

BIOMARIN PHARMACEUTICAL INC  
Form PRER14A  
May 16, 2005

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

(Amendment No. 1)

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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(1) Amount previously paid:

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(4) Date Filed:

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Preliminary Copy-Subject to Completion

**BIOMARIN PHARMACEUTICAL INC.**

**NOTICE OF ANNUAL MEETING OF STOCKHOLDERS**

**TO BE HELD ON \_\_\_\_\_, 2005**

TO THE STOCKHOLDERS OF BIOMARIN PHARMACEUTICAL INC.:

NOTICE IS HEREBY given that the 2005 Annual Meeting of the Stockholders (the Annual Meeting ) of BioMarin Pharmaceutical Inc. ( BioMarin or the Company ) will be held on \_\_\_\_\_, 2005 at 10:00 a.m. (California time), at the Embassy Suites Hotel located at 101 McInnis Parkway, San Rafael, California 94903 for the following purposes:

1. To elect 7 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, 2005; and
3. 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 20, 2005 as 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 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.

A group of stockholders affiliated with Caduceus Capital Master Fund Limited and Caduceus Capital II L.P., (together, the Caduceus Group ) has initiated a proxy solicitation to elect a slate of three persons selected by them to the Board of Directors. Your Board believes that this action is not in your best interests and urges you not to sign or return any proxy card the Caduceus Group may send you. If you have any questions or

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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:

G. Eric Davis  
*Vice President, Corporate Counsel and Assistant  
Secretary  
Novato, California*

May , 2005

**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**

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**Proxy Statement for 2005 Annual Meeting of Stockholders**

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**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 2005 Annual Meeting of Stockholders of the Company (the Annual Meeting) to be held on \_\_\_\_\_, 2005, 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 Embassy Suites Hotel located at 101 McInnis Parkway, San Rafael, California 94903. 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 \_\_\_\_\_, 2005. The Company's Annual Report, which includes its Form 10-K for the year ended December 31, 2004, as filed with the Securities and Exchange Commission, is provided with this proxy statement.

**Record Date; Outstanding Securities**

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 20, 2005 are entitled to notice of and to vote at the Annual Meeting. On May 20, 2005, there were \_\_\_\_\_ 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.

**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, Corporate Counsel and Assistant Secretary) a written notice of revocation, or (ii) submitting a duly

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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. Such persons are listed on *Annex A* to this proxy statement.

The Company has retained Morrow & Co. 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 \$150,000, and has agreed to reimburse Morrow & Co. for reasonable out-of-pocket expenses for these services. The Company has agreed to indemnify Morrow & Co. against certain liabilities and expenses. Morrow & Co. will employ approximately 30 people to solicit proxies from the Company's stockholders.

Because of the solicitation of opposition proxies as a result of the matters discussed below under the heading *Dissident Stockholder Solicitation*, the expenses related to its solicitation of proxies from its stockholders will exceed those normally spent for an annual meeting. Such expenses excluding the cost of litigation, if any, are expected to aggregate \$600,000, of which approximately \$175,000 has been spent to date. Such additional solicitation costs are expected to include the increased fee payable to Morrow (approximately \$150,000 this year as compared to \$10,000 in 2004), fees of outside counsel to advise the Company in connection with a contested solicitation of proxies, increased mailing costs, such as the costs of additional mailings of solicitation materials to stockholders, including printing costs, mailing costs and the reimbursement of reasonable expenses of banks, brokerage houses and other agents incurred in forwarding solicitation materials to beneficial owners of the Company's stock, as described above, and the costs of retaining an independent inspector of election. The Company will bear the cost of these expenses.

### **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 7 nominees





for directors set forth herein; and (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, 2005. 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.

The Company believes that a broker will not have discretionary authority to vote shares for Proposal One: Election of Directors. 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.

### **Election of Directors**

Election of the director nominees named in Proposal No. 1 requires the affirmative vote of a plurality of the shares of our common stock present in person or represented by proxy at the Annual Meeting and entitled to vote. The 7 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 Independent Registered Public Accounting Firm**

Ratification of the selection of KPMG LLP as the independent registered public accounting firm for the year ending December 31, 2005, as specified in Proposal No. 2, requires the affirmative vote of a majority of the shares of our 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 this proposal and will have the same effect as a vote against this proposal. Broker non-votes, if any, will have no effect on the vote for this proposal.

### **Submission of Stockholder Proposals for the 2006 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 2006 Annual Meeting of Stockholders ( 2006 Annual Meeting ) must deliver or mail a notice to the Assistant 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 \_\_\_\_\_, 2006 and no later than \_\_\_\_\_, 2006. In the event that the 2006 Annual Meeting is called for a date that is not within 25 days before or after \_\_\_\_\_, 2006, then the stockholder's notice must be received by the Assistant Secretary no later than the close of

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business on the 10th day following the day on which notice of the date of the 2006 Annual Meeting was mailed or public announcement of such date was made by the Company, whichever first occurs. With respect to the 2005 Annual Meeting, a stockholder's notice pursuant to the Company's Bylaws had to be delivered to and received by the Company by April 18, 2005.

The notice must contain a brief description of the business desired to be brought before the 2006 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 2006 Annual Meeting to bring such business before the 2006 Annual Meeting.

Stockholders interested in submitting a proposal for inclusion in the proxy materials for the 2006 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 Assistant Secretary of the Company no later than \_\_\_\_\_, 2006, or a reasonable time before the Company begins to print and mail its proxy materials if the 2006 Annual Meeting is changed by more than 30 days from the anniversary date of the 2005 Annual Meeting. Proposals should be sent to the Assistant 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**

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**PROPOSAL ONE: ELECTION OF DIRECTORS**

The Company has a Board currently consisting of 7 directors. At the Annual Meeting, stockholders will elect 7 directors. Unless otherwise instructed, the proxy holders will vote the proxies represented by the enclosed WHITE Proxy Card FOR the 7 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 a director. The Company has received notice that Caduceus Capital Master Fund Limited and Caduceus Capital II L.P. (together, the Caduceus Group), intend to nominate and through their advisors, OrbiMed Advisors LLC and OrbiMed Capital LLC (together, OrbiMed), intend to support three opposition candidates at the Annual Meeting and may solicit your proxy for those nominees. See *Dissident Stockholder Solicitation*, below. The term of office of each person elected as a director shall continue until the next annual meeting of stockholders and until that person's successor has been elected.

If a quorum is present, the 7 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
Jean-Jacques Bienaimé	51	Chief Executive Officer and Director
Franz L. Cristiani(2)(3)	63	Director
Elaine J. Heron, Ph.D.(2)(4)	57	Director
Pierre Lapalme (1)(3)	64	Director, Chairman of the Board
Erich Sager(1)	47	Director
John Urquhart, M.D.(1)(4)	71	Director
Gwynn R. Williams(2)(3)	71	Director

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

Each of the Board's nominees to the Board has consented to being named in this proxy statement and to serve as a director if elected. The Board has determined that each of the above nominees, other than Mr. Bienaimé, are independent as defined by Nasdaq and Securities and Exchange Commission rules. There is no family relationship between any director, the above nominees for director, or any executive officer of the Company.

**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 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. 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 Aventis). Mr. Bienaimé currently serves on the boards of the Biotechnology

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Industry Organization (BIO), Aerogen, Inc., NeurogesX, Inc., and Saegis Pharmaceuticals, Inc., and is a member of the advisory board of Bellevue Asset Management's BioVentures II fund. He received an MBA from the Wharton School at the University of Pennsylvania and an undergraduate degree in economics from the Ecole Supérieure de Commerce de Paris.

**Franz L. Cristiani** joined the BioMarin Board in June 2002 and serves as chairman of its Audit Committee. From 1964 to 1999, he was with Arthur Andersen, LLP as partner since 1976, with clients in high technology, life sciences, manufacturing, mining, forest products, distribution, publishing and food products industries. Since 1999, he has provided consulting services to various companies, served as a director of various companies and served as an adjunct professor at the University of San Francisco. Mr. Cristiani serves as a director of MTI Technology, Inc., Vitasoy USA, Accuray Incorporated and Nature's Sunshine Products. He holds a B.A. from San Francisco State University and is a C.P.A.

**Elaine J. Heron, Ph.D.** joined the BioMarin Board in July 2002. 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 Applera Corporation, including stints 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.

**Pierre Lapalme** joined the BioMarin Board in January 2004 and was named as Chairman in August 2004, upon the resignation of Fredric D. Price, BioMarin's former Chief Executive Officer and Chairman of the Board. 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 Ethicals, 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 International Wex Technologies. Mr. Lapalme studied at the University of Western Ontario and INSEAD France. Mr. Lapalme also served as non-executive chairman of the board of Diagnostics, a biotechnology company from 1995 to 2005.

**Erich Sager** joined the BioMarin Board in November 1997. Since 2004, Mr. Sager has acted as Chairman of Calltrade Carrier Services AG, a European wholesale phone operator. From September 1996 until August 2004, Mr. Sager served as the chairman of LaMont Asset Management SA, a private investment management firm. From April 1994 to August 1996, Mr. Sager served as senior vice president, head of Private Banking for Dresdner Bank (Switzerland) Ltd. From September 1991 to March 1994, Mr. Sager served as vice president, Private Banking-Head German Desk for Deutsche Bank (Switzerland) Ltd. From 1981 to 1989, Mr. Sager held various positions at a number of banks in Switzerland. Mr. Sager serves as a director of Calltrade Carrier Services AG and Zecotek Medical Systems Inc. Mr. Sager received a business degree from the School of Economics and Business Administration in Zurich, Switzerland.

**John Urquhart, M.D.**, joined the BioMarin Board in July 2003 and serves as chairman of its Compensation Committee and Science Committee and has been a member of the BioMarin Science Advisory Board since 2002. Since September 1995, Dr. Urquhart has been the chief scientist of AARDEX Ltd., acting as the chief technical officer of this pharmaceutical packaging company, and the chief executive officer of APREX, also a pharmaceutical packaging company and subsidiary of AARDEX. Since 1991, he has been a member of the board of directors of the Cayman subsidiary of HBM Bioventures, as Swiss-based venture capital firm. Dr. Urquhart also served, as director and non-executive vice chairman of the board of directors of Inveresk Research Group, Inc. of Cary, North Carolina, formerly Inveresk Research Group Ltd. of Edinburgh, Scotland until its acquisition in November 2004 by Charles River Laboratories, Inc. Since November 2004, he has served as a director of Charles River Laboratories, Inc. In April 2004, Dr. Urquhart retired from his position as Extraordinary Professor of Pharmacoepidemiology at Maastricht University in the Netherlands where he served since 1986 with a concentration on graduate education and research. He has been Adjunct Professor of Biopharmaceutical Sciences at University of California, San Francisco Medical Center since 1986. In addition, he occasionally serves as a

paid consultant to the pharmaceutical industry. Dr. Urquhart received his M.D. from Harvard Medical School and his B.A. from Rice University.

**Gwynn R. Williams** joined the BioMarin Board upon its incorporation in 1996 and serves as Chairman of its Corporate Governance and Nominating Committee. Mr. Williams is also a trustee of the United Kingdom Transplant Trust. Previously, Mr. Williams was a partner at Andersen Consulting where he specialized in the design and installation of financial control systems. Additionally, he served as a mathematician with General Motors Research, an internal consultant with Canadian International Paper Company, and a statistician with British Steel. He was a founder of numerous companies including, Xoma Inc., Murex Inc., AstroMed Ltd. and Astroscan Ltd. Mr. Williams was a major stockholder of Glyko Biomedical Ltd. (which was acquired by BioMarin in 2002) and its predecessor Glyko, Inc. Mr. Williams received a B.Sc. in theoretical physics from the University of Wales and is a Chartered Physicist and Fellow of the Institute of Physics in the United Kingdom.

### **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, a Corporate Governance and Nominating Committee and a Science Committee whose functions are briefly described below.

The Board held a total of 19 meetings during the year ended December 31, 2004 and took action by unanimous written consent on 2 occasions. During fiscal 2004, 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. Cristiani, Chairman, Dr. Heron, and Mr. Williams. 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](http://www.bmrn.com). Information on our website is not incorporated by reference in this proxy statement. During 2004, the Audit Committee met on 8 occasions.

**Compensation Committee.** The Compensation Committee sets general compensation policy for BioMarin and has final approval power over compensation of executive officers. 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. Urquhart, Chairman, Mr. Lapalme and Mr. Sager. Dr. Urquhart and Mr. Lapalme are each independent directors as defined by Nasdaq and Securities and Exchange Commission rules. Mr. Sager was not independent in 2004 due to consulting services he provided to BioMarin in 2001, but he was determined to be independent in 2005. The committee charter is available in the investors section of the Company's website at [www.bmrn.com](http://www.bmrn.com). Information on our website is not incorporated by reference in this proxy statement. During 2004, 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 our 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 Mr. Williams, Chairman, Mr. Cristiani and Mr. Lapalme. The committee charter and the Corporate Governance Guidelines are available in the investors section of the Company's website at [www.bmrn.com](http://www.bmrn.com). Information on our website is not incorporated by reference in this proxy statement. The Committee met on 8 occasions during 2004.

*Science Committee.* In 2002, the Company formed a Scientific Advisory Board ( SAB ). The SAB provides the Company with technical advice on its research and development programs. Dr. Urquhart is a member of the SAB and collaborates with Dr. Heron as a liaison between the SAB and the Board. Dr. Urquhart chairs the Science Committee with Dr. Heron as a Science Committee member.

#### **Compensation Committee Interlocks and Insider Participation**

The Compensation Committee consists (and consisted during all of fiscal 2004) of three of the Company's independent directors: Dr. Urquhart (chair) and Messrs. Lapalme and Sager. None of the members of the Compensation Committee (i) was an officer or employee of the Company or any of its subsidiaries during the 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 our 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 our board of directors or Compensation Committee.

**2004 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.

<u>Director Position</u>	<u>Annual Compensation</u>
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
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	\$10,000
Science Committee (Non-Chair)	\$5,000

From January 1, 2004 through August 19, 2004, Mr. Cristiani served as lead director. In August 2004, the Board amended the Company's Corporate Governance Principles to eliminate the position of lead director in connection with the adoption of the requirement to have the role of Chief Executive Officer and Chairman of the Board held by different persons. Mr. Cristiani received \$3,750 for serving as lead director in 2004. Directors are also reimbursed for their reasonable expenses in attending meetings of the Board and while performing services for the Company.

*2004 Director Compensation*

The following table lists actual compensation paid to each of the directors during 2004.

<u>Director Name</u>	<u>Director Fees</u>	<u>Option Grant Date</u>	<u>Number of Options Granted</u>	<u>Exercise Price Per Share</u>	<u>Option Expiry</u>
Franz L. Cristiani	\$ 37,500	6/14/04	30,000	\$ 5.97	6/14/14
Elaine J. Heron, Ph.D.	\$ 29,375	7/12/04	30,000	\$ 5.12	7/12/14
Pierre Lapalme	\$ 31,250	1/27/04	30,000	\$ 8.47	1/27/14
Erich Sager	\$ 25,000	11/30/04	30,000	\$ 5.33	11/30/14
John Urquhart, M.D.	\$ 32,500	7/28/04	30,000	\$ 5.57	7/28/14
Gwynn R. Williams	\$ 31,875	11/30/04	30,000	\$ 5.33	11/30/14

*1998 Director Option Plan*

The 1998 Director Option Plan was adopted by the Board in December 1999. It was approved by the Company'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 900,000 shares

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of the Company's 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 2004, options to purchase, in the aggregate, 180,000 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 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.

Except as set forth in this proxy statement or in *Annex A* hereto, to the best knowledge of the Company, none of the directors, nor any of the Board's nominees for election, nor any other persons participating in this solicitation on behalf of the Company, or with respect to items (ii), (viii) and (ix) of this paragraph, any associate (within the meaning of Rule 14a-1 of the Exchange Act) of the foregoing persons (i) have any substantial interest, direct or indirect, in any matter to be acted upon at the Annual Meeting, other than the interest of the Board's nominees in being elected and each director's interest in the operations of the Company, (ii) own beneficially, directly or indirectly, any securities of the Company, (iii) own any securities of the Company of record but not beneficially, (iv) have purchased or sold any securities of the Company within the past two years, (v) have incurred indebtedness for the purpose of acquiring or holding securities of the Company within the past two years, (vi) are or have within the past year been a party to any contract, arrangement or understanding with any person with respect to any securities of the Company, except with respect to any employment agreement, stock option grant or similar agreement described in the proxy statement or in *Annex A* hereto, (vii) own beneficially, directly or indirectly, any securities of any parent or subsidiary of the Company, (viii) have since the beginning of the Company's last fiscal year, been indebted to the Company or any of its subsidiaries in excess of \$60,000 or (ix) have any arrangement or understanding with respect to future employment by the Company or its affiliates or with respect to any future transactions to which the Company or any of its affiliates will or may be a party. In addition, except as set forth in this proxy statement or in *Annex A* hereto, to the best knowledge of the Company, none of the directors, nor any of the Board's nominees for election, nor any other person participating in this solicitation on behalf of the Company, nor any associates of any of the foregoing persons, have had or are to have a direct or indirect material interest in any transaction or proposed transaction with the Company in which the amount involved exceeds \$60,000 since the beginning of the Company's last fiscal year.

**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.**

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**IF YOU HAVE ANY QUESTIONS, OR REQUIRE ASSISTANCE IN VOTING YOUR SHARES, PLEASE CALL THE FIRM ASSISTING THE COMPANY IN THE SOLICITATION OF PROXIES, MORROW & CO., INC., TOLL-FREE AT (800) 607-0088.**

### **Dissident Shareholder Solicitation**

On April 18, 2005, the Caduceus Group, the beneficial owners of an aggregate of 2,110,000 shares (3.2%) of the Company's Common Stock, notified the Company of their intention to nominate 3 persons for election as directors of the Company through their advisors, OrbiMed Advisors LLC and OrbiMed Capital LLC (together, OrbiMed). OrbiMed also advised the Company that it would support the nominations through a number of entities through which it controls an additional 3,196,300 shares (5.0%), of the Company's Common Stock. The currently-proposed Caduceus Group nominees are: Richard Brewer, Joseph Klein III and Alan J. Lewis.

The following discussion summarizes the interactions between the Company, OrbiMed and the proposed Caduceus Group nominees.

In late 2004 the Committee contacted Richard B. Brewer about the possibility of serving as the Company's Chief Executive Officer. Mr. Brewer indicated then that he was unwilling to serve as the Chief Executive Officer, but that he was interested in serving as Chairman of the Board of Directors and had ideas regarding potential candidates for the position of Chief Executive Officer. Accordingly, members of the Committee met with Mr. Brewer personally, and the Committee held a number of formal and informal meetings during which Mr. Brewer's candidacy as a director as well as his proposals for the Company were discussed.

As a result of these meetings, the Committee determined that one of Mr. Brewer's primary proposals was based on a fundamental misunderstanding on his part of the composition and manufacture of Aldurazyme. In this regard, the Committee believed that Mr. Brewer failed to understand key differences between Aldurazyme and a simpler commercial product for which he had a broader understanding based on his past experience. Furthermore, the Committee believed that following their discussions, Mr. Brewer may have contacted OrbiMed, which in turn contacted one of the Company's corporate partners, without authorization, to pursue one of Mr. Brewer's proposals for the Company. Management, as well as members of the Board of Directors, believed that this unauthorized contact needlessly harmed the Company's relationship with that corporate partner.

The Committee recognized that Mr. Brewer's opinions with respect to this and other strategic issues were necessarily based in part on limited information available to him as an outsider. But as discussions ensued, the Committee determined that Mr. Brewer in fact was unwilling to alter his opinions through further discussion and learning about the Company and its products. For these and other reasons, the Committee believed that Mr. Brewer's approach could make it considerably more difficult, among other things, to hire a qualified Chief Executive Officer—an objective that remained of primary importance for the Company. Accordingly, the Committee concluded that Mr. Brewer's nomination was not in the best interests of the Company and advised him of its determination in January 2005.

Subsequently, in January, 2005, Samuel D. Isaly of OrbiMed contacted the Company to request that three potential candidates, including Mr. Brewer, Joseph Klein, III, and a third individual, be considered for nomination by the Board for election at the Company's upcoming annual meeting. In light of this request and to assist the Committee in its evaluation process, the Committee referred Mr. Klein and the third individual to Spencer Stuart, an executive search firm retained by the Company, to perform reference checks. In February, 2005, Committee members Pierre Lapalme and Gwynn R. Williams met with Mr. Isaly to discuss his request and the reasons for these specific candidates. As the Committee had already duly considered Mr. Brewer as a candidate for the Board, it decided not to pursue his nomination further and the Committee advised Mr. Isaly accordingly. Mr. Isaly then indicated that OrbiMed was agreeable to pursuing the other two candidates nominations and abandoning Mr. Brewer's nomination.



Upon receipt of reference information on the remaining proposed candidates, the Committee met by telephone on March 21, 2005, and decided to schedule interviews with Mr. Klein and the other candidate. Ultimately, the Committee was unable to interview the other candidate, after attempting unsuccessfully several times to schedule a mutually convenient meeting time. Messrs. Lapalme and Williams were able to interview Mr. Klein, although Mr. Williams was only able to do so by traveling at considerable inconvenience to Park City, Utah, where Mr. Klein then was vacationing.

Following the meeting with Mr. Klein, on April 1, 2005 the Board met to further consider the two remaining proposed candidates' qualifications in light of the Company's needs and objectives. With respect to the third proposed candidate, the Board determined to defer any decision and instead to continue reviewing his candidacy along with other candidates proposed by Spencer Stuart, in part as a result of the Committee's inability to meet in person with the third proposed candidate. Based on the Committee's recommendation, the Board determined that this third proposed candidate's qualifications appeared to be inferior to those of the other candidates then available for consideration. Finally, the Committee and the Board were influenced by the seeming lack of personal interest in the Company by the third proposed candidate. On April 5, 2005, the third proposed candidate formally withdrew his candidacy and was not among the candidates subsequently proposed by the Caduceus Group on April 15, 2005.

By contrast, the Board decided to move forward with Mr. Klein's nomination. On April 8, 2005, Mr. Lapalme informed Mr. Isaly of the Committee's decision to move forward with Mr. Klein's nomination and to retain for future consideration the third proposed candidate. Mr. Isaly expressed his pleasure at the potential nomination of Mr. Klein and regret at the Board's decision on the other proposed candidates.

On April 9, 2005, Mr. Klein accepted his proposed nomination by the Committee. However, he elected not to review and comment on the relevant portions of the Company's proxy statement then provided to him. Neither Mr. Klein nor Mr. Isaly thereafter made any further attempt to communicate with the Company until April 15, 2005.

On April 15, 2005, the Caduceus Group served the Company with notice of its intention to propose a slate of three candidates for nomination in opposition to the Company's nominees. The notice named Messrs. Brewer, Klein and Alan J. Lewis, a new candidate previously unknown to the Company, as the proposed Caduceus Group nominees. Concurrent with the notice, OrbiMed also filed a Schedule 13D with the SEC referencing the same candidates. On April 18, 2005, each of the Committee and the Board convened to discuss the Caduceus Group notice. Thereafter, the Committee directed Messrs. Lapalme and Cristiani to contact Mr. Isaly to determine what the Committee might do to better satisfy his expectations as a stockholder. When contacted, Mr. Isaly advised the Committee that he desired that all three of the Caduceus Group candidates be nominated for election to the Board, including Mr. Brewer.

On April 19, 2005, each of the Committee and the Board again convened to consider the Caduceus Group notice and the OrbiMed Schedule 13D. At this meeting, the Committee revisited its prior extensive discussions regarding possible director candidates. It noted that Mr. Brewer's candidacy had been previously discussed at length and accordingly reaffirmed its prior decision to decline his nomination. It then discussed the qualifications of the new proposed director, Mr. Lewis, in light of the Company's needs and determined that at that time, another nominee then being evaluated appeared to be better qualified. In doing so, the Committee noted that there was not enough time prior to the proposed filing date of the Company's proxy statement to complete the requisite due diligence necessary to include Mr. Lewis on the Company's slate of nominees. Accordingly, the Committee was unable to recommend his nomination to the Board at that time.

While the Committee initially supported the nomination of Mr. Klein to fill the current vacancy on the Board, the Committee withdrew its support when Mr. Klein announced his intention to participate with other Caduceus Group candidates that the Company had previously deferred or rejected. In doing so, the Committee believed, among other things, that so significant and fundamental a change to the Board of Directors could significantly disrupt or delay the CEO search, which urgently needed to be completed, as well as corporate





partnering and financing opportunities. The Committee was also influenced in part by the apparent lack of candor of Mr. Klein, as suggested by his decision not to advise the Committee of his joint objectives with the other Caduceus Group nominees, his apparent interest in serving only on a conditional basis with Mr. Brewer and his decision to run with the Caduceus Group candidates in opposition to the candidates approved by the Committee, without regard for the systems and procedures implemented by the Committee on behalf of the Company's stockholders. Accordingly, the Committee recommended that none of the Caduceus Group candidates be nominated by the Board for election at the Annual Meeting.

Stockholders should further be advised that the indenture governing the Company's 3.5% convertible subordinated notes provides that each holder of the notes may require the Company to repurchase all or part of its notes upon a change of control. As defined in the indenture, a change of control will occur if at any time at least a majority of the Company's directors are not comprised of members of the Board as it existed at the time of execution of the indenture (June 23, 2003) or nominated by a majority of those directors or the successors to those directors that were so nominated. While all of the Company's nominees satisfy this requirement under the indenture, the nominees by the Caduceus Group do not. Accordingly, a change of control requiring the repurchase of the notes will occur if all of the Caduceus Group nominees are elected and one or more of the other Board members resigns or dies, unless prior to his or her resignation or death, a new director is elected by both a majority of the Board, including the Caduceus Group nominees, and a majority of the directors comprised of or nominated by the existing Board, excluding the Caduceus Group nominees. As of March 31, there was \$125 million in aggregate principal amount of the notes outstanding.

THE BOARD WILL OPPOSE THE CADUCEUS GROUP CANDIDATES AND UNANIMOUSLY RECOMMENDS A VOTE AGAINST THE CADUCEUS GROUP CANDIDATES. YOU MAY RECEIVE A PROXY CARD SOLICITING YOUR VOTE FOR THE CADUCEUS GROUP CANDIDATES. WE URGE YOU TO DISREGARD ANY SUCH PROXY SOLICITATION AND TO SIGN AND DATE THE ENCLOSED WHITE PROXY CARD AND RETURN IT TODAY IN THE ENCLOSED PRE-ADDRESSED POSTAGE-PAID ENVELOPE.

## 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 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. John Urquhart and Gwynn Williams, current directors and nominees for the Board at the Annual Meeting, have separately indicated that in accordance with the corporate governance principles adopted by the Company, upon reaching age 72, they intend to submit their resignation for consideration by the Committee. The Committee expects to recommend that the Board decline such resignations and that Dr. Urquhart and Mr. Williams continue to serve on the Board through the end of their terms.

As described above, the Committee received three proposed nominations from the Caduceus Group. While the Committee initially supported the nomination of one such candidate, Mr. Klein, the Committee withdrew its support when Mr. Klein announced his intention to participate with a Caduceus Group nominee which the Committee previously deferred or rejected in opposition to the Committee's approved nominees.



In light of the determination not to nominate Mr. Klein to fill the current vacancy of the Board of Directors and the desire to fill that vacancy and have the CEO also serve as a director, the Committee recommended that Mr. Drapeau, former acting CEO, be submitted to the stockholders for consideration as a nominee for election at the Annual Meeting. On May 11, 2005, Jean-Jacques Bienaimé was appointed Chief Executive Officer and director, thereby filling the vacancy on the Board. In keeping with the Board's desire to have the CEO serve as a director, the Committee recommended that Mr. Bienaimé be submitted to stockholders for consideration as a nominee for election at the Annual Meeting, and, consequently, withdrew Mr. Drapeau's nomination.

In 2003, the independent Board members appointed Franz Cristiani as its Lead Director. The Lead Director was responsible for:

approving Board meeting schedules and meeting agendas;

approving Board meeting materials;

servicing 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.

Upon the resignation of Fredric D. Price as Chief Executive Officer and Chairman of the Board on August 12, 2004, the Lead Director position was eliminated and Mr. Lapalme was appointed Chairman of the Board. The responsibilities of the Lead Director were assumed by the Chairman of the Board.

#### **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 Assistant Secretary and Vice President, Corporate Counsel. Stockholders who wish to contact Audit Committee members directly to report complaints or concerns about accounting, internal controls or auditing matters, may do so anonymously by directing their correspondence to Franz Cristiani, Audit Committee Chairman, PO Box 130A, San Carlos, CA 94070.

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 2004 Annual Meeting of Stockholders attended such meeting.

#### **STANDARDS OF BUSINESS CONDUCT AND ETHICS**

The Board has adopted Standards of Business Conduct and Ethics which 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](http://www.bmrn.com). Information on our 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, 2005, 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.

<i>Description of Services Provided by KPMG LLP</i>	<i>Year Ended</i>	<i>Year Ended</i>
	<i>December 31, 2004</i>	<i>December 31, 2003</i>
<i>Audit Fees</i>	\$ 777,137	\$ 214,428
<i>Audit Related Fees:</i> These services primarily involved review and assurance services associated with certain regulatory filings (for fees incurred in 2004) the Company's debt and equity offerings and related regulatory filings (for fees incurred in 2003).	\$ 23,401	\$ 183,000
<i>Tax Compliance Fees:</i> These services related to the preparation of federal, state and foreign tax returns and other filings.	\$ 51,246	\$ 53,860
<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	\$ 9,100
<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 UNANIMOUSLY RECOMMENDS VOTING FOR THE RATIFICATION OF KPMG LLP AS THE COMPANY'S INDEPENDENT REGISTERED PUBLIC ACCOUNTING FIRM.**



**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 April 1, 2005 as to (i) each person, or group of affiliated persons, who is known by us to own beneficially more than 5% of the Company's common stock; (ii) each of the directors or nominees; (iii) each of the Company's executive officers named in the Summary Compensation Table set forth herein under the caption "Executive Compensation" (the "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.

<u>Name of Beneficial Owner</u>	<u>Number of Shares Beneficially Owned(1)</u>	<u>Number of Shares Subject To Options(2)</u>	<u>Percentage of Common Stock</u>
PRIMECAP Management Company(3)(4)	6,552,587		10.1%
Orbimed Advisors LLC / Orbimed Capital LLC(5)(6)	5,306,300		8.2%
Barclays Global Investors, NA(7)(8)	3,813,440		5.9%
Palo Alto Investors, LLC(9)(10)	3,367,666		5.2%
Jean-Jacques Bienaimé(11)			
Franz L. Cristiani	67,500	67,500	*
Elaine J. Heron, Ph.D.	72,500	72,500	*
Pierre Lapalme	37,500	37,500	*
Erich Sager	197,444	194,135	*
John Urquhart, M.D.	54,500	54,500	*
Gwynn R. Williams	932,565	120,000	1.4%
Louis Drapeau(12)	135,199	124,269	*
Robert Baffi	331,110	312,617	*
Emil D. Kakkis, M.D., Ph.D.(13)	349,595	321,561	*
Christopher M. Starr, Ph.D.	561,361	256,185	*
Stuart J. Swiedler, M.D., Ph.D.	212,107	202,107	*
Fredric D. Price(12)	907,255	842,970	1.4%
Jeffrey I. Landau	122,061	121,249	*
All current executive officers and directors as a group (12 persons)	3,015,665	1,762,873	4.7%

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

- (1) The "Number of Shares Beneficially Owned" column is based on 64,511,159 shares of common stock outstanding at April 1, 2005. Shares of common stock subject to options or warrants that are exercisable within 60 days of April 1, 2005 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.
- (2) 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 April 1, 2005. These shares are included in the amounts shown under the "Number of Shares Beneficially Owned," as provided in the preceding



note.

- (3) Information is based upon statements filed on Schedule 13G/A with the Securities and Exchange Commission on March 31, 2005.
- (4) The mailing address for PRIMECAP Management Company is 225 South Lake Avenue, #400, Pasadena, CA 91101.
- (5) Information based upon statements filed on Schedule 13D with the Securities and Exchange Commission on April 15, 2005 (the OrbiMed Schedule 13D ). According to information contained in the OrbiMed Schedule 13D, OrbiMed Advisors LLC/OrbiMed Capital LLC hold discretionary investment management authority over the shares held by the Caduceus Group.
- (6) The mailing address for OrbiMed Advisors LLC/OrbiMed Capital LLC is 767 Third Avenue, 30th Floor, New York NY 10017.
- (7) Information based upon statements filed on Schedule 13G with the Securities and Exchange Commission on February 14, 2005.
- (8) The mailing address for Barclays Global Investors, NA is 45 Fremont Street, San Francisco, CA 94105.
- (9) Information based upon statements filed on Schedule 13G with the Securities and Exchange Commission on February 10, 2005.
- (10) The mailing address for Palo Alto Investors, LLC is 470 University Avenue, Palo Alto, CA 94301.
- (11) Mr. Bienaimé was appointed Chief Executive Officer on May 11, 2005.
- (12) Effective August 12, 2004, Mr. Price resigned as Chief Executive Officer and Chairman.
- (13) Includes 3,000 shares of common stock held by Dr. Kakkis' spouse.

### Executive Officers and other Significant Employees

The following table sets forth certain information concerning the Company's executive officers other than Mr. Bienaimé, who was elected Chief Executive Officer on May 11, 2005 and other significant employees as of April 1, 2005. Information concerning Mr. Bienaimé is set forth under the caption "Nominees For Director."

Name	Age	Position with BioMarin
Louis Drapeau	61	Senior Vice President, Finance, Chief Financial Officer and Secretary
Robert A. Baffi, Ph.D.	50	Senior Vice President, Technical Operations
Emil D. Kakkis, M.D., Ph.D.	44	Senior Vice President, Business Operations
Christopher M. Starr, Ph.D.	52	Co-founder, Senior Vice President and Chief Scientific Officer
Stuart J. Swiedler, M.D., Ph.D.	49	Senior Vice President, Clinical Affairs
Jeffrey H. Cooper, C.P.A.	49	Vice President, Contoller
G. Eric Davis	34	Vice President, Corporate Counsel and Assistant Secretary
Daniel P. Maher	49	Vice President, Program Management
Kim R. Tsuchimoto, C.P.A.	42	Vice President, Treasurer

**Louis Drapeau** serves as BioMarin's Senior Vice President, Finance, Chief Financial Officer and Secretary. From August 2004 to May 2005, Mr. Drapeau served as BioMarin's Acting Chief Executive Officer, a position to which he was appointed upon the resignation of Frederic D. Price, BioMarin's former Chief Executive Officer. Mr. Drapeau joined BioMarin as Vice President, Finance, Chief Financial Officer, and Secretary in August 2002. Prior to joining BioMarin, Mr. Drapeau spent 31 years at Arthur Andersen LLP, including fourteen years as managing partner of Assurance and Consulting for Northern California, supervising more than 400 professionals. He has a strong international focus and he also has in-depth capital market experience as well as an extensive knowledge of best practices in the areas of financial controls and administration. From 1966 to 1969, he was a Naval Officer serving with U.S. Naval Civil Engineering Corps principally in Seabee units in the Republic of Vietnam. Mr. Drapeau is a California certified public accountant and holds an M.B.A. and a B.S. in Mechanical Engineering from Stanford University.

**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.

**Emil D. Kakkis, M.D., Ph.D.**, Senior Vice President, Business Operations

Dr. Kakkis serves as BioMarin's Senior Vice President, Business Operations and has been a 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.

**Christopher M. Starr, Ph.D.**, Co-founder, Senior Vice President and Chief Scientific Officer

Dr. Starr co-founded BioMarin in 1996 and serves as Senior Vice President and Chief Scientific Officer. Before BioMarin, Dr. Starr directed the research programs of Glyko, Inc., a subsidiary of BioMarin, the operations of which have been discontinued. He joined Glyko as director of research and development in 1991 and served as vice president of research and development from 1992 to 1998. Dr. Starr was a National Research Council Associate at the National Institutes of Health (NIH). He has published numerous peer-reviewed articles, including research papers on Fluorophore-Assisted Carbohydrate Electrophoresis (FACE) in the diagnosis of lysosomal storage diseases and in the identification of patients with MPS I. His work in the development of diagnostic tests for lysosomal storage diseases has been funded by several grants from the NIH and other institutions. Dr. Starr holds a Ph.D. in biochemistry and molecular biology from the State University of New York Health Science Center and a B.S. from Syracuse University.

**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 University School of Medicine, and a B.S. from the State University of New York at Albany.

**Jeffrey H. Cooper, C. P. A.**, Vice President, Controller

Mr. Cooper serves as BioMarin's Vice President, Controller. From August 2004 to May 2005, Mr. Cooper served as BioMarin's Acting Chief Financial Officer since the appointment of Mr. Drapeau (BioMarin's Chief Financial Officer until August 2004) to Acting Chief Executive Officer in August 2004. Mr. Cooper joined BioMarin as Vice President, Controller, in October 2003. Prior to joining BioMarin, Mr. Cooper

worked at Matrix Pharmaceutical for six years, serving most recently as vice president, finance. Earlier in 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 certified public accountant 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, Corporate Counsel and Assistant Secretary

Mr. Davis joined BioMarin in March 2004, and serves as Vice President, Corporate Counsel and Assistant 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, Program Management

Mr. Maher joined BioMarin as Vice President, Program Management in August 2003, bringing with him over 20 years of biotechnology industry experience. From 1998 to 2003, Mr. Maher worked at Aradigm Corporation serving most recently as vice president, program management, overseeing the pulmonary insulin project. Prior to this time, he served as director of clinical operations at Covance, Inc., and as vice president of operations at Spectra BioMedical, Inc. From 1993 to 1995, 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 project management. 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.

**Kim R. Tsuchimoto, C.P.A.**, Vice President, Treasurer

Ms. Tsuchimoto joined BioMarin in 1997 and currently serves as Vice President, Treasurer, a position she was appointed to in July 2003 after serving as Vice President, Controller. Prior to BioMarin, she worked as controller for three years at Dodd Smith Dann Inc., a fundraising and marketing consultant firm for non-profit organizations, and before that, as controller at Partech International, a venture capital firm affiliated with Banque Paribas (currently known as BNP Paribas). Ms. Tsuchimoto is a licensed California certified public accountant and received a B.S. in business administration 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) Fredric D. Price, the Company's former Chairman and Chief Executive Officer and Louis Drapeau, the Company's former Acting Chief Executive Officer and current Chief Financial Officer; (b) Robert A. Baffi, Ph.D., Emil D. Kakkis, M.D., Ph.D., Christopher M. Starr, Ph.D. and Stuart J. Swiedler, M.D., Ph.D., the four most highly-compensated officers other than the Chief Executive Officer who were serving as officers at the end of fiscal year 2004 and whose salary and bonus exceeded \$100,000; and (c) Jeffrey I. Landau, who would have been one of the four most highly compensated officers, but for the fact

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that he resigned on December 31, 2004. These individuals are referred as the Named Executive Officers. Jean-Jacques Bienaimé was appointed Chief Executive Officer on May 11, 2005. Accordingly, information concerning Mr. Bienaimé is not included below.

Except as noted for Mr. Price, and Mr. Landau, 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	All Other Compensation(\$)
		Salary(\$)	Bonus(\$)	Other Annual Compensation(\$)	Securities Underlying Options/SARs(#)(1)	
Louis Drapeau	2004	258,320	40,000(3)		25,000	5,318
	2003	250,025	10,000		40,000	5,600
Chief Financial Officer(2)	2002	102,885			150,000	290
Robert A. Baffi, Ph.D.	2004	249,871	80,000(3)			6,345
	2003	242,266	30,000		50,000	8,543
Senior Vice President, Technical Operations	2002	231,000				3,522
Emil D. Kakkis, M.D., Ph.D.	2004	270,402	160,000(3)(4)			4,913
	2003	270,027	110,000		125,000	4,974
Senior Vice President, Business Operations	2002	257,250				4,522
Christopher M. Starr, Ph.D.	2004	275,830	110,000(3)			6,417
	2003	275,454	60,000		125,000	4,992
Senior Vice President, Chief Scientific Officer	2002	262,500				4,522
Stuart J. Swiedler, M.D., Ph.D.	2004	256,652	80,000(3)			6,226
	2003	256,223	42,500		80,000	4,923
Senior Vice President, Clinical Affairs	2002	241,500				4,522
Fredric D. Price	2004	422,746	400,700			3,094,606
	2003	507,082	225,000		625,000	6,400
Former Chairman and Chief Executive Officer(5)	2002	458,129				2,496
Jeffrey I. Landau	2004	240,435	110,000(3)			220,661
	2003	249,355	42,500	4,000	115,000	6,100
Former Senior Vice President, Administration(6)	2002	168,750			125,000	5,174

- (1) For 2004 performance, on January 5, 2005, the Company granted the Named Executive Officers options for the purchase of the following number of shares of BioMarin common stock: Mr. Drapeau 100,000 shares; Dr. Baffi 140,000 shares; Dr. Kakkis, 100,000 shares; Dr. Starr, 60,000 shares and Dr. Swiedler, 80,000 shares. These grants are not reflected in the table.
- (2) Mr. Drapeau served as Acting Chief Executive Officer from August 2004 to May 2005. On August 19, 2004, upon his appointment as Acting Chief Executive Officer, Mr. Drapeau received options for the purchase of 25,000 shares of common stock.
- (3) The bonus represents amounts paid in February 2004 in recognition of performance in 2003. No officer received a cash bonus for performance in 2004.
- (4) Dr. Kakkis' bonus includes 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, as required by Dr. Kakkis' employment agreement.
- (5) In December 2003, the Board awarded a bonus of \$400,000 to Mr. Price for his performance in 2003, which was paid in January 2004. In connection with Mr. Price's termination of employment, on August 12, 2004, Mr. Price received a severance package of approximately \$2.9 million. This amount is included with All Other Compensation. Mr. Price resigned effective August 12, 2004.
- (6) Mr. Landau retired from the Company on December 31, 2004. Upon his resignation, Mr. Landau's loan plus accrued interest totaling \$116,092 was forgiven and the Company paid the tax on such loan forgiveness in the amount of \$99,093. This amount is included with All Other Compensation.





## Option Grants in Last Fiscal Year

Name	Individual Grants				Potential Realizable Value at Assumed Annual Rates of Stock Price Appreciation for Option Term\$(3)		First Exercisable
	Number of Securities Underlying Options Granted(#)	Percent of Total Options Granted to Employees in Fiscal Year(1)	Exercise or Base Price (\$/Share)(2)	Expiration Date	5%	10%	
Louis Drapeau(4)	25,000	1%	5.15	8/19/14	80,970	205,194	2/19/05

- (1) Based on an aggregate of 1,795,800 shares subject to options granted during fiscal year 2004 to employees.
- (2) Options were granted at an exercise price equal to the closing price of the 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 term, 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.
- (4) Options were granted to Mr. Drapeau on August 19, 2004 upon his appointment as Acting Chief Executive Officer. Mr. Drapeau currently serves as the Company's Chief Financial Officer.

## Fiscal 2004 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 2004.

## Options Exercised During Fiscal Year 2004

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
			Louis Drapeau		\$	99,790
Robert A. Baffi, Ph.D.			301,506	51,737		
Emil D. Kakkis, M.D., Ph.D.			304,895	90,105	466,050	
Christopher M. Starr, Ph.D.	5,570	21,828	239,520	90,104	249,588	
Stuart J. Swiedler, M.D., Ph.D.			190,910	59,323	250,950	
Fredric D. Price						
Jeffrey I. Landau			121,249	118,751		

- (1) Based on closing price on December 31, 2004 of \$6.39 per share less exercise price per share.



*Employment Agreements*

The Company is party to an employment agreement with Mr. Bienaimé which provides for a base salary of \$468,000 during his first year of employment and not less than \$568,000 (as determined by the Board of Directors) each year thereafter. The agreement also provides for a one-time sign-on bonus of \$100,000 and a potential annual performance bonus of up to 100% of base salary (including for purposes of the first year, the sign-on bonus). The Company granted Mr. Bienaimé an option to purchase 325,000 shares of the Company's common stock and will grant an additional (i) 162,500 options on the sixth month anniversary of the date of the agreement, (ii) 287,500 options on the first anniversary of the date of the agreement, and (iii) 125,000 options on each anniversary of the date of the agreement thereafter. All such options will be non-qualified stock options, have an exercise price equal to the closing price of the Company's common stock on the grant date, and vest in equal amounts on the first, second and third anniversary of the date of grant.

Mr. Bienaimé is entitled to participate in the Company's retirement, stock and stock option, stock purchase, pension, insurance, deferred compensation and similar plans as in effect from time to time. The Company will also maintain a insurance policy for Mr. Bienaimé and pay for financial planning assistance, relocation costs and expenses, and temporary housing costs. Upon the occurrence of specified termination events, Mr. Bienaimé will be entitled to receive cash severance payments equal to his then current annual base salary for a period of 18 months (or 24 months if Mr. Bienaimé has been employed for more than 3 years), a possible cash bonus equal to 100 percent of his target bonus for such year, continuation of all Company-paid health benefits for a period of 18 months (or 24 months if Mr. Bienaimé has been employed for more than 3 years), reimbursement of up to \$18,000 for outplacement services and automatic vesting of all unvested options granted to Mr. Bienaimé.

Moreover, upon certain change in control events, Mr. Bienaimé will be entitled to receive a cash payment equal to his then current annual base salary that he would have collected over the 24 months period (or 30 month period if Mr. Bienaimé has been employed for more than 3 years) following the date of termination, a possible cash bonus equal to 100 percent (250 percent if employed for more than 3 years) of his target bonus for such year, continuation of all Company-paid health benefits for a period of 24 months (or 30 months if Mr. Bienaimé has been employed for more than 3 years), plus cash payments for outplacement services and tax preparation, maintenance of his life insurance policy, contribution to his 401k plan for the year of termination, and automatic vesting of all unvested options granted to Mr. Bienaimé.

The Company is party to employment agreements with the Named Executive Officers, other than Mr. Price and Mr. Landau, on the terms set forth in the chart below. Each of these employment agreements is terminable without cause by the Company upon six months' prior written notice to the officer, or by the officer upon three months' prior written notice to the Company. The Company is obligated to pay the officer's salary and benefits until termination. In addition, with respect to Messrs. Baffi, Kakkis, Starr 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. The Company is a party to a Severance Agreement with Mr. Price as described below.

In January 2004, the Company instituted a Severance Plan and Summary Plan Description that is applicable to each of the officers listed in the table below. 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, the Named Executive Officers are entitled to receive one year of benefit continuation and a severance payment equal to one year of base salary and 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.

Name of Executive Officer	2004 Annual Salary Rate(1)	Annual Bonus	Initial Grant of Right to Purchase Equity Securities(2)	Agreement Termination Date
Louis Drapeau	\$ 275,000	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
Robert A. Baffi, Ph.D.	\$ 250,000	Annual bonus, payable in cash and/or stock.	Options to purchase up to 210,000 shares of the Company's common stock at a purchase price of \$22.00 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 the Company's common stock at a purchase price of \$4.00 per share.	None
Christopher M. Starr, Ph.D.	\$ 275,500	Annual bonus, payable in cash and/or stock.	400,000 shares of the Company's common stock at a purchase price of \$1.00 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 the Company's common stock at a purchase price of \$4.00 per share.	None

(1) 2004 Annual Salary Rate reflected in the above table reflects 2004 annual salary rate as of December 31, 2004. The table entitled *Summary Compensation Table* reflects actual salaries paid in 2004 and includes mid-year salary adjustments.

(2) Options in this table were granted on commencement of employment for each individual.

Effective August 12, 2004, Mr. Price terminated his employment with the Company. Mr. Price was paid a severance package that included: a cash payment of approximately \$2.9 million, acceleration of the full unvested portion of his 39,285 share restricted stock grant (and a registration right with respect to such shares) and the extension of the exercise period for all his vested stock options to one year following termination. The cash payment was netted against the amount of principal and interest due under the Company's loan to Mr. Price.

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 the Company's equity compensation plans in effect as of December 31, 2004.

**Equity Compensation Plan Information**

<b>Plan Category</b>	<b>Number of securities to be issued upon exercise of outstanding options, warrants and rights(1)</b>	<b>Weighted average exercise price of outstanding options, warrants and rights</b>	<b>Number of securities remaining available for future issuance under equity compensation plans(2)</b>
Equity compensation plans approved by stockholders	10,008,025	\$ 9.96	3,289,891
Equity compensation plans not approved by stockholders			
<b>Total</b>	<b>10,008,025</b>	<b>\$ 9.96</b>	<b>3,289,891</b>

- (1) Does not include any shares of the Company's 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 the Company'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 of the Company's fiscal years 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 Company's then outstanding capital stock, (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 Company's then outstanding capital stock, or (iii) a lower amount set by the Company's Board.

**Certain Relationships and Related Transactions**

Since January 1, 2004, 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, any of the Board's nominees for election as 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 proxy statement, and (ii) the transaction described below.

Dr. Emil Kakkis, one of BioMarin's Senior Vice Presidents, holds 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 \$300,000 in 2004, primarily for research. The Company's joint venture with

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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 the sales level of the Company 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 based on sales of Aldurazyme during 2004.

*Indebtedness of Directors and Executive Officers*

Other than as described below, none of the directors, Board nominees for director, executive officers of the Company, or any such person's respective affiliates or associates is or at any time since January 1, 2004 has been indebted to the Company.

In April 2001, the Company loaned Mr. Price, Chairman and Chief Executive Officer, \$860,000 to purchase a residential property and received a promissory note secured by the property, pursuant to the terms of his employment agreement with the Company. The note was to mature on October 31, 2006 and bore interest at the Federal mid-term rate. Pursuant to Mr. Price's severance package and based on the terms of his employment agreement with the Company, the note plus accrued interest totaling \$982,927 was repaid in August 2004 out of the amounts due to Mr. Price under his severance package.

In March 2002, the Company entered into an employment agreement with Mr. Landau that entitled him to loans from the Company of up to \$100,000 to be applied to the purchase of a home or up to \$36,000 annually if a purchase of a home was not completed. Pursuant to the terms of that agreement, in each of June 2002, June 2003 and June 2004, the Company loaned Mr. Landau \$36,000, which bore interest at 6.0%, 3.5% and 4% per annum, respectively. Such loans were due upon termination of his employment with the Company. Upon Mr. Landau's retirement from the Company on December 31, 2004, Mr. Landau's loans plus interest totaling \$116,092 were forgiven, again pursuant to the terms of that agreement.



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.

**Performance Graph**

The following graph compares the cumulative total stockholder return with the cumulative total return of the Nasdaq Stock Market (U.S.) and the Nasdaq Pharmaceutical Index of stocks in Standard Industry Code (SIC) 283, encompassing primarily biotechnology, pharmaceutical and medical specialty companies, and the Nasdaq Biotechnology Index, assuming a \$100 investment in common stock on December 31, 1999 and reinvestment of dividends during the period.

	<u>BioMarin</u>	<u>Nasdaq Stock Market (U.S.)</u>	<u>Nasdaq Pharmaceutical Index</u>	<u>Nasdaq Biotech Index</u>
12/31/1999	100	100	100	100
12/31/2000	82	60	125	123
12/31/2001	114	48	106	103
12/31/2002	60	33	69	56
12/31/2003	66	49	101	82
12/31/2004	54	54	107	87

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## 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 2004, the Compensation Committee was composed of three members of the Board.

### 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 pharmaceutical and biotechnology companies in the San Francisco Bay Area.
- (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 priced related compensation, such as its Employee Stock Purchase Program.

### 2004 Compensation

In 2004, the Compensation Committee hired an independent consultant to provide expertise in various matters that came before the Committee, including the review and analysis of executive compensation at BioMarin based upon the San Francisco Bay Area employment market and on milestone achievements at the Company. 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 small biotechnology companies in the San Francisco Bay Area.

Increases in base salary for 2004 were made effective January 1, 2004, primarily based on the progress and achievements of BioMarin during 2003 and competitive conditions in the job marketplace for biotechnology expertise in the San Francisco Bay Area marketplace.

The Compensation Committee took particular note of 2003 achievements, including the marketing approval and commercial launch of the Company's first commercial product, Aldurazyme, the successful Phase 2 trial and subsequent initiation of the Phase 3 trial of rhASB (formerly known as Aryplase), the raising of \$94 million in equity financings and the closing of a \$125 million convertible debt financing. Based on the

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Compensation Committee's judgment as to the value of these events and other less visible internal developments, the Compensation Committee awarded cash bonuses to the executives along with long-term compensation stock option grants to the executives and staff of BioMarin. Grants were pro-rated for the portion of the year that the employee was in the service of BioMarin.

Salary compensation for the staff below the rank of officer was increased by an average of 6.5%, which approximated the reported average salary increase in the biotechnology industry in the San Francisco Bay Area and which was also pro-rated for time in service during the year. The Compensation Committee deemed that these compensation actions were appropriate for the progress made by BioMarin in 2003 and maintained a competitive balance with biotechnology companies of similar size and state of development in the region.

With respect to officer's salaries, the Compensation Committee determined that, based on the factors described above, a modest increase was appropriate and elected to raise salaries by an average of 2.4%.

In February 2004, the Company paid the following performance bonuses to the Named Executive Officers (other than Mr. Price) for their service and achievements in 2003: Mr. Drapeau, \$40,000; Dr. Baffi, \$80,000; Dr. Starr, \$110,000; Dr. Kakkis, \$110,000; Mr. Landau, \$110,000; and Dr. Swiedler, \$80,000. These bonuses were in recognition of the Company's milestone achievements in 2003. In 2004, pursuant to his employment agreement, Dr. Kakkis also received a \$50,000 cash bonus upon the filing of an Investigational New Drug Application for BioMarin's product candidate Phenoptin.

### **Chief Executive Officer Compensation**

In August 2004, the Board granted long-term compensation stock options to Mr. Drapeau for the purchase of 25,000 shares of BioMarin common stock and elected to increase Mr. Drapeau's annual salary from \$250,000 to \$275,000, effective for such time as he continues to serve as Acting Chief Executive Officer. The Compensation Committee believed that such grant and increase is appropriate due to the increased responsibilities and commitments associated with Mr. Drapeau assuming the role of Acting Chief Executive Officer.

The compensation of the former Chief Executive Officer was established pursuant to an employment agreement executed in March 2003. Under this agreement, Mr. Price was eligible to receive a base salary of \$550,000 in 2004, a cash bonus of up to \$687,500 and long-term incentive stock options for the purchase of up to 200,000 shares of BioMarin common stock. Based upon the achievements of 2003, the Compensation Committee determined that Mr. Price had amply demonstrated that his skills merited his base salary remuneration. In particular, the Compensation Committee noted the Company's milestone achievements in 2003, including the marketing approval and commercial launch of the Company's first commercial product, Aldurazyme, the successful Phase 2 trial and subsequent initiation of the Phase 3 trial of rhASB, the raising of \$94 million in equity financings and the closing of a \$125 million convertible debt financing. In recognition for these successes, in December 2003, the Board awarded Mr. Price a bonus of \$400,000, which was paid in January 2004.

In August 2004, Mr. Price resigned from the Company and received a severance package totaling approximately \$2.9 million.

### **2005 Compensation**

In December 2004, the Compensation Committee considered the possibility of granting performance bonuses for and salary merit increases to the Company's officers. The Compensation Committee determined, in concurrence with the recommendation of the Company's officers, that it was appropriate to not grant bonuses or salary merit increases based on the Company's performance in 2004. However, partially in-lieu of bonuses and salary increases, in January 2005, for 2004 performance, the Company granted long-term incentive stock options to the officers. These options were rewarded in recognition of 2004 achievements including the regulatory approval of BioMarin's manufacturing facility as a multi-product plant, the filing for marketing approval in the U.S. and E.U. of rhASB, the initiation of a Phase 2 trial for Phenoptin, including the successful negotiation of supply agreements for the contract manufacturing of Phenoptin, the successful funding and completion of a new research and development facility and corporate headquarters and the completion of a transaction to purchase Orapred, including the integration

of a 60-person sales force into BioMarin.

The Compensation Committee believes that the option awards are an appropriate recognition for the Company's achievements and that the actions related to bonuses and salary increases are appropriate based on the challenges the Company experienced in 2004. The Compensation Committee 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 April 12, 2005 by the members of the Compensation Committee of the Board of Directors:

John Urquhart, M.D., Chairman

Pierre Lapalme

Erich Sager

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 2004, 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 2004 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, 2004, 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, 2005.

Respectfully submitted on April 12, 2005 by the members of the Audit Committee of the Board of Directors:

Franz L. Cristiani, Chairman

Elaine J. Heron, Ph.D.

Gwynn R. Williams

**SECTION 16(A) BENEFICIAL OWNERSHIP REPORTING COMPLIANCE**

Section 16(a) of the Exchange Act requires the Company's directors and officers and persons who own more than that 10% of a registered class of the Company's equity securities (the Reporting Persons) to file reports of ownership and reports of changes in the ownership with the Securities and Exchange Commission. Reporting Persons are required by the Securities and Exchange Commission to furnish the Company with copies of all Section 16(a) forms they file.

Except for the following transaction, to the best of the Company's knowledge, 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, 2004, all Reporting Persons complied with all Section 16(a) filing requirements. On February 11, 2005, Dr. Urquhart filed a Form 5, which reported a sale that should have been reported in July 2004 on a Form 4.

**OTHER MATTERS**

Except as otherwise indicated, information contained herein is given as of \_\_\_\_\_, 2005. 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, including the possible nomination of the Caduceus Group's nominees to the Board of Directors. 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     , day of May, 2005 at Novato, California

G. Eric Davis

Vice President, Corporate Counsel and Assistant Secretary

**INFORMATION CONCERNING PARTICIPANTS IN THE SOLICITATION  
OF PROXIES BY BIOMARIN PHARMACEUTICAL INC.**

Under applicable SEC regulations, directors, the Board's nominees for election and certain officers and employees of the Company may be deemed to be participants with respect to the Company's solicitation of proxies in connection with its 2005 annual meeting of stockholders. The following sets forth the name and the present principal occupation or employment, and the name and principal business address of the corporation or other organization in which such employment is carried on, if any, of the persons who may be deemed participants.

**DIRECTORS AND NOMINEES**

The principal occupations of the Company's directors and director nominees who are participants in the Company's solicitation are set forth in Proposal No. 1 under the Election of Directors section of this proxy statement. The name and business addresses of the organizations of employment of the Company's directors and director nominees are as follows:

<u>Name</u>	<u>Business Address</u>
Jean-Jacques Bienaimé	BioMarin Pharmaceutical Inc. 105 Digital Drive Novato, CA 94949
Franz L. Cristiani	N/A
Louis Drapeau	BioMarin Pharmaceutical Inc. 105 Digital Drive Novato, CA 94949
Elaine J. Heron	Labcyte Inc. 1190 Borregas Avenue Sunnyvale CA 94089
Pierre Lapalme	N/A
Erich Sager	Belmont Investments AG Gessnerallee 38 P.O. Box 7328 Zurich 8032

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SWITZERLAND

John Urquhart

AARDEX Ltd.

975 Hamilton Avenue

Palo Alto CA 94301

Gwynn Williams

Scarus Ltd.

Salisbury House 3rd Floor

15 Victoria Street

Douglas

Isle of Man IM1 2LW

United Kingdom

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**OFFICERS AND EMPLOYEES**

The principal occupations of the Company's executive officers and other officers and employees who may be deemed participants in the Company's solicitation of proxies are set forth below. The principal occupation refers to such person's position with the Company, and the business address is BioMarin Pharmaceutical Inc, 105 Digital Drive, Novato, CA 94949.

<u>Name</u>	<u>Principal Occupation</u>
Jean-Jacques Bienaimé	Chief Executive Officer
Louis Drapeau	Chief Financial Officer
Joshua A. Grass	Director, Investor Relations
Emil D. Kakkis	Senior Vice President, Business Operations

**INFORMATION REGARDING OWNERSHIP OF BIOMARIN SECURITIES BY PARTICIPANTS**

Except as described in this *Annex A* or the proxy statement, none of the persons listed above under Directors and Nominees and Officers and Employees owns any BioMarin securities of record which they do not own beneficially. The number of shares of the Company common stock held by directors, the Board's nominees for election, the named executive officers as of April 1, 2005, and any associate of the foregoing persons, is set forth in the Security Ownership of Certain Beneficial Owners and Management section of the proxy statement. The number of shares of the Company's common stock held by the other officers and employees listed above under Officers and Employees as of that date is set forth below. (The information includes shares that may be acquired by the exercise of stock options within 60 days of such date.)

<u>Name</u>	<u>Share Ownership</u>
Joshua A. Grass	9,311(1)

(1) Represents 9,311 shares subject to options exercisable within 60 days of April 1, 2005.

**INFORMATION REGARDING TRANSACTIONS IN BIOMARIN SECURITIES BY PARTICIPANTS**

The following table sets forth purchases and sales during the past two years of shares of the Company's common stock by the persons listed above under Directors and Nominees and Officers and Employees. All transactions were in the public market or pursuant to the Company's equity compensation plans, and none of the purchase price or market value of those shares is represented by funds borrowed or otherwise obtained for the purpose of acquiring or holding such securities. Except where otherwise indicated, the transaction were effected by means on the open market.

**PURCHASES AND SALES OF BIOMARIN COMMON STOCK BY JEAN-JACQUES BIENAIMÉ(1) :**

<b>Date</b>	<b>Transaction Type</b>	<b>Number of Shares</b>
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<u>Date</u>	<u>Transaction Type</u>	<u>Number of Shares</u>
04/01/05	Sale	500
03/30/05	Purchase	500

(1) Mr. Bienaimé was appointed Chief Executive Officer of the Company on May 11, 2005.

PURCHASES AND SALES OF BIOMARIN COMMON STOCK BY FRANZ L. CRISTIANI:

<u>Date</u>	<u>Transaction Type</u>	<u>Number of Shares</u>
06/14/04	Purchase (1)	30,000
07/21/03	Purchase (1)	10,000

(1) Stock option grant.

PURCHASES AND SALES OF BIOMARIN COMMON STOCK BY LOUIS DRAPEAU:

<u>Date</u>	<u>Transaction Type</u>	<u>Number of Shares</u>
04/29/05	Purchase (1)	4,004
01/07/05	Purchase (2)	100,000
10/31/04	Purchase (1)	3,687
08/19/04	Purchase (2)	25,000
05/03/04	Purchase (1)	2,192
12/31/03	Purchase (2)	30,000
11/03/03	Purchase (1)	2,360

- (1) Common Stock purchased pursuant to the Company's 1998 Employee Stock Purchase Plan.  
(2) Stock option grant.

PURCHASES AND SALES OF BIOMARIN COMMON STOCK BY JOSHUA A. GRASS:

<u>Date</u>	<u>Transaction Type</u>	<u>Number of Shares</u>
05/02/05	Sale(1)	1,885
04/29/05	Purchase (2)	1,885
11/02/04	Sale	1,467
10/31/04	Purchase (2)	1,467
06/07/04	Sale	868
04/30/04	Purchase (2)	868
11/06/03	Sale	868
10/31/03	Purchase (2)	868

- (1) Transaction pursuant to a 10b5-1 plan entered into on March 9, 2005.  
(2) Common Stock purchased pursuant to the Company's 1998 Employee Stock Purchase Plan.

PURCHASES AND SALES OF BIOMARIN COMMON STOCK BY ELAINE J. HERON:

<u>Date</u>	<u>Transaction Type</u>	<u>Number of Shares</u>
07/12/04	Purchase (1)	30,000
07/11/03	Purchase (1)	30,000

- (1) Stock option grant.

PURCHASES AND SALES OF BIOMARIN COMMON STOCK BY EMIL D. KAKKIS:

<u>Date</u>	<u>Transaction Type</u>	<u>Number of Shares</u>
04/29/05	Purchase (1)	1,968
01/07/05	Purchase (2)	100,000
10/31/04	Purchase (1)	1,924
05/06/04	Purchase	10,000
05/06/04	Purchase	5,000
12/31/03	Purchase (2)	75,000
12/29/03	Purchase (3)(4)	10,187
12/29/03	Sale (4)	10,187





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- (1) Common Stock purchased pursuant to the Company's 1998 Employee Stock Purchase Plan.
  - (2) Stock option grant.
  - (3) Exercise of stock option.
  - (4) Transaction pursuant to a 10b5-1 plan entered into on May 7, 2003.

PURCHASES AND SALES OF BIOMARIN COMMON STOCK BY PIERRE LAPALME:

<u>Date</u>	<u>Transaction Type</u>	<u>Number of Shares</u>
01/27/05	Purchase (1)	30,000
01/27/04	Purchase (1)	30,000

- (1) Stock option grant.

PURCHASES AND SALES OF BIOMARIN COMMON STOCK BY ERICH SAGER:

<u>Date</u>	<u>Transaction Type</u>	<u>Number of Shares</u>
11/30/04	Purchase (1)	30,000
01/02/04	Purchase (2)(3)	20,000
01/02/04	Sale (3)	20,000
11/28/03	Purchase (1)	30,000

- (1) Stock option grant.
- (2) Exercise of stock option.
- (3) Transaction pursuant to a 10b5-1 plan entered into on May 7, 2003.

PURCHASES AND SALES OF BIOMARIN COMMON STOCK BY JOHN URQUHART:

<u>Date</u>	<u>Transaction Type</u>	<u>Number of Shares</u>
08/11/04	Sale	1,000
07/28/04	Purchase (1)	30,000
07/28/03	Purchase (1)	30,000

- (1) Stock option grant.

PURCHASES AND SALES OF BIOMARIN COMMON STOCK BY GWYNN WILLIAMS:

<u>Date</u>	<u>Transaction Type</u>	<u>Number of Shares</u>
11/30/04	Purchase (1)	30,000
11/28/03	Purchase (1)	30,000
07/28/03	Sale (2)	68,700
07/24/03	Sale (2)	2,500
07/23/03	Sale (2)	16,300
07/18/03	Sale (2)	2,500
07/17/03	Sale (2)	15,000
07/16/03	Sale (2)	15,000
07/15/03	Sale (2)	15,000
07/14/03	Sale (2)	15,000

- (1) Stock option grant.
- (2) Transaction pursuant to a 10b5-1 plan.

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**MISCELLANEOUS INFORMATION CONCERNING PARTICIPANTS**

Except as described in this *Annex A* or the proxy statement, neither any participant nor any of their respective associates or affiliates (together, the Participant Affiliates), is either a party to any transaction or series of transactions since December 31, 2003, or has knowledge of any currently proposed transaction or series of proposed transactions, (i) to which the Company or any of its subsidiaries was or is to be a party, (ii) in which the amount involved exceeds \$60,000, and (iii) in which any participant or Participant Affiliate had, or will have, a direct or indirect material interest. Furthermore, except as described in this *Annex A* or the proxy statement, no participant or Participant Affiliate directly or indirectly beneficially owns any securities of the Company or any securities of any subsidiary of the Company.

Except as described in this *Annex A* or the proxy statement, no participant or Participant Affiliate has entered into any agreement or understanding with any person respecting any future employment by the Company or any of its affiliates or any future transactions to which the Company or any of its affiliates will or may be a party. Except as described in this *Annex A* or the proxy statement, there are no contracts, arrangements or understandings by any participant or Participant Affiliate within the past year with any person with respect to any securities of the Company.

***Stock Trading Plans***

In accordance with our stock trading policy, the following participants have established written sales plans or arrangements as permitted under Rule 10b5-1 of the Exchange Act ( Rule 10b5-1 ): Louis Drapeau, Joshua A. Grass, Emil D. Kakkis, Erich Sager and Gwynn Williams.

Pursuant to Mr. Drapeau's current 10b5-1 sales plan in effect with UBS Financial Services Inc. ( UBS ), dated February 27, 2004, he instructed his broker to sell a specified amount of shares of the Company's common stock during the period from November 1, 2004 through April 30, 2005, subject to the trading price of the Company's common stock, and subject to applicable securities laws, including Rule 144.

Pursuant to Mr. Grass' current 10b5-1 sales plan in effect with UBS, dated March of 2005, he instructed his broker to sell a specified amount of shares of the Company's common stock during the period from May 1, 2005 through February 28, 2006, subject to the trading price of the Company's common stock, and subject to applicable securities laws, including Rule 144.

Mr. Kakkis previously had three 10b5-1 sales plans in effect, one with UBS PaineWebber Inc. ( UBS PaineWebber ) dated May 7, 2003 and effective from May 22, 2003 through May 22, 2004, the second with UBS dated February 27, 2004 and effective from May 1, 2004 until his cancellation of the plan in May of 2004, and the third with UBS dated August 26, 2004 and effective from November 10, 2004 through December 31, 2004.

Mr. Sager previously had two 10b5-1 sales plans in effect, one with UBS PaineWebber dated May 7, 2003 and effective from May 22, 2003 through March 22, 2004, the other with UBS dated February 27, 2004 and effective from May 1, 2004 through January 26, 2005.

Mr. Williams previously had a 10b5-1 sales plan in effect with Merrill Lynch, Pierce, Fenner & Smith Incorporated ( Merrill Lynch ) dated May 7, 2003 and effective from July 14, 2003 to August 29, 2003. Pursuant to Mr. Williams' current 10b5-1 sales plan in effect with Merrill Lynch, he instructed his broker to sell a specified amount of shares of the Company's common stock during the period from May 10, 2004 through April 30, 2005, subject to the trading price of the Company's common stock and subject to applicable securities laws, including Rule 144.

*Employment Agreements*

The employment agreements held by BioMarin's executive officers and other officers and employees who may be deemed participants are set forth in the Executive Compensation section of the proxy statement.

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**FORM OF PROXY**  
**BIOMARIN PHARMACEUTICAL INC.**  
**ANNUAL MEETING OF STOCKHOLDERS**

, 2005

**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 \_\_\_\_\_, 2005, and hereby appoints Louis Drapeau, 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 \_\_\_\_\_, 2005 at the Embassy Suites Hotel, located at 101 McInnis Parkway, San Rafael, California 94903 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)

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**BioMarin's Board of Directors recommends a vote FOR Proposals 1 and 2. If not otherwise directed, this proxy will be voted FOR Proposals 1 and 2 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, Franz L. Cristiani, Elaine J. Heron, Pierre Lapalme, Erich Sager, John Urquhart and Gwynn R. Williams.

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, 2005.

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.