an unforeseeable emergency of the Participant, as hereinafter defined, the Participant may apply to the Committee for the distribution of all or any part of his or her vested Account. The Committee shall consider the circumstances of each such case, and the best interests of the Participant and his or her family, and shall have the right, in its sole discretion, if applicable, to allow such distribution, or, if applicable, to direct a distribution of part of the amount requested, or to refuse to allow any distribution. Upon a finding of unforeseeable emergency, the Committee shall make the appropriate distribution to the Participant from amounts under the Participant's vested Account. In no event shall the aggregate amount of the distribution exceed either the full value of the Participant's vested Account or the amount determined by the Committee to be necessary to alleviate the Participant's financial hardship (which financial hardship may be considered to include any taxes due because of the distribution occurring because of this **Section 5.2**) caused by the unforeseeable emergency, and which is not reasonably available from other resources of the Participant. For purposes of this **Section 5.2**, the value of the Participant's vested Account shall be determined as of the date of the distribution. "Unforeseeable Emergency" shall mean an unanticipated emergency that is caused by an event beyond the control of the Participant that would result in severe financial hardship to the Participant resulting from (i) a sudden and unexpected illness or accident of the Participant, or the Participant's spouse, Beneficiary, or dependent (as defined in Code Section 152, without regard to Code Section 152(b)(1), (b)(2), and (d)(1)(B)), (ii) a loss of the Participant's property due to casualty, or (iii) another similar extraordinary and unforeseeable circumstance arising as a result of events beyond the control of the Participant, all as determined in the sole discretion of the Committee, consistent with Treasury Regulation Section 1.409A-3(i)(3).

5.3 Limitation On Distributions To Covered Employees. If the Company reasonably anticipates that the Company's deduction with respect to any distribution from this Plan would be limited or eliminated by application of Code Section 162(m), then to the extent permitted by Treasury Regulation Section 1.409A-2(b)(7)(i), payment shall be delayed as deemed necessary to ensure that the entire amount of any distribution from this Plan is deductible. Any amounts for which distribution is delayed pursuant to this Section shall continue to be credited or debited with additional amounts in accordance with Section 4. The delayed amounts (as adjusted for any amounts credited or debited thereon) shall be distributed to the Participant (or his or her Beneficiary in the event of the Participant's death) at the earliest date the Company reasonably anticipates that the deduction of the payment of the amount will not be limited or eliminated by application of Code Section 162(m). Notwithstanding the foregoing, distribution of a Participant's Account shall be made without regard to the deductibility limitation of Code section 162(m) if the time for distribution is accelerated pursuant to **Section 9.3** or **Section 10.3**.

6. DISTRIBUTION OF BENEFITS.

6.1 Amount. The value of the Participant's (or his or her Beneficiary's) distribution shall be equal to the vested value of the Participant's Account as of the Valuation Date or such other date as the Committee may specify, each as adjusted for Compensation Deferrals, Discretionary Employer Contributions, and/or withdrawals which have been subsequently credited thereto or made therefrom prior to the distribution date.

6.2 Timing Of Distribution. Distributions shall be paid (or, payments shall commence in installments) within ninety (90) days after the earlier of:

- (a) the fixed payment date designated by the Participant; or
- (b) the date within ninety (90) days after the Participant's Separation from Service, death, or Disability.

6.3 Method Of Distribution. A Participant's Account shall be paid in one of the following methods, as specified in his or her Election:

- (a) as to all or any designated portion of Participant's Account, a single lump sum payment;
- (b) if, and only if, the Participant's employment was terminated at a time when the Participant was a Qualified Employee or within twelve (12) months after a Change of Control that qualifies as a "change in control event" within the meaning of Treasury Regulation Section 1.409A-3(i)(5), and if elected by the Participant in his or her most recent effective Election made in accordance with **Section 5.1**, in annual installment payments of substantially equal amounts over a period of up to fifteen (15) years;
- (c) a Participant may amend his or her Election so as to select installments upon Separation from Service at a time when the Participant is a Qualified Employee by filing an amended Election provided, however, that such Election to so change to installment distributions upon such Separation of Service is made by the Participant at least twelve (12) months prior to the separation date, the election will not take effect until at least twelve (12) months after the date on which the election is made, and the new postponed distribution date is at least five (5) years from the original Separation from Service by the Qualified Employee; provided that, in no event shall any such distribution date be accelerated to a date earlier than that initially selected by the Participant; and
- (d) the Employer (or its designee) may establish from time to time limitations on the Participant's ability to select the time and method of payment of his Account based upon the amount in the Participant's Account; provided further that, unless and until changed

by the Employer (or its designee), any Account that has a total vested balance of less than \$5,000 at the time of distribution shall be paid in a lump sum regardless of an election by the Participant to be paid in installments.

- (e) If, at the time of Participant's Separation from Service, the Participant is a "specified employee" (within the meaning of Code Section 409A and Treas. Reg. Section 1.409A-3(i)(2)), the Company will not pay or provide any "Specified Benefits" (as defined herein) during the six-month period (the "409A Suspension Period") beginning immediately after the Participant's Separation from Service. For purposes of this Agreement, "Specified Benefits" are any amounts or benefits that would be subject to Code Section 409A penalties if the Company were to pay them, pursuant to this Plan, on account of the Participant's Separation from Service. During the 409A Suspension Period, the Participant's Account will continue to be credited or debited in accordance with **Section 4.1** until the Participant's Account is distributed. Within fourteen (14) calendar days following the end of the 409A Suspension Period, the Participant shall be paid a lump sum payment in cash equal to any Specified Benefits delayed during the 409A Suspension Period.

7. **BENEFICIARIES; PARTICIPANT DATA.**

- 7.1 **Designation Of Beneficiaries.** Each Participant from time to time may designate any person or persons (who may be named contingently or successively) to receive such benefits as may be payable under the Plan upon or after the Participant's death, and such designation may be changed from time to time by the Participant by filing a new designation. Each designation will revoke all prior designations by the same Participant, shall be in a form prescribed by the Employer, and will be effective only when filed in writing with the Employer during the Participant's lifetime.

In the absence of a valid Beneficiary designation, or if, at the time any benefit payment is due to a Beneficiary, there is no living Beneficiary validly named by the Participant, the Employer shall pay any such benefit payment to the Participant's spouse, if then living, but otherwise to the Participant's then living descendants, if any, per stirpes, but, if none, to the Participant's estate. In determining the existence or identity of anyone entitled to a benefit payment, the Employer may rely conclusively upon information supplied by the Participant's personal representative, executor or administrator. If a question arises as to the existence or identity of anyone entitled to receive a benefit payment as aforesaid, or if a dispute arises with respect to any such payment, then, notwithstanding the foregoing, the Employer, in its sole discretion, may distribute such payment to the Participant's estate without liability for any tax or other consequences which might flow therefrom, or may take such other action as the Employer deems to be appropriate.

- 7.2 **Information To Be Furnished By Participants And Beneficiaries; Inability To Locate Participants Or Beneficiaries.** Any communication, statement or notice addressed to a Participant or to a Beneficiary at his or her last post office address as shown on the Employer's records shall be binding on the Participant or Beneficiary for all purposes of the Plan. The Committee shall not be obliged to search for any Participant or Beneficiary beyond the sending of notice to such last known address. If the Committee notifies any Participant or Beneficiary that he or she is entitled to an amount under the Plan and the Participant or Beneficiary fails to claim such amount or make his or her location known to the Committee within three (3) years thereafter, then, except as otherwise required by law, if the location of one or more of the next of kin of the Participant is known to the Committee, the Committee may direct distribution of such amount to any one or more or all of such next of kin, and in such proportions as the Committee determines. If the location of none of the foregoing persons can be determined, the Committee shall have the right to direct that the amount payable shall be deemed to be a forfeiture, except that the dollar amount of the forfeiture, unadjusted for deemed gains or losses in the interim, shall be paid by the Committee if a claim for the benefit subsequently is made by the Participant or the Beneficiary to whom it was payable. If a benefit payable to an unlocated Participant or Beneficiary is subject to escheat pursuant to applicable state law, the Committee shall not be liable to any person for any payment made in accordance with such law.

8. ADMINISTRATION.

- 8.1 Committee Powers And Responsibilities. Other than the powers reserved for the Board, the Committee shall have the complete control of the administration of the Plan herein set forth with all the powers necessary to enable it to properly carry out its duties in that respect. Not in limitation, but in amplification of the foregoing, the Committee shall have the power and authority to:
- (a) Construe and interpret the Plan and determine all questions that shall arise as to the interpretations of the Plan's provisions including determination of which individuals are Eligible Employees and the determination of the amounts credited to a Participant's Account, and the appropriate timing and method of distributions.
 - (b) Adopt such rules of procedure and regulations as in its opinion may be necessary for the proper and efficient administration of the Plan and as are consistent with the Plan.
 - (c) Implement the Plan in accordance with its terms and the rules and regulations adopted as above.
 - (d) Appoint any persons or firms, or otherwise act to secure specialized advice or assistance, as it deems necessary or desirable in connection with the administration and operation of the Plan, and the Committee shall be entitled to rely conclusively upon, and shall be fully protected in any action or omission taken by it in good faith reliance upon, the advice or opinion of such firms or persons, The Committee may authorize one or more persons to execute any certificate or document on behalf of the Company, an Employer or the Committee, in which event any person notified by the Committee of such authorization shall be entitled to accept and conclusively rely upon any such certificate or document executed by such person as representing action by the Committee until such notified person shall have been notified of the revocation of such authority.
 - (e) Subject to **Section 9** , adopt amendments to the Plan document which are deemed necessary or desirable to facilitate administration of the Plan and/or to bring the Plan into compliance with all applicable laws and regulations, provided that the Committee shall not have the authority to adopt any Plan amendment that will result in substantially increased costs to the Company unless such amendment is either expressly authorized by the Board or contingent upon ratification by the Board before becoming effective.
 - (f) Select, review and retain or change any deemed investment fund under the Plan.
 - (g) Compile and maintain all records it determines to be necessary, appropriate or convenient in connection with the administration of the Plan.
 - (h) Direct the investment of the assets of the Trust.
 - (i) Review the performance of the Trustee and any other advisor or service provider to the Plan.
 - (j) Take such other action as may be necessary or appropriate to the management and investment of the Plan assets and administration of the Plan.
- 8.2 Litigation. Except as may be otherwise required by law, in any action or judicial proceeding affecting the Plan, no Participant or Beneficiary shall be entitled to any notice or service of process, and any final judgment entered in such action shall be binding on all persons interested in, or claiming under, the Plan.

8.3 Indemnification. To the extent permitted by law, the Company shall indemnify each member of the Committee, and any other employee or member of the Board with duties under the Plan, against losses and expenses (including any amount paid in settlement) reasonably incurred by such person in connection with any claims against such person by reason of such person's conduct in the performance of duties under the Plan, except in relation to matters as to which such person has acted fraudulently or in bad faith in the performance of duties. Notwithstanding the foregoing, the Company shall not indemnify any person for any expense incurred through any settlement or compromise of any action unless the Company consents in writing to the settlement or compromise.

8.4 Claims Procedure.

- (a) Initial Claim. A Participant or Beneficiary who believes he or she is entitled to any Benefit (a "Claimant") under this Plan may file a claim with the Committee. The Committee shall review the claim itself or appoint an individual or an entity to review the claim.
- (i) Benefit Claim. The Claimant shall be notified within ninety (90) days after the claim is filed whether the claim is allowed or denied, unless the Claimant receives written notice from the Committee or from an appointee of the Committee before the end of the ninety (90) day period stating that special circumstances require an extension of the time for decision. Any such extension will not extend beyond one hundred eighty (180) days after the day the claim is filed.
- (ii) Manner and Content of Denial of Initial Claims. If the Plan Administrator denies a claim, it must provide to the Claimant, in writing or by electronic communication:
1. the specific reasons for the denial;
 2. a reference to the Plan provision upon which the denial is based;
 3. a description of any additional information or material that the Claimant must provide in order to perfect the claim;
 4. an explanation of why such additional material or information is necessary;
 5. notice that the Claimant has a right to request a review of the claim denial and information on the steps to be taken if the Claimant wishes to request a review of the claim denial; and
 6. a statement of the participant's right to bring a civil action under ERISA §502(a) following a denial on review of the initial denial.
- (b) Review Procedures.
- (i) Benefit Claims. A request for review of a denied claim must be made in writing to the Committee within sixty (60) days after receiving notice of denial. The decision upon review will be made within sixty (60) days after the Committee's receipt of a request for review, unless special circumstances

require an extension of time for processing, in which case a decision will be rendered not later than one hundred twenty (120) days after receipt of a request for review. A notice of such an extension must be provided to the Claimant within the initial sixty (60) day period and must explain the special circumstances and provide an expected date of decision. The reviewer shall afford the Claimant an opportunity to review and receive, without charge, all relevant documents, information and records and to submit issues and comments in writing to the Committee. The reviewer shall take into account all comments, documents, records and other information submitted by the Claimant relating to the claim regardless of whether the information was submitted or considered in the initial benefit determination.

- (ii) Manner and Content of Notice of Decision on Review. Upon completion of its review of an adverse initial claim determination, the Committee will give the Claimant, in writing or by electronic notification, a notice containing:
1. its decision;
 2. the specific reasons for the decision;
 3. the relevant Plan provisions on which its decision is based;
 4. a statement that the Claimant is entitled to receive, upon request and without charge, reasonable access to, and copies of, all documents, records and other information in the Plan's files which is relevant to the Claimant's claim for benefits;
 5. a statement describing the Claimant's right to bring an action for judicial review under ERISA §502(a); and
 6. if an internal rule, guideline, protocol or other similar criterion was relied upon in making the adverse determination on review, a statement that a copy of the rule, guideline, protocol or other similar criterion will be provided without charge to the Claimant upon request.
- (c) Calculation of Time Periods. For purposes of the time periods specified in this **Section 8.5**, the period of time during which a benefit determination is required to be made begins at the time a claim is filed in accordance with the Plan procedures without regard to whether all the information necessary to make a decision accompanies the claim. If a period of time is extended due to a Claimant's failure to submit all information necessary, the period for making the determination shall be tolled from the date the notification is sent to the Claimant until the date the Claimant responds.
- (d) Failure of Plan to Follow Procedures. If the Plan fails to follow the claims procedures required by this **Section 8.5**, a Claimant shall be deemed to have exhausted the administrative remedies available under the Plan and shall be entitled to pursue any available remedy under ERISA section 502(a) on the basis that the Plan has failed to provide a reasonable claims procedure that would yield a decision on the merits of the claim. If the Claimant fails to follow the claims procedures required by this **Section 8.5**, the Claimant shall not be entitled to pursue any further legal action, claim or remedy until such time as the Claimant, to the extent applicable, exhausts the administrative remedies available under the Plan.

9. AMENDMENT.

- 9.1 Right To Amend. The Committee or the Company, by action of the Board, shall have the right to amend the Plan, at any time and with respect to any provisions hereof, and all parties hereto or claiming any interest hereunder shall be bound by such amendment; provided, however, that no such amendment shall deprive a Participant or a Beneficiary of a right accrued hereunder prior to the date of the amendment unless such an amendment is required by applicable law or deemed necessary to preserve the preferred tax treatment of the Plan.
- 9.2 Amendments To Ensure Proper Characterization Of Plan. Notwithstanding the provisions of **Section 9.1**, the Plan may be amended by the Committee or the Company, by action of its Board, at any time, retroactively if required, if found necessary, in the opinion of the Committee or the Board, in order to ensure that the Plan is characterized as a “top-hat” plan of deferred compensation maintained for a select group of management or highly compensated employees as described under ERISA sections 201(2), 301(a)(3), and 401(a)(1), and to conform the Plan to the provisions and requirements of any applicable law (including specifically Section 409A of the Code, and other applicable portions of ERISA and the Code). No such amendment shall be considered prejudicial to any interest of a Participant or a Beneficiary hereunder.
- 9.3 Changes In Law Affecting Taxability.
- (a) Operation. This **Section 9.3** shall become operative upon the enactment of any change in applicable statutory law or the promulgation by the Internal Revenue Service of a final regulation or other pronouncement having the force of law, which statutory law, as changed, or final regulation or pronouncement, as promulgated, would cause any Participant to include in his or her federal gross income amounts accrued by the Participant under the Plan on a date (an “Early Taxation Event”) prior to the date on which such amounts are made available to him or her hereunder.
- (b) Affected Right or Feature Nullified. Notwithstanding any other Section of this Plan to the contrary (but subject to **Section 9.3(c)**), as of an Early Taxation Event, the feature or features of this Plan that would cause the Early Taxation Event shall be null and void, to the extent, and only to the extent, required to prevent the Participant from being required to include in his or her federal gross income amounts accrued by the Participant under the Plan prior to the date on which such amounts are made available to him or her hereunder. If only a portion of a Participant’s Account is impacted by the change in the law, then only such portion shall be subject to this **Section 9.3**, with the remainder of the Account not so affected being subject to such rights and features as if the law were not changed. If the law only impacts Participants who have a certain status with respect to the Employer, then only such Participants shall be subject to this **Section 9.3**.
- (c) Tax Distribution. If the Plan fails to satisfy the requirements of Code Section 409A with respect to a Participant, the Participant’s Account may be distributed to the Participant in an amount that is not greater than the amount required to be included in the Participant’s income as a result of the Plan’s failure to comply with Code Section 409A.

10. TERMINATION.

- 10.1 Employer’s Right To Terminate Or Suspend Plan. Subject to **Section 10.3** hereof, the Employer reserves the right to terminate the Plan and/or its obligation to make further credits to Plan Accounts, by action of its Board of Directors. The Employer also reserves the right to suspend the operation of the Plan for a fixed or indeterminate period of time, by action of its Board of Directors.

- 10.2 Suspension Of Deferrals. In the event of a suspension of the Plan, the Employer shall continue all aspects of the Plan, other than Compensation Deferrals and Discretionary Employer Contributions, during the period of the suspension, in which event payments hereunder will continue to be made during the period of the suspension in accordance with **Sections 5 and 6**. Notwithstanding the foregoing, Compensation Deferrals may not be discontinued in the middle of a Plan Year.
- 10.3 Limits on Plan Termination. The Plan may be terminated at any time in accordance with one of the following circumstances set forth in (a) through (c) below in accordance with Treasury Regulation Section 1.409A-3(j)(4)(ix).
- (a) The Company may terminate the Plan if the termination and liquidation is not proximate to a downturn in the Company's financial health and:
 - (i) The Plan and all other plans maintained by the Company that would be aggregated with the Plan under Treasury Regulation Section 1.409A-1(c) are irrevocably terminated;
 - (ii) No payments other than payments that would otherwise be payable under the terms of the Plan are made within twelve (12) months following the date the Company takes all necessary actions to terminate and liquidate the Plan;
 - (iii) Except with respect to the Participants who became entitled to benefits under the terms of the Plan and any other plan maintained by the Employer that would be aggregated with the Plan under Treasury Regulation Section 1.409A-1(c) within the first twelve (12) months following the date such plans are irrevocably terminated, all payments to the Participants due under the terms of such plans must be made between the first day of the 13th month and the last day of the 24th month following the date such plans terminated; and
 - (iv) The Company does not adopt a plan that would be aggregated with this Plan under Treasury Regulation Section 1.409A-1(c) within three years following the date the Plan is terminated.
 - (b) The Company terminates and liquidates the Plan pursuant to irrevocable action taken within thirty (30) days preceding or twelve (12) months following a "change in control event" (defined below), provided that the Plan and all other plans maintained by the Company that would be aggregated with the Plan under Treasury Regulation Section 1.409A-1(c) are terminated on the same date with respect to each participant in such plans that experienced the "change in control event," and all such participants receive all benefits payable under such plans within twelve (12) months following the termination date. For purposes of this **Section 10.1** (b), "change in control event" shall have the meaning set forth in Treasury Regulation Section 1.409A-3(i)(5).
 - (c) The Company terminates and liquidates the Plan within twelve (12) months of a corporate dissolution taxed under Code Section 331, or with the approval of a bankruptcy court pursuant to 11 U.S.C. § 503(b)(1)(A), provided that all benefits payable under the Plan are distributed to Participants during the earlier of (i) the taxable year in which the amount is actually or constructively received, or (ii) the latest of the calendar year in which (a) the Plan is terminated and liquidated; (b) the benefits are no longer subject to a substantial risk of forfeiture; or (c) the payment first becomes administratively practicable.

11. THE TRUST.

- 11.1 Establishment Of Trust. The Employer, in its sole and absolute discretion, may establish a Trust with a qualified trustee pursuant to such terms and conditions as are set forth in a Trust agreement to be entered into between the Employer and such Trustee. Or, the Employer may cause to be maintained one or more separate subaccounts in an existing Trust maintained with the Trustee with respect to one or more other plans of the Employer, which subaccount or subaccounts represent Participants' interests in the Plan. The Employer shall have the discretion to make contributions to such Trust that correspond to credits to Participants' Accounts and/or to invest Trust assets in a manner that corresponds to Participants' selected deemed investments in order to provide a source of funds with which the Employer shall pay Plan benefits as they become due.

Any amounts held in a Trust established under this **Section 11.1** shall be the sole property of the Employer and will not be held as collateral security for fulfillment of the Employer's obligation under the Plan. Any such Trust shall be intended to be treated as a "grantor trust" under the Code and the establishment of the Trust or the utilization of any existing Trust for Plan benefits, as applicable, shall not be intended to cause any Participant to realize current income on amounts contributed thereto, and the Trust shall be so interpreted. Any such funds will be subject to the claims of all bankruptcy or insolvency creditors of the Employer as provided in the Trust agreement, and no Participant or Beneficiary will have any vested interest or secured or preferred position with respect to such funds or have any claims against the Employer hereunder except as a general creditor.

12. MISCELLANEOUS.

- 12.1 Limitations On Liability Of Employer. Neither the establishment of the Plan nor any modification thereof, nor the creation of any account under the Plan, nor the payment of any benefits under the Plan shall be construed as giving to any Participant or other person any legal or equitable right against the Employer, or any officer or employer thereof except as provided by law or by any Plan provision. The Employer does not in any way guarantee any Participant's Account from loss or depreciation, whether caused by poor investment performance of a deemed investment or the inability to realize upon an investment due to an insolvency affecting an investment vehicle or any other reason. In no event shall the Employer, or any successor, employee, officer, director or stockholder of the Employer, be liable to any person on account of any claim arising by reason of the provisions of the Plan or of any instrument or instruments implementing its provisions, or for the failure of any Participant, Beneficiary or other person to be entitled to any particular tax consequences with respect to the Plan, or any credit or distribution hereunder. Participant shall be solely responsible for the satisfaction of any taxes with respect to the benefits payable to the Participant under this Plan (including, but not limited to, employment taxes imposed on employees and additional taxes on nonqualified deferred compensation). Although the Company intends and expects that the Plan and its payments and benefits will not give rise to taxes imposed under Section 409A of the Code, neither the Company, nor its employees, directors, or agents shall have any obligation to mitigate or to hold any Participant harmless from any or all of such taxes.
- 12.2 Construction. The Plan is intended to be and at all times shall be interpreted and administered so as to qualify as an unfunded deferred compensation plan, and no provision of the Plan shall be interpreted so as to give any individual any right in any assets of the Employer which right is greater than the rights of a general unsecured creditor of the Employer.
- 12.3 Spendthrift Provision. No amount payable to a Participant or a Beneficiary under the Plan will, except as otherwise specifically provided by law, be subject in any manner to anticipation, alienation, attachment, garnishment, sale, transfer, assignment (either at law or in equity), levy, execution, pledge, encumbrance, charge or any other legal or equitable process, and any attempt to do so will be void; nor will any benefit be in any manner liable for or subject to the debts, contracts, liabilities, engagements or torts of the person entitled thereto. Further, (i) the withholding of taxes from Plan benefit payments, (ii) the recovery under the Plan of overpayments of benefits previously made to a Participant or Beneficiary, (iii) if applicable, the transfer of

benefit rights from the Plan to another plan, or (iv) the direct deposit of benefit payments to an account in a banking institution (if not actually part of an arrangement constituting an assignment or alienation) shall not be construed as an assignment or alienation.

In the event that any Participant's or Beneficiary's benefits hereunder are garnished or attached by order of any court, the Employer or Trustee may bring an action or a declaratory judgment in a court of competent jurisdiction to determine the proper recipient of the benefits to be paid under the Plan. During the pendency of said action, any benefits that become payable shall be held as credits to the Participant's or Beneficiary's Account or, if the Employer or Trustee prefers, paid into the court as they become payable, to be distributed by the court to the recipient as the court deems proper at the close of said action.

- 12.4 Tax Withholding. Distribution and withdrawal payments under this Plan shall be subject to all applicable withholding requirements for state and federal income taxes and to any other federal, state or local taxes that may be applicable to such payments. The Company shall have the right, but not the obligation, to deduct from any distribution from the Plan, that amount equal to all or any part of the federal, state, local and foreign taxes, if any, required by law to be withheld by the Company with respect to such distributions. Alternatively or in addition, in its discretion, the Company shall have the right to require a Participant, through payroll withholding, cash payment or otherwise, to make adequate provision for any such tax withholding obligations of the Company arising in connection with any distribution from the Plan. The Trustee shall have no obligation to distribute amounts from the Trust until the Company's tax withholding obligations have been satisfied by the Participant.
- 12.5 No Employment Agreement. Nothing contained herein shall be construed as conferring upon any Participant the right to continue in the employ of the Employer as an employee.
- 12.6 Attorney's Fees. If the Employer, the Participant, any Beneficiary, any beneficiary under an insurance policy purchased under the Trust, and/or a successor in interest to any of the foregoing, brings legal action to enforce any of the provisions of this Plan, the prevailing party in such legal action shall be reimbursed by the other party, the prevailing party's costs of such legal action including, without limitation, reasonable fees of attorneys, accountants and similar advisors and expert witnesses.
- 12.7 Governing Law. This Plan shall be construed in accordance with and governed by any applicable provisions of ERISA and the laws of the State of Delaware.
- 12.8 Entire Agreement. This Plan constitutes the entire understanding and agreement with respect to the subject matter contained herein, and there are no agreements, understandings, restrictions, representations or warranties among any Participant and the Employer other than those as set forth or provided for herein.
- 12.9 Severability. If any provision of this Plan is determined, by the Committee or any governmental agency or court decision, to be unenforceable or invalid under any applicable law, such unenforceability or invalidity shall not render this Plan unenforceable or invalid as a whole, and such provision shall be changed and interpreted by the Committee, in its sole discretion, so as to best accomplish the objectives of such unenforceable or invalid provision within the limits of applicable law or applicable court decisions.

CERTIFICATION

I, Jean-Jacques Bienaimé, certify that:

1. I have reviewed this Quarterly Report on Form 10-Q of BioMarin Pharmaceutical Inc.;
2. Based on my knowledge, this report does not contain any untrue statement of a material fact or omit to state a material fact necessary to make the statements made, in light of the circumstances under which such statements were made, not misleading with respect to the period covered by this report;
3. Based on my knowledge, the financial statements, and other financial information included in this report, fairly present in all material respects the financial condition, results of operations and cash flows of the registrant as of, and for, the periods presented in this report;
4. The registrant's other certifying officer(s) and I are responsible for establishing and maintaining disclosure controls and procedures (as defined in Exchange Act Rules 13a-15(e) and 15d-15(e)) and internal control over financial reporting (as defined in Exchange Act Rules 13a-15(f) and 15d-15(f)) for the registrant and have:
 - a) Designed such disclosure controls and procedures, or caused such disclosure controls and procedures to be designed under our supervision, to ensure that material information relating to the registrant, including its consolidated subsidiaries, is made known to us by others within those entities, particularly during the period in which this report is being prepared;
 - b) Designed such internal control over financial reporting, or caused such internal control over financial reporting to be designed under our supervision, 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;
 - c) Evaluated the effectiveness of the registrant's disclosure controls and procedures and presented in this report our conclusions about the effectiveness of the disclosure controls and procedures, as of the end of the period covered by this report based on such evaluation; and
 - d) Disclosed in this report any change in the registrant's internal control over financial reporting that occurred during the registrant's most recent fiscal quarter (the registrant's fourth fiscal quarter in the case of an annual report) that has materially affected, or is reasonably likely to materially affect, the registrant's internal control over financial reporting; and
5. The registrant's other certifying officer(s) and I have disclosed, based on our most recent evaluation of internal control over financial reporting, to the registrant's auditors and the audit committee of the registrant's board of directors (or persons performing the equivalent functions):
 - a) All significant deficiencies and material weaknesses in the design or operation of internal control over financial reporting which are reasonably likely to adversely affect the registrant's ability to record, process, summarize and report financial information; and
 - b) Any fraud, whether or not material, that involves management or other employees who have a significant role in the registrant's internal control over financial reporting.

May 2, 2014

/s/ JEAN-JACQUES BIENAIMÉ

Jean-Jacques Bienaimé
Chief Executive Officer

CERTIFICATION

I, Daniel Spiegelman, certify that:

1. I have reviewed this Quarterly Report on Form 10-Q of BioMarin Pharmaceutical Inc.;
2. Based on my knowledge, this report does not contain any untrue statement of a material fact or omit to state a material fact necessary to make the statements made, in light of the circumstances under which such statements were made, not misleading with respect to the period covered by this report;
3. Based on my knowledge, the financial statements, and other financial information included in this report, fairly present in all material respects the financial condition, results of operations and cash flows of the registrant as of, and for, the periods presented in this report;
4. The registrant's other certifying officer(s) and I are responsible for establishing and maintaining disclosure controls and procedures (as defined in Exchange Act Rules 13a-15(e) and 15d-15(e)) and internal control over financial reporting (as defined in Exchange Act Rules 13a-15(f) and 15d-15(f)) for the registrant and have:
 - a) Designed such disclosure controls and procedures, or caused such disclosure controls and procedures to be designed under our supervision, to ensure that material information relating to the registrant, including its consolidated subsidiaries, is made known to us by others within those entities, particularly during the period in which this report is being prepared;
 - b) Designed such internal control over financial reporting, or caused such internal control over financial reporting to be designed under our supervision, 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;
 - c) Evaluated the effectiveness of the registrant's disclosure controls and procedures and presented in this report our conclusions about the effectiveness of the disclosure controls and procedures, as of the end of the period covered by this report based on such evaluation; and
 - d) Disclosed in this report any change in the registrant's internal control over financial reporting that occurred during the registrant's most recent fiscal quarter (the registrant's fourth fiscal quarter in the case of an annual report) that has materially affected, or is reasonably likely to materially affect, the registrant's internal control over financial reporting; and
5. The registrant's other certifying officer(s) and I have disclosed, based on our most recent evaluation of internal control over financial reporting, to the registrant's auditors and the audit committee of the registrant's board of directors (or persons performing the equivalent functions):
 - a) All significant deficiencies and material weaknesses in the design or operation of internal control over financial reporting which are reasonably likely to adversely affect the registrant's ability to record, process, summarize and report financial information; and
 - b) Any fraud, whether or not material, that involves management or other employees who have a significant role in the registrant's internal control over financial reporting.

May 2, 2014

/s/ DANIEL SPIEGELMAN

Daniel Spiegelman

Executive Vice President and Chief Financial Officer

**CERTIFICATION PURSUANT TO
18 U.S.C. SECTION 1350,
AS ADOPTED PURSUANT TO
SECTION 906 OF THE SARBANES-OXLEY ACT OF 2002**

We, Jean-Jacques Bienaimé and Daniel Spiegelman hereby certify, pursuant to 18 U.S.C. §1350, as adopted pursuant to §906 of the Sarbanes-Oxley Act of 2002, that BioMarin Pharmaceutical Inc.'s Quarterly Report on Form 10-Q for the period ended March 31, 2014, fully complies with the requirements of Section 13(a) or 15(d) of the Securities Exchange Act of 1934 and the information contained in such Form 10-Q fairly presents, in all material respects, the financial condition and results of operations of BioMarin Pharmaceutical Inc.

/s/ JEAN-JACQUES BIENAIMÉ

Jean-Jacques Bienaimé
Chief Executive Officer

May 2, 2014

/s/ DANIEL SPIEGELMAN

Daniel Spiegelman
Executive Vice President and Chief Financial Officer

May 2, 2014