

BIOMARIN PHARMACEUTICAL INC
Form DEF 14A
May 16, 2006

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

SCHEDULE 14A

Proxy Statement Pursuant to Section 14(a) of the
Securities Exchange Act of 1934

Filed by the Registrant Filed by a Party other than the Registrant

Check the appropriate box:

- Preliminary Proxy Statement
- Confidential, for Use of the Commission Only** (as permitted by Rule 14a-6(e)(2))
- Definitive Proxy Statement
- Definitive Additional Materials
- Soliciting Material Pursuant to Section 240.14a-12

BioMarin Pharmaceutical Inc.

(Name of Registrant as Specified In Its Charter)

(Name of Person(s) Filing Proxy Statement if other than the Registrant)

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BIOMARIN PHARMACEUTICAL INC.

NOTICE OF ANNUAL MEETING OF STOCKHOLDERS

TO BE HELD ON JUNE 21, 2006

TO THE STOCKHOLDERS OF BIOMARIN PHARMACEUTICAL INC.:

NOTICE IS HEREBY given that the 2006 Annual Meeting of the Stockholders (the Annual Meeting) of BioMarin Pharmaceutical Inc. (BioMarin or the Company) will be held on June 21, 2006 at 10:00 a.m. (California time), at the Courtyard By Marriott Hotel at 1400 N. Hamilton Parkway, Novato, California 94949 for the following purposes:

1. to elect 6 directors of the Company;
2. to ratify the selection of KPMG LLP as the independent registered public accounting firm for BioMarin for the year ending December 31, 2006;
3. to approve the 2006 Share Incentive Plan (the SIP) for directors and employees;
4. to approve the Amended and Restated 2006 Employee Stock Purchase Plan (the ESPP); and
5. to transact such other business as properly may be brought before the Annual Meeting or any adjournment thereof.

The foregoing items of business are more fully described in the proxy statement accompanying this notice.

The Board of Directors has fixed the close of business on May 5, 2006 as the record date (the Record Date) for determining the stockholders entitled to receive notice of, and to vote at, the Annual Meeting or any adjournment thereof. A complete list of such stockholders will be available for examination by any stockholder for any purpose germane to the meeting during ordinary business hours at the Company s executive offices at 105 Digital Drive, Novato, California 94949, for a period of ten days before the Annual Meeting.

All stockholders are cordially invited to attend the Annual Meeting. To ensure your representation at the Annual Meeting, you are urged to sign and date the enclosed WHITE Proxy Card and return it today in the enclosed pre-addressed postage-paid envelope. Any stockholder attending the Annual Meeting may vote in person even if that stockholder has returned a proxy.

If you have any questions or need assistance in voting your shares, please call the firm assisting the Company in the solicitation of proxies:

Morrow & Co., Inc.

445 Park Avenue - 5th Floor

New York, NY 10022

1-800-607-0088

By Order of the Board of Directors

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G. Eric Davis

Vice President, General Counsel and Secretary

Novato, California

May 5, 2006

YOUR VOTE IS IMPORTANT. IN ORDER TO ASSURE YOUR REPRESENTATION AT THE ANNUAL MEETING, YOU ARE REQUESTED TO SIGN AND DATE THE ENCLOSED WHITE PROXY CARD AND RETURN IT TODAY IN THE ENCLOSED PRE-ADDRESSED POSTAGE-PAID ENVELOPE.

BioMarin Pharmaceutical Inc.

105 Digital Drive

Novato, California 94949

Proxy Statement for 2006 Annual Meeting of Stockholders

INFORMATION CONCERNING SOLICITATION OF PROXIES AND VOTING

General

This proxy statement is furnished in connection with the solicitation of proxies by the Board of Directors of the Company (the Board) for use at the 2006 Annual Meeting of Stockholders of the Company (the Annual Meeting) to be held on June 21, 2006, or at any adjournment or postponement of the Annual Meeting, for the purpose set forth herein and in the foregoing Notice of Annual Meeting of Stockholders. The Annual Meeting will be held at 10:00 a.m. local time at the Courtyard by Marriott Hotel located at 1400 N. Hamilton Parkway, Novato, California 94949. Copies of this solicitation material and the Company's Annual Report are being mailed to stockholders entitled to vote at the Annual Meeting and furnished to brokerage houses, fiduciaries and custodians to forward to beneficial owners of common stock of the Company held in their names on or about May 16, 2006. The Company's Annual Report, which includes its Form 10-K for the year ended December 31, 2005, as filed with the Securities and Exchange Commission, is provided with this proxy statement.

Record Date; Outstanding Shares

The voting securities of the Company entitled to vote at the Annual Meeting consist of only shares of common stock. Only stockholders of record at the close of business on May 5, 2006 are entitled to notice of and to vote at the Annual Meeting. On May 5, 2006, there were 85,358,801 shares of BioMarin common stock, par value \$0.001 per share, issued and outstanding and entitled to vote. Each share of common stock is entitled to one vote. As of May 5, 2006, an additional 19,326,886 shares of common stock were reserved for issuance upon conversion of the Company's outstanding convertible debt. The convertible debt does not include any right to vote on the matters to be considered at the Annual Meeting.

Revocability of Proxies

A stockholder who signs and returns a WHITE Proxy Card will have the power to revoke it at any time before it is voted. A proxy represented by a WHITE Proxy Card may be revoked by (i) delivering to the Company at the Company's mailing address appearing at the top of this page (Attention: G. Eric Davis, Vice President, General Counsel and Secretary) a written notice of revocation, or (ii) submitting a duly executed proxy card bearing a later date, or (iii) by appearing at the Annual Meeting and voting in person. Attendance at the meeting in and of itself, without voting in person at the meeting, will not cause your previously granted proxy to be revoked. For shares you hold in street name, you may change your vote by submitting new voting instructions to your broker, bank or other nominee or, if you have obtained a legal proxy from your broker, bank or other nominee giving you the right to vote your shares at the annual meeting, by attending the meeting and voting in person.

Voting

Each stockholder is entitled to one vote for each share of common stock held. This includes shares held directly by stockholders of record and shares held through a stock broker, bank or nominee.

Solicitation of Proxies

This solicitation of proxies is made by the Company and all related costs, including expenses in connection with preparing and mailing this proxy statement, will be borne by the Company. Copies of solicitation material will be furnished to brokerage firms and other persons representing beneficial owners of shares. In addition, if asked, the Company will reimburse brokerage firms and other persons representing beneficial owners of shares for their expenses in forwarding solicitation material to such beneficial owners. The Company has requested brokerage firms and other persons representing beneficial owners of shares to forward all solicitation materials to the beneficial owners of the shares they hold of record. Proxies may also be solicited by certain of the Company's directors, officers and regular employees, without additional compensation. The original solicitation by mail may also be supplemented by telephone, facsimile, e-mail and personal solicitation by these directors, officers and employees, but stockholders will not be asked to submit proxies by telephone or the Internet.

The Company has retained Morrow & Co., Inc. to assist it in the solicitation of proxies. The Company has agreed to pay customary fees to Morrow & Co. for its services in soliciting proxies, which are estimated to be \$10,000, and has agreed to reimburse Morrow & Co. for reasonable out-of-pocket expenses for these services.

Quorum; Abstentions; Broker Non-Votes

The Company's Bylaws provide that a majority of all the shares of the common stock entitled to vote, whether present in person or by proxy, shall constitute a quorum for the transaction of business at the Annual Meeting. Broker non-votes are shares held in street name for which the broker has not received instructions from the beneficial owners or other persons entitled to vote and the broker does not have discretionary voting authority. Abstentions and broker non-votes will be counted as shares present for purposes of determining whether a quorum is present. If a quorum is not present or represented, then either the chairman of the Annual Meeting or the stockholders entitled to vote at the Annual Meeting, present in person or represented by proxy, will have the power to adjourn the Annual Meeting from time to time, without notice other than an announcement at the Annual Meeting, until a quorum is present. At any adjourned Annual Meeting at which a quorum is present, any business may be transacted that might have been transacted at the Annual Meeting as originally notified. If the adjournment is for more than 30 days, or if after that adjournment a new record date is fixed for the adjourned Annual Meeting, a notice of the adjourned Annual Meeting shall be given to each stockholder of record entitled to vote at the adjourned Annual Meeting.

When proxies are properly executed and returned, the shares represented by such proxies will be voted at the Annual Meeting in accordance with the instructions of the stockholder. If no specific instructions are given, the shares represented by an executed WHITE Proxy Card will be voted (i) FOR the election of the 6 nominees for directors set forth herein; (ii) FOR the ratification of the selection of KPMG LLP as the independent registered public accounting firm for BioMarin for the year ending December 31, 2006; (iii) FOR the approval of the new SIP; and (iv) FOR the approval of the new ESPP. The Board knows of no other business that will be presented at the meeting. If, however, other matters are properly presented, the persons named in the enclosed WHITE Proxy Card will vote the shares represented thereby in accordance with their best judgment pursuant to the discretionary authority granted in the proxy.

If your shares are held in street name, your broker, bank or nominee will include a voting instruction card with this proxy statement. You should vote your shares by following the instructions provided on the voting instruction card.

Attendance at Meeting

If you plan to attend the Annual Meeting, please note that attendance will be limited to stockholders as of the record date. Admission will be on a first-come, first-served basis. Each stockholder may be asked to present

valid picture identification, such as a driver's license or passport. Stockholders holding stock in brokerage accounts or by a bank or other nominee may be required to show a brokerage statement or account statement reflecting stock ownership as of the record date. Cameras, recording devices and other electronic devices will not be permitted at the Annual Meeting.

Required Vote

Election of the director nominees named in Proposal No. 1 requires the affirmative vote of a plurality of the shares of the Company's common stock present in person or represented by proxy at the Annual Meeting and entitled to vote. The 6 candidates receiving the highest number of affirmative votes of the shares entitled to be voted will be elected directors. Shares represented by executed WHITE Proxy Cards will be voted, if authority to do so is not withheld, FOR the election of the director nominees named in Proposal No. 1. Votes may be cast in favor of or withheld with respect to all of the director nominees, or any of them. Broker non-votes, if any, will not be counted as having been voted and will have no effect on the outcome of the vote on the election of directors. Stockholders may not cumulate votes in the election of directors.

Ratification of the selection of KPMG LLP as the independent registered public accounting firm for the year ending December 31, 2006, as specified in Proposal No. 2, approval of the 2006 Share Incentive Plan, as specified in Proposal No. 3, and approval of the Amended and Restated 2006 Employee Stock Purchase Plan, as specified in Proposal No. 4, requires the affirmative vote of a majority of the shares of the Company's common stock present or represented by proxy at the Annual Meeting and entitled to vote. Abstentions will be counted toward the tabulation of votes cast on these proposals and will have the same effect as votes against these proposals. Broker non-votes, if any, will have no effect on the vote for these proposals.

Submission of Stockholder Proposals for the 2007 Annual Meeting

The Company's Bylaws provide a formal procedure for bringing business before an annual meeting of stockholders. Stockholders who intend to present a proposal at the 2007 Annual Meeting of Stockholders (2007 Annual Meeting) must deliver or mail a notice to the Secretary of the Company, and the notice must be received at the Company's executive offices at 105 Digital Drive, Novato, California, 94949 no earlier than February 28, 2007 and no later than March 30, 2007. In the event that the 2007 Annual Meeting is called for a date that is not within 30 days before or after June 21, 2007, then the stockholder's notice must be received by the Secretary no later than the close of business on the 10th day following the day on which notice of the date of the 2007 Annual Meeting was mailed or public announcement of such date was made by the Company, whichever first occurs. With respect to the 2006 Annual Meeting, a stockholder's notice pursuant to the Company's Bylaws had to be delivered to and received by the Company by March 20, 2006.

The notice must contain a brief description of the business desired to be brought before the 2007 Annual Meeting, the reasons for conducting such business, the name and address of the stockholder, the class or series and number of shares of the Company's capital stock which are owned beneficially or of record by the stockholder, a description of all arrangements or understandings between such stockholder and any other person in connection with the proposal of such business and any material interest of the stockholder in such business and a representation that the stockholder intends to appear in person or by proxy at the 2007 Annual Meeting to bring such business before the 2007 Annual Meeting.

Stockholders interested in submitting a proposal for inclusion in the proxy materials for the 2007 Annual Meeting may do so by following the procedures prescribed in Rule 14a-8 under the Securities Exchange Act of 1934, as amended (the Exchange Act). To be eligible for inclusion, stockholder proposals must be received by the Secretary of the Company no later than February 21, 2007, or at least 60 days before the Company begins to print and mail its proxy materials if the 2007 Annual Meeting is changed by more than 30 days from the

anniversary date of the 2006 Annual Meeting. Proposals should be sent to the Secretary of the Company at the Company's executive offices at 105 Digital Drive, Novato, California, 94949.

YOUR VOTE IS EXTREMELY IMPORTANT, NO MATTER HOW MANY OR HOW FEW SHARES YOU OWN. PLEASE SIGN AND DATE THE ENCLOSED WHITE PROXY CARD AND RETURN IT TODAY IN THE ENCLOSED PRE-ADDRESSED POSTAGE-PAID ENVELOPE.

IMPORTANT: If your shares are held in the name of a brokerage firm, bank, nominee or other institution, only it can sign a WHITE Proxy Card with respect to your shares and only upon specific instructions from you. Please return the enclosed WHITE Proxy Card to your broker or bank and contact the person responsible for your account to ensure that a WHITE Proxy Card is voted on your behalf.

If you have any questions or need assistance in voting your shares, please call the firm assisting the Company in the solicitation of proxies:

Morrow & Co., Inc.

445 Park Avenue 5th Floor

New York, NY 10022

1-800-607-0088

PROPOSAL ONE: ELECTION OF DIRECTORS

The Company has a Board currently consisting of 7 Directors. On May 2, 2006, pursuant to the Company's Bylaws, the Board fixed the number of Directors at 6, provided that all 7 current Directors will continue to serve until the Annual Meeting. At the Annual Meeting, stockholders will elect 6 directors. All Directors serve until their successors are duly appointed at the next annual meeting, or until they resign or are otherwise removed. Unless otherwise instructed, the proxy holders will vote the proxies represented by the enclosed WHITE Proxy Card FOR the 6 nominees named below, all of whom are presently Directors of the Company. If any of the Board's nominees are unable or decline to serve as Director, the proxies will be voted for any substitute nominee who shall be designated by the Board. It is not expected that any of the Board's nominees will be unable to or will decline to serve as Director. Mr. Franz Cristiani served as a Director during all of 2005, but will not stand for re-election in 2006.

If a quorum is present, the 6 nominees receiving the highest number of affirmative votes of the votes cast shall be elected as directors.

Nominees For Director

Set forth below are the Board's nominees to the Board and certain information regarding each nominee:

| Name | Age | Position with BioMarin | Director Since |
|------------------------------|------------|-----------------------------------|-----------------------|
| Jean-Jacques Bienaimé | 52 | Director, Chief Executive Officer | May 2005 |
| Michael Grey(1)(2) | 53 | Director | December 2005 |
| Elaine J. Heron, Ph.D.(2)(3) | 58 | Director | July 2002 |
| Joseph Klein, III(2)(3) | 45 | Director | July 2005 |
| Pierre Lapalme(1) | 65 | Director, Chairman of the Board | January 2004 |
| Alan Lewis, Ph.D.(1)(3) | 60 | Director | June 2005 |

- (1) Member of BioMarin's Compensation Committee
(2) Member of BioMarin's Audit Committee
(3) Member of BioMarin's Corporate Governance and Nominating Committee

There is no family relationship between any Director and any executive officer of BioMarin.

Jean-Jacques Bienaimé joined the BioMarin Board in May 2005, at the same time he became BioMarin's Chief Executive Officer. Prior to joining BioMarin, Mr. Bienaimé served as Chief Executive Officer and President of Genencor International, a biotechnology company, from November 2002 to May 2005 and its Chairman from April 2003 to May 2005. Prior to joining Genencor, Mr. Bienaimé was Chairman, President and Chief Executive Officer of SangStat Medical Corporation, another biotechnology company. He became President of SangStat Medical Corporation in 1998 and Chief Executive Officer in 1999. Prior to joining SangStat Medical Corporation, Mr. Bienaimé held various management positions from 1992 to 1998, including Senior Vice President of Corporate Marketing and Business Development, and Vice President and General Manager of the advanced therapeutic and oncology division, with Rhône-Poulenc Rorer Pharmaceuticals (now known as Sanofi/Aventis). Mr. Bienaimé currently serves on the boards of NeurogesX, Inc., Saegis Pharmaceuticals, Inc., and Ensemble Discovery and is a member of the advisory board of Bellevue Asset Management's BioVentures II fund. He received an M.B.A. from the Wharton School at the University of Pennsylvania and an undergraduate degree in economics from the Ecole Supérieure de Commerce de Paris.

Michael Grey joined the BioMarin Board in December 2005. Mr. Grey has served as President and Chief Executive Officer of SGX Pharmaceuticals, Inc., a public biotechnology company, where he previously served as President from June 2003 to January 2005 and as Chief Business Officer from April 2001 until June 2003. Between January 1999 and September 2001, he served as President and Chief Executive Officer of Trega

Biosciences, Inc., a biotechnology company. Prior to joining Trega, Mr. Grey served as President of BioChem Therapeutics, Inc., a division of BioChem Pharma, Inc., a pharmaceutical company, from November 1994 to August 1998. During 1994, Mr. Grey served as President and Chief Operating Officer of Ansan, Inc., a biopharmaceutical company. From 1974 to 1993, Mr. Grey served in various roles with Glaxo, Inc. and Glaxo Holdings, plc, a pharmaceutical company, culminating in his position as Vice President, Corporate Development. Mr. Grey is as a member of the board of directors of IDM Pharma, Inc. and Achillion Pharmaceuticals, Inc. and SGX Pharmaceuticals. Mr. Grey's appointment to the BioMarin Board was made pursuant to Section 1(b) of the Agreement between BioMarin and Caduceus Capital Master Fund Limited, Caduceus Capital II, L.P., OrbiMed Advisers LLC and OrbiMed Capital LLC.

Elaine J. Heron, Ph.D. joined the BioMarin Board in July 2002 and serves as the chairman of its Corporate Governance and Nominating Committee. Dr. Heron has served as Chairman and Chief Executive Officer of Labcyte Inc. since July 2001. Prior to that time, she spent six years in increasingly responsible positions at the Applied Biosystems Group of Applied Biosystems Corporation, including as General Manager and Vice President of Sales and Marketing. She was Vice President, Marketing, at Affymetrix, Inc., from 1995 to 1996. Dr. Heron has a B.S. in chemistry with Highest Distinction and a Ph.D. in analytical biochemistry, both from Purdue University, and an M.B.A. from Pepperdine University.

Joseph Klein, III joined the BioMarin Board in June 2005 and serves as the chairman of its Audit Committee. Mr. Klein is currently Managing Director of Gauss Capital Advisors, LLC, a financial consulting and investment advisory firm focused on biopharmaceuticals, which he founded in March 1998. In addition to his investment advisory and financial consulting activities, Mr. Klein currently serves as a Venture Partner of Red Abbey Venture Partners, LLC, the general partner of several life science investment partnerships. From September 2001 to September 2002, Mr. Klein was a Venture Partner of MPM Capital, a healthcare venture capital firm. From June 1999 to September 2000 when it merged with WebMD Corporation, Mr. Klein served as Vice President, Strategy for Medical Manager Corporation, a leading developer of physician office management information systems. Mr. Klein serves on the board of directors of three other publicly held biotechnology companies: Clinical Data, Inc., ISIS Pharmaceuticals and NPS Pharmaceuticals, Inc.; as well as two private health care companies: Keel Pharmaceuticals, Inc. and the Maryland Medical Research Institute. Mr. Klein received a B.A. *summa cum laude* in economics from Yale University, where he was elected to Phi Beta Kappa, and an M.B.A. from the Stanford Graduate School of Business. Mr. Klein's appointment to the BioMarin Board was made pursuant to Section 1(b) of the Agreement between BioMarin and Caduceus Capital Master Fund Limited, Caduceus Capital II, L.P., OrbiMed Advisers LLC and OrbiMed Capital LLC.

Pierre Lapalme joined the BioMarin Board in January 2004 and was named as Chairman in August 2004. Throughout his career, Mr. Lapalme has held numerous senior management positions in the pharmaceutical industry, including Chief Executive Officer and Chairman of the Board of Rhône-Poulenc Pharmaceuticals, Inc. in Canada, and Senior Vice President and General Manager of North America Ethical, a division of Rhône-Poulenc Rorer, Inc., where he oversaw the development of the ethical pharmaceutical business in the United States, Canada, Mexico, and Central America. Mr. Lapalme served on the board of the National Pharmaceutical Council and was a board member of the Pharmaceutical Manufacturers Association of Canada, where he played a leading role in reinstating patent protection for pharmaceuticals. Mr. Lapalme currently serves as director of First Horizon Pharmaceuticals, Bioxel Pharma Inc. and Wex Pharmaceuticals Inc. Mr. Lapalme was Chairman of Diagnostics Inc. from 1995 to 2005. He remains as special advisor to the President. Mr. Lapalme studied at the University of Western Ontario and INSEAD France.

Alan J. Lewis, Ph.D. joined the BioMarin Board in June 2005 and serves as the chairman of its Compensation Committee. Since February 2006, Dr. Lewis has been the President and CEO of Novocell, Inc., a privately held regenerative disease biotechnology company. Prior to this, starting in 2000, he was President of Celgene Signal Research, a wholly owned subsidiary of the Celgene Corporation, an integrated pharmaceutical company. Previously, he was the President and Chief Executive Officer of Signal Research, a company that focused on the discovery and development of drugs that regulate genes associated with diseases. Previously, he

was the President and Chief Executive Officer of Signal Pharmaceuticals, Inc., where he guided the company to its successful acquisition by Celgene. Before joining Signal Pharmaceuticals, Dr. Lewis held a number of positions at Wyeth-Ayerst Research and its predecessor Wyeth Laboratories, Inc. from 1979 to 1994, including Vice President of Research at Wyeth-Ayerst. Dr. Lewis currently serves as a director of Discovery Partners International. Dr. Lewis has published over 120 full manuscripts and has written and edited seven books and 100 abstracts. Dr. Lewis was a Research Associate at Yale University from 1972 to 1973. Dr. Lewis received a B.Sc. in Physiology and Biochemistry from Southampton University, Southampton, Hampshire, U.K. and a Ph.D. in Pharmacology from the University of Wales, Cardiff, U.K. Dr. Lewis appointment to the BioMarin Board was made pursuant to Section 1(b) of the Agreement between BioMarin and Caduceus Capital Master Fund Limited, Caduceus Capital II, L.P., OrbiMed Advisers LLC and OrbiMed Capital LLC.

Board Meetings and Board Committees

The Board manages the business of the Company. It establishes overall policies and standards for the Company and reviews the performance of management. In addition, the Board has established an Audit Committee, a Compensation Committee, and a Corporate Governance and Nominating Committee whose functions are briefly described below.

The Board held a total of 14 meetings during the year ended December 31, 2005 and took action by unanimous written consent on 3 occasions. During 2005, no Director participated in fewer than 75% of all such meetings of the Board and the committees upon which Directors served.

Audit Committee. The Audit Committee provides oversight of (i) the financial reporting process, the system of internal financial controls and the audit process of BioMarin and (ii) BioMarin's independent registered public accounting firm. The Audit Committee also recommends to the Board the appointment of the independent registered public accounting firm for BioMarin. The members of the Audit Committee are Mr. Klein, Chairman, Dr. Heron, and Mr. Grey. Each of the members of the Audit Committee is an audit committee financial expert as defined by Nasdaq and Securities and Exchange Commission rules and, as required by the Audit Charter, is an independent director. The committee charter is available in the investors section of the Company's website at www.bmrn.com. Information on the Company's website is not incorporated by reference in this proxy statement. During 2005, the Audit Committee met on 9 occasions.

Compensation Committee. The Compensation Committee sets general compensation policy for BioMarin and recommends compensation of executive officers for approval by the full Board. The Compensation Committee also has final approval power over guidelines and criteria for officers' bonuses and administers BioMarin's 1997 Stock Plan and 1998 Director Option Plan. The members of the Compensation Committee are Dr. Lewis, Chairman, and Messrs. Lapalme and Grey. Each of the members of the compensation committee are independent directors as defined by Nasdaq and Securities and Exchange Commission rules. The committee charter is available in the investors section of the Company's website at www.bmrn.com. Information on the Company's website is not incorporated by reference in this proxy statement. During 2005, the Compensation Committee met on 4 occasions.

Corporate Governance and Nominating Committee. The Corporate Governance and Nominating Committee (the Committee) is responsible for overseeing the selection of qualified candidates to serve as members of the Board of Directors and guiding the corporate governance philosophy and practices of the Company. To that end, the Committee is responsible for identifying individuals to fill vacancies on the Board, recommending nominees to be voted upon at the annual meeting of stockholders, recommending to the Board appointees to serve on committees of the Board and overseeing the development and implementation of BioMarin's corporate governance policies and code of ethics. A detailed discussion of Committee procedures appears under the caption *Procedure of the Corporate Governance and Nominating Committee*.

The Committee also oversees policies including, but not limited to:

adoption of corporate governance guidelines;

establishing written charters for each Board committee;

maintaining independence standards for each Board member;

requiring sessions of Board meetings without management present;

mandating execution of a standard of business ethics for every employee and Board member;

implementing, in conjunction with the Audit Committee, the independent audit function, in order to oversee the Company's financial processes and internal controls report to the Audit Committee on a regular basis; and

establishing a toll-free telephone number for employees to anonymously report complaints relating to financial fraud, environmental hazards, illegal or unfair employment practices and unethical behavior.

The members of the Committee are Dr. Heron, Chair, Dr. Lewis and Mr. Klein. The committee charter and the Corporate Governance Guidelines are available in the investors section of the Company's website at www.bmrn.com. Information on the Company's website is not incorporated by reference in this proxy statement. The Committee met on 5 occasions during 2005.

Compensation Committee Interlocks and Insider Participation

During all of 2005, and through the date of this report, the Compensation Committee consisted of three of the Company's independent Directors. From January 2005 through June 2005, the members of the Committee were Dr. Urquhart (Chair) and Messrs. Sager and Lapalme. At the end of June, 2005, following the Annual Meeting of Stockholders, the members of the Committee were Mr. Sager (Chair), Mr. Lapalme and Dr. Lewis. Effective March 1, 2006, the Compensation Committee consists of Dr. Lewis (Chair) and Messrs. Grey and Lapalme. None of the members of the Compensation Committee: (i) was an officer or employee of the Company or any of its subsidiaries during the last or current fiscal year; (ii) was formerly an officer of the Company or any of its subsidiaries; or (iii) had any relationships requiring disclosure by the Company under the SEC's rules requiring disclosure of certain relationships and related party transactions. None of the Company's executive officers serves as a member of the board of directors or compensation committee of any entity that has one or more executive officers serving on the Board or Compensation Committee.

2005 Director Compensation

The following table is a summary of the compensation paid to the Directors. Each applicable line item is an additional element of compensation.

| Director Position | Annual Compensation |
|-------------------------|--|
| All Independent Members | \$25,000 |
| | Options to purchase 30,000 shares of the |
| | Company's common stock |
| Chairman of the Board | \$30,000 |
| Audit Committee Chair | \$15,000 |

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| | |
|---|----------|
| Audit Committee (Non-Chair) | \$7,500 |
| Compensation Committee Chair | \$10,000 |
| Compensation Committee (Non-Chair) | \$5,000 |
| Corporate Governance and Nominating Committee Chair | \$10,000 |
| Corporate Governance and Nominating Committee (Non-Chair) | \$5,000 |
| Science Committee Chair(1) | \$10,000 |
| Science Committee (Non-Chair)(1) | \$5,000 |

(1) The Science Committee was disbanded in June 2005.

Directors are also reimbursed for their reasonable expenses in attending meetings of the Board and while performing services for BioMarin.

2005 Director Compensation

The following table lists actual compensation paid to each of the Directors during 2005.

| Director Name | Director Fees | Option Grant Date | Number of Options Granted | Exercise Price Per Share | Option Expiry |
|-------------------------|---------------|-------------------|---------------------------|--------------------------|---------------|
| Franz L. Cristiani | \$ 45,000 | 6/14/2005 | 30,000 | \$ 6.50 | 6/13/2015 |
| Michael Grey(1) | \$ | 12/14/2005 | 30,000 | \$ 10.33 | 12/13/2015 |
| Elaine J. Heron, Ph.D. | \$ 38,750 | 7/12/2005 | 30,000 | \$ 7.46 | 7/11/2015 |
| Joseph Klein, III(2) | \$ 9,375 | 6/28/2005 | 30,000 | \$ 7.72 | 6/28/2015 |
| Pierre Lapalme | \$ 63,750 | 1/27/2005 | 30,000 | \$ 5.92 | 1/26/2015 |
| Alan J. Lewis, Ph.D.(2) | \$ 8,750 | 6/28/2005 | 30,000 | \$ 7.72 | 6/28/2015 |
| Erich Sager(3) | \$ 31,250 | 11/30/05 | 30,000 | \$ 9.74 | 11/30/2015 |
| John Urquhart, M.D.(4) | \$ 40,750 | | | \$ | |
| Gwynn R. Williams(4) | \$ 31,875 | | | \$ | |

- (1) Mr. Grey was appointed to the Board on December 14, 2005.
- (2) Mr. Klein and Dr. Lewis were elected to the Board on June 28, 2005.
- (3) Mr. Sager resigned from the Board on February 28, 2006.
- (4) Pursuant to an agreement between BioMarin and Caduceus Capital Master Fund Limited, Caduceus Capital II, L.P., OrbiMed Advisors LLC and OrbiMed Capital LLC, Dr. Urquhart and Mr. Williams did not stand for reelection at BioMarin's 2005 Annual Meeting of Stockholders.

1998 Director Option Plan

The 1998 Director Option Plan was adopted by the Board in December 1999. It was approved by BioMarin's stockholders as of January 15, 2000. The 1998 Director Option Plan provides for the grant of nonstatutory stock options to non-employee Directors. A total of 1,489,650 shares of BioMarin common stock have been reserved for issuance under the 1998 Director Option Plan. The 1998 Director Option Plan also provides for an annual increase in this number of shares equal to the lesser of: (i) 0.5% of BioMarin's outstanding common stock; (ii) 200,000 shares; or (iii) a lesser amount determined by the Board.

In fiscal year 2005, options to purchase, in the aggregate, 210,000 shares were issued to Directors under the 1998 Director Option Plan.

The 1998 Director Option Plan, as amended in 2003 and May 2004, provides that each non-employee Director shall automatically be granted an initial option and subsequent annual options to purchase 30,000 shares of BioMarin common stock on the date that such person first becomes a non-employee Director and on each anniversary of the date of the non-employee Director's appointment to the Board. These options have a term of 10 years. The shares subject to these options vest quarterly over one year. These options continue to vest only while the Director serves. The exercise price per share of each of these options is 100% of the fair market value of a share of BioMarin's common stock at the date of the grant of the option.

In the event of a merger or the sale of substantially all of the assets of BioMarin, each option issued under the 1998 Director Option Plan may be assumed or substituted by the successor corporation. If an option is assumed or substituted, it shall continue to vest as provided in the 1998 Director Option Plan. However, if a non-employee Director's status as a Director of BioMarin or the successor corporation, as applicable, is terminated, other than upon a voluntary resignation by the non-employee Director, the option immediately becomes fully vested and exercisable. If the successor corporation does not agree to assume or substitute options, each option becomes fully vested and exercisable for a period of 30 days from the date the Board notifies the optionee of the option's full exercisability, after which period the option terminates.

Options granted under the 1998 Director Option Plan are required to be exercised within three months of the end of the optionee's tenure as a Director, or within 12 months after termination by death or disability, or within the original option expiration if the Director retires from the Board after having served 4.5 years, but in no event later than the expiration of the option's ten-year term. No option granted under the 1998 Director Option Plan is transferable by the optionee other than by will or the laws of descent or distribution. Each option is exercisable, during the lifetime of the optionee, only by the optionee. Unless sooner terminated by the Board, the 1998 Director Option Plan will terminate automatically 10 years from the effective date of the 1998 Director Option Plan.

THE BOARD OF DIRECTORS UNANIMOUSLY RECOMMENDS THAT THE STOCKHOLDERS VOTE FOR THE ELECTION OF ALL NOMINEES NAMED ABOVE. IF YOU SIGN AND RETURN THE ENCLOSED WHITE PROXY CARD, UNLESS YOU DIRECT TO THE CONTRARY ON THAT CARD, THE SHARES REPRESENTED BY THAT PROXY CARD WILL BE VOTED FOR THE ELECTION OF ALL NOMINEES LISTED ABOVE.

PROCEDURES OF THE CORPORATE GOVERNANCE AND NOMINATING COMMITTEE

The Corporate Governance and Nominating Committee of the Board of Directors (the Committee) is responsible for overseeing the selection of qualified candidates to serve as members of the Board of Directors and guiding the corporate governance philosophy and practices of the Company. The Committee is composed of three Directors each of whom is independent under the listing qualifications of the Nasdaq National Market. The Committee operates according to a charter that complies with the guidelines established by Nasdaq.

The Committee has not adopted formal procedures in selecting individuals to serve as members of the Board of Directors. Instead, it utilizes general guidelines that allow it to adjust the process to best satisfy the objectives it is attempting to accomplish in any director search. The first step in the general process is to identify the type of candidate the Committee may desire for a particular opening. This may involve locating candidates with specific backgrounds, skill sets or experiences. Once identified, the Committee looks to the best method of finding a candidate who satisfies the specified criteria. The Committee may consider candidates recommended by management, by other members of the Committee or the Board of Directors, by stockholders or it may engage a third party to conduct a search for possible candidates. In considering candidates submitted by stockholders, the Committee will take into consideration the needs of the Board and the qualifications of the candidate. To have a candidate considered by the Committee, a stockholder should submit a written recommendation that includes (i) the name of the stockholder and evidence of the stockholder's ownership of Company stock, including the number of shares owned and the length of time of ownership, and (ii) the name of the candidate, the candidate's resume or a listing of his or her qualifications to be a director of the Company and the person's consent to be named as a director if selected by the Committee and nominated by the Board. Stockholder recommendations should be addressed to the Corporate Governance and Nominating Committee in care of the Secretary of the Company at the address set forth under the heading *Stockholder Communications with the Board of Directors*.

Once candidates are identified, the Committee conducts an evaluation of the candidate. The evaluation generally includes interviews and background and reference checks. There is no difference in the evaluation process of a candidate recommended by a stockholder as compared to the evaluation process of a candidate identified by any of the other means described above. While the Committee has not established minimum criteria for a candidate, it has established important factors to consider in evaluating a candidate. These factors include: strength of character, mature judgment, business experience, availability and level of interest, attendance, career specialization, relevant technical skills, diversity and the extent to which the candidate would fill a present need on the Board of Directors.

If the Committee determines that a candidate should be nominated as a candidate for election to the Board, the candidate's nomination is then recommended to the Board of Directors, and the Directors may in turn conduct their own review to the extent it deems appropriate. When the Board of Directors has agreed upon a candidate, such candidate is recommended to the stockholders for election at an Annual Meeting of stockholders.

All of the current Directors, other than Mr. Cristiani, have been recommended by the Committee to the Board of Directors for election as Directors of the Company at the Annual Meeting, and the Board has approved such recommendations.

In 2005, the independent Board members appointed Mr. Lapalme as Chairman of the Board. The Chairman of the Board is responsible for:

approving Board meeting schedules and meeting agendas;

approving Board meeting materials;

serving as the liaison between the then executive-Chairman and the independent Board members;

leading executive sessions of the independent Board members;

setting meetings of independent Board members; and

being available for consultation with major stockholders.

STOCKHOLDER COMMUNICATIONS WITH THE BOARD OF DIRECTORS

The Board has adopted a process for stockholders and others to send communications to the Board or any Director. All such communications should be sent by mail to 105 Digital Drive, Novato, CA 94949, c/o G. Eric Davis, the Company's Vice President, General Counsel and Secretary.

It is the Company's policy to request that all Board members attend the Annual Meeting of Stockholders. However, the Company also recognizes that personal attendance by all Directors is not always possible. All of the Directors serving at the time of the 2005 Annual Meeting of Stockholders attended such meeting, except for Dr. Urquhart and Mr. Williams, who were not standing for reelection.

STANDARDS OF BUSINESS CONDUCT AND ETHICS

The Board has adopted Standards of Business Conduct and Ethics that are applicable to all employees and Directors, including the Company's Chief Executive Officer, Chief Financial Officer, other executive officers and senior financial personnel. A copy of the Company's Standards of Business Conduct and Ethics is available on the Company's website at www.bmrn.com. Information on the Company's website is not incorporated by reference in this proxy statement. The Company intends to post any waiver of, or material changes to, these Standards, if any, to its website within five business days of such event.

PROPOSAL TWO: RATIFICATION OF THE INDEPENDENT REGISTERED PUBLIC ACCOUNTING FIRM FOR BIOMARIN

The Audit Committee of the Board has selected KPMG LLP, an independent registered public accounting firm, to audit the financial statements of the Company for the year ending December 31, 2006, and recommends that stockholders vote for ratification of such appointment. Although stockholder ratification is not required by the Company's Bylaws or otherwise, the Board has determined that it is desirable to request approval of this selection by the stockholders as a matter of good corporate practice. Notwithstanding this selection, the Audit Committee, in its discretion, may direct the appointment of a new independent registered public accounting firm at any time during the year if the Audit Committee feels that such a change would be in the best interests of the Company and its stockholders. In the event of a negative vote on ratification, the Audit Committee may reconsider its selection.

Independent Registered Public Accounting Firm

Since June 11, 2002, KPMG LLP served as the Company's independent registered public accounting firm.

Representatives of KPMG LLP plan to attend the Annual Meeting and will be available to answer appropriate questions from stockholders and, although they do not expect to do so, they will have the opportunity to make a statement if they so desire.

The following is a summary of the fees and services provided for the fiscal years 2005 and 2004.

| <i>Description of Services Provided by KPMG LLP</i> | <i>Year Ended December 31, 2005</i> | <i>Year Ended December 31, 2004</i> |
|---|---|---|
| <i>Audit Fees</i> | \$ 607,000 | \$ 777,137 |
| <i>Audit Related Fees:</i> These services primarily involved review and assurance services associated with certain regulatory filings (for fees incurred in 2005) the Company's debt and equity offerings and related regulatory filings (for fees incurred in 2004). | \$ 54,800 | \$ 23,401 |
| <i>Tax Compliance Fees:</i> These services related to the preparation of federal, state and foreign tax returns and other filings. | none | \$ 51,246 |
| <i>Tax Consulting and Advisory Services:</i> These services primarily relate to the area of tax strategy and minimizing Federal, state, local and foreign taxes. | none | none |
| <i>All Other Fees</i> | none | none |

As provided in the Audit Committee charter, the Audit Committee pre-approves all services provided by its independent registered public accounting firm. All of the above services and estimates of the expected fees were reviewed and approved by the Audit Committee before the respective services were rendered.

The Audit Committee has considered the nature and amount of the fees billed by KPMG LLP and believes that the provision of the services for activities unrelated to the audit is compatible with maintaining KPMG LLP's independence.

THE BOARD OF DIRECTORS UNANIMOUSLY RECOMMENDS THAT THE STOCKHOLDERS VOTE FOR THE RATIFICATION OF KPMG LLP AS THE COMPANY'S INDEPENDENT REGISTERED PUBLIC ACCOUNTING FIRM. IF YOU SIGN AND RETURN THE ENCLOSED WHITE PROXY CARD, UNLESS YOU DIRECT TO THE CONTRARY ON THAT CARD, THE SHARES REPRESENTED BY THAT PROXY CARD WILL BE VOTED FOR THE RATIFICATION OF KPMG LLP AS THE COMPANY'S INDEPENDENT REGISTERED PUBLIC ACCOUNTING FIRM.

PROPOSAL THREE: APPROVAL OF THE 2006 SHARE INCENTIVE PLAN

On May 2, 2006, the Board of Directors of the Company adopted, subject to Stockholder approval at this Annual Meeting, the BioMarin Pharmaceutical Inc. 2006 Share Incentive Plan (the "SIP"). Below is a summary of the principal provisions of the SIP and its operation. A copy of the SIP is set forth in full in *Appendix A* to this proxy statement, and the following description of the SIP is qualified in its entirety by reference to *Appendix A*.

Purpose of Proposal

The SIP is designed to provide an incentive to eligible employees, consultants, Directors and advisors of the Company and its affiliates, to encourage proprietary interest in the Company by such persons, to encourage such persons to remain in the service of the Company and its affiliates and to attract new employees and Directors with outstanding qualifications. The Company is proposing approval of the SIP because the Company's 1997 Stock Plan (the "1997 Plan") will expire in April 2008 and the Company's 1998 Director Option Plan (the "1998 Plan") will expire in December 2009. It is necessary for the Company to continue to grant stock incentive awards to employees, consultants, Directors and advisors as part of their compensation to provide appropriate incentives. If the SIP is approved by the stockholders, no new grants will be made under either the 1997 Plan or the 1998 Plan. Further, recent regulatory changes, including the adoption of Section 409A under the Internal Revenue Code of 1986, may impact certain provisions of the 1997 Plan. The Company believes that the SIP has been drafted in a manner fully compliant and consistent with such recent regulatory developments.

Description of the 2006 Share Incentive Plan

The following summary is not intended to be complete and reference should be made to *Appendix A* for a complete statement of the terms and provisions of the SIP. Capitalized terms used in this summary and not otherwise defined will have the meanings ascribed to such terms in the SIP.

Purpose. The purpose of the SIP is to attract, retain and motivate select employees, officers, directors, advisors and consultants of the Company and its affiliates (referred to collectively as "Eligible Persons") and to provide incentives and rewards for superior performance.

Shares Subject to the SIP. The SIP provides that no more than 15,000,000 shares of Common Stock may be issued pursuant to Awards under the SIP. If the SIP is approved by the stockholders, the Company shall not make additional awards under the 1997 Plan or the 1998 Plan. These shares shall be authorized but unissued shares. The number of shares available for Awards, as well as the terms of outstanding Awards, are subject to adjustment as provided in the SIP for stock splits, stock dividends, recapitalizations and other similar events.

Shares of Common Stock that are subject to any Award that expires, or is forfeited, cancelled or becomes unexercisable will again be available for subsequent Awards, except as prohibited by law. In addition, shares that the Company refrains from delivering pursuant to an Award as payment of either the exercise price of an Award or applicable withholding and employment taxes will be available for subsequent Awards.

Administration. Either the Board of Directors or a committee appointed by the Board will administer the SIP. The Board of Directors and any committee exercising discretion under the SIP from time to time are referred to as the "Committee." Unless otherwise provided by the Board, the Compensation Committee will serve as the administrator of the SIP. The Board of Directors may at any time appoint additional members to the Committee, remove and replace members of the Committee with or without cause, and fill vacancies on the Committee. To the extent permitted by law, the Committee may authorize one or more persons who are reporting persons for purposes of Rule 16b-3 under the Securities Exchange Act of 1934, as amended (or other officers), to make Awards to eligible persons who are not reporting persons for purposes of Rule 16b-3 under the Securities Exchange Act of 1934, as amended (or other officers whom the Company has specifically authorized to make Awards). With respect to decisions involving an Award intended to satisfy the requirements of Section 162(m) of

the Code, the Committee is to consist of two or more directors who are outside directors for purposes of that Code section. The Committee may delegate administrative functions to individuals who are reporting persons for purposes of Rule 16b-3 of the Exchange Act, officers or employees of the Company or its affiliates.

Subject to the terms of the SIP, the Committee has express authority to determine the Eligible Persons who will receive Awards, the number of shares of Common Stock or units to be covered by each Award, and the terms and conditions of Awards. The Committee has broad discretion to prescribe, amend and rescind rules relating to the SIP and its administration, to interpret and construe the SIP and the terms of all Award agreements, and to take all actions necessary or advisable to administer the SIP. Within the limits of the SIP, the Committee may accelerate the vesting of any Award, allow the exercise of unvested Awards, and may modify, replace, cancel or renew them.

The SIP provides that the Company and its affiliates will indemnify members of the Committee and their delegates against any claims, liabilities or costs arising from the good faith performance of their duties under the SIP. The SIP releases these individuals from liability for good faith actions associated with the SIP's administration.

Eligibility. The Committee may grant options that are intended to qualify as incentive stock options (ISOs) only to employees, and may grant all other Awards to Eligible Persons. The SIP and the discussion below use the term Participant to refer to an Eligible Person who has received an Award.

Options. Options granted under the SIP provide Participants with the right to purchase shares of Common Stock at a predetermined exercise price. The Committee may grant options that are intended to qualify as ISOs or options that are not intended to so qualify (Non-ISOs). The SIP also provides that ISO treatment may not be available for options that become first exercisable in any calendar year to the extent the value of the underlying shares that are the subject of the option exceed \$100,000 (based upon the fair market value of the shares of Common Stock on the option grant date).

Exercise Price for Options. The exercise price of Options may not be less than 100% of the fair market value on the grant date of the shares of Common Stock subject to the Award. The exercise price of ISOs may not be less than 110% of the fair market value on the grant date of the underlying shares of Common Stock subject to the Award for Participants who own more than ten percent of the Company's shares of Common Stock on the grant date. Neither the Company nor the Committee shall, without shareholder approval, allow for a repricing within the meaning of the federal securities laws applicable to proxy statement disclosures.

Exercise of Options. To the extent exercisable in accordance with the agreement granting them, an Option may be exercised in whole or in part, and from time to time during its term, subject to earlier termination relating to a holder's termination of employment or service. With respect to Options, the Committee has the discretion to accept payment of the exercise price in any of the following forms (or combination of them): cash or check in U.S. dollars, certain shares of Common Stock, and cashless exercise under a program the Committee approves.

The term over which Participants may exercise Options may not exceed ten years from the date of grant (five years in the case of ISOs granted to employees who, at the time of grant, own more than 10% of the Company's outstanding shares of Common Stock).

Prohibition on Repricing. The SIP provides that the exercise price of an Award may not be reduced, whether directly or indirectly, such as through a surrender and regrant, unless such repricing is approved by the stockholders.

Restricted Shares, Restricted Share Units, Unrestricted Shares and Deferred Share Units. Under the SIP, the Committee may grant (i) restricted shares that are forfeitable until certain vesting requirements are met, (ii) restricted share units which represent the right to receive shares of Common Stock after certain vesting

requirements are met, and (iii) unrestricted shares as to which the Participant's interest is immediately vested. For restricted Awards, the SIP provides the Committee with discretion to determine the terms and conditions under which a Participant's interests in such Awards become vested. The SIP provides for deferred share units in order to permit certain directors, consultants or select members of management to defer their receipt of compensation payable in cash or shares of Common Stock (including shares that would otherwise be issued upon the vesting of restricted shares and restricted share units). Deferred share units represent a future right to receive shares of Common Stock.

Whenever shares of Common Stock are released pursuant to these Awards, the Participant will be entitled to receive additional shares of Common Stock that reflect any stock dividends that the Company's stockholders received between the date of the Award and issuance or release of the shares of Common Stock. Likewise, a Participant will be entitled to receive a cash payment reflecting cash dividends paid to the Company's stockholders during the same period. Such cash dividends will accrue interest, at 5% per annum, from their payment date to the Company's stockholders until paid in cash when the shares of Common Stock to which they relate are either released from restrictions in the case of restricted shares or issued in the case of restricted share units.

Performance Awards. The SIP authorizes the Committee to grant performance-based awards in the form of Performance Units that the Committee may or may not designate as Performance Compensation Awards that are intended to be exempt from Code section 162(m) limitations. In either case, Performance Awards vest and become payable based upon the achievement, within the specified period of time, of performance objectives applicable to the individual, the Company or any affiliate. The Committee decides the length of performance periods, but the periods may not be less than one fiscal year of the Company.

With respect to Performance Compensation Awards, the SIP requires that the Committee specify in writing the performance period to which the Award relates, and an objective formula by which to measure whether and the extent to which the Award is earned on the basis of the level of performance achieved with respect to one or more performance measures. Once established for a performance period, the performance measures and performance formula applicable to the Award may not be amended or modified in a manner that would cause the compensation payable under the Award to fail to constitute performance-based compensation under Code Section 162(m).

Under the SIP, the possible performance measures for Performance Compensation Awards include basic, diluted or adjusted earnings per share; sales or revenue; earnings before interest, taxes and other adjustments (in total or on a per share basis); basic or adjusted net income; returns on equity, assets, capital, revenue or similar measure; economic value added; working capital; total stockholder return; and product development, product market share, research, licensing, litigation, human resources, information services, mergers, acquisitions, and sales of assets of affiliates or business units. Each measure will be, to the extent applicable, determined in accordance with generally accepted accounting principles as consistently applied by the Company (or such other standard applied by the Committee) and, if so determined by the Committee, and in the case of a Performance Compensation Award, to the extent permitted under Code section 162(m), adjusted to omit the effects of extraordinary items, gain or loss on the disposal of a business segment, unusual or infrequently occurring events and transactions and cumulative effects of changes in accounting principles. Performance measures may vary from performance period to performance period, and from Participant to Participant, and may be established on a stand-alone basis, in tandem or in the alternative.

Income Tax Withholding. As a condition for the issuance of shares of Common Stock pursuant to Awards, the SIP requires satisfaction of any applicable federal, state, local or foreign withholding tax obligations that may arise in connection with the Award or the issuance of shares of Common Stock.

Transferability. Awards may not be sold, pledged, assigned, hypothecated, transferred or disposed of other than by will or the laws of descent and distribution, except to the extent the Committee permits lifetime transfers to charitable institutions, certain family members or related trusts or as otherwise approved by the Committee.

Certain Corporate Transactions. The Committee shall equitably adjust the number of shares covered by each outstanding Award, and the number of shares that have been authorized for issuance under the SIP but as to which no Awards have yet been granted or that have been returned to the SIP upon cancellation, forfeiture or expiration of an Award, as well as the price per share covered by each such outstanding Award, to reflect any increase or decrease in the number of issued shares resulting from a stock split, reverse stock split, stock dividend, combination, recapitalization or reclassification of the shares of Common Stock, or any other increase or decrease in the number of issued shares effected without receipt of consideration by the Company. In the event of any such transaction or event, the Committee may provide in substitution for any or all outstanding Options under the SIP such alternative consideration (including securities of any surviving entity) as it may in good faith determine to be equitable under the circumstances and may require in connection therewith the surrender of all Options so replaced. In any case, such substitution of securities will not require the consent of any person who is granted options pursuant to the SIP.

In addition, in the event or in anticipation of a Change in Control, the Committee may at any time in its sole and absolute discretion and authority, without obtaining the approval or consent of the Company's stockholders or any Participant with respect to his or her outstanding Awards (except to the extent an Award provides otherwise), take one or more of the following actions: (a) arrange for or otherwise provide that each outstanding Award will be assumed or substituted with a substantially equivalent award by a successor corporation or a parent or subsidiary of such successor corporation; (b) accelerate the vesting of Awards for any period (and may provide for termination of unexercised Options at the end of that period) so that Awards shall vest (and, to the extent applicable, become exercisable) as to the shares of Common Stock that otherwise would have been unvested and provide that repurchase rights of the Company with respect to shares of Common Stock issued upon exercise of an Award shall lapse as to the shares of Common Stock subject to such repurchase right; (c) arrange or otherwise provide for payment of cash or other consideration to Participants in exchange for the satisfaction and cancellation of outstanding Awards; or (d) terminate upon the consummation of the transaction, provided that the Committee may in its sole discretion provide for vesting of all or some outstanding Awards in full as of a date immediately prior to consummation of the Change of Control. To the extent that an Award is not exercised prior to consummation of a transaction in which the Award is not being assumed or substituted, such Award shall terminate upon such consummation.

Notwithstanding the above, in the event a Participant holding an Award assumed or substituted by the successor corporation in a Change in Control is Involuntarily Terminated by the successor corporation in connection with, or within 12 months following consummation of, the Change in Control, then any assumed or substituted Award held by the terminated Participant at the time of termination shall accelerate and become fully vested (and exercisable in full in the case of Options), and any repurchase right applicable to any shares of Common Stock shall lapse in full. The acceleration of vesting and lapse of repurchase rights provided for in the previous sentence shall occur immediately prior to the effective date of the Participant's termination.

In the event of any distribution to the Company's stockholders of securities of any other entity or other assets (other than dividends payable in cash or stock of the Company) without receipt of consideration by the Company, the Committee may, in its discretion, appropriately adjust the price per share covered by each outstanding Award to reflect the effect of such distribution. Finally, if the Company dissolves or liquidates, all Awards will immediately terminate, subject to the ability of the Board to exercise any discretion that the Board may exercise in the case of a Change in Control.

Term of SIP; Amendments and Termination. The term of the SIP is ten years from the date of stockholder approval. The Board of Directors may from time to time, amend, alter, suspend, discontinue or terminate the SIP; provided that no amendment, suspension or termination of the SIP shall materially and adversely affect Awards already granted unless it relates to an adjustment pursuant to certain transactions that change the Company's capitalization or it is otherwise mutually agreed between the Participant and the Committee. In addition, the Committee may not cancel an outstanding option that is out-of-the-money for the purpose of reissuing the option to the participant at a lower exercise price or granting a replacement award of a different type. Notwithstanding

the foregoing, the Committee may amend the SIP to eliminate provisions which are no longer necessary as a result of changes in tax or securities laws or regulations, or in the interpretation thereof.

Termination, Rescission and Recapture. Each Award under the SIP is intended to align the Participant's long-term interest with those of the Company. If the Participant engages in certain activities (such as disclosure of confidential or proprietary information without Company authorization, or breach of certain agreements relating to the protection of the Company's intellectual property), either during employment or after employment with the Company terminates for any reason, the Participant is deemed to be acting contrary to the long-term interests of the Company. In such cases, except as otherwise expressly provided in the Award Agreement, the Company may terminate any outstanding, unexercised, unexpired, unpaid, or deferred Awards, rescind any exercise, payment or delivery pursuant to the Award, or recapture any Common Stock (whether restricted or unrestricted) or proceeds from the Participant's sale of Shares issued pursuant to the Award.

Expected U.S. Federal Income Tax Consequences. The following is a brief summary of certain tax consequences of certain transactions under the SIP. This summary is not intended to be complete and does not describe state or local tax consequences. Special rules may apply to the Company's officers, directors or greater than ten percent stockholders. Participants in the SIP should review the current tax treatment with their individual tax advisors at the time of grant, exercise or any other transaction relating to an Award or the underlying shares.

Under the United States Internal Revenue Code, the Company will generally be entitled to a deduction for federal income tax purposes at the same time and in the same amount as the ordinary income that Participants recognize pursuant to Awards (subject to the Participant's overall compensation being reasonable, and to the discussion below with respect to Code section 162(m)). For Participants, the expected U.S. federal income tax consequences of Awards are as follows:

Non-ISOs. A Participant will not recognize income at the time a Non-ISO is granted. At the time a Non-ISO is exercised, the Participant will recognize ordinary income in an amount equal to the excess of (a) the fair market value of the shares of Common Stock issued to the Participant on the exercise date over (b) the exercise price paid for the shares. At the time of sale of shares acquired pursuant to the exercise of a Non-ISO, the appreciation (or depreciation) in value of the shares after the date of exercise will be treated either as short-term or long-term capital gain (or loss) depending on how long the shares have been held.

ISOs. A Participant will not recognize income upon the grant of an ISO. There are generally no tax consequences to the Participant upon exercise of an ISO (except the amount by which the fair market value of the shares at the time of exercise exceeds the option exercise price is a tax preference item possibly giving rise to an alternative minimum tax). If the shares of Common Stock are not disposed of within two years from the date the ISO was granted or within one year after the ISO was exercised, any gain realized upon the subsequent disposition of the shares will be characterized as long-term capital gain and any loss will be characterized as long-term capital loss. If both of these holding period requirements are not met, then a disqualifying disposition occurs and (a) the Participant recognizes gain in the amount by which the fair market value of the shares at the time of exercise exceeded the exercise price for the ISO and (b) any remaining amount realized on disposition (except for certain wash sales, gifts or sales to related persons) will be characterized as capital gain or loss.

Restricted Shares, Restricted Share Units, Defined Share Units, and Performance Awards. In general, a Participant will not recognize income at the time of grant of restricted shares, restricted share units, defined share units or Performance Awards, unless the Participant elects with respect to restricted shares or restricted share units to accelerate income taxation to the date of the Award. In this event, a Participant would recognize ordinary income equal to the excess of the market value of the restricted shares over any amount the Participant pays for them (in which case subsequent gain or loss would be capital in nature). In the absence of an election to accelerate income taxation to the date of an Award, a Participant must recognize taxable compensation income equal to the value of any cash or shares of Common Stock that the Participant receives when the Award vests. The same tax consequences apply to Performance Awards.

Special Tax Provisions. Under certain circumstances, the accelerated vesting, cash-out or accelerated lapse of restrictions on Awards in connection with a change in control of the Company might be deemed an excess parachute payment for purposes of the golden parachute tax provisions of Code section 280G, and the Participant may be subject to a 20% excise tax and the Company may be denied a tax deduction. Furthermore, the Company may not be able to deduct the aggregate compensation in excess of \$1,000,000 attributable to Awards that are not performance-based within the meaning of Code section 162(m) in certain circumstances.

Income Taxes and Deferred Compensation. The SIP provides that participants are solely responsible and liable for the satisfaction of all taxes and penalties that may arise in connection with Awards (including any taxes arising under Section 409A of the Code), and that the Company will not have any obligation to indemnify or otherwise hold any Participant harmless from any or all of such taxes. Nevertheless, the SIP authorizes the Committee to organize any deferral program, to require deferral election forms, and to grant or to unilaterally modify any Award in a manner that (a) conforms with the requirements of Section 409A of the Code; (b) that voids any Participant election to the extent it would violate Section 409A of the Code; and (c) for any distribution election that would violate Section 409A of the Code, to make distributions pursuant to the Award at the earliest to occur of a distribution event that is allowable under Section 409A of the Code or any distribution event that is both allowable under Section 409A of the Code and is elected by the Participant, with the Committee's consent, in accordance with Section 409A.

New Plan Benefits. The Committee will grant Awards under the SIP at its discretion. Consequently, it is not possible to determine at this time the amount or dollar value of Awards to be provided under the SIP, other than to note that the Committee has not granted Awards that are contingent upon the approval of the SIP.

THE BOARD OF DIRECTORS UNANIMOUSLY RECOMMENDS THAT THE STOCKHOLDERS VOTE FOR THE APPROVAL OF THE BIOMARIN PHARMACEUTICAL INC. 2006 SHARE INCENTIVE PLAN. IF YOU SIGN AND RETURN THE ENCLOSED WHITE PROXY CARD, UNLESS YOU DIRECT TO THE CONTRARY ON THAT CARD, THE SHARES REPRESENTED BY THAT PROXY CARD WILL BE VOTED FOR THE APPROVAL OF THE BIOMARIN PHARMACEUTICAL INC. 2006 SHARE INCENTIVE PLAN. The affirmative vote of holders of a majority of the shares of Common Stock which are present in person or by proxy at the Annual Meeting, entitled to vote on this proposal and which have actually voted is required for approval of this proposal.

PROPOSAL FOUR: APPROVAL OF THE AMENDED AND RESTATED 2006 EMPLOYEE STOCK PURCHASE PLAN

Background

Subject to stockholder approval, the Board of Directors has adopted the BioMarin Pharmaceutical Inc. Amended and Restated 2006 Employee Stock Purchase Plan (the 2006 ESPP). The 2006 ESPP is an amendment and restatement of the BioMarin Pharmaceutical Inc. 1998 Employee Stock Purchase Plan (the 1998 ESPP), which shall remain in full force and effect (including all Offering Periods, as defined below), except as modified by the 2006 ESPP, in the event the 2006 ESPP receives stockholder approval. The 2006 ESPP is amending and restating the 1998 ESPP primarily in order to replenish the reserve of shares that are available to allow eligible employees and officers of the Company and its designated affiliates to purchase, through payroll deductions, shares of the Company's Common Stock. Below is a summary of the principal provisions of the 2006 ESPP and its operation. A copy of the 2006 ESPP is set forth in full in *Appendix B* to this proxy statement, and the following description of the 2006 ESPP is qualified in its entirety by reference to *Appendix B*. Terms below that appear in initial capital letters have the special meaning set forth either above or in the 2006 ESPP.

The 2006 ESPP is primarily designed to retain and motivate U.S. employees of the Company and its designated affiliates by encouraging them to acquire ownership in the Company on a tax-favored basis. In particular, the 2006 ESPP is intended to be an employee stock purchase plan within the meaning of Section 423 of the Code and thereby to allow participating employees to defer recognition of taxes when purchasing common stock at a discount under such a purchase plan. The maximum number of shares available to participants in the 2006 ESPP for purchase in offering periods ending after April 30, 2006 is 2,500,000 shares. Such shares may be newly-issued or treasury shares, or instead purchased by a designated broker who will be purchasing shares for participants on the open market. To the extent the Company offers to sell shares at a price below fair market value, the Company will make cash payments to the broker to subsidize the discount.

Summary of the Amended and Restated 2006 Employee Stock Purchase Plan

Administration. The 2006 ESPP will be administered by the Board of Directors or a person or committee appointed from time to time by the Board of Directors (the Administrator). The Board of Directors or the Administrator, if one has been appointed, is vested with full authority to construe, interpret and apply the terms of the 2006 ESPP, to determine eligibility, to adjudicate all disputed claims under the 2006 ESPP, to adopt, amend and rescind any rules deemed appropriate for the administration of the 2006 ESPP, and to make all other determinations necessary or advisable for the administration of the 2006 ESPP. Determinations by the Board of Directors or the Administrator, as to the interpretation and operation of the 2006 ESPP, will be final and binding on all parties. The Company's Compensation Committee currently administers the 1998 ESPP, and will initially act as Administrator of the 2006 ESPP if it receives approval at the Annual Meeting.

Offering Periods And Purchase Dates. Similar to the terms of the 1998 ESPP, the 2006 ESPP includes overlapping Offering Periods, which are twenty-four months each (the Offering Period or Offering), with a new Offering Period generally commencing on the first business day on or after May 1 or November 1 of each year (or on such other day as the Administrator shall determine), and ending on the last day of the calendar month that is approximately twenty-four (24) months following the first day of the Offering Period. The first Offering Period under the 2006 ESPP will be the Offering Period that began on May 1, 2005. The 2006 ESPP's first new six-month purchase period, which is the period beginning on the first business day after the last day of the prior purchase period (the Purchase Period), shall begin on May 1, 2006. The Administrator or the Board of Directors may change the Purchase Period associated with future Offerings to up to 27 months, without stockholder approval, but with notice to participants of at least fifteen days prior to the first Purchase Period to be affected.

Eligibility. All employees of the Company and its Designated Employers (including designated related entities for sub-plans) will be eligible to participate in the 2006 ESPP, except persons whose customary employment is less than 20 hours per week or five months or less per year, and persons who have been employed by the Company for less than six months as of the date 90 days before the first business day of the Purchase Period. Persons who are deemed for purposes of Section 423(b)(3) of the Code to own shares of Common Stock possessing 5% or more of the total combined voting power or value of all classes of Common Stock or shares of a subsidiary will be ineligible to participate in the 2006 ESPP. In addition, if an option granted pursuant to the 2006 ESPP would permit a person's rights to purchase shares of Common Stock to accrue at a rate that exceeds \$25,000 of the fair market value (Fair Market Value) of such Common Stock for any calendar year, such person will not be eligible to participate in the 2006 ESPP. In addition, the Board of Directors or the Administrator, in its sole discretion, may permit Company employees who are 5% or more stockholders, and consultants to participate in certain sub-plans which are not designed to qualify as Code Section 423 plans. As of the Record Date, the Company and its designated affiliates had no directors or consultants who were eligible to participate in the 2006 ESPP and approximately 370 employees who were eligible to participate in the 2006 ESPP.

Participation. Eligible employees may elect to participate in one or more of the Offering Periods, by electing to make payroll deductions during the Offering Period. The amount of the payroll deductions must not be less than one (1%) percent or more than ten (10%) percent of a participant's compensation on each payday of the Offering Period, subject to both adjustment for capital changes and to the discretion of the Board of Directors or the Administrator to revise these limits for any Offering before it commences. All sums deducted from the regular compensation of participants will be credited to a share purchase account established for each participant on the Company's books, but prior to use of these funds for the purchase of shares of Common Stock in accordance with the 2006 ESPP, the Company may use these funds for any valid corporate purpose. The Company will not be under any obligation to pay interest on funds credited to a participant's stock purchase account in any event.

Price. The price per share of Common Stock sold under the 2006 ESPP during an Offering will be 85% of the Fair Market Value of a Share of the Company's Common Stock on the Offering Date (the first day of each Offering Period) or the Purchase Date (the last day of a Purchase Period), whichever is lower; provided that the Administrator may establish an alternative purchase price so long as it does not result in a lower purchase price than is allowed under Code Section 423. Each participating employee will receive an option, effective on the first day of the Offering, to purchase shares of Common Stock on the last day of the Offering. The number of shares which a participant may purchase under the option during each Offering will be the quotient of the aggregate payroll deductions in the Offering authorized by the participant, and not withdrawn, divided by the applicable purchase price; provided, however, that the maximum number of shares a participant may purchase shall not exceed 5,000 shares per Purchase Period. The Administrator may increase or decrease the number of shares a participant may purchase each Purchase Period of a future Offering Period.

Purchase of Shares. A participant's option to purchase Common Stock pursuant to the 2006 ESPP will be automatically exercised on the Purchase Date. Before that date, a participant may terminate his or her participation in the 2006 ESPP by providing written notice to the Company or its designated broker (Designated Broker) at least five days prior to the last day of the Offering. A participant who terminates his or her participation in the 2006 ESPP during an Offering will receive a refund of his or her 2006 ESPP contributions. If a participant fails to work at least twenty hours per week during an Offering, the participant shall be deemed to have terminated his or her participation in the 2006 ESPP. A participant who withdraws from an offering may enroll in a subsequent Purchase Period within the same calendar year, but only once within the same calendar year for the 2006 ESPP or the 1998 ESPP. Other than terminating his or her participation in the 2006 ESPP altogether, once an Offering begins, a participant may only increase or decrease how much he or she has elected to contribute to the 2006 ESPP during the Offering in accordance with the rules that the Administrator establishes before the Offering begins.

The Administrator shall determine for each Purchase Period whether and the extent to which the shares shall be newly issued or treasury shares, or purchased on the open market through a Designated Broker. To the extent

that the purchase price for the shares is below Fair Market Value for any Purchase Period, the Company will pay the Designated Broker the amounts necessary to subsidize the purchase price for shares purchased on the open market.

Transferability. Options under the 2006 ESPP may not be assigned, transferred, pledged or otherwise disposed of except by will or in accordance with the laws of descent and distribution.

Employment Termination. If a participant's employment terminates for any reason, his or her payroll deductions or contributions will be refunded, and the participant will have up to thirty days to transfer Common Stock from the 2006 ESPP to himself or herself, a designated beneficiary or a broker. If the participant's shares of Common Stock are not so transferred, a share certificate will be issued and mailed to the participant.

Duration of 2006 ESPP. If approved at the Annual Meeting, the 2006 ESPP will expire on May 2, 2016, unless the Board exercises its discretion to terminate it on an earlier date.

Amendment or Termination of the 2006 ESPP. The Company's Board of Directors may at any time amend or terminate the 2006 ESPP, subject to stockholder approval to the extent the Board of Directors or the Administrator determines that such approval is appropriate, for example, to conform the 2006 ESPP with Section 423 of the Code (currently, for example, the approval of the stockholders of the Company is required to increase the number of shares of Common Stock authorized for purchase under the 2006 ESPP or to change the class of employees eligible to receive options under the 2006 ESPP, other than to designate additional affiliates as eligible subsidiaries for the 2006 ESPP).

Change in Company Capital Structure. If there is any change in the shares of the Company as a result of a merger, consolidation, reorganization, recapitalization, declaration of stock dividends, stock split, combination of shares, exchange of shares, change in corporate structure or similar event, appropriate adjustments will be made to the class and number of shares that the 2006 ESPP may issue the class and number of shares each participant may purchase, and the class and number of shares and the price per share under each outstanding purchase right.

U.S. Federal Income Tax Consequences. No taxable income will be recognized by a participant until the sale or other disposition of the shares of Common Stock acquired under the 2006 ESPP. At that time, a participant generally will recognize ordinary income and capital gains. When the shares are disposed of by a participant two years or more after the beginning of the Offering in which the shares were purchased, he or she will recognize ordinary income equal to the lesser of (a) the excess of the Fair Market Value of the shares on the purchase date over the purchase price (the Discount) or (b) the excess of the Fair Market Value of the shares at disposition over the purchase price. When shares are disposed of after less than two years (in what is known as a disqualifying disposition), the participant must recognize ordinary income in the amount of the Discount, even if the disposition is a gift or is at a loss. In the event of a participant's death while owning shares acquired under the 2006 ESPP, ordinary income must be recognized in the year of death as though the shares had been sold.

In the cases discussed above (other than death), the amount of ordinary income recognized by a participant is added to the purchase price paid by the participant, and this amount becomes the tax basis for determining the amount of the capital gain or loss from the disposition of the shares. Additional gain, if any, will be short-term or long-term capital gain depending on whether the holding period is 12 months or less, or more than 12 months.

Net capital gains from the disposition of capital stock held more than 12 months are currently taxed at a maximum federal income tax rate of 15% and net capital gains from the disposition of stock held not more than 12 months is taxed as ordinary income (maximum rate of 35%). However, limitations on itemized deductions and the phase-out of personal exemptions may result in effective marginal tax rates higher than 15% for net capital gains and 35% for ordinary income.

The Company is entitled to tax deductions for shares issued under the 2006 ESPP only in the event of disqualifying dispositions. For disqualifying dispositions, the Company is allowed a deduction to the extent of the amount of ordinary income includable in gross income by such participant for the taxable year as a result of the premature disposition of the shares. The 2006 ESPP will not meet the requirements in Section 162(m) of the Code, which means that there may be no Company deductions for disqualifying dispositions by the Company's Chief Executive Officer and four most highly paid other executive officers.

THE BOARD OF DIRECTORS UNANIMOUSLY RECOMMENDS THAT THE STOCKHOLDERS VOTE FOR THE APPROVAL OF THE BIOMARIN PHARMACEUTICAL INC. AMENDED AND RESTATED 2006 EMPLOYEE STOCK PURCHASE PLAN. IF YOU SIGN AND RETURN THE ENCLOSED WHITE PROXY CARD, UNLESS YOU DIRECT TO THE CONTRARY ON THAT CARD, THE SHARES REPRESENTED BY THAT PROXY CARD WILL BE VOTED FOR THE APPROVAL OF THE BIOMARIN PHARMACEUTICAL INC. AMENDED AND RESTATED 2006 EMPLOYEE STOCK PURCHASE PLAN. The affirmative vote of holders of a majority of the shares of Common Stock which are present in person or by proxy at the Annual Meeting, entitled to vote on this proposal and which have actually voted is required for approval of this proposal.

OTHER INFORMATION RELATED TO BIOMARIN, THE DIRECTORS

AND EXECUTIVE OFFICERS

Security Ownership of Certain Beneficial Owners

The following table sets forth certain information with respect to the beneficial ownership of the Company's common stock as of March 14, 2006 as to: (i) each person, or group of affiliated persons, who is known by the Company to own beneficially more than 5% of the Company's common stock; (ii) each of the Directors; (iii) each of the Company's Named Executive Officers; and (iv) all of the Directors and current executive officers as a group.

Except as otherwise noted, the persons or entities in this table have sole voting and investing power with respect to all the shares of BioMarin's common stock beneficially owned by them, subject to community property laws, where applicable. The information with respect to each person specified was supplied or confirmed by such person or based upon statements filed with the Securities and Exchange Commission. Except as otherwise indicated, the mailing address for each stockholder in the table below is c/o BioMarin Pharmaceutical Inc., 105 Digital Drive, Novato, California 94949.

| Name of Beneficial Owner | Number of Shares Beneficially Owned | Number of Shares Subject To Options(1) | Percentage of Common Stock(2) |
|--|-------------------------------------|--|-------------------------------|
| FMR Corp.(3) | 11,137,550 | | 14.9% |
| PRIMECAP Management Company(4) | 6,815,887 | | 9.1% |
| OrbiMed Advisors LLC / OrbiMed Capital LLC(5) | 5,218,100 | | 7% |
| Vanguard Horizon Funds Vanguard Capital Opportunity Fund(6) | 4,670,500 | | 6.2% |
| Franz L. Cristiani | 97,500 | 97,500 | * |
| Michael Grey | 7,500 | 7,500 | * |
| Elaine Heron | 102,500 | 102,500 | * |
| Joseph Klein, II | 34,500 | 22,500 | * |
| Pierre Lapalme | 71,000 | 67,500 | * |
| Alan Lewis, Ph.D. | 22,500 | 22,500 | * |
| Jean-Jacques Bienaimé | 123,834 | 108,334 | * |
| Jeffrey H. Cooper | 78,092 | 78,092 | * |
| Robert Baffi, Ph.D. | 401,824 | 379,075 | * |
| Emil D. Kakkis, M.D., Ph.D. | 425,073 | 392,499 | * |
| Christopher M. Starr, Ph.D. | 424,486 | 129,310 | * |
| Stuart J. Swiedler, M.D., Ph.D. | 221,065 | 211,065 | * |
| All current executive officers and Directors as a group (11 persons) | 1,565,378 | 1,489,055 | 2.1% |

* Represents less than 1% of BioMarin's outstanding common stock.

- (1) The Number of Shares Subject to Options enumerates for each 5% stockholder, director and Named Executive Officer and for all executive officers and directors in the aggregate, the shares of common stock subject to options exercisable within 60 days of March 14, 2006. These shares are included in the amounts shown under the Number of Shares Beneficially Owned, as provided in the note below.
- (2) The Percentage of Common Stock column is based on 74,748,424 shares of common stock outstanding at March 14, 2006. Shares of common stock subject to options or warrants that are exercisable within 60 days of March 14, 2006 are deemed to be outstanding and to be beneficially owned by the person holding the options or warrants for the purpose of computing the percentage ownership of the person but are not treated as outstanding for the purpose of computing the percentage ownership of any other person.
- (3) Information based upon statements filed on Schedule 13G with the Securities and Exchange Commission on February 14, 2006. The mailing address for FMR Corp. is 82 Devonshire Street, Boston, MA 02109.

- (4) Information is based upon statements filed on Schedule 13G/A with the Securities and Exchange Commission on February 8, 2006. The mailing address for PRIMECAP Management Company is 225 South Lake Avenue, #400, Pasadena, CA 91101.
- (5) Information based upon statements filed on Schedule 13G/A with the Securities and Exchange Commission on February 8, 2006. The mailing address for OrbiMed Advisors LLC/OrbiMed Capital LLC is 767 Third Avenue, 30th Floor, New York NY 10017.
- (6) Information based upon statements filed on Schedule 13G with the Securities and Exchange Commission on February 13, 2006. The mailing address for Vanguard Horizon Funds Vanguard Capital Opportunity Fund is 100 Vanguard Boulevard, Malvern, PA 19355.

Executive Officers and other Significant Employees

The following table sets forth certain information concerning BioMarin's executive officers and other significant employees as of May 1, 2005.

| Name | Age | Position with BioMarin |
|---------------------------------|-----|---|
| Jean-Jacques Bienaimé | 52 | Chief Executive Officer |
| Emil D. Kakkis, M.D., Ph.D. | 46 | Chief Medical Officer |
| Stephen Aselage | 55 | Senior Vice President, Global Commercial Operations |
| Robert A. Baffi, Ph.D. | 51 | Senior Vice President, Technical Operations |
| Stuart J. Swiedler, M.D., Ph.D. | 50 | Senior Vice President, Clinical Affairs |
| Jeff Ajer | 44 | Vice President, Sales and Marketing Operations |
| William E. Aliski | 59 | Vice President and General Manager, European Operations |
| Jeffrey H. Cooper, C.P.A. | 50 | Vice President, Chief Financial Officer |
| G. Eric Davis | 35 | Vice President, General Counsel and Secretary |
| Daniel P. Maher | 50 | Vice President, Product Development |

There is no family relationship between any Director and any executive officer of BioMarin.

Emil D. Kakkis, M.D., Ph.D., Chief Medical Officer

Dr. Kakkis serves as BioMarin's Chief Medical Officer and has been a Senior Vice President or Vice President at BioMarin since joining the Company in September 1998. Together with his colleague, Elizabeth F. Neufeld, Ph.D., of the University of California at Los Angeles (UCLA), Dr. Kakkis developed Aldurazyme, a recombinant form of alpha-L-iduronidase, the enzyme deficient in MPS I patients. Before joining BioMarin, Dr. Kakkis held the position of assistant professor at the Harbor-UCLA Medical Center, Division of Genetics, Department of Pediatrics. From 1991 to 1994, he completed a fellowship in genetics at the UCLA Intercampus Medical Genetics training program and, prior to that, conducted his pediatric residency at the Harbor-UCLA Medical Center. Dr. Kakkis is the author of numerous published articles and abstracts on MPS I and alpha-L-iduronidase. He holds an M.D. and a Ph.D. in biological chemistry from the Medical Scientist training program at the UCLA School of Medicine. He is board-certified in pediatrics and medical genetics.

Stephen Aselage, Senior Vice President, Global Commercial Operations

Mr. Aselage joined BioMarin in June 2005 and serves as Senior Vice President, Global Commercial Operations. From February 2004 to June 2005, Mr. Aselage served as Executive Vice President of Global Commercial Operations at Cell Therapeutics, a biotechnology company focused on cancer therapeutics. From September 2003 to February 2004, Mr. Aselage served as Senior Vice President of North American Sales and Marketing for Genzyme Corporation following Genzyme's acquisition of Sangstat Medical Corporation where he had worked since February 1999. While at Sangstat, Mr. Aselage restructured the company's sales, marketing and medical affairs groups. From 1996 through 1999, Mr. Aselage served as Director of Sales and Marketing at Advanced Tissue Sciences. Earlier in his career, Mr. Aselage held a variety of sales and sales management

positions at biotechnology and pharmaceutical companies including Rhône-Poulenc Rorer Pharmaceuticals (now Sanofi-Aventis), Genentech, Inc., and Bristol Laboratories. Mr. Aselage holds a B.S. in biology from the University of Notre Dame.

Robert A. Baffi, Ph.D., Senior Vice President, Technical Operations

Dr. Baffi joined BioMarin in May 2000, and serves as Senior Vice President, Technical Operations, responsible for overseeing the company's manufacturing, process development, and quality and analytical chemistry departments. From 1986 to 2000, Dr. Baffi served in a number of progressively more responsible positions at Genentech, primarily in the functional area of quality control. Prior to Genentech, Dr. Baffi worked for Cooper BioMedical as a research scientist and at Becton Dickinson Research Center as a post-doctoral fellow. Dr. Baffi has contributed to more than 20 major regulatory submissions for product approval in the United States and Europe and to more than 50 regulatory submissions for investigational new drug testing. Dr. Baffi received a Ph.D. in biochemistry, an M.B.A. from Regis University, as well as an M.Phil. and a B.S. in biochemistry from the City University of New York.

Stuart J. Swiedler, M.D., Ph.D., Senior Vice President, Clinical Affairs

Dr. Swiedler joined BioMarin in June 1998 and serves as Senior Vice President, Clinical Affairs. Before joining BioMarin, Dr. Swiedler served as Vice President, Research Programs at Glycomed. He is board-certified in anatomic pathology and has conducted extensive research in the molecular biology of carbohydrate enzymes. Dr. Swiedler holds five patents and is the author of 20 peer-reviewed journal articles. Dr. Swiedler holds a Ph.D. from the Johns Hopkins University School of Medicine, Biochemistry, Cellular, and Molecular Biology training program, an M.D. from the Johns Hopkins School of Medicine, and a B.S. from the State University of New York at Albany.

Jeff Ajer, Vice President, Sales and Marketing Operations

Mr. Ajer joined BioMarin in September 2005 and serves as Vice President, Sales and Marketing Operations. From September 2003 to August 2005, Mr. Ajer worked at Genzyme Corporation where he served most recently as Vice President, Global Transplant Operations. Prior to this, from 1999 to 2003, he served as Senior Director of Sales Operations, Bids and Contracts for SangStat Medical Corporation, until the company's acquisition by Genzyme. From June 1986 to July 1999, Mr. Ajer held positions with increasing responsibility at ICN Pharmaceuticals, Inc., including Director, Marketing and Sales Services, and Director, Purchasing Worldwide. Mr. Ajer received both a B.S. in chemistry and an M.B.A. from the University of California, Irvine.

William E. Aliski, Vice President and General Manager, European Operations

Mr. Aliski joined BioMarin in December 2005 and serves as Vice President and General Manager, European Operations. From August 1999 through October 2005, Mr. Aliski held the position of Vice President of Commercial Operations at Transkaryotic Therapies (TKT; acquired by Shire Pharmaceuticals Group in July 2005) where he managed worldwide drug distribution, inventory management and patient support for the company's lead product. From September 1991 to August 1999, Mr. Aliski worked at Genzyme Corporation where he most recently served as Vice President of Health Systems and Vice President of Patient and Product Services, responsible for overseeing the development and implementation of reimbursement planning and launch for multiple products. Mr. Aliski received an M.P.A. from the Kennedy School of Government at Harvard University and a B.S. degree in Economics and a Masters of Social Planning from Boston College.

Jeffrey H. Cooper, C.P.A., Vice President, Chief Financial Officer

Mr. Cooper joined BioMarin in October 2003 as Vice President, Controller and was promoted to the position of Vice President, Chief Financial Officer in August 2005. Prior to joining BioMarin, Mr. Cooper

worked at Matrix Pharmaceutical for six years, serving most recently as Vice President, Finance. During his career, Mr. Cooper served in numerous finance-related roles within the health care and pharmaceutical industries, including corporate controller at Foundation Health Systems, and Director of Business Analysis at Syntex Corporation, a company he worked for from 1983 to 1995. Mr. Cooper is a California C.P.A. and earned a B.A. in economics from the University of California, Los Angeles, and an M.B.A. from Santa Clara University.

G. Eric Davis, Vice President, General Counsel and Secretary

Mr. Davis joined BioMarin in March 2004, and serves as Vice President, General Counsel and Secretary. From 2000 to 2004, Mr. Davis worked in the San Francisco office of Paul, Hastings, Janofsky & Walker LLP, an international law firm, where he was an associate in the Corporate Department and served on the firm's national securities practice committee. Mr. Davis received a B.A. from the University of California, Berkeley, and a J.D. from the University of San Francisco.

Daniel P. Maher, Vice President, Product Development

Mr. Maher joined BioMarin as Vice President, Program Management in August 2003. He is currently responsible for program management and research. From 1998 to 2003, Mr. Maher worked at Aradigm Corporation serving as Vice President, Program Management. Prior to Aradigm, he served as Director of Clinical Operations at Covance, Inc., and as Vice President of Operations at Spectra BioMedical, Inc. Prior to Spectra, Mr. Maher acted as Director, Therapeutics Project Management, at Chiron Corporation. Mr. Maher started his career at Genentech, Inc. in 1981, serving in several capacities in operations, and as Manager of Product Development. Mr. Maher received an M.B.A. in health services management from Golden Gate University and a B.A. in biology from San Francisco State University.

Executive Compensation*Summary Compensation Table*

The following table discloses compensation paid by the Company during the last three fiscal years to (a) Jean-Jacques Bienaimé, the Company's Chief Executive Officer and Louis Drapeau, the Company's former Acting Chief Executive Officer, who retired from the Company effective October 31, 2005; (b) Robert A. Baffi, Ph.D., Emil D. Kakkis, M.D., Ph.D., Stuart J. Swiedler, M.D., Ph.D. and Jeffrey H. Cooper, the four most highly-compensated officers other than the Chief Executive Officer who were serving as officers at the end of fiscal year 2005 and whose salary and bonus exceeded \$100,000; and (c) Christopher M. Starr, Ph.D., who would have been one of the four most highly compensated officers, but for the fact that he resigned on December 31, 2005. These individuals are referred to as the Named Executive Officers.

The entries under the column *All Other Compensation* in the table represent the premiums paid for life insurance benefits, financial planning consultation, fitness benefits and vested 401(k) matching for each Named Executive Officer.

Summary Compensation Table

| Name and Principal Position | Year | Annual Compensation | | | Long-Term Compensation Awards Securities Underlying Options/SARs(#) | All Other Compensation(\$) |
|---|------|---------------------|---------------|-------------------------------|---|----------------------------|
| | | Salary(\$) | Bonus(\$) | Other Annual Compensation(\$) | | |
| Jean-Jacques Bienaimé | 2005 | 275,400 | 100,000(1) | | 487,500 | 40,234 |
| Chief Executive Officer | 2004 | | | | | |
| | 2003 | | | | | |
| Emil D. Kakkis, M.D., Ph.D. | 2005 | 270,250 | 100,000(3) | | 120,000 | 1,156 |
| Chief Medical Officer | 2004 | 270,402 | 160,000(2)(4) | | | 4,913 |
| | 2003 | 270,027 | 110,000 | | 125,000 | 4,974 |
| Robert A. Baffi, Ph.D. | | | | | | |
| Senior Vice President, Technical | 2005 | 250,000 | | | 140,000 | 11,564 |
| | 2004 | 249,871 | 80,000(2) | | | 6,345 |
| Operations | 2003 | 242,266 | 30,000 | | 50,000 | 8,543 |
| Stuart J. Swiedler, M.D., Ph.D. | | | | | | |
| Senior Vice President, Clinical | 2005 | 256,500 | | | 80,000 | 1,816 |
| | 2004 | 256,652 | 80,000(2) | | | 6,226 |
| Affairs | 2003 | 256,223 | 42,500 | | 80,000 | 4,923 |
| Jeffrey H. Cooper | 2005 | 217,538 | | | 60,000 | 19,327 |
| Chief Financial Officer | 2004 | 206,615 | 10,000(2) | | 10,000 | 18,135 |
| | 2003 | 42,308 | | | 110,000 | 145 |
| Louis Drapeau | | | | | | |
| Former Acting Chief Executive | 2005 | 267,471 | | | 100,000 | 4,242 |
| | 2004 | 258,320 | 40,000 | | 25,000 | 5,318 |
| Officer | 2003 | 250,025 | 10,000(2) | | 40,000 | 5,600 |
| Christopher M. Starr, Ph.D. | | | | | | |
| Senior Vice President, Chief Scientific | 2005 | 324,492 | | | 60,000 | 1,981 |
| | 2004 | 275,830 | 110,000(2) | | | 6,417 |
| Officer | 2003 | 275,474 | 60,000 | | 125,000 | 4,992 |

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- (1) Mr. Bienaimé received a one-time sign-on bonus in 2005 pursuant to his employment agreement with the Company.
 - (2) This bonus represents amounts paid in February 2004 in recognition of performance in 2003. No officer received a cash bonus for performance in 2004.
 - (3) Dr. Kakkis' bonus was related to the FDA approval of Naglazyme, pursuant to Dr. Kakkis' employment agreement.
 - (4) Dr. Kakkis' bonus included a \$110,000 general merit bonus and a \$50,000 bonus related to the filing of the Investigational New Drug Application for the Company's product candidate Phenoptin, pursuant to Dr. Kakkis' employment agreement.

Stock Option Grants

The following table sets forth certain information for each grant of options to purchase the Company's common stock during fiscal year 2005 to each of the Named Executive Officers.

Option Grants in Last Fiscal Year

| Name | Number of Securities Underlying Options Granted (#) | Individual Grants | | Exercise or Base Price (\$/Share)(2) | Expiration Date | Potential Realizable Value at Assumed Annual Rates of Stock Price Appreciation for Option Term \$(3) | | First Exercisable |
|---------------------------------|---|---|--|--------------------------------------|-----------------|--|--------------|-------------------|
| | | Percent of Total Options Granted to Employees in Fiscal Year(1) | | | | 5% | 10% | |
| Jean-Jacques Bienaimé | 325,000 | 12% | | \$ 6.46 | 5/11/2015 | \$ 1,320,364 | \$ 3,346,062 | 5/11/2006 |
| | 162,500 | 6% | | 9.86 | 11/11/2015 | 1,007,646 | 2,553,574 | 11/11/2006 |
| Emil D. Kakkis, M.D., Ph.D. | 20,000 | 1% | | 8.05 | 7/18/2015 | 101,252 | 256,593 | 1/18/2006 |
| | 100,000 | 4% | | 6.13 | 1/7/2015 | 385,512 | 974,711 | 7/7/2005 |
| Robert A. Baffi, Ph.D. | 140,000 | 5% | | 6.13 | 1/7/2015 | 539,717 | 1,364,596 | 7/7/2005 |
| Stuart J. Swiedler, M.D., Ph.D. | 80,000 | 3% | | 6.13 | 1/7/2015 | 308,410 | 779,769 | 7/7/2005 |
| Jeffrey H. Cooper | 60,000 | 2% | | 6.13 | 1/7/2015 | 231,307 | 584,827 | 7/7/2005 |
| Louis Drapeau | 100,000 | 4% | | 6.13 | 1/7/2015 | 385,512 | 974,711 | 7/7/2005 |
| Christopher M. Starr, Ph.D. | 60,000 | 2% | | 6.13 | 1/7/2015 | 231,307 | 584,827 | 7/7/2005 |

(1) Based on an aggregate of 2,712,471 shares subject to options granted by BioMarin during fiscal year 2005 to employees, consultants and the Named Executive Officers.

(2) Options were granted at an exercise price equal to the closing price of BioMarin's common stock on Nasdaq on the date of the grant.

(3) The 5% and 10% assumed annual rates of compounded stock price appreciation are mandated by rules of the Securities and Exchange Commission. The actual stock price over the option term could be substantially more or less than the assumed 5% and 10% levels included in the table. Unless the market price of the Company's common stock appreciates over the option period, no value will be realized from the option grants made to the Named Executive Officers. The potential realizable value is calculated by assuming that the closing price per share on the date of grant appreciates at the indicated rate for the entire term of the option and that the option is exercised at the exercise price and sold on the last day of its term at the appreciated price. The potential realizable value computation is net of the applicable exercise price, but does not take into account applicable federal or state income tax consequences and other expenses of option exercises. The values shown do not consider non-transferability or termination of the options upon termination of such employee's employment.

Fiscal 2005 Option Exercises and Option Value Table

The following table sets forth the number of shares covered by both exercisable and unexercisable stock options held by each of the Named Executive Officers and certain information with respect to options exercised by the Named Executive Officers during the Company's fiscal year 2005.

Options Exercised During Fiscal Year 2005

| Name | Shares Acquired on Exercise | Value Realized(\$) | Number of Securities Underlying Unexercised Options at Year-End | | Value of Unexercised In-the-Money Options at Year-End(1) | |
|---------------------------------|-----------------------------|--------------------|---|---------------|--|---------------|
| | | | Exercisable | Unexercisable | Exercisable | Unexercisable |
| Jean-Jacques Bienaimé | | \$ | | 487,500 | \$ | \$ 1,553,500 |
| Emil D. Kakkis, M.D., Ph.D. | | | 366,353 | 223,647 | 1,764,502 | 585,198 |
| Robert A. Baffi, Ph.D. | | | 359,284 | 208,959 | 273,637 | 577,314 |
| Stuart J. Swiedler, M.D., Ph.D. | 30,000 | 180,000 | 205,024 | 170,209 | 822,303 | 397,595 |
| Jeffrey H. Cooper | | | 76,249 | 163,751 | 242,967 | 390,533 |
| Louis Drapeau | 30,000 | 55,106 | 158,853 | | 548,187 | |
| Christopher M. Starr, Ph.D. | 11,142 | 66,852 | 280,669 | | 1,037,693 | |

(1) Based on the closing price of BioMarin's common stock on December 31, 2005 of \$10.78 per share less exercise price per share.
Employment Agreements

BioMarin is party to employment agreements with Mr. Bienaimé, Dr. Baffi, Dr. Kakkis and Dr. Swiedler on the terms set forth in the chart below. Each of these employment agreements, other than Mr. Bienaimé's agreement, is terminable without cause by BioMarin upon six months prior written notice to the officer, or by the officer upon three months' prior written notice to BioMarin. BioMarin is obligated to pay each of the officers' salary and benefits until termination. In addition, with respect to Drs. Baffi, Kakkis and Swiedler, if any of them is involuntarily terminated within one year of a change of control, he is entitled to receive from the Company a severance payment equal to six months of his then-current annual salary and fifty percent of the annual bonus that he would otherwise be entitled to receive for the calendar year in which he was involuntarily terminated, and fifty percent of the then-unvested portion of all of his options in the Company shall immediately vest. With respect to Mr. Cooper, his employment is terminable by the Company or by Mr. Cooper at will. Each of the Named Executive Officers other than Mr. Bienaimé is entitled to receive benefits under the Company's Severance Plan, described below.

With respect to Mr. Bienaimé's employment agreement, the Company can terminate the agreement at any time upon written notice to Mr. Bienaimé. If the Company terminates Mr. Bienaimé's employment without cause or if Mr. Bienaimé resigns for good reason or becomes permanently disabled while employed by the Company or if the Company files bankruptcy, Mr. Bienaimé will be entitled to receive: (i) cash compensation in an amount equal to his then current annual base salary as of the date of termination for a period of 18 months; (ii) a cash bonus equal to 100 percent of his target bonus for such year provided that the senior vice presidents of the Company are paid bonuses under the Company's bonus plan for the year of his termination, and provided that certain performance goals are met; (iii) a continuation of all Company-paid health benefits for a period of 18 months after the date of termination; (iv) reimbursement of up to \$18,000 for outplacement services; and (v) automatic vesting of all options granted to Mr. Bienaimé that have not vested as of the date of termination. If Mr. Bienaimé has been employed by the Company for more than 3 years as of the date of termination, the applicable period for purposes of clauses (i) and (iii) of this paragraph will be 24 months instead of 18 months.

If the Company terminates Mr. Bienaimé's employment without cause or if Mr. Bienaimé resigns for good reason or becomes permanently disabled while employed by the Company, in any such case following a change

of control of the Company, Mr. Bienaimé will be entitled to receive: (i) cash compensation in an amount equal to the present value of his then current annual base salary that he would have collected over the 24 month period following the date of termination; (ii) a cash bonus equal to 100 percent (250 percent if employed for more than 3 years) of his target bonus for such year provided that the senior vice presidents of the Company are paid bonuses under the Company's bonus plan for the year of his termination, and provided that certain performance goals are met; (iii) a continuation of all Company-paid health benefits for a period of 24 months after the date of termination; (iv) a cash payment of \$18,000 for outplacement services (plus an amount for taxes payable on such cash payment); (v) the fully-paid whole life insurance policy with a stated death benefit of \$500,000 maintained for Mr. Bienaimé (plus an amount for taxes payable on imputed income and such amount for taxes); (vi) a cash payment of up to \$5,000 for tax preparation (plus an amount for taxes payable on such cash payment); (vii) the Company's annual contribution to Mr. Bienaimé's 401k plan for the year of termination to the extent allowable; and (viii) automatic vesting of all options granted to Mr. Bienaimé that have not vested as of the date of termination. If Mr. Bienaimé has been employed by the Company for more than 3 years as of the date of termination, the applicable period for purposes of clauses (i) and (iii) of this paragraph will be 30 months instead of 24 months.

If amounts payable to Mr. Bienaimé as the result of a change of control of the Company would result in a parachute payment under Section 280G of the Internal Revenue Code of 1986, as amended (the Code), which would be subject to an excise tax under Code Section 4999, or interest or penalties are incurred with respect to such excise tax, the Company will pay Mr. Bienaimé an additional payment such that, after payment by Mr. Bienaimé of all taxes imposed upon this payment and any interest or penalties imposed with respect to such taxes, Mr. Bienaimé retains an amount equal to the sum of: (i) the excise tax (including interest and penalties) imposed; and (ii) the product of any income tax deductions disallowed to Mr. Bienaimé because of the inclusion of the payment in his adjusted gross income, and the highest applicable marginal rate of federal income taxation for the calendar year in which the payment is to be made.

| Name of Executive Officer | 2005 Annual Salary Rate(1) | Annual Bonus | Initial Grant of Right to Purchase Equity Securities(2) | Agreement Termination Date |
|---------------------------------|----------------------------|--|---|----------------------------|
| Jean-Jacques Bienaimé | \$ 468,000 | Annual bonus, payable in cash and/or stock. | Options to purchase up to 325,000 of BioMarin's common stock at a purchase price of \$6.46 per share. | None |
| Emil D. Kakkis, M.D., Ph.D. | \$ 270,250 | Eligible to receive a cash bonus based on achievement of milestones and an annual bonus, payable in cash and/or stock. | Options to purchase up to 200,000 shares of BioMarin's common stock at a purchase price of \$4.00 per share. | None |
| Robert A. Baffi, Ph.D. | \$ 250,000 | Annual bonus, payable in cash and/or stock. | Options to purchase up to 210,000 shares of BioMarin's common stock at a purchase price of \$22.00 per share. | None |
| Stuart J. Swiedler, M.D., Ph.D. | \$ 256,500 | Annual bonus, payable in cash and/or stock. | Options to purchase up to 150,000 shares of BioMarin's common stock at a purchase price of \$4.00 per share. | None |
| Jeffrey H. Cooper | \$ 230,000 | Annual bonus, payable in cash and/or stock | Options to purchase 100,000 shares of BioMarin's common stock at a purchase price of \$8.10 per share. | None |

| Name of Executive Officer | 2005 Annual Salary Rate(1) | Annual Bonus | Initial Grant of Right to Purchase Equity Securities(2) | Agreement Termination Date |
|--------------------------------|----------------------------|---|---|----------------------------|
| Louis Drapeau(3) | \$ 250,000 | Annual bonus, payable in cash and/or stock | Options to purchase up to 125,000 and 25,000 shares of the Company's common stock at a purchase price of \$9.25 and \$4.24, respectively. | None |
| Christopher M. Starr, Ph.D.(4) | \$ 275,500 | Annual bonus, payable in cash and/or stock. | 400,000 shares of BioMarin's common stock at a purchase price of \$1.00 per share. | None |

- (1) 2005 Annual Salary Rate reflected in the above table reflects 2005 annual salary rate as of December 31, 2005. The table entitled "Summary Compensation Table" reflects actual salaries paid in 2005 and includes mid-year salary adjustments.
- (2) Options in this table were granted on commencement of employment for each individual.
- (3) Mr. Drapeau retired on October 31, 2005.
- (4) Dr. Starr retired on December 31, 2005.

In January 2004, the Company instituted a Severance Plan that is applicable to each of the officers listed in the table above. This policy was amended and restated in March 2005. Under the plan, on a change of control, vesting of all employee options under the 1997 Option Plan is accelerated in full, which is unchanged from the plan adopted in January 2004. Further, upon an involuntary termination on or within one year of a change of control, employees are entitled to receive certain specified benefits consisting of cash severance and a continuation of employee benefits. Under the plan, Senior Vice Presidents and Vice Presidents who are not offered a position or who decline an offer of employment from the successor company are entitled to receive one year of benefit continuation and a severance payment equal to one year of base salary. If voluntarily terminated by the successor company, Vice Presidents are entitled to receive 10.5 months of benefit continuation and a severance payment equal to 10.5 months of base salary. Senior Vice Presidents and Vice Presidents are entitled to the greater of the actual bonus paid for the prior year or the target bonus payable for the current year. Benefits payable under this policy may be reduced for employees who are entitled to severance benefits under separate arrangements with the Company to avoid the payment of duplicate benefits.

On December 1, 2005, following approval by the Company's Compensation Committee and the Board on November 28, 2005, the Company entered into the BioMarin Pharmaceutical Inc. Nonqualified Deferred Compensation Plan (the "Deferred Compensation Plan"). The Deferred Compensation Plan allows members of management, other highly compensated employees and members of the Board to make voluntary irrevocable deferrals of the compensation that they would otherwise be paid by the Company to specified future dates, employment termination, hardship events, disability, retirement or death. Participants are permitted to defer up to 100% of salary, annual cash bonus and restricted stock awards, subject to limitations to allow the Company to make necessary withholding payments. Plan participants' deferred compensation is 100% vested under the Deferred Compensation Plan. The Company may make additional direct contributions to the Deferred Compensation Plan for the benefit of the participants, but any such Company contributions must be approved by the Board. Company contributions, if any, will become 100% vested after 3 years of service with the Company (or such other time as the Company designates at the time of the contribution), or upon a change of control, death or disability. Participants have an unsecured contractual commitment by the Company to pay the amounts that become due under the Deferred Compensation Plan. Deferred compensation may be held in trust and is deemed invested based on participant direction as allowed by the Deferred Compensation Plan. Any funds held in a trust will be the sole property of the Company, subject to any claims of general creditors in the event of bankruptcy, and plan participants will have no vested interest with respect to such trust fund.

The Company has adopted a form of indemnification agreement to be entered into between BioMarin and each of its executive officers and Directors. This indemnification agreement requires the Company, among other things, to indemnify executive officers and Directors against liabilities that may arise by reason of their status or performance of their duties as executive officers or Directors and to advance their expenses incurred as a result of

any proceeding against them as to which they could be indemnified. For a description of other transactions between BioMarin and its affiliates, see *Certain Relationships and Related Transactions*.

Section 162(m)

The Company has considered the potential future effects of Section 162(m) of the Internal Revenue Code on the compensation paid to its executive officers. Section 162(m) disallows a tax deduction for any publicly held corporation for individual compensation exceeding \$1.0 million in any taxable year for any of the Named Executive Officers, unless compensation is performance-based. The Company has adopted a policy that, where reasonably practicable, it will seek to qualify the variable compensation paid to its executive officers for an exemption from the deductibility limitations of Section 162(m).

Equity Compensation Plans

The following table provides certain information with respect to all of BioMarin's equity compensation plans in effect as of December 31, 2005.

Equity Compensation Plan Information

| Plan Category | Number of securities to be issued upon exercise of outstanding options, warrants and rights(1) | Weighted average exercise price of outstanding options, warrants and rights | Number of securities remaining available for future issuance under equity compensation plans(2) |
|--|---|--|--|
| Equity compensation plans approved by stockholders | 6,968,569 | \$ 8.60 | 7,896,769 |
| Equity compensation plans not approved by stockholders | | | |
| Total(3) | 6,968,569 | \$ 8.60 | 7,896,769 |

- (1) Does not include any shares of BioMarin common stock issuable under its 1998 Employee Stock Purchase Plan. The Company issues shares under this plan once every six months based on employee elections in the preceding six months. Pursuant to the terms of this plan, the number of shares to be issued and the price per share is not determined until immediately before the date of issuance.
- (2) Includes options and shares of common stock issuable pursuant to BioMarin's 1997 Stock Plan, as amended, 1998 Director Option Plan as amended and 1998 Employee Stock Purchase Plan. Pursuant to the terms of these plans, the number of shares of common stock available for future issuance under these plans increases on the first day of each fiscal year of the Company or January 1 of each year. On the first day of each of the Company's fiscal years, the common stock available for future issuance under the 1997 Stock Plan increases by the lesser of: (i) 4% of the then outstanding capital stock of BioMarin; (ii) 2,000,000 shares; or (iii) a lower amount set by the Company's Board. On the first day of each of the Company's fiscal years, the common stock available for future issuance under each of the 1998 Director Option Plan and the 1998 Employee Stock Purchase Plan increases by the lesser of: (i) 100,000 shares in the case of the 1998 Employee Stock Purchase Plan and 200,000 shares in the case of the 1998 Director Option Plan; (ii) 0.5% of the then outstanding capital stock of BioMarin; or (iii) a lower amount set by the Company's Board.
- (3) As of May 1, 2006, the number of securities to be issued upon exercise of outstanding options was 8,147,543 with a weighted average exercise price of \$9.46 per share and a weighted average remaining term of 7.4 years.

Certain Relationships And Related Transactions

Since January 1, 2005, there has not been nor is there currently proposed any transaction or series of similar transactions to which the Company was or is to be a party in which the amount involved exceeds \$60,000 and in which any Director, executive officer, holder of more than 5% of the common stock of the Company or any member of the immediate family of any of the foregoing persons had or will have a direct or indirect material interest other than: (i) compensation agreements and other arrangements, which are described elsewhere in this form; and (ii) the transactions described below.

Transactions with Directors, Executive Officers and 5% Stockholders

The Company's Chief Medical Officer, Emil D. Kakkis, M.D., Ph.D. held an adjunct faculty position with Harbor-UCLA Research Educational Institute (REI) for purposes of conducting research. REI licenses certain intellectual property and provides other research services to the Company. The Company is also obligated to pay REI royalties on future sales of products covered by the license agreement. Minimum annual royalties payable to REI are \$25,000. The Company paid REI approximately \$0.3 million and \$0.1 million in 2004 and 2005, respectively, primarily for research and certain related license fees. The Company's joint venture with Genzyme is subject to a second agreement with REI that requires the joint venture to pay REI a royalty on sales of products covered by the license agreement through November 2019, of which Dr. Kakkis is entitled to certain portions, based on net sales of Aldurazyme per the terms of the agreement. The license agreement was effective before Dr. Kakkis was an officer of the Company. Pursuant to these agreements, Dr. Kakkis was entitled to approximately \$498,000 and \$888,000 during 2004 and 2005, respectively.

In February 2006, BioMarin transferred the intellectual property associated with the former NeuroTrans program to a new company formed by BioMarin's former Chief Scientific Officer and founder, Dr. Christopher Starr. NeuroTrans is a platform technology intended to allow for large molecule therapeutics, such as proteins, to be transported across the blood-brain barrier. The terms of the transaction were negotiated after Dr. Starr had retired from the Company. In consideration for the assignment, BioMarin will be entitled to receive milestones and royalties on net sales, if the technology is successfully developed. The milestones are not material to BioMarin unless the technology completes a successful Phase 3 clinical trial.

Indebtedness Of Directors And Executive Officers

No Director or executive officer of the Company or associate of any Director or executive officer is or at any time since January 1, 2005 has been indebted to the Company.

Performance Graph

The following is not deemed filed with the Securities and Exchange Commission and is not to be incorporated by reference into any filing of the Company under the Securities Act of 1933 or the Securities Exchange Act of 1934, whether made before or after the date hereof and irrespective of any general incorporation by reference language in such filing.

The following graph compares the cumulative total stockholder return with the cumulative total return of the Nasdaq Stock Market (U.S.) and the Nasdaq Biotechnology Index, assuming a \$100 investment in BioMarin's common stock on December 31, 2000 and reinvestment of dividends during the period.

| | BioMarin | Nasdaq Stock Market (U.S.) | Nasdaq Biotech Index |
|------------|-----------------|---------------------------------------|---------------------------------|
| 12/31/2000 | 100 | 100 | 100 |
| 12/31/2001 | 139 | 79 | 84 |
| 12/31/2002 | 73 | 54 | 46 |
| 12/31/2003 | 80 | 81 | 67 |
| 12/31/2004 | 66 | 88 | 71 |
| 12/31/2005 | 111 | 89 | 73 |

The following is not deemed filed with the Securities and Exchange Commission and is not to be incorporated by reference into any filing of the Company under the Securities Act of 1933 or the Securities Exchange Act of 1934, whether made before or after the date hereof and irrespective of any general incorporation by reference language in such filing.

REPORT OF COMPENSATION COMMITTEE ON EXECUTIVE COMPENSATION

The Compensation Committee is responsible for setting general compensation goals and operational guidelines for BioMarin personnel, for setting all elements of the compensation of the executive officers of BioMarin, and for approving grants of stock options for officers of BioMarin. During 2005, the Compensation Committee was composed of three members of the Board. The Compensation Committee meets at regular intervals during the year, and holds additional meetings as necessary. The Compensation Committee met four times in 2005.

Compensation Goals and Policies

The goal of BioMarin's compensation policies is to provide compensation sufficient to attract, motivate and retain executives and staff of outstanding ability and potential. Compensation policies are intended to establish an appropriate relationship between executive compensation and the creation of stockholder value as measured by the equity markets. BioMarin uses the following principles to help achieve that goal:

- (1) BioMarin provides competitive compensation packages incorporating all compensation elements for executives and staff based upon BioMarin's internal policies and compensation packages at similarly situated U.S. pharmaceutical and biotechnology companies.
- (2) BioMarin rewards executives and senior staff for outstanding performance by the individual and by BioMarin.
- (3) BioMarin seeks to align the long-term interests of the stockholder and the executives and the senior staff through the use of employee stock options and other stock price related compensation, such as its Employee Stock Purchase Program.

2005 Compensation

In 2005, the Compensation Committee continued to use an independent consultant to provide expertise in various matters that come before the Committee, including the review and analysis of executive compensation at BioMarin. The levels of compensation at competitor companies, based upon compensation surveys, were used for comparison in establishing the Company's executive compensation and cash and long-term equity compensation bonus plans. The primary comparator companies consisted of biotechnology companies with market capitalization between \$500 million and \$1.5 billion, with emphasis given to companies located in the San Francisco Bay Area.

Increases in base salary for 2005 were made effective January 1, 2005 primarily based on the progress and achievements of BioMarin during 2004 and competitive conditions in the job marketplace for biotechnology expertise in the San Francisco Bay Area marketplace. The Compensation Committee noted the achievements during 2004, including the completion of the clinical program for Naglazyme and filing of the marketing application for Naglazyme and the very substantial progress achieved in the development of Phenoptin. However, the Compensation Committee also noted the disappointing results associated with Orapred sales and the overall slight decline in the Company's share price.

Based on the substantial efforts associated with the positive events, the Committee provided option grants to most staff below the rank of officer. Additionally, for this group, at the beginning of 2005, cash compensation was increased by an average of 6.2%, which was intended to bring compensation closer to the averages for the biotechnology industry in the San Francisco Bay Area. Additionally, in the middle of 2005, the Compensation

Committee approved additional salary increases for employees that were significantly below the industry average. This was deemed to be appropriate in order to ensure that the Company is able to hire and retain qualified individuals.

With respect to officer salary, the Compensation Committee determined, based on the factors described above, with the concurrence of the officers, that it was appropriate not to grant the officers any salary increase or cash bonus for performance during 2004 and therefore there were no changes to cash compensation for 2005. However, to better align the officer's compensation to creating shareholder value, the officers were given stock option grants in January 2005 appropriate for their positions.

Additionally, in order to make the Company's overall compensation structure more competitive with peer companies, during 2005, the Compensation Committee established a company-wide cash bonus program. The program had specific performance metrics approved by the entire Board, which included both product program development milestones and financial performance goals. The funding of the program was based on achieving these goals and, in March 2006, the Compensation Committee, in applying these metrics, funded 91% of the targeted bonus. The individual pay-outs were based on employee level and individual performance ratings. Employees with sub-standard performance ratings were not entitled to receive a bonus.

Actions Related to 2006 Compensation

In December 2005, the Compensation Committee considered officers' compensation packages. It was determined that the officers' total compensation packages were generally significantly below the average for peer companies. Further, the Compensation Committee noted that officers had led the company through very significant accomplishments during 2005, including the successful launch of Naglazyme, the creation of the partnership with Serono and the substantial progress with Phenoptin. Based on these factors, the Compensation Committee determined to grant the officers both salary increases and stock option awards effective in January 2006.

Further, the Compensation Committee believes that the cash bonus program was highly successful at motivating performance, reducing turnover and attracting qualified employees. Accordingly, the Compensation Committee and the full Board renewed the cash bonus program and updated the performance metrics based on goals for 2006.

Chief Executive Officer Compensation

In May 2005, the Company hired Jean-Jacques Bienaimé as the Company's Chief Executive Officer. His employment package was negotiated with the help of an independent compensation consultant. The Compensation Committee considered Mr. Bienaimé's extensive qualifications and demonstrated ability to create long term stockholder value at his prior companies. Based on these factors, the Compensation Committee and the full Board approved Mr. Bienaimé's employment agreement.

The Compensation Committee believes that the foregoing actions are recognition for and appropriate to the Company's achievements. The Compensation Committee further believes that the above authorized compensation actions based upon BioMarin achievements and competitive compensation levels will serve to help retain a highly qualified and motivated staff led by excellent senior management that is a requirement for the prosperity of BioMarin and the creation of stockholder value.

Respectfully submitted on March 3, 2006 by the members of the Compensation Committee of the Board of Directors:

Alan Lewis, Ph.D., Chairman

Michael Grey

Pierre Lapalme

The following is not soliciting material, is not deemed filed with the Securities and Exchange Commission and is not to be incorporated by reference into any filing of the Company under the Securities Act of 1933 or the Exchange Act, whether made before or after the date hereof and irrespective of any general incorporation by reference language in such filing.

REPORT OF THE AUDIT COMMITTEE

The Audit Committee reviews the Company's financial reporting process on behalf of the Board. Management has the primary responsibility for the financial statements and the reporting process, including the system of internal financial controls.

In this context, during fiscal year 2005, the Audit Committee met and held discussions with management and the independent registered public accounting firm for BioMarin. Management represented to the Audit Committee that the Company's consolidated financial statements were prepared in accordance with generally accepted accounting principles, and the Audit Committee has reviewed and discussed the consolidated financial statements with management and the independent registered public accounting firm. The Audit Committee discussed with the independent registered public accounting firm matters required to be discussed by Statement on Auditing Standards No. 61 (Communication With Audit Committees).

In addition, the Audit Committee has received from KPMG LLP the written disclosures and the letter required by the Independence Standards Board Standard No. 1 (Independence Discussions With Audit Committees) and has discussed with the independent registered public accounting firm, the firm's independence from the Company and its management.

The Audit Committee discussed with management and the independent registered public accounting firm for BioMarin the overall scope and plans for the 2005 annual audit. The Audit Committee met with management and with the independent registered public accounting firm, with and without management present, to discuss the results of their examination, the evaluations of the Company's internal financial controls, and the overall quality of the Company's financial reporting.

In reliance on the reviews and discussions referred to above, the Audit Committee recommended to the Board, and the Board has approved, that the audited financial statements be included in the Company's Annual Report on Form 10-K for the year ended December 31, 2005, for filing with the Securities and Exchange Commission and the SWX Swiss Exchange. The Audit Committee and the Board also have recommended the selection of KPMG LLP as the Company's independent registered public accounting firm for the year ending December 31, 2006.

Respectfully submitted on February 21, 2006 by the members of the Audit Committee of the Board of Directors:

Joseph Klein, III, Chairman

Elaine J. Heron, Ph.D.

Michael Grey

SECTION 16(A) BENEFICIAL OWNERSHIP REPORTING COMPLIANCE

Section 16(a) of the Securities Exchange Act of 1934, as amended (the Exchange Act), requires the Company's Directors and officers and persons who own more than 10% of a registered class of the Company's equity securities to file reports of ownership and reports of changes in the ownership with the Securities and Exchange Commission. Executive officers, Directors and greater than 10% stockholders are required by the Securities and Exchange Commission to furnish the Company with copies of all Section 16(a) forms they file.

To the best of the Company's knowledge, except as noted in the following sentence, based solely on a review of the copies of such reports furnished to the Company or written representation that no other reports were required, during the fiscal year ended December 31, 2005, all officers, Directors, and greater than 10% stockholders complied with all Section 16(a) filing requirements. Mr. Grey's initial Form 4 disclosing his option grant on appointment to the Board was filed approximately one week late.

OTHER MATTERS

Except as otherwise indicated, information contained herein is given as of May 5, 2006. The Company's management knows of no matters to come before the Annual Meeting other than the matters referred to in the Notice of Annual Meeting of Stockholders. The Board knows of no other business that will be presented at the meeting. The persons named in the enclosed proxy will vote the shares represented thereby in accordance with the recommendation of the Board as to any proposal properly presented at the Annual Meeting, or if no recommendation is made by the Board, then pursuant to the authority granted in the proxy.

The matters to be considered at the Annual Meeting are of great importance to the Company's stockholders. Accordingly, you are urged to read and carefully consider the information presented in this proxy statement, and to sign and date the enclosed WHITE Proxy Card and return it today in the enclosed pre-addressed postage-paid envelope.

IMPORTANT NOTE

YOUR VOTE IS IMPORTANT, NO MATTER HOW MANY OR HOW FEW SHARES YOU HOLD. Please vote only on the enclosed WHITE Proxy Card. Please sign and date the enclosed WHITE Proxy Card and return it today in the enclosed pre-addressed postage-paid envelope. Please do not complete any subsequently delivered proxy cards unless they are solicited by the Company. If your shares are held in street name, only your broker or bank can vote your shares and only upon receipt of your specific instructions. Please return the enclosed WHITE Proxy Card to your broker and contact the person responsible for your account to ensure that a WHITE Proxy Card is voted on your behalf. **IN ADDITION, TO ENSURE THAT THE PRESENCE OF A QUORUM AT THE MEETING MAY BE ASSURED, PLEASE SIGN AND DATE THE ENCLOSED WHITE PROXY CARD AND RETURN IT TODAY IN THE ENCLOSED PRE-ADDRESSED POSTAGE-PAID ENVELOPE.**

CONTACT FOR QUESTIONS AND ASSISTANCE IN VOTING

If you have any questions or need assistance in voting your shares, please call the firm assisting the Company in the solicitation of proxies:

Morrow & Co., Inc.

445 Park Avenue - 5th Floor

New York, NY 10022

1-800-607-0088

If you need additional copies of this proxy statement or voting materials, you should contact Morrow & Co., Inc. as described above.

APPROVAL

The contents of this proxy statement and the sending thereof to the stockholders have been authorized by the Board of Directors of the Company.

DATED this 5th day of May 2006 at Novato, California

G. Eric Davis

Vice President, General Counsel and Secretary

BIOMARIN PHARMACEUTICAL INC.
2006 SHARE INCENTIVE PLAN

As approved by the Board of
Directors on May 2, 2006

BIOMARIN PHARMACEUTICAL INC.

2006 SHARE INCENTIVE PLAN

PLAN DOCUMENT

1. *Establishment, Purpose, and Types of Awards*

BioMarin Pharmaceutical Inc. (the **Company**) hereby establishes this equity-based incentive compensation plan to be known as the BioMarin Pharmaceutical Inc. 2006 Share Incentive Plan (hereinafter referred to as the **Plan**), in order to provide incentives and awards to select employees, directors, consultants, and advisors of the Company and its Affiliates. The Plan permits grants of the following types of awards (**Awards**), according to the Sections of the Plan listed here:

| | |
|------------------|--|
| Section 6 | Options |
| Section 7 | Restricted Shares, Restricted Share Units, and Unrestricted Shares |
| Section 8 | Deferred Share Units |
| Section 9 | Performance Awards |

The Plan is not intended to affect and shall not affect any stock options, equity-based compensation, or other benefits that the Company or its Affiliates may have provided, or may separately provide in the future pursuant to any agreement, plan, or program that is independent of this Plan. Without limiting the foregoing, upon approval of the Plan by the stockholders of the Company, no further awards or grants shall be made under the Company's 1997 Stock Plan or the Company's 1998 Director Option Plan.

2. *Defined Terms*

Terms in the Plan that begin with an initial capital letter have the defined meaning set forth in **Appendix A**, unless defined elsewhere in this Plan or the context of their use clearly indicates a different meaning.

3. *Shares Subject to the Plan*

Subject to the provisions of **Section 12**, the maximum number of Shares that the Company may issue for all Awards is Fifteen Million (15,000,000) Shares. For all Awards, the Shares issued pursuant to the Plan may be authorized but unissued Shares, or Shares that the Company has reacquired or otherwise holds in treasury.

Shares that are subject to an Award that for any reason expires, is forfeited, is cancelled, or becomes unexercisable, and Shares that are for any other reason not paid or delivered under the Plan shall again, except to the extent prohibited by Applicable Law, be available for subsequent Awards under the Plan. Notwithstanding the foregoing, but subject to adjustments pursuant to **Section 12**, the number of Shares that are available for ISO Awards shall be determined, to the extent required under applicable tax laws, by reducing the number of Shares designated in the preceding paragraph by the number of Shares issued pursuant to Awards.

4. *Administration*

(a) *General.* The Committee shall administer the Plan in accordance with its terms, provided that the Board may act in lieu of the Committee on any matter. The Committee shall hold meetings at such times and places as it may determine and shall make such rules and regulations for the conduct of its business as it deems advisable. In the absence of a duly appointed Committee or if the Board otherwise chooses to act in lieu of the Committee, the Board shall function as the Committee for all purposes of the Plan.

(b) *Committee Composition.* The Board shall appoint the members of the Committee. If and to the extent permitted by Applicable Law, the Committee may authorize one or more Directors to make Awards to Eligible Persons who are not Reporting Persons. The Board may at any time appoint additional members to the

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Committee, remove and replace members of the Committee with or without Cause, and fill vacancies on the Committee however caused. Unless otherwise directed by the Board, the Committee shall be the Compensation Committee of the Board.

(c) *Powers of the Committee.* Subject to the provisions of the Plan, the Committee shall have the authority, in its sole discretion:

(i) to determine Eligible Persons to whom Awards shall be granted from time to time and the number of Shares or units to be covered by each Award;

(ii) to determine, from time to time, the Fair Market Value of Shares;

(iii) to determine, and to set forth in Award Agreements, the terms and conditions of all Awards, including any applicable exercise or purchase price, the installments and conditions under which an Award shall become vested (which may be based on performance), terminated, expired, cancelled, or replaced, and the circumstances for vesting acceleration or waiver of forfeiture restrictions, and other restrictions and limitations;

(iv) to approve the forms of Award Agreements and all other documents, notices and certificates in connection therewith which need not be identical either as to type of Award or among Participants;

(v) to construe and interpret the terms of the Plan and any Award Agreement, to determine the meaning of their terms, and to prescribe, amend, and rescind rules and procedures relating to the Plan and its administration;

(vi) in order to fulfill the purposes of the Plan and without amending the Plan, modify, cancel, or waive the Company's rights with respect to any Awards, to adjust or to modify Award Agreements for changes in Applicable Law, and to recognize differences in foreign law, tax policies, or customs; and

(vii) to make all other interpretations and to take all other actions that the Committee may consider necessary or advisable to administer the Plan or to effectuate its purposes.

Subject to Applicable Law and the restrictions set forth in the Plan, the Committee may delegate administrative functions to individuals who are Reporting Persons, officers, or Employees of the Company or its Affiliates.

(d) *Deference to Committee Determinations.* The Committee shall have the discretion to interpret or construe ambiguous, unclear, or implied (but omitted) terms in any fashion it deems to be appropriate in its sole discretion, and to make any findings of fact needed in the administration of the Plan or Award Agreements. The Committee's prior exercise of its discretionary authority shall not obligate it to exercise its authority in a like fashion thereafter. The Committee's interpretation and construction of any provision of the Plan, or of any Award or Award Agreement, shall be final, binding, and conclusive. The validity of any such interpretation, construction, decision or finding of fact shall not be given de novo review if challenged in court, by arbitration, or in any other forum, and shall be upheld unless clearly made in bad faith or materially affected by fraud.

(e) *Prohibition on Repricing.* Notwithstanding anything in this Plan to the contrary, unless the Company has obtained the consent of a majority of the Shareholders, in no event will the Committee or the Company authorize any amendment to the Plan, or to any Award under the Plan, that would effect a reduction in the exercise price of such Award, other than as a result of a stock split or other recapitalization as contemplated by **Section 12**. This restriction applies to both direct and indirect reductions, such as a surrender and regrant program.

(f) *No Liability; Indemnification.* Neither the Board nor any Committee member, nor any Person acting at the direction of the Board or the Committee, shall be liable for any act, omission, interpretation, construction or determination made in good faith with respect to the Plan, any Award or any Award Agreement. The Company and its Affiliates shall pay or reimburse any member of the Committee, as well as any Director, Employee, or Consultant who takes action in connection with the Plan, for all expenses incurred with respect to the Plan, and to the full extent allowable under Applicable Law shall indemnify each and every one of them for any claims, liabilities, and costs (including reasonable attorney's fees) arising out of their good faith performance of duties under the Plan. The Company and its Affiliates may obtain liability insurance for this purpose.

5. *Eligibility*

(a) *General Rule.* The Committee may grant ISOs only to Employees (including officers who are Employees) of the Company or an Affiliate that is a parent corporation or subsidiary corporation within the meaning of Section 424 of the Code, and may grant all other Awards to any Eligible Person. A Participant who has been granted an Award may be granted an additional Award or Awards if the Committee shall so determine, if such person is otherwise an Eligible Person and if otherwise in accordance with the terms of the Plan.

(b) *Grant of Awards.* Subject to the express provisions of the Plan, the Committee shall determine from the class of Eligible Persons those individuals to whom Awards under the Plan may be granted, the number of Shares subject to each Award, the price (if any) to be paid for the Shares or the Award and, in the case of Performance Awards, in addition to the matters addressed in **Section 9**, the specific objectives, goals and performance criteria that further define the Performance Award. Each Award shall be evidenced by an Award Agreement signed by the Company and, if required by the Committee, by the Participant. The Award Agreement shall set forth the material terms and conditions of the Award established by the Committee, and each Award shall be subject to the terms and conditions set forth in **Sections 22, 23, and 24** unless otherwise specifically provided in an Award Agreement.

(c) *Replacement Awards.* Subject to Applicable Laws (including any associated Shareholder approval requirements), the Committee may, in its sole discretion and upon such terms as it deems appropriate, require as a condition of the grant of an Award to a Participant that the Participant surrender for cancellation some or all of the Awards that have previously been granted to the Participant under this Plan or otherwise. An Award that is conditioned upon such surrender may or may not be the same type of Award, may cover the same (or a lesser or greater) number of Shares as such surrendered Award, may have other terms that are determined without regard to the terms or conditions of such surrendered Award, and may contain any other terms that the Committee deems appropriate. In the case of Options, these other terms may not involve an Exercise Price that is lower than the Exercise Price of the surrendered Option unless the Company's shareholders approve the grant itself or the program under which the grant is made pursuant to the Plan.

6. *Option Awards*

(a) *Types; Documentation.* Subject to **Section 5(a)**, the Committee may in its discretion grant Options pursuant to Award Agreements that are delivered to Participants. Each Option shall be designated in the Award Agreement as an ISO or a Non-ISO, and the same Award Agreement may grant both types of Options. At the sole discretion of the Committee, any Option may be exercisable, in whole or in part, immediately upon the grant thereof, or only after the occurrence of a specified event, or only in installments, which installments may vary. Options granted under the Plan may contain such terms and provisions not inconsistent with the Plan that the Committee shall deem advisable in its sole and absolute discretion.

(b) *ISO \$100,000 Limitation.* To the extent that the aggregate Fair Market Value of Shares with respect to which Options designated as ISOs first become exercisable by a Participant in any calendar year (under this Plan and any other plan of the Company or any Affiliate) exceeds \$100,000, such excess Options shall be treated as Non-ISOs. For purposes of determining whether the \$100,000 limit is exceeded, the Fair Market Value of the Shares subject to an ISO shall be determined as of the Grant Date. In reducing the number of Options treated as ISOs to meet the \$100,000 limit, the most recently granted Options shall be reduced first. In the event that Section 422 of the Code is amended to alter the limitation set forth therein, the limitation of this **Section 6(b)** shall be automatically adjusted accordingly.

(c) *Term of Options.* Each Award Agreement shall specify a term at the end of which the Option automatically expires, subject to earlier termination provisions contained in **Section 6(h)** hereof; provided, that, the term of any Option may not exceed ten years from the Grant Date. In the case of an ISO granted to an Employee who is a Ten Percent Holder on the Grant Date, the term of the ISO shall not exceed five years from the Grant Date.

(d) *Exercise Price.* The exercise price of an Option shall be determined by the Committee in its sole discretion and shall be set forth in the Award Agreement, provided that

(i) if an ISO is granted to an Employee who on the Grant Date is a Ten Percent Holder, the per Share exercise price shall not be less than 110% of the Fair Market Value per Share on the Grant Date, and

(ii) for all other Options, such per Share exercise price shall not be less than 100% of the Fair Market Value per Share on the Grant Date.

(e) *Exercise of Option.* The times, circumstances and conditions under which an Option shall be exercisable shall be determined by the Committee in its sole discretion and set forth in the Award Agreement. The Committee shall have the discretion to determine whether and to what extent the vesting of Options shall be tolled during any unpaid leave of absence; provided, however, that in the absence of such determination, vesting of Options shall be tolled during any such leave approved by the Company.

(f) *Minimum Exercise Requirements.* An Option may not be exercised for a fraction of a Share. The Committee may require in an Award Agreement that an Option be exercised as to a minimum number of Shares, provided that such requirement shall not prevent a Participant from purchasing the full number of Shares as to which the Option is then exercisable.

(g) *Methods of Exercise.* Prior to its expiration pursuant to the terms of the applicable Award Agreement, and subject to the times, circumstances and conditions for exercise contained in the applicable Award Agreement, each Option may be exercised, in whole or in part (provided that the Company shall not be required to issue fractional shares), by delivery of written notice of exercise to the secretary of the Company accompanied by the full exercise price of the Shares being purchased. In the case of an ISO, the Committee shall determine the acceptable methods of payment on the Grant Date and it shall be included in the applicable Award Agreement. The methods of payment that the Committee may in its discretion accept or commit to accept in an Award Agreement include:

(i) cash or check payable to the Company (in U.S. dollars);

(ii) other Shares that (A) are owned by the Participant who is purchasing Shares pursuant to an Option, (B) have a Fair Market Value on the date of surrender equal to the aggregate exercise price of the Shares as to which the Option is being exercised, (C) were not acquired by such Participant pursuant to the exercise of an Option, unless such Shares have been owned by such Participant for at least six months or such other period as the Committee may determine, (D) are all, at the time of such surrender, free and clear of any and all claims, pledges, liens and encumbrances, or any restrictions which would in any manner restrict the transfer of such shares to or by the Company (other than such restrictions as may have existed prior to an issuance of such Shares by the Company to such Participant), and (E) are duly endorsed for transfer to the Company;

(iii) a cashless exercise program that the Committee may approve, from time to time in its discretion, pursuant to which a Participant may concurrently provide irrevocable instructions (A) to such Participant's broker or dealer to effect the immediate sale of the purchased Shares and remit to the Company, out of the sale proceeds available on the settlement date, sufficient funds to cover the exercise price of the Option plus all applicable taxes required to be withheld by the Company by reason of such exercise, and (B) to the Company to deliver the certificates for the purchased Shares directly to such broker or dealer in order to complete the sale; or

(iv) any combination of the foregoing methods of payment.

The Company shall not be required to deliver Shares pursuant to the exercise of an Option until payment of the full exercise price therefore is received by the Company.

(h) *Termination of Continuous Service.* The Committee may establish and set forth in the applicable Award Agreement the terms and conditions on which an Option shall remain exercisable, if at all, following termination of a Participant's Continuous Service. The Committee may waive or modify these provisions at any time. To the extent that a Participant is not entitled to exercise an Option at the date of his or her termination of Continuous Service, or if the Participant (or other person entitled to exercise the Option) does not exercise the Option to the extent so entitled within the time specified in the Award Agreement or below (as applicable), the Option shall terminate and the Shares underlying the unexercised portion of the Option shall revert to the Plan and become available for future Awards. In no event may any Option be exercised after the expiration of the Option term as set forth in the Award Agreement.

The following provisions shall apply to the extent an Award Agreement does not specify the terms and conditions upon which an Option shall terminate when there is a termination of a Participant's Continuous Service:

(i) *Termination other than Upon Disability or Death or for Cause.* In the event of termination of a Participant's Continuous Service (other than as a result of Participant's death, disability, retirement or termination for Cause), the Participant shall have the right to exercise an Option at any time within 90 days following such termination to the extent the Participant was entitled to exercise such Option at the date of such termination.

(ii) *Disability.* In the event of termination of a Participant's Continuous Service as a result of his or her being Disabled, the Participant shall have the right to exercise an Option at any time within one year following such termination to the extent the Participant was entitled to exercise such Option at the date of such termination.

(iii) *Retirement.* In the event of termination of a Participant's Continuous Service as a result of Participant's retirement, the Participant shall have the right to exercise the Option at any time within six months following such termination to the extent the Participant was entitled to exercise such Option at the date of such termination.

(iv) *Death.* In the event of the death of a Participant during the period of Continuous Service since the Grant Date of an Option, or within thirty days following termination of the Participant's Continuous Service, the Option may be exercised, at any time within one year following the date of the Participant's death, by the Participant's estate or by a person who acquired the right to exercise the Option by bequest or inheritance, but only to the extent the right to exercise the Option had vested at the date of death or, if earlier, the date the Participant's Continuous Service terminated.

(v) *Cause.* If the Committee determines that a Participant's Continuous Service terminated due to Cause, the Participant shall immediately forfeit the right to exercise any Option, and it shall be considered immediately null and void.

(i) *Reverse Vesting.* The Committee in its sole discretion may allow a Participant to exercise unvested Options, in which case the Shares then issued shall be Restricted Shares having analogous vesting restrictions to the unvested Options.

7. Restricted Shares, Restricted Share Units, and Unrestricted Shares

(a) *Grants.* The Committee may in its sole discretion grant restricted shares (**Restricted Shares**) to any Eligible Person and shall evidence such grant in an Award Agreement that is delivered to the Participant and that sets forth the number of Restricted Shares, the purchase price for such Restricted Shares (if any), and the terms upon which the Restricted Shares may become vested. In addition, the Company may in its discretion grant to any Eligible Person the right to receive Shares after certain vesting requirements are met (**Restricted Share Units**), and shall evidence such grant in an Award Agreement that is delivered to the Participant and that sets forth the number of Shares (or formula, that may be based on future performance or conditions, for determining the number of Shares) that the Participant shall be entitled to receive upon vesting and the terms upon which the

Shares subject to a Restricted Share Unit may become vested and the delivery terms for such Shares. The Committee may condition any Award of Restricted Shares or Restricted Share Units to a Participant on receiving from the Participant such further assurances and documents as the Committee may require to enforce the restrictions. In addition, the Committee may grant Awards hereunder in the form of unrestricted shares (**Unrestricted Shares**), which shall vest in full upon the date of grant or such other date as the Committee may determine or which the Committee may issue pursuant to any program under which one or more Eligible Persons (selected by the Committee in its sole discretion) elect to pay for such Shares or to receive Unrestricted Shares in lieu of cash bonuses that would otherwise be paid.

(b) *Vesting and Forfeiture.* The Committee shall set forth in an Award Agreement granting Restricted Shares or Restricted Share Units, the terms and conditions under which the Participant's interest in the Restricted Shares or the Shares subject to Restricted Share Units will become vested and non-forfeitable. Except as set forth in the applicable Award Agreement or the Committee otherwise determines, upon termination of a Participant's Continuous Service for any other reason, the Participant shall forfeit his or her Restricted Shares and unvested Restricted Share Units; provided that if a Participant purchases the Restricted Shares and forfeits them for any reason, the Company shall return the purchase price to the Participant only if and to the extent set forth in an Award Agreement.

(c) *Issuance of Restricted Shares Prior to Vesting.* The Company shall issue stock certificates that evidence Restricted Shares pending the lapse of applicable restrictions, and that bear a legend making appropriate reference to such restrictions. Except as set forth in the applicable Award Agreement or the Committee otherwise determines, the Company or a third party that the Company designates shall hold such Restricted Shares and any dividends that accrue with respect to Restricted Shares pursuant to **Section 7(e)**.

(d) *Issuance of Shares upon Vesting.* As soon as practicable after vesting of a Participant's Restricted Shares (or Shares underlying Restricted Share Units) and the Participant's satisfaction of applicable tax withholding requirements, the Company shall release to the Participant, free from the vesting restrictions, one Share for each vested Restricted Share (or issue one Share free of the vesting restriction for each vested Restricted Share Unit), unless an Award Agreement provides otherwise. No fractional shares shall be distributed, and cash shall be paid in lieu thereof.

(e) *Dividends Payable on Vesting.* Unless otherwise provided in an Award Agreement, whenever unrestricted Shares are issued to a Participant pursuant to **Section 7(d)**, the Participant shall also receive, with respect to each Share issued, (i) a number of Shares equal to the stock dividends which were declared and paid to the holders of Shares between the Grant Date and the date such Share is issued, and (ii) a number of Shares having a Fair Market Value equal to any cash dividends that were paid to the holders of Shares based on a record date between the Grant Date and the date such Share is issued.

(f) *Section 83(b) Elections.* A Participant may make an election under Section 83(b) of the Code (the **Section 83(b) Election**) with respect to Restricted Shares. If a Participant who has received Restricted Share Units provides the Committee with written notice of his or her intention to make a Section 83(b) Election with respect to the Shares subject to such Restricted Share Units, the Committee may in its discretion convert the Participant's Restricted Share Units into Restricted Shares, on a one-for-one basis, in full satisfaction of the Participant's Restricted Share Unit Award. The Participant may then make a Section 83(b) Election with respect to those Restricted Shares. Shares with respect to which a Participant makes a Section 83(b) Election shall not be eligible for deferral pursuant to **Section 8**.

(g) *Deferral Elections.* At any time within the thirty-day period (or other shorter or longer period that the Committee selects in its sole discretion) in which a Participant who is a member of a select group of management or highly compensated employees (within the meaning of the Code) receives an Award of either Restricted Shares or Restricted Share Units, the Committee may permit the Participant to irrevocably elect, on a form provided by and acceptable to the Committee, to defer the receipt of all or a percentage of the Shares that would

otherwise be transferred to the Participant upon the vesting of such Award. If the Participant makes this election, the Shares subject to the election, and any associated dividends and interest, shall be credited to an account established pursuant to **Section 8** on the date such Shares would otherwise have been released or issued to the Participant pursuant to **Section 7(d)**.

8. *Deferred Share Units*

(a) *Elections to Defer.* The Committee may permit any Eligible Person who is a Director, Consultant or member of a select group of management or highly compensated employees (within the meaning of the Code) to irrevocably elect, on a form provided by and acceptable to the Committee (the **Election Form**), to forego the receipt of cash or other compensation (including the Shares deliverable pursuant to any Award other than Restricted Shares for which a Section 83(b) Election has been made), and in lieu thereof to have the Company credit to an internal Plan account (the **Account**) a number of deferred share units (**Deferred Share Units**) having a Fair Market Value equal to the Shares and other compensation deferred. These credits will be made at the end of each calendar month during which compensation is deferred. Each Election Form shall take effect on the first day of the next calendar year (or on the first day of the next calendar month in the case of an initial election by a Participant who first becomes eligible to defer hereunder) after its delivery to the Company, subject to **Section 7(g)** regarding deferral of Restricted Shares and Restricted Share Units and to **Section 9(e)** regarding deferral of Performance Awards, unless the Company sends the Participant a written notice explaining why the Election Form is invalid within five business days after the Company receives it. Notwithstanding the foregoing sentence: (i) Election Forms shall be ineffective with respect to any compensation that a Participant earns before the date on which the Company receives the Election Form, and (ii) the Committee may unilaterally make awards in the form of Deferred Share Units, regardless of whether or not the Participant foregoes other compensation.

(b) *Vesting.* Unless an Award Agreement expressly provides otherwise, each Participant shall be 100% vested at all times in any Shares subject to Deferred Share Units.

(c) *Issuances of Shares.* The Company shall provide a Participant with one Share for each Deferred Share Unit in five substantially equal annual installments that are issued before the last day of each of the five calendar years that end after the date on which the Participant's Continuous Service terminates, *unless*

(i) the Participant has properly elected a different form of distribution, on a form approved by the Committee, that permits the Participant to select any combination of a lump sum and annual installments that are completed within ten years following termination of the Participant's Continuous Service, and

(ii) the Company received the Participant's distribution election form at the time the Participant elects to defer the receipt of cash or other compensation pursuant to **Section 8(a)**, provided that such election may be changed through any subsequent election that (i) is delivered to the Company at least one year before the date on which distributions are otherwise scheduled to commence pursuant to the Participant's election, and (ii) defers the commencement of distributions by at least five years from the originally scheduled commencement date.

Fractional shares shall not be issued, and instead shall be paid out in cash.

(d) *Crediting of Dividends.* Unless otherwise provided in an Award Agreement, whenever Shares are issued to a Participant pursuant to **Section 8(c)**, the Participant shall also receive, with respect to each Share issued, (i) a number of Shares equal to any stock dividends which were declared and paid to the holders of Shares between the Grant Date and the date such Share is issued, and (ii) a number of Shares having a Fair Market Value equal to any cash dividends that were paid to the holders of Shares based on a record date between the Grant Date and the date such Share is issued.

(e) *Emergency Withdrawals.* In the event a Participant suffers an unforeseeable emergency within the contemplation of this **Section 8(e)** and Section 409A of the Code, the Participant may apply to the Company for

an immediate distribution of all or a portion of the Participant's Deferred Share Units. The unforeseeable emergency must result from a sudden and unexpected illness or accident of the Participant, the Participant's spouse, or a dependent (within the meaning of Section 152(a) of the Code) of the Participant, casualty loss of the Participant's property, or other similar extraordinary and unforeseeable conditions beyond the control of the Participant. Examples of purposes which are not considered unforeseeable emergencies include post-secondary school expenses or the desire to purchase a residence. In no event will a distribution be made to the extent the unforeseeable emergency could be relieved through reimbursement or compensation by insurance or otherwise, or by liquidation of the Participant's nonessential assets to the extent such liquidation would not itself cause a severe financial hardship. The amount of any distribution hereunder shall be limited to the amount necessary to relieve the Participant's unforeseeable emergency plus amounts necessary to pay taxes reasonably anticipated as a result of the distribution. The Committee shall determine whether a Participant has a qualifying unforeseeable emergency and the amount which qualifies for distribution, if any. The Committee may require evidence of the purpose and amount of the need, and may establish such application or other procedures as it deems appropriate.

(f) *Unsecured Rights to Deferred Compensation.* A Participant's right to Deferred Share Units shall at all times constitute an unsecured promise of the Company to pay benefits as they come due. The right of the Participant or the Participant's duly-authorized transferee to receive benefits hereunder shall be solely an unsecured claim against the general assets of the Company. Neither the Participant nor the Participant's duly-authorized transferee shall have any claim against or rights in any specific assets, shares, or other funds of the Company.

9. *Performance Awards*

(a) *Performance Units.* The Committee may in its discretion grant Performance Units to any Eligible Person and shall evidence such grant in an Award Agreement that is delivered to the Participant which sets forth the terms and conditions of the Award.

(b) *Performance Compensation Awards.* The Committee may, at the time of grant of a Performance Unit, designate such Award as a Performance Compensation Award (payable in cash or Shares) in order that such Award constitutes qualified performance-based compensation under Code Section 162(m), in which event the Committee shall have the power to grant such Performance Compensation Award upon terms and conditions that qualify it as qualified performance-based compensation within the meaning of Code Section 162(m). With respect to each such Performance Compensation Award, the Committee shall establish, in writing within the time required under Code Section 162(m), a Performance Period, Performance Measure(s), and Performance Formula(e) (each such term being hereinafter defined).

A Participant shall be eligible to receive payment in respect of a Performance Compensation Award only to the extent that the Performance Measure(s) for such Award is achieved and the Performance Formula(e) as applied against such Performance Measure(s) determines that all or some portion of such Participant's Award has been earned for the Performance Period. As soon as practicable after the close of each Performance Period, the Committee shall review and certify in writing whether, and to what extent, the Performance Measure(s) for the Performance Period have been achieved and, if so, determine and certify in writing the amount of the Performance Compensation Award to be paid to the Participant and, in so doing, may use negative discretion to decrease, but not increase, the amount of the Award otherwise payable to the Participant based upon such performance.

(c) *Definitions.*

(i) **Performance Formula** means, for a Performance Period, one or more objective formulas or standards established by the Committee for purposes of determining whether or the extent to which an Award has been earned based on the level of performance attained or to be attained with respect to one or more Performance Measure(s). Performance Formulae may vary from Performance Period to Performance Period and from Participant to Participant and may be established on a stand-alone basis, in tandem or in the alternative.

(ii) **Performance Measure** means one or more of the following selected by the Committee to measure Company, Affiliate, and/or business unit performance for a Performance Period, whether in absolute or relative terms (including, without limitation, terms relative to a peer group or index): basic, diluted, or adjusted earnings per share; sales or revenue; earnings before interest, taxes, and other adjustments (in total or on a per share basis); basic or adjusted net income; returns on equity, assets, capital, revenue or similar measure; economic value added; working capital; total shareholder return; and product development, product market share, research, licensing, litigation, human resources, information services, mergers, acquisitions, sales of assets of Affiliates or business units. Each such measure shall be, to the extent applicable, determined in accordance with generally accepted accounting principles as consistently applied by the Company (or such other standard applied by the Committee) and, if so determined by the Committee, and in the case of a Performance Compensation Award, to the extent permitted under Code Section 162(m), adjusted to omit the effects of extraordinary items, gain or loss on the disposal of a business segment, unusual or infrequently occurring events and transactions and cumulative effects of changes in accounting principles. Performance Measures may vary from Performance Period to Performance Period and from Participant to Participant, and may be established on a stand-alone basis, in tandem or in the alternative.

(iii) **Performance Period** means one or more periods of time (of not less than one fiscal year of the Company), as the Committee may designate, over which the attainment of one or more Performance Measure(s) will be measured for the purpose of determining a Participant's rights in respect of an Award.

(d) *Deferral Elections.* At any time prior to the date that is at least six months before the close of a Performance Period (or shorter or longer period that the Committee selects) with respect to an Award of either Performance Units or Performance Compensation, the Committee may permit a Participant who is a member of a select group of management or highly compensated employees (within the meaning of the Code) to irrevocably elect, on a form provided by and acceptable to the Committee, to defer the receipt of all or a percentage of the cash or Shares that would otherwise be transferred to the Participant upon the vesting of such Award. If the Participant makes this election, the cash or Shares subject to the election, and any associated interest and dividends, shall be credited to an account established pursuant to **Section 8** on the date such cash or Shares would otherwise have been released or issued to the Participant pursuant to **Section 9(a)** or **Section 9(b)**.

10. **Taxes**

(a) *General.* As a condition to the issuance or distribution of Shares pursuant to the Plan, the Participant (or in the case of the Participant's death, the person who succeeds to the Participant's rights) shall make such arrangements as the Company may require for the satisfaction of any applicable federal, state, local or foreign withholding tax obligations that may arise in connection with the Award and the issuance of Shares. The Company shall not be required to issue any Shares until such obligations are satisfied. If the Committee allows the withholding or surrender of Shares to satisfy a Participant's tax withholding obligations, the Committee shall not allow Shares to be withheld in an amount that exceeds the minimum statutory withholding rates for federal and state tax purposes, including payroll taxes.

(b) *Default Rule for Employees.* In the absence of any other arrangement, an Employee shall be deemed to have directed the Company to withhold or collect from his or her cash compensation an amount sufficient to satisfy such tax obligations from the next payroll payment otherwise payable after the date of the exercise of an Award.

(c) *Special Rules.* In the case of a Participant other than an Employee (or in the case of an Employee where the next payroll payment is not sufficient to satisfy such tax obligations, with respect to any remaining tax obligations), in the absence of any other arrangement and to the extent permitted under Applicable Law, the Participant shall be deemed to have elected to have the Company withhold from the Shares or cash to be issued pursuant to an Award that number of Shares having a Fair Market Value determined as of the applicable Tax Date (as defined below) or cash equal to the amount required to be withheld. For purposes of this **Section 10**, the

Fair Market Value of the Shares to be withheld shall be determined on the date that the amount of tax to be withheld is to be determined under the Applicable Law (the **Tax Date**).

(d) *Surrender of Shares.* If permitted by the Committee, in its discretion, a Participant may satisfy the minimum applicable tax withholding and employment tax obligations associated with an Award by surrendering Shares to the Company (including Shares that would otherwise be issued pursuant to the Award) that have a Fair Market Value determined as of the applicable Tax Date equal to the amount required to be withheld. In the case of Shares previously acquired from the Company that are surrendered under this **Section 10**, such Shares must have been owned by the Participant for more than six months on the date of surrender (or such longer period of time the Company may in its discretion require).

(e) *Income Taxes and Deferred Compensation.* Participants are solely responsible and liable for the satisfaction of all taxes and penalties that may arise in connection with Awards (including any taxes arising under Section 409A of the Code), and the Company shall not have any obligation to indemnify or otherwise hold any Participant harmless from any or all of such taxes. The Committee shall have the discretion to organize any deferral program, to require deferral election forms, and to grant or to unilaterally modify any Award in a manner that (i) conforms with the requirements of Section 409A of the Code with respect to compensation that is deferred and that vests after December 31, 2004, (ii) that voids any Participant election to the extent it would violate Section 409A of the Code, and (iii) for any distribution election that would violate Section 409A of the Code, to make distributions pursuant to the Award at the earliest to occur of a distribution event that is allowable under Section 409A of the Code or any distribution event that is both allowable under Section 409A of the Code and is elected by the Participant, subject to any valid second election to defer, provided that the Committee permits second elections to defer in accordance with Section 409A(a)(4)(C). The Committee shall have the sole discretion to interpret the requirements of the Code, including Section 409A, for purposes of the Plan and all Awards.

11. *Non-Transferability of Awards*

(a) *General.* Except as set forth in this **Section 11**, or as otherwise approved by the Committee, Awards may not be sold, pledged, assigned, hypothecated, transferred or disposed of in any manner other than by will or by the laws of descent or distribution, or in the case of an option other than an ISO, pursuant to a domestic relations order as defined under Rule 16a-12 under the Exchange Act. The designation of a beneficiary by a Participant will not constitute a transfer. An Award may be exercised, during the lifetime of the holder of an Award, only by such holder, the duly-authorized legal representative of a Participant who is Disabled, a transferee permitted by this **Section 11**, or except as would cause an ISO to lose such status, by a bankruptcy trustee.

(b) *Limited Transferability Rights.* Notwithstanding anything else in this **Section 11**, the Committee may in its discretion provide in an Award Agreement that an Award relating to non-ISOs, Restricted Shares, or Performance Shares may be transferred, on such terms and conditions as the Committee deems appropriate, either (i) by instrument to the Participant's Immediate Family (as defined below), (ii) by instrument to an inter vivos or testamentary trust (or other entity) in which the Award is to be passed to the Participant's designated beneficiaries, or (iii) by gift to charitable institutions. Each share of restricted stock shall be non-transferable until such share becomes non-forfeitable. Any transferee of the Participant's rights shall succeed and be subject to all of the terms of the applicable Award Agreement and the Plan. Immediate Family means any child, stepchild, grandchild, parent, stepparent, grandparent, spouse, former spouse, sibling, niece, nephew, mother-in-law, father-in-law, son-in-law, daughter-in-law, brother-in-law, or sister-in-law, and shall include adoptive relationships.

12. *Adjustments Upon Changes in Capitalization, Merger or Certain Other Transactions*

(a) *Changes in Capitalization.* The Committee shall equitably adjust the number of Shares covered by each outstanding Award, and the number of Shares that have been authorized for issuance under the Plan but as

to which no Awards have yet been granted or that have been returned to the Plan upon cancellation, forfeiture, or expiration of an Award, as well as the price per Share covered by each such outstanding Award, to reflect any increase or decrease in the number of issued Shares resulting from a stock-split, reverse stock-split, stock dividend, combination, recapitalization or reclassification of the Shares, or any other increase or decrease in the number of issued Shares effected without receipt of consideration by the Company. In the event of any such transaction or event, the Committee may provide in substitution for any or all outstanding Options under the Plan such alternative consideration (including securities of any surviving entity) as it may in good faith determine to be equitable under the circumstances and may require in connection therewith the surrender of all Options so replaced. In any case, such substitution of securities shall not require the consent of any person who is granted Options pursuant to the Plan. Except as expressly provided herein, or in an Award Agreement, if the Company issues for consideration shares of stock of any class or securities convertible into shares of stock of any class, the issuance shall not affect, and no adjustment by reason thereof shall be required to be made with respect to the number or price of Shares subject to any Award.

(b) *Dissolution or Liquidation.* In the event of the dissolution or liquidation of the Company other than as part of a Change of Control, each Award will terminate immediately prior to the consummation of such action, subject to the ability of the Committee to exercise any discretion authorized in the case of a Change in Control.

(c) *Change in Control.* In the event of a Change in Control, the Committee may in its sole and absolute discretion and authority, without obtaining the approval or consent of the Company's shareholders or any Participant with respect to his or her outstanding Awards, take one or more of the following actions:

(i) arrange for or otherwise provide that each outstanding Award shall be assumed or a substantially similar award shall be substituted by a successor corporation or a parent or subsidiary of such successor corporation (the *Successor Corporation*);

(ii) accelerate the vesting of Awards so that Awards shall vest (and, to the extent applicable, become exercisable) as to the Shares that otherwise would have been unvested and provide that repurchase rights of the Company with respect to Shares issued upon exercise of an Award shall lapse as to the Shares subject to such repurchase right;

(iii) arrange or otherwise provide for the payment of cash or other consideration to Participants in exchange for the satisfaction and cancellation of outstanding Awards;

(iv) terminate upon the consummation of the transaction, provided that the Committee may in its sole discretion provide for vesting of all or some outstanding Awards in full as of a date immediately prior to consummation of the Change of Control. To the extent that an Award is not exercised prior to consummation of a transaction in which the Award is not being assumed or substituted, such Award shall terminate upon such consummation; or

(v) make such other modifications, adjustments or amendments to outstanding Awards or this Plan as the Committee deems necessary or appropriate, subject however to the terms of **Section 14(a)**.

Notwithstanding the above, in the event a Participant holding an Award assumed or substituted by the Successor Corporation in a Change in Control is Involuntarily Terminated by the Successor Corporation in connection with, or within 12 months following consummation of, the Change in Control, then any assumed or substituted Award held by the terminated Participant at the time of termination shall accelerate and become fully vested (and exercisable in full in the case of Options), and any repurchase right applicable to any Shares shall lapse in full, unless an Award Agreement provides for a more restrictive acceleration or vesting schedule or more restrictive limitations on the lapse of repurchase rights or otherwise places additional restrictions, limitations and conditions on an Award. The acceleration of vesting and lapse of repurchase rights provided for in the previous sentence shall occur immediately prior to the effective date of the Participant's termination, unless an Award Agreement provides otherwise.

(d) *Certain Distributions.* In the event of any distribution to the Company's shareholders of securities of any other entity or other assets (other than dividends payable in cash or stock of the Company) without receipt of consideration by the Company, the Committee may, in its discretion, appropriately adjust the price per Share covered by each outstanding Award to reflect the effect of such distribution.

13. *Time of Granting Awards.*

The date of grant (**Grant Date**) of an Award shall be the date on which the Committee makes the determination granting such Award or such other date as is determined by the Committee and set forth in the Award Agreement, provided that in the case of an ISO, the Grant Date shall be the later of the date on which the Committee makes the determination granting such ISO or the date of commencement of the Participant's employment relationship with the Company.

14. *Modification of Awards and Substitution of Options.*

(a) *Modification, Extension, and Renewal of Awards.* Within the limitations of the Plan, the Committee may modify an Award to accelerate the rate at which an Option may be exercised (including without limitation permitting an Option to be exercised in full without regard to the installment or vesting provisions of the applicable Award Agreement or whether the Option is at the time exercisable, to the extent it has not previously been exercised), to accelerate the vesting of any Award, to extend or renew outstanding Awards or to accept the cancellation of outstanding Awards to the extent not previously exercised. However, the Committee may not cancel an outstanding option that is underwater for the purpose of reissuing the option to the participant at a lower exercise price or granting a replacement award of a different type. Notwithstanding the foregoing provision, no modification of an outstanding Award shall materially and adversely affect such Participant's rights thereunder, unless either the Participant provides written consent or there is an express Plan provision permitting the Committee to act unilaterally to make the modification.

(b) *Substitution of Options.* Notwithstanding any inconsistent provisions or limits under the Plan, in the event the Company or an Affiliate acquires (whether by purchase, merger or otherwise) all or substantially all of outstanding capital stock or assets of another corporation or in the event of any reorganization or other transaction qualifying under Section 424 of the Code, the Committee may, in accordance with the provisions of that Section, substitute Options for options under the plan of the acquired company provided (i) the excess of the aggregate fair market value of the shares subject to an option immediately after the substitution over the aggregate option price of such shares is not more than the similar excess immediately before such substitution and (ii) the new option does not give persons additional benefits, including any extension of the exercise period.

15. *Term of Plan.*

The Plan shall continue in effect for a term of ten (10) years from its effective date as determined under **Section 19**, unless the Plan is sooner terminated under **Section 16**.

16. *Amendment and Termination of the Plan.*

(a) *Authority to Amend or Terminate.* Subject to Applicable Laws, the Board may from time to time amend, alter, suspend, discontinue, or terminate the Plan.

(b) *Effect of Amendment or Termination.* No amendment, suspension, or termination of the Plan shall materially and adversely affect Awards already granted unless either it relates to an adjustment pursuant to **Section 12**, or it is otherwise mutually agreed between the Participant and the Committee, which agreement must be in writing and signed by the Participant and the Company. Notwithstanding the foregoing, the Committee may amend the Plan to eliminate provisions which are no longer necessary as a result of changes in tax or securities laws or regulations, or in the interpretation thereof.

17. Conditions Upon Issuance of Shares.

Notwithstanding any other provision of the Plan or any agreement entered into by the Company pursuant to the Plan, the Company shall not be obligated, and shall have no liability for failure, to issue or deliver any Shares under the Plan unless such issuance or delivery would comply with Applicable Law, with such compliance determined by the Company in consultation with its legal counsel.

18. Reservation of Shares.

The Company, during the term of this Plan, will at all times reserve and keep available such number of Shares as shall be sufficient to satisfy the requirements of the Plan.

19. Effective Date.

This Plan shall become effective on the date of its approval by the Board; provided that this Plan shall be submitted to the Company's stockholders for approval, and if not approved by the stockholders in accordance with Applicable Laws (as determined by the Committee in its sole discretion) within one year from the date of approval by the Board, this Plan and any Awards shall be null, void, and of no force and effect. Awards granted under this Plan before approval of this Plan by the shareholders shall be granted subject to such approval, and no Shares shall be distributed before such approval.

20. Controlling Law.

All disputes relating to or arising from the Plan shall be governed by the internal substantive laws (and not the laws of conflicts of laws) of the State of Delaware, to the extent not preempted by United States federal law. If any provision of this Plan is held by a court of competent jurisdiction to be invalid and unenforceable, the remaining provisions shall continue to be fully effective.

21. Laws And Regulations.

(a) *U.S. Securities Laws.* This Plan, the grant of Awards, and the exercise of Options under this Plan, and the obligation of the Company to sell or deliver any of its securities (including, without limitation, Options, Restricted Shares, Restricted Share Units, Deferred Share Units, and Shares) under this Plan shall be subject to all Applicable Law. In the event that the Shares are not registered under the Securities Act of 1933, as amended (the "Act"), or any applicable state securities laws prior to the delivery of such Shares, the Company may require, as a condition to the issuance thereof, that the persons to whom Shares are to be issued represent and warrant in writing to the Company that such Shares are being acquired by him or her for investment for his or her own account and not with a view to, for resale in connection with, or with an intent of participating directly or indirectly in, any distribution of such Shares within the meaning of the Act, and a legend to that effect may be placed on the certificates representing the Shares.

(b) *Other Jurisdictions.* To facilitate the making of any grant of an Award under this Plan, the Committee may provide for such special terms for Awards to Participants who are foreign nationals or who are employed by the Company or any Affiliate outside of the United States of America as the Committee may consider necessary or appropriate to accommodate differences in local law, tax policy or custom. The Company may adopt rules and procedures relating to the operation and administration of this Plan to accommodate the specific requirements of local laws and procedures of particular countries. Without limiting the foregoing, the Company is specifically authorized to adopt rules and procedures regarding the conversion of local currency, taxes, withholding procedures and handling of stock certificates which vary with the customs and requirements of particular countries. The Company may adopt sub-plans and establish escrow accounts and trusts as may be appropriate or applicable to particular locations and countries.

22. No Shareholder Rights. Neither a Participant nor any transferee of a Participant shall have any rights as a shareholder of the Company with respect to any Shares underlying any Award until the date of issuance of a share certificate to a Participant or a transferee of a Participant for such Shares in accordance with the

Company's governing instruments and Applicable Law. Prior to the issuance of Shares pursuant to an Award, a Participant shall not have the right to vote or to receive dividends or any other rights as a shareholder with respect to the Shares underlying the Award, notwithstanding its exercise in the case of Options. No adjustment will be made for a dividend or other right that is determined based on a record date prior to the date the stock certificate is issued, except as otherwise specifically provided for in this Plan.

23. **No Employment Rights.** The Plan shall not confer upon any Participant any right to continue an employment, service or consulting relationship with the Company, nor shall it affect in any way a Participant's right or the Company's right to terminate the Participant's employment, service, or consulting relationship at any time, with or without Cause.

24. **Termination, Rescission and Recapture of Awards.** Notwithstanding any other provision of the Plan, but subject to any contrary terms set forth in any Award Agreement, this **Section 24** shall only apply to a Participant who is, on the Award Date, an Employee of the Company or its Affiliates, and shall automatically cease to apply to any Participant from and after his or her termination of Continuous Service after a Change in Control.

(a) Each Award under the Plan is intended to align the Participant's long-term interest with those of the Company. If the Participant engages in certain activities discussed below, either during employment or after employment with the Company terminates for any reason, the Participant is acting contrary to the long-term interests of the Company. Accordingly, except as otherwise expressly provided in the Award Agreement, the Company may terminate any outstanding, unexercised, unexpired, unpaid, or deferred Awards (**Termination**), rescind any exercise, payment or delivery pursuant to the Award (**Rescission**), or recapture any Common Stock (whether restricted or unrestricted) or proceeds from the Participant's sale of Shares issued pursuant to the Award (**Recapture**), if the Participant does not comply with the conditions of subsections (b) and (c) hereof (collectively, the **Conditions**).

(b) A Participant shall not, without the Company's prior written authorization, disclose to anyone outside the Company, or use in other than the Company's business, any proprietary or confidential information or material, as those or other similar terms are used in any applicable patent, confidentiality, inventions, secrecy, or other agreement between the Participant and the Company with regard to any such proprietary or confidential information or material.

(c) Pursuant to any agreement between the Participant and the Company with regard to intellectual property (including but not limited to patents, trademarks, copyrights, trade secrets, inventions, developments, improvements, proprietary information, confidential business and personnel information), a Participant shall promptly disclose and assign to the Company or its designee all right, title, and interest in such intellectual property, and shall take all reasonable steps necessary to enable the Company to secure all right, title and interest in such intellectual property in the United States and in any foreign country.

(d) Upon exercise, payment, or delivery of cash or Common Stock pursuant to an Award, the Participant shall certify on a form acceptable to the Company that he or she is in compliance with the terms and conditions of the Plan and, if a severance of Continuous Service has occurred for any reason, shall state the name and address of the Participant's then-current employer or any entity for which the Participant performs business services and the Participant's title, and shall identify any organization or business in which the Participant owns a greater-than-five-percent equity interest.

(e) If the Company determines, in its sole and absolute discretion, that (i) a Participant has violated any of the Conditions or (ii) during his or her Continuous Service, or within 2 years after its termination for any reason, a Participant (a) has rendered services to or otherwise directly or indirectly engaged in or assisted, any organization or business that, in the judgment of the Company in its sole and absolute discretion, is or is working to become competitive with the Company; (b) has solicited any non-administrative employee of the Company to

terminate employment with the Company; or (c) has engaged in activities which are materially prejudicial to or in conflict with the interests of the Company, including any breaches of fiduciary duty or the duty of loyalty, then the Company may, in its sole and absolute discretion, impose a Termination, Rescission, and/or Recapture with respect to any or all of the Participant's relevant Awards, Shares, and the proceeds thereof.

(f) Within ten days after receiving notice from the Company of any such activity, the Participant shall deliver to the Company the Shares acquired pursuant to the Award, or, if Participant has sold the Shares, the gain realized, or payment received as a result of the rescinded exercise, payment, or delivery; provided, that if the Participant returns Shares that the Participant purchased pursuant to the exercise of an Option (or the gains realized from the sale of such Common Stock), the Company shall promptly refund the exercise price, without earnings, that the Participant paid for the Shares. Any payment by the Participant to the Company pursuant to this **Section 24** shall be made either in cash or by returning to the Company the number of Shares that the Participant received in connection with the rescinded exercise, payment, or delivery. It shall not be a basis for Termination, Rescission or Recapture if after termination of a Participant's Continuous Service, the Participant purchases, as an investment or otherwise, stock or other securities of such an organization or business, so long as (i) such stock or other securities are listed upon a recognized securities exchange or traded over-the-counter, and (ii) such investment does not represent more than a five percent (5%) equity interest in the organization or business.

(g) Notwithstanding the foregoing provisions of this Section, the Company has sole and absolute discretion not to require Termination, Rescission and/or Recapture, and its determination not to require Termination, Rescission and/or Recapture with respect to any particular act by a particular Participant or Award shall not in any way reduce or eliminate the Company's authority to require Termination, Rescission and/or Recapture with respect to any other act or Participant or Award. Nothing in this Section shall be construed to impose obligations on the Participant to refrain from engaging in lawful competition with the Company after the termination of employment that does not violate subsections (b) or (c) of this Section, other than any obligations that are part of any separate agreement between the Company and the Participant or that arise under applicable law.

(h) All administrative and discretionary authority given to the Company under this Section shall be exercised by the most senior human resources executive of the Company or such other person or committee (including without limitation the Committee) as the Committee may designate from time to time.

(i) Notwithstanding any provision of this Section, if any provision of this Section is determined to be unenforceable or invalid under any applicable law, such provision will be applied to the maximum extent permitted by applicable law, and shall automatically be deemed amended in a manner consistent with its objectives to the extent necessary to conform to any limitations required under applicable law. Furthermore, if any provision of this Section is illegal under any applicable law, such provision shall be null and void to the extent necessary to comply with applicable law.

Biomarin Pharmaceutical Inc.

SHARE INCENTIVE PLAN

Appendix A: Definitions

As used in the Plan, the following definitions shall apply:

Affiliate means, with respect to any Person (as defined below), any other Person that directly or indirectly controls or is controlled by or under common control with such Person. For the purposes of this definition, control, when used with respect to any Person, means the possession, direct or indirect, of the power to direct or cause the direction of the management and policies of such Person or the power to elect directors, whether through the ownership of voting securities, by contract or otherwise; and the terms affiliated, controlling and controlled have meanings correlative to the foregoing.

Applicable Law means the legal requirements relating to the administration of options and share-based plans under applicable U.S. federal and state laws, the Code, any applicable stock exchange or automated quotation system rules or regulations, and the applicable laws of any other country or jurisdiction where Awards are granted, as such laws, rules, regulations and requirements shall be in place from time to time.

Award means any award made pursuant to the Plan, including awards made in the form of an Option, a Restricted Share, a Restricted Share Unit, an Unrestricted Share, a Deferred Share Unit, and a Performance Award, or any combination thereof, whether alternative or cumulative, authorized by and granted under this Plan.

Award Agreement means any written document setting forth the terms of an Award that has been authorized by the Committee. The Committee shall determine the form or forms of documents to be used, and may change them from time to time for any reason.

Board means the Board of Directors of the Company.

Cause for termination of a Participant's Continuous Service will have the meaning set forth in any unexpired employment agreement between the Company and the Participant. In the absence of such an agreement, Cause will exist if the Participant is terminated from employment or other service with the Company or an Affiliate for any of the following reasons: (i) the Participant's willful failure to substantially perform his or her duties and responsibilities to the Company or deliberate violation of a material Company policy; (ii) the Participant's commission of any material act or acts of fraud, embezzlement, dishonesty, or other willful misconduct; (iii) the Participant's material unauthorized use or disclosure of any proprietary information or trade secrets of the Company or any other party to whom the Participant owes an obligation of nondisclosure as a result of his or her relationship with the Company; or (iv) Participant's willful and material breach of any of his or her obligations under any written agreement or covenant with the Company.

The Committee shall in its discretion determine whether or not a Participant is being terminated for Cause. The Committee's determination shall, unless arbitrary and capricious, be final and binding on the Participant, the Company, and all other affected persons. The foregoing definition does not in any way limit the Company's ability to terminate a Participant's employment or consulting relationship at any time, and the term Company will be interpreted herein to include any Affiliate or successor thereto, if appropriate.

Change in Control means any of the following:

(i) **Acquisition of Controlling Interest.** Any Person becomes the Beneficial Owner, directly or indirectly, of securities of the Company representing 50% or more of the combined voting power of the Company's then outstanding securities. In applying the preceding sentence, (i) securities acquired directly from the Company or its Affiliates by or for the Person shall not be taken into account, and (ii) an agreement to vote securities shall be disregarded unless its ultimate purpose is to cause what would otherwise be Change of Control, as reasonably determined by the Board.

(ii) *Change in Board Control.* During a consecutive 2-year period commencing after the date of adoption of this Plan, individuals who constituted the Board at the beginning of the period (or their approved replacements, as defined in the next sentence) cease for any reason to constitute a majority of the Board. A new Director shall be considered an approved replacement Director if his or her election (or nomination for election) was approved by a vote of at least a majority of the Directors then still in office who either were Directors at the beginning of the period or were themselves approved replacement Directors, but in either case excluding any Director whose initial assumption of office occurred as a result of an actual or threatened solicitation of proxies or consents by or on behalf of any Person other than the Board.

(iii) *Merger.* The Company consummates a merger, or consolidation of the Company with any other corporation unless: (a) the voting securities of the Company outstanding immediately before the merger or consolidation would continue to represent (either by remaining outstanding or by being converted into voting securities of the surviving entity) at least 50% of the combined voting power of the voting securities of the Company or such surviving entity outstanding immediately after such merger or consolidation; and (b) no Person becomes the Beneficial Owner, directly or indirectly, of securities of the Company representing 50% or more of the combined voting power of the Company's then outstanding securities.

(iv) *Sale of Assets.* The stockholders of the Company approve an agreement for the sale or disposition by the Company of all, or substantially all, of the Company's assets.

(v) *Liquidation or Dissolution.* The stockholders of the Company approve a plan or proposal for liquidation or dissolution of the Company.

Notwithstanding the foregoing, a Change in Control shall not be deemed to have occurred by virtue of the consummation of any transaction or series of integrated transactions immediately following which the record holders of the common stock of the Company immediately prior to such transaction or series of transactions continue to have substantially the same proportionate ownership in an entity which owns all or substantially all of the assets of the Company immediately following such transaction or series of transactions.

Code means the U.S. Internal Revenue Code of 1986, as amended.

Committee means one or more committees or subcommittees of the Board appointed by the Board to administer the Plan in accordance with Section 4 above. With respect to any decision involving an Award intended to satisfy the requirements of Section 162(m) of the Code, the Committee shall consist of two or more Directors of the Company who are outside directors within the meaning of Section 162(m) of the Code. With respect to any decision relating to a Reporting Person, the Committee shall consist of two or more Directors who are disinterested within the meaning of Rule 16b-3.

Company means BioMarin Pharmaceutical Inc., a Delaware corporation; provided, however, that in the event the Company reincorporates to another jurisdiction, all references to the term Company shall refer to the Company in such new jurisdiction.

Consultant means any person, including an advisor, who is engaged by the Company or any Affiliate to render services and is compensated for such services.

Continuous Service means the absence of any interruption or termination of service as an Employee, Director, or Consultant. Continuous Service shall not be considered interrupted in the case of: (i) sick leave; (ii) military leave; (iii) any other leave of absence approved by the Committee, provided that such leave is for a period of not more than 90 days, unless reemployment upon the expiration of such leave is guaranteed by contract or statute, or unless provided otherwise pursuant to Company policy adopted from time to time; (iv) changes in status from Director to advisory director or emeritus status; or (v) in the case of transfers between locations of the Company or between the Company, its Affiliates or their respective successors. Changes in status between service as an Employee, Director, and a Consultant will not constitute an interruption of Continuous Service.

Deferred Share Units mean Awards pursuant to **Section 8**.

Director means a member of the Board, or a member of the board of directors of an Affiliate.

Disabled means a condition under which a Participant

(a) is unable to engage in any substantial gainful activity 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 12 months, or

(b) 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 12 months, received income replacement benefits for a period of not less than 3 months under an accident or health plan covering employees of the Company.

Eligible Person means any Consultant, Director or Employee and includes non-Employees to whom an offer of employment has been extended.

Employee means any person whom the Company or any Affiliate classifies as an employee (including an officer) for employment tax purposes, whether or not that classification is correct. The payment by the Company of a director's fee to a Director shall not be sufficient to constitute employment of such Director by the Company.

Exchange Act means the Securities Exchange Act of 1934, as amended.

Fair Market Value means, as of any date (the **Determination Date**): (i) the closing price of a Share on the New York Stock Exchange or the American Stock Exchange (collectively, the **Exchange**), on the Determination Date, or, if shares were not traded on the Determination Date, then on the nearest preceding trading day during which a sale occurred; or (ii) if such stock is not traded on the Exchange but is quoted on NASDAQ or a successor quotation system, (A) the last sales price (if the stock is then listed as a National Market Issue under The Nasdaq National Market System) or (B) the mean between the closing representative bid and asked prices (in all other cases) for the stock on the Determination Date as reported by NASDAQ or such successor quotation system; or (iii) if such stock is not traded on the Exchange or quoted on NASDAQ but is otherwise traded in the over-the-counter, the mean between the representative bid and asked prices on the Determination Date; or (iv) if subsections (i)-(iii) do not apply, the fair market value established in good faith by the Board.

Grant Date has the meaning set forth in **Section 13**.

Incentive Share Option or ISO hereinafter means an Option intended to qualify as an incentive stock option within the meaning of Section 422 of the Code, as designated in the applicable Award Agreement.

Involuntary Termination means termination of a Participant's Continuous Service under the following circumstances occurring on or after a Change in Control: (i) termination without Cause by the Company or an Affiliate or successor thereto, as appropriate; or (ii) voluntary termination by the Participant within 60 days following (A) a material reduction in the Participant's job responsibilities, provided that neither a mere change in title alone nor reassignment to a substantially similar position shall constitute a material reduction in job responsibilities; (B) an involuntary relocation of the Participant's work site to a facility or location more than 50 miles from the Participant's principal work site at the time of the Change in Control; or (C) a material reduction in Participant's total compensation other than as part of a reduction by the same percentage amount in the compensation of all other similarly-situated Employees, Directors or Consultants.

Non-ISO means an Option not intended to qualify as an ISO, as designated in the applicable Award Agreement.

Option means any stock option granted pursuant to **Section 6**.

Participant means any holder of one or more Awards, or the Shares issuable or issued upon exercise of such Awards, under the Plan.

Performance Awards mean Performance Units and Performance Compensation Awards granted pursuant to **Section 9**.

Performance Compensation Awards mean Awards granted pursuant to **Section 9(b)**.

Performance Unit means Awards granted pursuant to **Section 9(a)**, which may be paid in cash, in Shares, or such combination of cash and Shares as the Committee in its sole discretion shall determine.

Person means any natural person, association, trust, business trust, cooperative, corporation, general partnership, joint venture, joint-stock company, limited partnership, limited liability company, real estate investment trust, regulatory body, governmental agency or instrumentality, unincorporated organization or organizational entity.

Plan means this BioMarin Pharmaceutical Inc. 2006 Share Incentive Plan.

Reporting Person means an officer, Director, or greater than ten percent shareholder of the Company within the meaning of Rule 16a-2 under the Exchange Act, who is required to file reports pursuant to Rule 16a-3 under the Exchange Act.

Restricted Shares mean Shares subject to restrictions imposed pursuant to **Section 7**.

Restricted Share Units mean Awards pursuant to **Section 7**.

Rule 16b-3 means Rule 16b-3 promulgated under the Exchange Act, as amended from time to time, or any successor provision.

Share means a share of common stock of the Company, as adjusted in accordance with **Section 12**.

Ten Percent Holder means a person who owns stock representing more than ten percent (10%) of the combined voting power of all classes of stock of the Company or any Affiliate.

Unrestricted Shares mean Shares awarded pursuant to **Section 7**.

BIOMARIN PHARMACEUTICAL INC.
AMENDED AND RESTATED
2006 EMPLOYEE STOCK PURCHASE PLAN

BIOMARIN PHARMACEUTICAL INC.

AMENDED AND RESTATED

2006 EMPLOYEE STOCK PURCHASE PLAN

The following constitutes the provisions of the BioMarin Pharmaceutical Inc. Amended and Restated 2006 Employee Stock Purchase Plan of BioMarin Pharmaceutical Inc. (the Company), which is an amendment and restatement of the Company's 1998 Employee Stock Purchase Plan (which shall remain in full force and effect (including all Offering Periods, as defined below, in effect there under).

1. **Purpose.** The purpose of the Plan is to provide employees of the Company and its Designated Subsidiaries with an opportunity to purchase Common Shares of the Company. It is the intention, but not the obligation, of the Company to have the Plan qualify as an employee stock purchase plan under Section 423 of the Code. The provisions of the Plan shall, accordingly, be construed so as to extend and limit participation in a manner consistent with the requirements of that section of the Code.

2. **Definitions.**

(a) **Administrator** means (i) any person or committee to whom the Board delegates administrative discretion under the Plan, and (ii) the Board, which may exercise any and all administrative powers associated with the Plan.

(b) **Board** means the Board of Directors of the Company.

(c) **Code** means the Internal Revenue Code of 1986, as amended.

(d) **Common Shares** means shares of common stock, par value \$.001 per share, of the Company.

(e) **Company** means BioMarin Pharmaceutical Inc., a Delaware corporation.

(f) **Compensation** means the sums of the types and amounts of compensation determined from time to time by the Administrator in its sole discretion to be eligible to be taken into account under the Plan, provided that no such determination shall include or exclude any type or amount of compensation contrary to the requirements of Section 423 of the Code, including the equal treatment of participants having the same employer corporation.

(g) **Continuous Status as an Employee** means the absence of any interruption or termination of service as an Employee. Continuous Status as an Employee shall not be considered interrupted in the case of (i) sick leave; (ii) military leave; (iii) any other leave of absence approved by the Administrator, provided that such leave is for a period of not more than 90 days, unless reemployment upon the expiration of such leave is guaranteed by contract or statute, or unless provided otherwise pursuant to Company policy adopted from time to time; or (iv) in the case of transfers between locations of the Company or between the Company and its Designated Subsidiaries.

(h) **Contributions** means all amounts credited to the account of a participant pursuant to the Plan.

(i) **Corporate Transaction** means a sale of all or substantially all of the Company's assets, or a merger, consolidation, or other capital reorganization of the Company with or into another corporation, or any other transaction or series of related transactions in which the Company's shareholders immediately prior thereto own less than 50% of the voting shares of beneficial interest of the Company (or its successor or parent) immediately thereafter.

(j) **Designated Subsidiaries** means the Subsidiaries (or other entities with respect to sub-plans established under **Section 19(d)**) that have been designated by the Board from time to time in its sole discretion as eligible to participate in the Plan.

(k) **Employee** means any person, including an Officer, whom the Company or one of its Designated Subsidiaries classifies as an employee for payroll tax purposes and who (i) is customarily employed by the Company or one of its Designated Subsidiaries for at least 20 hours per week, (ii) is customarily employed

by the Company or one of its Designated Subsidiaries for more than five months in a calendar year, and (iii) has been employed by the Company or one of its Designated Subsidiaries for at least six (6) months.

(l) **Exchange Act** means the Securities Exchange Act of 1934, as amended.

(m) **Offering Date** means the first business day of each Offering Period (and shall thereby be the grant date for each Offering Period).

(n) **Offering Period** means a period of approximately twenty-four (24) months, commencing on the first business day on or after May 1 and November 1 of each year and terminating on the last business day of the periods ending twenty-four months later (or such other period that the Administrator may determine in its sole discretion before an Offering Date); provided that the first Offering Period under the Plan, as amended and restated herein, shall begin on May 1, 2005 and shall end on April 30, 2007.

(o) **Officer** means a person who is an officer of the Company within the meaning of Section 16 of the Exchange Act and the rules and regulations promulgated there under.

(p) **Plan** means this Amended and Restated 2006 Employee Stock Purchase Plan.

(q) **Purchase Date** means the last day of each Purchase Period of the Plan, provided, however, that if such date is not a business day, the Purchase Date shall mean the immediately preceding business day.

(r) **Purchase Period** means a period of six calendar months (or such other period of up to 27 consecutive months that the Administrator may determine in its sole discretion before an Offering Date), beginning on the day after each Purchase Date and ending on the next Purchase Date, except that the first Purchase Period of any Offering Period shall commence on the Enrollment Date and end with the next Purchase Date; provided, however, that the first Purchase Period under the Plan as amended and restated shall commence on May 1, 2006.

(s) **Purchase Price** means with respect to a Purchase Period an amount equal to 85% of the Fair Market Value (as defined in **Section 7(b)**) of a Share on the Offering Date or the Purchase Date, whichever is lower; provided, however, that the Administrator may before any Offering Date establish a different formula for determining the Purchase Price so long as the formula does not result in a lower Purchase Price than is allowable under Section 423(b)(6) of the Code.

(t) **Share** means one Common Share, as adjusted in accordance with **Section 18**.

(u) **Subsidiary** means a corporation (or an unincorporated entity of which the Company is a co-employer of its employees), domestic or foreign, of which not less than 50% of the voting shares are held by the Company or a Subsidiary, whether or not such corporation now exists or is hereafter organized or acquired by the Company or a Subsidiary.

3. Eligibility.

(a) Any person who is an Employee as of the date 30 days before the Offering Date of a given Offering Period shall be eligible to participate in such Offering Period, subject to the requirements of **Section 5(a)** and the limitations imposed by Section 423(b) of the Code.

(b) Any provisions of the Plan to the contrary notwithstanding other than **Section 3(c)**, no Employee shall be granted an option under the Plan

(i) if, immediately after the grant, such Employee (or any other person whose stock would be attributed to such Employee pursuant to Section 424(d) of the Code) would own shares of beneficial ownership of the Company and/or hold outstanding options to purchase Shares possessing five percent (5%) or more of the total combined voting power or value of all classes of Shares of the Company or shares of common stock of any Subsidiary of the Company, or (ii) if such option would permit his or her rights to purchase Shares under all employee stock purchase plans (described in Section 423 of the Code) of the Company and its Subsidiaries to accrue at a rate that exceeds Twenty-Five Thousand Dollars (\$25,000) of the Fair Market Value of such Shares (determined at the time such option is granted) for each calendar year in which such option is outstanding at any time.

(c) Employees of affiliates of the Company that are not corporate Subsidiaries, and Employees who are ineligible to participate pursuant to **Section 3(b)(i)** may, in the sole discretion of the Administrator, be eligible to participate in any Company sub-plan or sub-plans that the Administrator may establish in accordance with **Section 19(d)**.

4. **Offering Periods.** The Plan shall be implemented by consecutive, overlapping Offering Periods with a new Offering Period generally commencing on the first business day on or after May 1 or November 1 of each year (or on such other day as the Administrator shall determine), ending on the last day of the calendar month that is approximately twenty-four (24) months after the Purchase Period begins, and continuing thereafter until terminated in accordance with **Section 20**; provided, however, that the first Offering Period under the Plan shall be the Offering Period that commenced May 1, 2005. The Administrator shall have the power to change the duration and/or frequency of Offering Periods and Purchase Periods with respect to future purchases without stockholder approval, provided that the Administrator shall announce any such change at least fifteen (15) days prior to the scheduled beginning of the first Purchase Period to be affected.

5. **Participation.**

(a) An eligible Employee may become a participant in the Plan by completing a subscription agreement (the *Subscription Agreement*) provided by the Company in the form attached hereto as Exhibit A and filing it with the Company's Human Resources Department or the stock brokerage or other financial services firms designated or approved by the Administrator from time to time (each, a *Designated Broker*) prior to the applicable Offering Date, unless a later time for filing the Subscription Agreement is set by the Board for all eligible Employees with respect to a given Purchase Period. The Subscription Agreement shall set forth the percentage of the participant's Compensation (subject to **Section 6(a)**) to be paid as Contributions pursuant to the Plan.

(b) Payroll deductions shall commence on the first full payroll following the Offering Date and shall end on the last payroll paid on or prior to the last Purchase Period to which the Subscription Agreement is applicable, unless sooner terminated by the participant as provided in **Section 10**.

(c) A participant's Subscription Agreement shall remain in effect for successive Purchase Periods unless modified as provided in **Section 6** or terminated as provided in **Section 10**.

6. **Method of Payment of Contributions.**

(a) Subject to the limitation set forth in **Section 6(c)**, a participant shall elect to have payroll deductions made on each payday during the Purchase Period in an amount not less than one percent (1%) nor more than ten percent (10%) of such participant's Compensation on each payday during the Offering Period. All payroll deductions made by a participant shall be credited to his or her account under the Plan. A participant may not make any additional payments into such account.

(b) A participant may discontinue his or her participation in the Plan as provided in **Section 10**, and may increase or decrease the rate of his or her Contributions with respect to the Purchase Period only in accordance with rules that the Administrator establishes before the Offering begins. Any change in rate shall be effective as of the beginning of the next calendar month following the date of filing of the new Subscription Agreement, if the agreement is filed at least five (5) business days prior to such date and, if not, as of the beginning of the next succeeding calendar month.

(c) Notwithstanding the foregoing, to the extent necessary to comply with Section 423(b)(8) of the Code and **Section 3(b)**, a participant's payroll deductions may be decreased during any Purchase Period scheduled to end during the current calendar year to 0%. Payroll deductions shall re-commence at the rate provided in such participant's Subscription Agreement at the beginning of the first Purchase Period that is scheduled to end in the following calendar year, unless terminated by the participant as provided in **Section 10**.

7. Grant of Option.

(a) On the Offering Date of each Offering Period, each eligible Employee participating in such Offering Period shall be granted an option to purchase on each Purchase Date for each Purchase Period within the Offering Period a number of Shares determined by dividing such Employee's Contributions accumulated during such Purchase Period and retained in the participant's account as of the Purchase Date by the applicable Purchase Price; provided however that the maximum number of Shares an Employee may purchase during each Purchase Period shall not exceed 5,000 Shares (subject to adjustment pursuant to **Section 18**), and provided further that such purchase shall be subject to the limitations set forth in **Sections 3(b)** and **12**. The Board may, for future Offering Periods, increase or decrease, in its absolute discretion, the maximum number of shares of the Company's Common Stock that an eligible Employee may purchase during each Purchase Period of such Offering Period. Exercise of the option shall occur as provided in **Section 8**, unless the participant has withdrawn pursuant to **Section 10**. The option shall expire on the last day of the Offering Period.

(b) The fair market value of the Company's Common Shares on a given date (the *Fair Market Value*) shall be

(i) the closing sales price of the Common Shares for such date (or, in the event that the Common Shares are not traded on such date, on the immediately preceding trading date), as reported by the New York Stock Exchange or the American Stock Exchange, or, if such price is not reported, then on the nearest preceding trading day during which a sale occurred; or

(ii) if such stock is not traded on either exchange but is quoted on NASDAQ or a successor quotation system (A) the last sales price (if the stock is then listed as a National Market Issue under The Nasdaq National Market System or any successor system) or (B) the mean of the bid and asked prices per-share of the Common Shares as reported by the NASDAQ or successor; or

(iii) in the event the Common Shares are not listed on a stock exchange or quoted on NASDAQ but is otherwise traded in the over-the-counter market, the Fair Market Value per share shall be the mean between the most recent representative bid and asked prices; or

(iv) if subsections (i)-(iii) do not apply, the fair market value established in good faith by the Board.

8. Exercise of Option. Unless a participant withdraws from the Plan as provided in **Section 10**, his or her option for the purchase of Shares will be exercised automatically on each Purchase Date of a Purchase Period, and the maximum number of full Shares subject to the option will be purchased at the applicable Purchase Price with the accumulated Contributions in his or her account. No fractional Shares shall be sold or issued pursuant to the Plan. Any payroll deductions accumulated in a participant's account that are not sufficient to purchase a full Share shall be retained in the participant's account for the subsequent Purchase Period, subject to earlier withdrawal by the participant as provided in **Section 10**. Any other amounts left over in a participant's account after a Purchase Date shall be returned to the participant. The Shares purchased upon exercise of an option hereunder shall be deemed to be transferred to the participant on the Purchase Date. During his or her lifetime, a participant's option to purchase Shares hereunder is exercisable only by him or her.

9. Delivery. As promptly as practicable after each Purchase Date of each Purchase Period, the number of Shares purchased by each participant upon exercise of his or her option shall be deposited into an account established in the participant's name with a Designated Broker.

10. Voluntary Withdrawal; Termination of Employment.

(a) Subject to applicable securities law restrictions (e.g., the Company's insider trading policy), a participant may withdraw all but not less than all the Contributions credited to his or her account under the Plan at any time prior to each Purchase Date by giving the Company or the Designated Broker, a notice of withdrawal (the *Notice of Withdrawal*) in the form attached hereto as Exhibit B, at least five (5) days

prior to the Purchase Date. All of the participant's Contributions credited to his or her account will be paid to him or her promptly after receipt of his or her Notice of Withdrawal and his or her option for the current period will be automatically terminated, and no further Contributions for the purchase of Shares will be made during the Purchase Period.

(b) Upon termination of the participant's Continuous Status as an Employee prior to the Purchase Date of a Purchase Period for any reason, including retirement or death, the Contributions credited to his or her account will be returned to him or her or, in the case of his or her death, to the person or persons entitled thereto under **Section 13**, and his or her option will be automatically terminated. A participant will have up to 30 days to transfer, to himself, to a designated beneficiary, or to a designated broker, any Shares that the Company or the Designated Broker holds for the benefit of the Participant (using a form that the Administrator provides). If within 30 days, the participant's Shares are not transferred, the Administrator may, but shall not be obligated to, issue and mail a stock certificate for the Shares to the participant.

(c) In the event an Employee fails to remain in Continuous Status as an Employee of the Company for at least twenty (20) hours per week during the Purchase Period in which the employee is a participant, he or she will be deemed to have elected to withdraw from the Plan and the Contributions credited to his or her account will be returned to him or her and his or her option terminated.

(d) If a participant withdraws from a Purchase Period, the participant may enroll in a subsequent purchase period but only once within the same calendar year for this Plan or any succeeding employee stock purchase plan or any similar plan which may hereafter be adopted by the Company and for which such participant is otherwise eligible, subject to applicable securities law restrictions. If a participant withdraws from a Purchase Period, payroll deductions shall not resume at the beginning of any succeeding Purchase Period for which the participant is eligible to enroll unless the participant delivers a new Subscription Agreement to the Company.

11. **Interest.** No interest shall accrue on the Contributions of a participant in the Plan.

12. **Shares.**

(a) Subject to **Section 18**, the maximum number of Shares that participants may purchase pursuant to the Plan for Offering Periods beginning on or after May 1, 2005 shall be Two Million Five Hundred Thousand (2,500,000) Shares. The Shares that participants purchase pursuant to the Plan shall be newly-issued Shares, treasury Shares, or Shares purchased by the Designated Broker on the open market provided, however, that no more than 2,500,000 Shares, (as adjusted pursuant to **Section 18**) shall be purchased pursuant to options under the Plan. In the latter case, to the extent the Purchase Price for Shares is below their Fair Market Value for any Purchase Period, the Company shall pay the Designated Broker such amounts as are necessary to subsidize the Purchase Price for Shares purchased on the open market.

(b) The participant shall have no interest (including no right to receive any dividends) or voting right in Shares covered by his or her option until such option has been exercised.

(c) Shares to be delivered to a participant under the Plan will be registered in the name of the participant or, if directed by the participant in writing, in the name of the participant and his or her spouse.

13. **Administration.** The Administrator shall supervise and administer the Plan, and shall have full and exclusive discretionary authority to construe, interpret, and apply the terms of the Plan, to determine eligibility, to adjudicate all disputed claims under the Plan, to adopt, amend and rescind any rules deemed appropriate for the administration of the Plan, and to make all other determinations necessary or advisable for the administration of the Plan. Every finding, decision, and determination made by the Administrator shall, to the full extent permitted by law, be final and binding upon all parties. No person acting individually or jointly as the Administrator shall be liable for any action or determination made in good faith with respect to the Plan or any participant.

14. Designation of Beneficiary.

(a) A participant may designate a beneficiary who is to receive any Shares and cash, if any, from the participant's account under the Plan in the event of such participant's death subsequent to the end of a Purchase Period but prior to delivery to him or her of such Shares and cash. In addition, a participant may designate a beneficiary who is to receive any cash from the Participant's account under the Plan in the event of such participant's death prior to the Purchase Date. If a participant is married and the designated beneficiary is not the spouse, spousal consent shall be required for such designation to be effective. Beneficiary designations under this **Section 14(a)** shall be made in the form and in the manner as directed by the Company's Human Resources Department.

(b) Such designation of beneficiary may be changed by the participant (and his or her spouse, if any) at any time by written notice in accordance with **Section 14(a)**. In the event of the death of a participant and in the absence of a beneficiary validly designated under the Plan who is living at the time of such participant's death, the Company shall deliver such Shares and/or cash to the executor or administrator of the estate of the participant, or if no such executor or administrator has been appointed (to the knowledge of the Company), the Company, in its discretion, may deliver such Shares and/or cash to the spouse or to any one or more dependents or relatives of the participant, or if no spouse, dependent or relative is known to the Company, then to such other person as the Company may designate.

15. Transferability. Neither Contributions credited to a participant's account nor any rights with regard to the exercise of an option or to receive Shares under the Plan may be assigned, transferred, pledged or otherwise disposed of in any way (other than by will, the laws of descent and distribution, or as provided in **Section 14**) by the participant. Any such attempt at assignment, transfer, pledge or other disposition shall be without effect, except that the Company may treat such act as an election to withdraw funds in accordance with **Section 10**.

16. Use of Funds. All Contributions received or held by the Company under the Plan may be used by the Company for any corporate purpose, and the Company shall not be obligated to segregate such Contributions.

17. Reports. Individual recordkeeping accounts will be maintained for each participant in the Plan. Statements of account will be provided to participating Employees at least annually by the Designated Broker, which statements will set forth the amounts of Contributions, the per Share Purchase Price, the number of Shares purchased, and the remaining cash balance, if any.

18. Adjustments Upon Corporate Transactions.

(a) In the event of a proposed dissolution or liquidation of the Company, any Purchase Period then in progress will terminate immediately prior to the consummation of such action, unless otherwise provided by the Board. In the event of a Corporate Transaction, each option outstanding under the Plan shall be assumed or an equivalent option shall be substituted by the successor corporation or a parent or Subsidiary of such successor corporation. In the event that the successor corporation refuses to assume or substitute for outstanding options, each Purchase Period then in progress shall be shortened and a new Purchase Date shall be set (the *New Purchase Date*), as of which date any Purchase Period then in progress will terminate. The New Purchase Date shall be on or before the date of consummation of the transaction and the Board shall notify each participant in writing, at least ten (10) days prior to the New Purchase Date, that the Purchase Date for his or her option has been changed to the New Purchase Date and that his or her option will be exercised automatically on the New Purchase Date, unless prior to such date he or she has withdrawn from the Purchase Period as provided in **Section 10**.

(b) For purposes of this **Section 18**, an option granted under the Plan shall be deemed to be assumed, without limitation, if, at the time of issuance of the stock or other consideration upon a Corporate Transaction, each holder of an option under the Plan would be entitled to receive upon exercise of the option the same number and kind of shares of stock or the same amount of property, cash or securities as such holder would have been entitled to receive upon the occurrence of the transaction if the holder had been,

immediately prior to the transaction, the holder of the number of Shares covered by the option at such time (after giving effect to any adjustments in the number of Shares covered by the option as provided for in this Section 18); provided, however, that if the consideration received in the transaction is not solely common stock of the successor corporation or its parent (as defined in Section 424(e) of the Code), the Board may, with the consent of the successor corporation, provide for the consideration to be received upon exercise of the option to be solely common stock of the successor corporation or its parent equal in Fair Market Value to the per Share consideration received by holders of Common Shares in the transaction.

(c) The Administrator shall equitably adjust the number of Shares covered by each outstanding option, and the number of Shares that may be purchased pursuant to options under the Plan, as well as the price per Share covered by each such outstanding option, to reflect any increase or decrease in the number of issued Shares resulting from a stock-split, reverse stock-split, stock dividend, combination, recapitalization or reclassification of the Shares, or any other increase or decrease in the number of issued Shares effected without receipt of consideration by the Company. Except as expressly provided herein, no issuance by the Company of shares of stock of any class, or securities convertible into shares of stock of any class, shall affect, and no adjustment by reason thereof shall be required to be made with respect to, the number or price of Shares subject to any option.

19. Amendment or Termination.

(a) The Board may at any time and for any reason terminate or amend the Plan. Except as provided in **Section 18**, no such termination of the Plan may affect options previously granted, provided that the Plan or a Purchase Period may be terminated by the Board on a Purchase Date or by the Board setting a new Purchase Date with respect to a Purchase Period then in progress if the Board determines that termination of the Plan and/or the Purchase Period is in the best interests of the Company and the shareholders, or if continuation of the Plan and/or the Purchase Period would cause the Company to incur adverse accounting charges as a result of a change after the effective date of the Plan in the generally accepted accounting rules applicable to the Plan. Except as provided in **Section 18** and in this **Section 19**, no amendment to the Plan shall make any change in any option previously granted that adversely affects the rights of any participant. In addition, to the extent the Administrator considers it appropriate to conform the Plan with Rule 16b-3 under the Exchange Act, Section 423 of the Code, or any other applicable law, regulation, or stock exchange rule, the Company shall obtain stockholder approval in such a manner and to such a degree as so required.

(b) Without shareholder consent and without regard to whether any participant rights may be considered to have been adversely affected, the Board (or its committee) shall be entitled to change the Purchase Periods, to limit the frequency and/or number of changes in the amount withheld during a Purchase Period, to establish the exchange ratio applicable to amounts withheld in a currency other than U.S. dollars, to permit payroll withholding in excess of the amount designated by a participant in order to adjust for delays or mistakes in the Company's processing of properly completed withholding elections, to establish reasonable waiting and adjustment periods and/or accounting and crediting procedures to ensure that amounts applied toward the purchase of Common Shares for each participant properly correspond with amounts withheld from the participant's Compensation, and to establish such other limitations or procedures as the Board (or its committee) determines in its sole discretion advisable that are consistent with the Plan.

(c) The Company may adopt rules or procedures relating to the operation and administration of the Plan to accommodate the specific requirements of local laws and procedures. Without limiting the generality of the foregoing, the Company specifically authorizes the Administrator to adopt rules and procedures regarding handling of payroll deductions, payment of interest, conversion of local currency, payroll tax, withholding procedures and handling of stock certificates which vary with local requirements.

(d) The Administrator may also adopt sub-plans applicable to the Company or to particular Subsidiaries, or locations, which sub-plans may be designed to be outside the scope of Code Section 423. The rules of such sub-plans may take precedence over other provisions of this Plan, but unless otherwise superseded by the specific terms of such sub-plan, the provisions of this Plan shall govern the operation of such sub-plan. In addition, the Administrator may adopt rules or procedures relating to the operation and

administration of the Plan to accommodate the specific requirements of local laws and procedures. Without limiting the generality of the foregoing, the Company is specifically authorized to adopt rules and procedures regarding handling of payroll deductions, payment of interest, conversion of local currency, payroll tax, withholding procedures and handling of stock certificates which vary with local requirements.

20. **Notices.** All notices or other communications by a participant to the Company under or in connection with the Plan shall be deemed to have been duly given when received in the form specified by the Company at the location, or by the person, designated by the Company for the receipt thereof.

21. **Conditions Upon Issuance of Shares.** Shares shall not be issued with respect to an option unless the exercise of such option and the issuance and delivery of such Shares pursuant thereto shall comply with all applicable provisions of law, domestic or foreign, including, without limitation, the Securities Act of 1933, as amended, the Exchange Act, the rules and regulations promulgated thereunder, applicable state securities laws, and the requirements of any stock exchange upon which the Shares may then be listed, and shall be further subject to the approval of counsel for the Company with respect to such compliance.

As a condition to the exercise of an option, the Company may require the person exercising such option to represent and warrant at the time of any such exercise that the Shares are being purchased only for investment and without any present intention to sell or distribute such Shares if, in the opinion of counsel for the Company, such a representation is required by any of the aforementioned applicable provisions of law.

22. **Term of Plan; Effective Date.** The Plan, as amended and restated, shall become effective on May 2, 2006, provided that the Plan's effectiveness shall be contingent on its receipt of approval in 2006 by a vote of a majority of the votes cast at a duly held meeting of the Company's shareholders (or by such other shareholder vote that the Administrator determines to be sufficient for the issuance of Shares or stock options according to the Company's governing documents and applicable state law). The Plan shall continue in effect for a term of ten (10) years unless sooner terminated under **Section 19**. In the event that the Plan does not receive shareholder approval in 2006, this amended and restated Plan will be null and void, and the Plan as in effect immediately before this amendment and restatement will continue in full force and effect.

23. **Additional Restrictions of Rule 16b-3.** The terms and conditions of options granted hereunder to, and the purchase of Shares by, persons subject to Section 16 of the Exchange Act shall comply with the applicable provisions of Rule 16b-3. This Plan shall be deemed to contain, and such options shall contain, and the Shares issued upon exercise thereof shall be subject to, such additional conditions and restrictions as may be required by Rule 16b-3 to qualify for the maximum exemption from Section 16 of the Exchange Act with respect to Plan transactions.

24. **Notice of Disqualifying Dispositions.** By electing to participate in the Plan, each participant agrees to notify the Company in writing immediately after the participant sells, transfers or otherwise disposes of any Shares acquired under the Plan, if such disposition occurs within the earlier of (i) two (2) years of the Offering Date, or (ii) one (1) year of the Purchase Date, associated with such Shares. Each participant further agrees to provide any information about a disposition of Shares as may be requested by the Company to assist it in complying with any applicable tax laws.

25. **Withholding of Taxes.** Each participant must make adequate provision for all applicable federal, state, or other tax withholding obligations which may arise upon the exercise of any option or the disposition of any Shares.

26. **No Employment Rights.** The Plan does not create, directly or indirectly, any right for the benefit of any employee or class of employees to purchase any Shares from the Company (other than as expressly provided in, and subject to the terms and conditions of, the Plan), or create in any employee or class of employees any right with respect to continuation of employment by the Company or any Subsidiary, and it shall not be deemed to

interfere in any way with the Company's or any Subsidiary's right to terminate, or otherwise modify, an employee's employment at any time.

27. **Offsets.** To the extent permitted by law, the Company shall have the absolute right to withhold any amounts payable to any participant under the terms of the Plan to the extent of any amount owed for any reason by such participant to the Company or any Subsidiary and to set off and apply the amounts so withheld to payment of any such amount owed to the Company or any Subsidiary, whether or not such amount shall then be immediately due and payable and in such order or priority as among such amounts owed as the Board or its committee, in its sole discretion, shall determine.

28. **Captions.** The captions of the sections and paragraphs of this Plan have been inserted solely as a matter of convenience and in no way define or limit the scope or intent of any provision of the Plan. References to sections herein are to the specified sections of this Plan unless another reference is specifically stated. Wherever used herein, a singular number shall be deemed to include the plural unless a different meaning is required by the context.

29. **Governing Law.** The internal laws of the State of Delaware shall govern all matters relating to this Plan except to the extent superseded by the laws of the United States.

BIOMARIN PHARMACEUTICAL INC.
AMENDED AND RESTATED
2006 EMPLOYEE STOCK PURCHASE PLAN

SUBSCRIPTION AGREEMENT

Reason for Agreement:

_____ Original Application: for the Offering Date of _____ 1, _____ (must be a date at least five days after the date of this election).

_____ Change in Payroll Deduction Rate

_____ Change of Beneficiary(ies)

1. I, _____, hereby elect to participate in the BioMarin Pharmaceutical Inc. Amended and Restated 2006 Employee Stock Purchase Plan (the "ESPP"), and hereby subscribe to purchase Common Shares of the Company in accordance with this Subscription Agreement and the ESPP. Terms in this Subscription Agreement that begin with initial capital letters have the specially-defined meanings that are either set forth herein or in the ESPP (unless the context clearly indicates a different meaning).

2. Subject to the ESPP's terms and conditions and those below, I hereby authorize payroll deductions from each paycheck in the amount of _____ % of my Compensation on each payday (at least 1% and not to exceed 10%) during the Offering Period. (Please note that no fractional percentages are permitted.)

3. I understand that said payroll deductions shall be accumulated for the purchase of Common Shares at the applicable Purchase Price determined in accordance with the ESPP. I understand that if I do not withdraw from an Offering Period, any accumulated payroll deductions will be used to automatically exercise my option.

4. I have received a copy of the complete 2006 Employee Stock Purchase Plan. I understand that my participation in the ESPP is in all respects subject to the terms of the Plan. I understand that the grant of the option by the Company under this Subscription Agreement may be subject to obtaining stockholder approval of the ESPP.

5. Shares purchased for me under the ESPP should be issued only in my name (or, if this space _____ is checked, in the name(s) of me and my spouse, whose name is _____).

6. I understand that if I dispose of any shares received by me pursuant to the Plan within two years after the Enrollment Date (the first day of the Purchase Period during which I purchased such shares) or within one year of the Purchase Date, I will be treated for Federal income tax purposes as having received ordinary income at the time of such disposition in an amount equal to the excess of the fair market value of the shares at the time such shares were purchased by me over the price which I paid for the shares.

I HEREBY AGREE TO NOTIFY THE COMPANY IN WRITING WITHIN 30 DAYS AFTER THE DATE OF ANY DISPOSITION OF SHARES AND I WILL MAKE ADEQUATE PROVISION FOR FEDERAL, STATE OR OTHER TAX WITHHOLDING OBLIGATIONS, IF ANY, WHICH ARISE UPON THE DISPOSITION OF THE COMMON STOCK.

The Company may, but will not be obligated to, withhold from my compensation the amount necessary to meet any applicable withholding obligation including any withholding necessary to make available to the Company any tax deductions or benefits attributable to sale or early disposition of Common Shares by me. If I dispose of such shares at any time after the expiration of the 2-year and 1-year holding periods described above, I understand that I will be treated for Federal income tax purposes as having received income only at the time of such disposition, and that such income will be taxed as ordinary income only to the extent of an amount equal to the lesser of (1) the excess of the fair market value of the shares at the time of such disposition over the purchase price which I paid for the shares, or (2) 15% of the fair market value of the shares on the first day of the Offering Period. The remainder of the gain, if any, recognized on such disposition will be taxed as capital gain.

I UNDERSTAND THAT NOTHING IN THIS AGREEMENT CONSTITUTES TAX ADVICE, AND I ACKNOWLEDGE THAT THE COMPANY HAS ENCOURAGED ME TO CONSULT MY OWN TAX ADVISOR WITH REGARD TO THE TAX CONSEQUENCES OF PARTICIPATING IN THE 2006 EMPLOYEE STOCK PURCHASE PLAN.

7. In the event of my death, I hereby designate the following as my beneficiary(ies) to receive all payments and Shares due me under the ESPP:

PRIMARY BENEFICIARY:

(First) (Middle) (Last)

Relationship

(Address)

SECONDARY BENEFICIARY:

(First) (Middle) (Last)

Relationship

(Address)

8. I hereby agree to be bound by the terms of the ESPP, as modified by this Subscription Agreement. The effectiveness of this Subscription Agreement is dependent upon my eligibility to participate in the ESPP.

I UNDERSTAND THAT THIS SUBSCRIPTION AGREEMENT SHALL REMAIN IN EFFECT THROUGHOUT SUCCESSIVE PURCHASE PERIODS UNLESS TERMINATED BY ME, AND I HEREBY CONFIRM THAT THE FOLLOWING INFORMATION IS TRUE AND CORRECT.

Employee s Social

Security Number:

Employee s Address:

Dated: _____

Signature of Employee

Dated: _____

Spouse s Signature

(If beneficiary other than spouse)

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BIOMARIN PHARMACEUTICAL INC.
2006 EMPLOYEE STOCK PURCHASE PLAN

NOTICE OF WITHDRAWAL

I, the undersigned participant in the BioMarin Pharmaceutical Inc. 2006 Employee Stock Purchase Plan (the ESPP), hereby notify Independence Holding Company (the Company) that I hereby withdraw from the participation in the ESPP for the purchase period which began on , 2006 (the Enrollment Date). I hereby direct the Company to pay to me, as promptly as practicable, all the payroll deductions credited to my account with respect to such Purchase Period. I understand and agree that my purchase option for such Purchase Period will be automatically terminated.

I understand further that no further payroll deductions will be made for the purchase of shares in the current Purchase Period, and that I shall be eligible to participate in succeeding Purchase Periods only by delivering to the Company a new Subscription Agreement for a future Purchase Period beginning after the date of this withdrawal but only once for re-commencing participation within the same calendar year.

Name and Address of Participant:

Signature:

Date: _____

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FORM OF PROXY

BIOMARIN PHARMACEUTICAL INC.

ANNUAL MEETING OF STOCKHOLDERS

June 21, 2006

THIS PROXY IS SOLICITED ON BEHALF OF THE BOARD OF DIRECTORS

OF

BIOMARIN PHARMACEUTICAL INC.

The undersigned stockholder of BioMarin Pharmaceutical Inc., a Delaware corporation (BioMarin), hereby acknowledges receipt of the Notice of Annual Meeting of Stockholders and the Proxy Statement, each dated May 5, 2006, and hereby appoints Jean-Jacques Bienaimé, Jeffrey Cooper and G. Eric Davis, or any of them, as proxy and attorney-in-fact, with full power of substitution, on behalf and in the name of the undersigned, to represent the undersigned at the annual meeting of the stockholders of BioMarin to be held on June 21, 2006 at the Courtyard by Marriott Hotel, located at 1400 N. Hamilton Parkway, Novato, California 94949 at 10:00 a.m., California time (the Annual Meeting), and at any adjournment or postponement thereof, and to vote all shares of common stock of BioMarin, which the undersigned would be entitled to vote, if then and there personally present, at the Annual Meeting on the matters set forth on the reverse side and according to the discretion of the proxy holders for any other matters that may properly come before the meeting or any postponement or adjournment thereof.

(PLEASE DATE AND SIGN ON REVERSE SIDE)

BioMarin's Board of Directors recommends a vote FOR Proposals 1 through 4. If not otherwise directed, this proxy will be voted FOR Proposals 1 through 4 and according to the discretion of the proxy holders for any other matters that may properly come before the meeting or any postponement or adjournment thereof.

1. Election of directors. Nominees: Jean-Jacques Bienaimé, Michael Grey, Elaine J. Heron, Joseph Klein, III, Pierre Lapalme, and Alan Lewis.

FOR all nominees (except as marked to the contrary) WITHHOLD AUTHORITY to vote for all nominees
INSTRUCTION: To withhold authority to vote for any individual nominee, mark the FOR box above and write that nominee's name in the space provided below:

2. Proposal to ratify the selection by the Board of Directors of KPMG LLP as the Company's independent registered public accounting firm for the year ending December 31, 2006.

FOR AGAINST ABSTAIN

3. Proposal to approve the 2006 Share Incentive Plan for Directors and employees.

FOR AGAINST ABSTAIN

4. Proposal to approve the Amended and Restated 2006 Employee Stock Purchase Plan.

FOR AGAINST ABSTAIN

Signature(s):

Date:

This proxy should be marked, dated and signed by the stockholder(s) exactly as his or her name appears hereon, and returned promptly in the enclosed envelope. Persons signing in fiduciary capacity should so indicate. If shares are held by joint tenants or as community property, both must sign. When signing as executor, administrator, trustee or other representative, please give your full title.

PLEASE DATE, SIGN AND MAIL YOUR PROXY PROMPTLY IN THE ENCLOSED ENVELOPE.