

BIOMARIN PHARMACEUTICAL INC
Form DEF 14A
April 01, 2004

SCHEDULE 14A

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

Filed by the Registrant

Filed by a Party other than the Registrant

Check the appropriate box:

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| <input type="checkbox"/> | Preliminary Proxy Statement | <input type="checkbox"/> | Confidential, for Use of the Commission Only (as permitted by Rule 14a-6(e)(2)) |
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BioMarin Pharmaceutical Inc.

(Name of Registrant as Specified In Its Charter)

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

Payment of Filing Fee (Check the appropriate box):

- No fee required.
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(1) Title of each class of securities to which transaction applies:

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(2) Aggregate number of securities to which transaction applies:

(3) Per unit price or other underlying value of transaction computed pursuant to Exchange Act Rule 0-11 (set forth the amount on which the filing fee is calculated and state how it was determined):

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(1) Amount previously paid:

(2) Form, Schedule or Registration Statement No.:

(3) Filing Party:

(4) Date Filed:

BIOMARIN PHARMACEUTICAL INC.

371 Bel Marin Keys Boulevard, Suite 210

Novato, California 94949

Dear BioMarin Pharmaceutical Inc. Stockholder:

You are cordially invited to attend the annual meeting of the stockholders of BioMarin Pharmaceutical Inc. (BioMarin) to be held on May 5, 2004 at 10:00 a.m. (California time), at the Embassy Suites Hotel located at 101 McInnis Parkway, San Rafael, California 94903.

The purpose of the annual meeting is to consider and vote upon proposals to: (i) elect seven Directors of BioMarin; (ii) ratify the selection by the Board of Directors of KPMG LLP as BioMarin's independent auditors for the year ending December 31, 2004; and (iii) transact such other business as properly may be brought before the BioMarin annual meeting and any adjournment or postponement thereof.

The attached document provides detailed information concerning BioMarin, the proposals to be considered and voted upon at the annual meeting. Please give all of the information contained in the attached document your careful attention.

BioMarin's Board of Directors recommends that you vote FOR the election of each of the nominees to the Board of Directors and FOR the ratification of KPMG LLP as BioMarin's independent auditors.

YOUR VOTE IS IMPORTANT. Even if you do not attend the annual meeting, it is important that your shares be voted. Please vote by promptly completing and mailing the enclosed BioMarin proxy card so that your shares will be represented at the annual meeting and voted as you wish.

Sincerely,

Fredric D. Price

Chairman and Chief Executive Officer

This document is dated March 29, 2004 and is being first mailed to the stockholders of BioMarin on or about April 1, 2004.

BIOMARIN PHARMACEUTICAL INC.

NOTICE OF ANNUAL MEETING OF STOCKHOLDERS

TO BE HELD ON MAY 5, 2004

TO THE STOCKHOLDERS OF BIOMARIN PHARMACEUTICAL INC.:

NOTICE IS HEREBY given that the 2004 Annual Meeting of the stockholders (the Annual Meeting) of BioMarin Pharmaceutical Inc. (BioMarin or the Company) will be held on May 5, 2004 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 seven Directors of the Company;
2. To ratify the selection by the Board of Directors of KPMG LLP as the Company s independent auditors for the year ending December 31, 2004; 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 Proxy).

The Board of Directors has fixed the close of business on March 19, 2004 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 371 Bel Marin Keys Boulevard, Suite 210, Novato, California 94949, for 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 complete, date, sign and return the enclosed proxy as promptly as possible. A postage-prepaid envelope is enclosed for that purpose. Any stockholder attending the Annual Meeting may vote in person even if that stockholder has returned a proxy.

By Order of the Board of Directors:

Louis Drapeau

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Vice President, Finance, Chief Financial Officer and Secretary

Novato, California

March 29, 2004

YOUR VOTE IS IMPORTANT. IN ORDER TO ASSURE YOUR REPRESENTATION AT THE ANNUAL MEETING, YOU ARE REQUESTED TO COMPLETE, SIGN AND DATE THE ENCLOSED PROXY AS PROMPTLY AS POSSIBLE AND RETURN IT IN THE ENCLOSED ENVELOPE.

BioMarin Pharmaceutical Inc.

371 Bel Marin Keys Boulevard, Suite 210

Novato, California 94949

Proxy Statement For 2004 Annual Meeting of Stockholders

INFORMATION CONCERNING SOLICITATION OF PROXIES AND VOTING

General

This Proxy is furnished in connection with the solicitation of proxies by the Board of Directors of the Company (the Board) for use at the Annual Meeting of Stockholders of the Company (the Annual Meeting) to be held on Wednesday, May 5, 2004, or at any adjournment 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 the Embassy Suites Hotel located at 101 McInnis Parkway, San Rafael, California 94903. Copies of this solicitation material and the Company's Annual Report will be 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 April 1, 2004. The Company's Annual Report, which includes its Form 10-K for the year ended December 31, 2003, as filed with the Securities and Exchange Commission, is provided with this Proxy.

Record Date; Outstanding Securities

The voting securities of the Company entitled to vote at the Annual Meeting consist only of shares of common stock. Only stockholders of record at the close of business on March 19, 2004 are entitled to notice of and to vote at the Annual Meeting. On March 19, 2004, there were 64,270,101 shares of BioMarin's common stock, par value \$0.001 per share, issued and outstanding. Each share of common stock is entitled to one vote. As of the record date, there were 113.676 shares of the Company's Series A Preferred Stock outstanding and are all held by an indirect wholly-owned subsidiary of BioMarin. The Series A Preferred Stock does not have any voting rights with respect to matters to be acted upon at the Annual Meeting.

Revocability of Proxies

A stockholder who signs and returns a proxy will have the power to revoke it at any time before it is voted. A proxy may be revoked by (i) filing with the Company (Attention: Louis Drapeau, Vice President, Finance, Chief Financial Officer and Secretary) a written revocation, or (ii) submitting a duly executed proxy bearing a later date, or (iii) by appearing at the Annual Meeting and electing to vote in person.

Voting

Each stockholder is entitled to one vote for each share of common stock held.

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, will be borne by the Company. In addition, 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. Proxies may also be solicited by certain of the Company's Directors, officers and regular employees, without additional compensation, in person or by telephone, facsimile or e-mail.

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

Quorums; 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. 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.

All shares represented by valid proxies received prior to the Annual Meeting will be voted and, where a stockholder specifies by means of the proxy a choice with respect to any matter to be acted upon, the shares will be voted in accordance with the specification so made.

Votes cast by proxy or in person at the Annual Meeting will be tabulated by the Inspector of Elections (the "Inspector") who will be an employee of the Company's transfer agent. The Inspector will also determine whether or not a quorum is present. Other than for the election of Directors, the affirmative vote of a majority of shares present in person or represented by proxy at a duly held Annual Meeting at which a quorum is present is required for approval of each of the matters to be acted on at the Annual Meeting. For the election of Directors, the seven nominees receiving the highest number of votes "FOR" will be elected as Directors. This number is called a plurality.

The Inspector will treat shares that are voted "WITHHELD" or "ABSTAIN" as being present and entitled to vote for purposes of determining the presence of a quorum but such shares will not be treated as votes in favor of approving any matter submitted to the stockholders for a vote. When proxies are properly dated, 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 will be voted (i) FOR the election of the seven nominees for Directors set forth herein; and (ii) FOR the ratification of the selection by the Board of KPMG LLP as the Company's independent auditors for the year ending December 31, 2004. The Company is not seeking proxies for the authority to vote on any other business as may properly come before the Annual Meeting or any adjournment thereof.

If a broker indicates on the enclosed proxy or its substitute that such broker does not have discretionary authority as to certain shares to vote on a particular matter ("Broker Non-Votes"), those shares will not be considered as present with respect to that matter. The Company believes that the tabulation procedure to be followed by the Inspector is consistent with the general statutory requirements in Delaware concerning voting of shares and determination of a quorum.

Submission of Stockholder Proposals for 2005 Annual Meeting

Stockholders who intend to submit a proposal for inclusion in the Company's Proxy materials for the 2005 Annual Meeting of Stockholders must submit the proposal to the Company no later than January 15, 2005. Stockholders who intend to present a proposal at the 2005 Annual Meeting are required to provide notice of such proposal to the Company no later than March 15, 2005. The Company reserves the right to reject, rule out of order, or take other appropriate action with respect to any proposal that does not comply with these and other applicable requirements.

PROPOSAL ONE: ELECTION OF DIRECTORS

The Company has a Board currently consisting of eight Directors. However, effective as of the time of the Annual Meeting, the size of the Board will be reduced to seven Directors. At the Annual Meeting, stockholders will elect seven Directors. The proxy holders may not vote the proxies for a greater number of people than the number of nominees named. Unless otherwise instructed, the proxy holders will vote the proxies received by them FOR the seven nominees named below, all of whom are presently Directors of the Company. If any nominee is unable or declines to serve as a Director at the time of the Annual Meeting, the proxies will be voted for any nominee who shall be designated by the present Board to fill the vacancy. It is not expected that any nominee will be unable to or will decline to serve as a Director. If stockholders nominate additional persons for election as Directors, the proxy holder will vote all proxies received by him to assure the election of as many of the Board's nominees as possible with the proxy holder making any required selection of specific nominees to be voted for. 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.

In addition to the Directors nominated for re-election, Dr. Phyllis Gardner served as a Director during 2003 until her resignation on March 21, 2003. On January 27, 2004, Mr. Vijay Samant tendered his resignation as a Director, to be effective May 4, 2004, and declined to stand for re-election to the Board for 2004. Neither of these individuals has any continuing relationship with the Company. On July 28, 2003 and January 27, 2004, the Board appointed Dr. John Urquhart and Mr. Pierre Lapalme, respectively, as Directors to serve until the Annual Meeting.

If a quorum is present, the seven 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 nominees to the Board and certain information regarding each nominee:

<u>Name</u>	<u>Age</u>	<u>Position with BioMarin</u>	<u>Director Since</u>
Fredric D. Price	58	Chief Executive Officer and Chairman of the Board	October 2000
Franz L. Cristiani(2)(3)	62	Director	June 2002
Elaine J. Heron, Ph.D.(2)(4)	56	Director	July 2002
Pierre Lapalme (1)(3)	63	Director	January 2004
Erich Sager(1)	46	Director	November 1997
John Urquhart, M.D.(1)(4)	70	Director	July 2003
Gwynn R. Williams(2)(3)	70	Director	October 1996

- (1) Member of BioMarin's Compensation Committee effective April 1, 2004.
(2) Member of BioMarin's Audit Committee.
(3) Member of BioMarin's Corporate Governance and Nominating Committee effective April 1, 2004.
(4) Member of BioMarin's Science Committee.

Each nominee to the Board has consented to being named in the Proxy and to serve as a Director if elected. There is no family relationship between any Director and any executive officer of BioMarin.

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Fredric D. Price was elected Chairman of the Board and Chief Executive Officer of BioMarin in October 2000. From September 1994 to September 2000, he was President, Chief Executive Officer, and a member of the board of Applied Microbiology/AMBI, a biotechnology and nutrition company. From July 1991 to September 1994, he was Vice President Finance & Administration and Chief Financial Officer of Regeneron Pharmaceuticals. From March 1986 to July 1991, he was a pharmaceuticals and biotechnology industry strategy

consultant. For the 13 previous years, he was employed by Pfizer Pharmaceuticals where he was a Vice President with both staff and line responsibilities. Mr. Price received a B.A. from Dartmouth College and an M.B.A. from the Wharton School of the University of Pennsylvania.

Franz L. Cristiani joined the BioMarin Board in June 2002 and serves as Chairman of its Audit Committee and its Lead Director. Mr. Cristiani provided consulting services to BioMarin from January 2002 to May 2002. From 1999 to 2001, Mr. Cristiani devoted his time to personal affairs. 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. Mr. Cristiani serves as a director of MTI Technology, Inc. and Vitasoy USA. He holds a B.A. from San Francisco State University.

Elaine J. Heron, Ph.D., has served as a Director of BioMarin since July 2002. Prior to her present position as Chairman and Chief Executive Officer of Labcyte Inc., which she assumed in July 2001, she spent six years in increasingly responsible positions at the Applied Biosystems Group of Applied Biosystems Corporation, including the positions of 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 has served as a Director of BioMarin since January 2004. From 1995 until his retirement in 2003, he served as the President and Chief Executive Officer of North America Ethypharm, Inc., a drug delivery company. Throughout his career, Mr. Lapalme 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 non-executive Chairman of the Board of Diagnocure, a biotechnology company, as director of First Horizon Pharmaceuticals, and is a board member of EXIMIAS Pharmaceutical Corporation and International Wex Technologies. Mr. Lapalme studied at the University of Western Ontario and INSEAD France.

Erich Sager has served as a Director of BioMarin since November 1997. Since September 1996, Mr. Sager has 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 Belmont Holdings AG, Belmont Invest AG, Beverage Metrics LLC, Telinvest Holding AG and Calltrade Carrier Services AG. Mr. Sager received a business degree from the School of Economics and Business Administration in Zurich, Switzerland.

Gwynn R. Williams has served as a Director of BioMarin since its incorporation in October 1996 and serves as Chairman of its Corporate Governance and Nominating Committee. He was a founder of numerous companies including, Xoma Inc., Murex Inc., AstroMed Ltd. and Astroscan Ltd. 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. Mr. Williams is currently a trustee of the United Kingdom Transplant Trust. Mr. Williams was a major stockholder of Glyko Biomedical Ltd. and its predecessor, Glyko, Inc. He earned 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.

John Urquhart, M.D., was appointed to the Company's Board of Directors in July 2003, filling a vacancy, and serves as Chairman of its Compensation and Science Committees 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 the pharmaceutical packaging company, and the Chief Executive Officer of APREX, also a pharmaceutical packaging company and a subsidiary of AARDEX. Since 1991, he has been a member of the board of directors of the Cayman subsidiary of HBM Bioventures, a Swiss-based venture capital firm. Dr. Urquhart also serves as a non-executive Vice Chairman of the board of directors of Inveresk Research Group, Inc. and serves as a director of Inveresk Research Group Limited. Dr. Urquhart has been Extraordinary Professor of Pharmacoepidemiology at Maastricht University in the Netherlands since 1991, concentrating on graduate education and research, and an 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 Bachelor's degree from Rice University.

Board Meetings and Board Committees

The Board of Directors 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, a Science Committee and an Employee Option Committee whose functions are briefly described below.

The Board of Directors of the Company held a total of nine meetings during the year ended December 31, 2003 and took action by unanimous written consent on four occasions. No Director participated in fewer than 75% of all such meetings of the Board and the committees held during fiscal 2003, if any, upon which such Directors served.

Audit Committee. The Audit Committee provides oversight of (i) the financial reporting process, the system of internal controls and the audit process of BioMarin and (ii) BioMarin's independent auditors. The Audit Committee also recommends to the Board the appointment of BioMarin's independent auditors. As required by the Audit Charter, each of the members of the Audit Committee is an independent director as defined by the Nasdaq rules. The members of the Audit Committee are Mr. Cristiani, Chairman, Dr. Heron, and Mr. Williams. The Board has determined that Mr. Cristiani is a financial expert as such term is defined in Item 401(h)(2) of Regulation S-K. During 2003, the Audit Committee met on nine occasions.

Compensation Committee. The Compensation Committee sets general compensation policy for BioMarin and has final approval power over the 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. Effective on April 1, 2004, Dr. Urquhart will replace Mr. Sager (who will continue to serve as a Compensation Committee member) as Chairman of the Compensation Committee, Mr. Lapalme will replace Mr. Samant and Dr. Heron will no longer be a member of the Compensation Committee. During 2003, the Compensation Committee met on three occasions.

Corporate Governance and Nominating Committee. The Corporate Governance and Nominating Committee is responsible for identifying individuals to fill vacancies on the Board, recommending nominees to be voted upon at the annual meeting of stockholders, and 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. The Corporate Governance and Nominating Committee will consider nominees recommended by stockholders if such nominations have been submitted in writing, accompanied both by a description of the proposed nominee's qualifications and an indication of the consent of the proposed nominee and relevant biographical information. The recommendation should be addressed to the Corporate Governance and Nominating Committee in care of the Secretary of the Company. Recommendations for consideration for the 2005 Annual Meeting must be received by the Company by January 15, 2005. Effective April 1, 2004, Mr. Williams will continue to chair the Corporate Governance and Nominating Committee and Mr. Lapalme will replace Mr. Sager on the Committee. Mr. Cristiani will continue to serve as a member of the

Corporate Governance and Nominating Committee. Also effective April 1, 2004, the Board amended the Corporate Governance and Nominating Charter. A copy of the amended Charter is attached hereto as Annex A. The Corporate Governance and Nominating Committee met five times during 2003.

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 of Directors. Dr. Urquhart chairs the Science Committee with Dr. Heron as a member.

Employee Option Committee. The Employee Option Committee, which consists of Mr. Price, determines grants of stock options pursuant to BioMarin's 1997 Stock Plan to employees of BioMarin below the level of vice president. The option grants by the Employee Option Committee may be made only within the guidelines and parameters established by the Compensation Committee.

Compensation Committee Interlocks and Insider Participation

None of the members of the Compensation Committee is currently or had been, at any time since the formation of the Company, an officer or employee of the Company. No member of the Compensation Committee serves as a member of the board or compensation committee of any entity that has one or more executive officers serving as a member of the Company's Board or Compensation Committee.

Director Compensation

In 2003, the Board approved an increase in the amount of initial and annual subsequent options issued under the 1998 Director Option Plan from options to purchase 20,000 and 15,000 shares of BioMarin common stock, respectively to options to purchase 30,000 shares of BioMarin common stock each. In March 2003 the Board approved an increase in the annual cash compensation from \$7,500 to \$20,000 for each non-employee Director and an additional \$10,000 (a \$7,500 increase) for serving as a chair of a Board committee. In June 2003, the Board approved an increase in the annual cash compensation from \$20,000 to \$25,000 for each non-employee Director and increased the compensation of the Audit Committee Chairman to \$15,000 annually. Additionally, Audit Committee members receive \$7,500 annually, and other committee members receive \$5,000 for each non-Audit Committee position and the Lead Director receives \$5,000 annually. Directors are also reimbursed for their reasonable expenses in attending meetings of the Board and while performing services for BioMarin.

2003 Director Compensation

Director Name	Director Fees	Option Grant Date	Number of Options Granted	Exercise Price Per Share	Option Expiry
Franz L. Cristiani	\$ 25,000	6/13/03	20,000	\$ 12.20	6/13/13
		7/21/03	10,000	\$ 9.90	7/21/13
Elaine J. Heron, Ph.D.	\$ 19,375	7/11/03	30,000	\$ 10.38	7/11/13
Pierre Lapalme					
Erich Sager	\$ 22,500	11/28/03	30,000	\$ 7.35	11/28/13
Vijay Samant	\$ 16,250				

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		6/13/03	20,000	\$	12.20	8/3/04
		7/21/03	10,000	\$	9.90	8/3/04
John Urquhart, M.D.	\$ 10,000	7/28/03	30,000	\$	10.34	7/28/13
Gwynn R. Williams	\$ 23,125	11/28/03	30,000	\$	7.35	11/28/13

1998 Director Option Plan

The 1998 Director Option Plan was adopted by the Board in December 1999. It was approved by BioMarin's stockholders as of January 15, 2000. The 1998 Director Option Plan provides for the grant of nonstatutory stock options to non-employee Directors. A total of 700,000 shares of BioMarin common stock have been reserved for issuance under the 1998 Director Option Plan. The 1998 Director Option Plan also provides for an annual increase in this number of shares equal to the lesser of: (i) 0.5% of the BioMarin's outstanding common stock, (ii) 200,000 shares, or (iii) a lesser amount determined by the Board.

In 2003, options to purchase, in the aggregate, 180,000 shares of BioMarin common stock were issued to Directors under the 1998 Director Option Plan.

The 1998 Director Option Plan, as amended in 2003, 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 shall 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 shall be 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.

The Board unanimously recommends voting FOR each of the foregoing nominees for Director.

REPORT OF THE CORPORATE GOVERNANCE AND NOMINATING COMMITTEE

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

The Nominating 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 Nominating Committee may desire for a particular opening. This may involve locating someone with a specific background, skill set or experiences. Once identified, the Nominating Committee next looks to the best method of finding a candidate who satisfies the specified criteria. The Nominating Committee may consider candidates recommended by management, by other members of the Nominating Committee or the Board, by stockholders or it may engage a third party to conduct a search for possible candidates. The Nominating Committee accepts recommendations for director candidates from stockholders and information regarding the submission of candidates is included in this Proxy under the heading Stockholder Communications with the Board .

Once candidates are identified, the Nominating 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 Nominating 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, career specialization, relevant technical skills, diversity and the extent to which the candidate would fill a present need on the Board.

If the Nominating Committee determines that a candidate should be nominated as a candidate in the Proxy, the candidate s nomination is then recommended to the Board, who may in turn conduct their own review to the extent they deem it appropriate. When the Board has agreed upon a candidate, they are then recommended to the stockholders for election at an Annual Meeting of Stockholders.

Other than Mr. Samant, who has decided to resign, all of the current directors have been recommended by the Nominating Committee to the Board of Directors for re-election as directors of the Company at the Annual Meeting and the Board has approved the recommendations. The Company did not engage a third party search firm to assist with the selection of the director candidates for the Annual Meeting. The Company received no recommendation for directors from any stockholders.

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

approving Board meeting schedules and meeting agendas

approving Board meeting materials

serving as the liaison between the Chairman and the independent board members

leading executive sessions of the independent board members

setting meetings of independent board members

being available for consultation with major stockholders

STOCKHOLDER COMMUNICATIONS WITH THE BOARD

The Company has not adopted a formal process for stockholders to communicate directly with members of the Board. In the past, the Board has felt there was not a need for a formal policy, as stockholders could contact the Board through the Chairman and Chief Executive Officer, who is located at the Company's headquarters in Novato. While the Board will review the need for a formal policy, stockholders who wish to contact the Board may continue to do so through the Chairman and Chief Executive Officer. Stockholders who wish to contact Board members other than the Chairman may do so by directing their correspondence to such member in care of Franz Cristiani, Audit Committee Chairman and Lead Director, PO Box 130A, San Carlos, CA 94070.

It is the Company's policy to request that all Board members attend the Annual Meeting. However, the Company also recognizes that personal attendance by all directors is not always possible. All of the directors attended the 2003 annual meeting of stockholders.

PROPOSAL TWO: RATIFICATION OF AUDITORS

The Board has selected KPMG LLP, independent auditors, to audit the financial statements of the Company for the year ending December 31, 2004, and recommends that stockholders vote for ratification of such appointment. Although action by stockholders is not required by law, the Board has determined that it is desirable to request approval of this selection by the stockholders. Notwithstanding this selection, the Board, in its discretion, may direct the appointment of new independent auditors at any time during the year, if the Board feels that such a change would be in the best interest of the Company and its stockholders. In the event of a negative vote on ratification, the Board will reconsider its selection.

Auditors

Since June 11, 2002, KPMG LLP has been BioMarin's independent auditors. Arthur Andersen LLP, certified public accountants, were the independent auditors of BioMarin from BioMarin's inception on March 21, 1997 until June 11, 2002. During that time, Arthur Andersen LLP audited BioMarin's financial statements annually.

Effective as of June 11, 2002, BioMarin dismissed Arthur Andersen LLP as its independent auditors. BioMarin's Audit Committee recommended the dismissal of Arthur Andersen LLP and the entire Board participated in and approved such dismissal. Effective as of the same date, BioMarin engaged KPMG LLP as its new independent auditors. BioMarin's Audit Committee recommended this action, which was approved unanimously by its Board and was ratified by the stockholders in 2003.

During fiscal years 2000 and 2001 and through June 11, 2002, BioMarin had not consulted with KPMG LLP regarding either:

the application of accounting principles to a specified transaction, either completed or proposed; or the type of audit opinion that might be rendered on BioMarin's financial statements, and either a written report was provided to BioMarin or oral advice was provided that KPMG LLP concluded was an important factor considered by BioMarin in reaching a decision as to the accounting, auditing of financial reporting issue; or

any matter that was either the subject of a disagreement, as that term is defined in Item 304(a)(1)(iv) of Regulation S-K under the Securities Act and the related instructions to Item 304 of Regulation S-K under the Securities Act, or a reportable event, as that term is defined in Item 304(a)(1)(v) of Regulation S-K under the Securities Act.

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

Audit Fees. For the year ended December 31, 2002, Arthur Andersen LLP billed BioMarin \$20,000 for services related to the audit of the Company's financial statements and review of financial statements included in the Company's Form 10-Q. For the years ended December 31, 2003 and 2002, KPMG LLP billed the Company \$214,428 and \$115,000 for such services, respectively.

Audit Related Fees. For the year ended December 31, 2002, Arthur Andersen LLP billed BioMarin \$16,000 for assurance and other services related to the performance of the audit of the Company's financial statements and review of financial statements. These services primarily involved review and assurance services associated with the Company's equity offerings and related regulatory filings. For the years ended

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December 31, 2003 and 2002, KPMG LLP billed \$183,000 and \$0 for assurance and other services related to the performance of the audit of the Company's financial statements and review of financial statements. These services primarily involved review and assurance services associated with the Company's debt and equity offerings and related regulatory filings.

Tax fees:

Tax compliance fees. For the year ended December 31, 2002, Arthur Andersen LLP billed BioMarin \$7,698 for services related to tax compliance. For the years ended December 31, 2003 and 2002, KPMG LLP billed the Company \$53,860 and \$50,500 for such services, respectively. In each instance, these services related to the preparation of Federal, state and foreign tax returns and other filings.

Tax consulting and advisory services. For the year ended December 31, 2003 KPMG LLP provided tax consulting or advisory services and has billed the Company \$9,100. For the year ended December 31, 2003, the Company retained Deloitte & Touche LLP as its tax consultants in the area of tax strategy and minimizing Federal, state and local taxes and has billed the Company \$63,444 for such services.

All Other Fees. For the year ended December 31, 2002, Arthur Andersen LLP billed BioMarin \$562,197 for services other than those described above. The services in 2002 included \$412,354 related to the implementation of the Company's financial reporting system and \$149,843 related to consultation on various accounting matters, including acquisitions of assets and other entities. For the years ended December 31, 2003 and 2002, KPMG LLP did not perform services other than those described above.

As provided in the Audit Committee Charter the Audit Committee pre-approves all services provided by its independent auditors. All of the above services and 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 auditors for the year ending December 31, 2004.

**OTHER INFORMATION RELATED TO BIOMARIN, THE DIRECTORS
AND EXECUTIVE OFFICERS**

Security Ownership Of Certain Beneficial Owners

The following table sets forth certain information with respect to the beneficial ownership of the Company's common stock as of March 15, 2004 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; (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 is as supplied or confirmed by such person, based upon statements filed with the Securities and Exchange Commission, or based upon the actual knowledge of BioMarin. Except as otherwise indicated, the mailing address for each stockholder in the table below is c/o BioMarin Pharmaceutical Inc., 371 Bel Marin Keys Blvd., Suite 210, Novato, California 94949.

Name of Beneficial Owner	Number of Shares Beneficially Owned(1)	Number of Shares Subject To Options(2)	Percentage of Common Stock
PRIMECAP Management Company(3)	6,205,500		9.7%
Fredric D. Price	845,906	781,621	1.3%
Franz L. Cristiani	37,500	37,500	*
Elaine J. Heron, Ph.D.	42,500	42,500	*
Pierre Lapalme	7,500	7,500	*
Erich Sager	159,944	156,635	*
Vijay B. Samant	42,500	42,500	*
John Urquhart, M.D.	24,500	24,500	*
Gwynn R. Williams	895,065	82,500	1.4%
Emil D. Kakkis, M.D., Ph.D.	323,154	312,044	*
Jeffrey I. Landau	79,975	78,438	*
Christopher M. Starr, Ph.D.	572,476	252,300	*
Stuart J. Swiedler, M.D., Ph.D.	208,311	198,311	*
All current executive officers and Directors as a group (13 persons)	3,386,391	2,163,850	5.2%

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

- (1) The "Number of Shares Beneficially Owned" column is based on shares of common stock outstanding at March 15, 2004. Shares of common stock subject to options or warrants that are exercisable within 60 days of March 15, 2004 (the "Number of Shares Beneficially Owned") 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 March 15, 2004. These shares are in the calculation of the "Number of Shares Beneficially Owned."
- (3) The mailing address for PRIMECAP Management Company is 225 South Lake Avenue #400, Pasadena, CA 91101.

Executive Officers and Other Significant Employees of BioMarin

The following table sets forth certain information concerning BioMarin's executive officers (other than Fredric D. Price) and other significant employees as of March 15, 2004.

<u>Name</u>	<u>Age</u>	<u>Position with BioMarin</u>
Robert A. Baffi, Ph.D.	49	Senior Vice President, Technical Operations
Emil D. Kakkis, M.D., Ph.D.	43	Senior Vice President, Business Operations
Jeffrey I. Landau	61	Senior Vice President, Administration
Christopher M. Starr, Ph.D.	51	Co-founder, Senior Vice President and Chief Scientific Officer
Jeffrey H. Cooper, C.P.A.	48	Vice President, Controller
Louis Drapeau, C.P.A.	60	Vice President, Finance, Chief Financial Officer and Secretary
Robert A. Heft, Ph.D.	49	Vice President, Corporate Planning
Andria G.M. Langenberg, M.D.	47	Vice President, Clinical Development and Operations
Daniel P. Maher	48	Vice President, Program Management
Matthew R. Patterson	32	Vice President, Commercial Planning and Government Affairs
Stuart J. Swiedler, M.D., Ph.D.	48	Vice President, Clinical Affairs
Kim R. Tsuchimoto, C.P.A.	40	Vice President, Treasurer

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

Dr. Baffi joined BioMarin in May 2000, and currently 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.

Jeffrey I. Landau, Senior Vice President, Administration

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Mr. Landau joined BioMarin as Vice President, Administration in April 2002 and was promoted to Senior Vice President, Administration in January 2003. Prior to joining BioMarin, Mr. Landau served as the Chief Operating Officer for Transition 1 Management Accounting Systems, a consulting company, from 2000 to 2002, and served as the Chief Operating Officer of GlobeNet Software from 1998 to 1999. He has also served as Vice President of Operations and Vice President of Information Technology at a number of major corporations including Pfizer Pharmaceuticals, The Franklin Mint, NutriSystem, and the Walt Disney Corporation. Mr. Landau received a M.S. in computer and operations research, and a B.S. in mathematics from New York University.

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

Dr. Starr co-founded BioMarin in 1997 and currently serves as Senior Vice President and Chief Scientific Officer. Before BioMarin, Dr. Starr directed Glyko Inc.'s research programs. 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.

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

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 an M.B.A. from Santa Clara University and a B.A. in economics from the University of California, Los Angeles.

Louis Drapeau, C.P.A. Vice President, Finance, Chief Financial Officer and Secretary

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. His skill set includes working with businesses that have 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. in Finance and a B.S. in mechanical engineering from Stanford University.

Robert A. Heft, Ph.D., Vice President, Corporate Planning

Dr. Heft, Vice President, Corporate Planning, joined BioMarin in October 2001, following the Company's acquisition of the pharmaceutical assets of IBEX Technologies Inc. Dr. Heft founded IBEX in 1986 and served as its President and Chief Scientist until joining BioMarin. He has directed the preclinical and clinical development of several enzyme therapies, including enzymes for metabolic disorders, and has been awarded several patents and authored papers related to this work. He is a past member of the Board of Directors of the Canadian Genetic Diseases Network and the Industrial Biotechnology Association of Canada. Dr. Heft holds a Ph.D. from the Massachusetts Institute of Technology in genetic engineering/radiological sciences, a master's degree in nuclear engineering from Cornell University, and a bachelor of mechanical engineering from McGill University.

Andria G.M. Langenberg, M.D., Vice President, Clinical Development and Operations

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Dr. Langenberg joined BioMarin in November 2003, as Vice President, Clinical Development and Operations, bringing with her approximately 14 years of domestic and international clinical research and development experience. Prior to joining BioMarin, Dr. Langenberg worked at Chiron Corporation for over 11 years, serving most recently as Senior Director of Clinical Development and Head of Clinical Operations. Dr. Langenberg is board-certified in both internal medicine and dermatology, and has an Associate Clinical

Professor appointment in the dermatology department at the University of California, San Francisco. She earned an M.D. from the University of Nebraska Medical Center, Omaha, and a B.S in life sciences at the University of Nebraska, Lincoln.

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.

Matthew R. Patterson, Vice President, Commercial Planning and Government Affairs

Mr. Patterson joined BioMarin in August 1998, and currently serves as Vice President, Commercial Planning and Government Affairs, a position he was appointed to in July 2003. Since joining the Company, he has played a critical role in guiding BioMarin's program development strategy and managing corporate relationships. Prior to BioMarin, Mr. Patterson worked for Genzyme Corporation for five years, initially in manufacturing and subsequently in regulatory affairs. While at Genzyme, he contributed to the development and global licensing of multiple biotechnology products. Mr. Patterson received a B.A. in biochemistry from Bowdoin College.

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

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

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 California certified public accountant and received a B.S. in business administration from San Francisco State University.

Executive Compensation

Summary Compensation Table

The Company is required by the SEC to disclose compensation awarded to, earned by, or paid for services rendered to the Company in all capacities during the last three fiscal years to (a) the Company's Chief Executive Officer; (b) the Company's four most highly compensated officers, other than the Chief Executive Officer, who were serving as officers at the end of fiscal year 2003; and (c) up to two additional individuals for whom such

disclosure would have been provided but for the fact that the individual was not serving as an executive officer of the Company at the end of fiscal year 2003; provided, however, that no disclosure need be provided for any executive officer, other than the CEO, whose total annual salary and bonus does not exceed \$100,000.

Accordingly, the following table discloses compensation paid by the Company during the last three fiscal years to (a) Fredric D. Price, the Company's Chairman and Chief Executive Officer; (b) Christopher M. Starr, Ph.D., Emil D. Kakkis, M.D., Ph.D., Jeffrey Landau, 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 2003 and whose salary and bonus exceeded \$100,000. These individuals are referred to as the Named Executive Officers.

The entries under the column "All Other Compensation" in the table represent the premiums paid for life insurance benefits, financial planning consultation, 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)	
Fredric D. Price	2003	507,082	400,000(3)		625,000	6,400
Chairman and Chief Executive Officer	2002	458,129	225,000(2)			2,496
	2001	408,269	278,500		175,000	989
Christopher M. Starr, Ph.D.	2003	275,454	110,000(3)		125,000	4,992
Senior Vice President and Chief Scientific Officer	2002	262,500	60,000(2)			4,522
	2001	250,000			25,000	2,294
Emil D. Kakkis, M.D., Ph.D.	2003	270,027	110,000(3)		125,000	4,974
Senior Vice President, Business Operations	2002	257,250	110,000(2)(4)			4,522
	2001	245,000			25,000	1,869
Jeffrey I. Landau	2003	249,355	110,000(3)		115,000	6,100
Senior Vice President, Administration	2002	168,750	42,500(2)	4,000	125,000	5,174
	2001					
Stuart J. Swiedler, M.D., Ph.D.	2003	256,223	80,000(3)		80,000	4,923
Vice President, Clinical Affairs	2002	241,500	42,500(2)			4,522
	2001	230,000			20,000	1,869

(1) For 2002 performance, the Company granted on January 6, 2003 to the Named Executive Officers options for the purchase of the following number of shares of BioMarin common stock: Mr. Price, 125,000 shares; Dr. Starr, 50,000 shares; Dr. Kakkis, 50,000 shares; Mr. Landau, 40,000 shares; and Dr. Swiedler, 30,000 shares. On October 31, 2003, pursuant to an employment agreement, the Company granted to Mr. Price options for the purchase of 500,000 shares. For 2003 performance, on December 31, 2003, the Company granted to

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the Named Executive Officers options for the purchase of the following number of shares of BioMarin common stock: Dr. Starr, 75,000 shares; Dr. Kakkis, 75,000 shares; Mr. Landau, 75,000 shares; and Dr. Swiedler, 50,000 shares.

- (2) In the first quarter of 2003, the Board awarded Mr. Price and each of the Named Executive Officers cash bonuses for their respective performance in 2002. The bonuses were paid in 2003, shortly after they were awarded.
- (3) Bonuses awarded for 2003 performance were paid in the first quarter of 2004.
- (4) In addition to Dr. Kakkis' 2002 performance bonus of \$60,000, Dr. Kakkis earned a \$50,000 bonus for the marketing approval of Aldurazyme, pursuant to his employment agreement.

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%	
Fredric D. Price	125,000(4)	4%	\$ 6.43	1/6/13	505,474	1,280,971	2/1/03
	500,000(5)	14%	7.00	10/31/13	2,201,131	5,578,099	11/30/03
Christopher M. Starr, Ph.D.	50,000(4)	1%	6.43	1/6/13	202,190	512,388	6/30/03
	75,000(6)	2%	7.76	12/31/13	366,017	927,558	6/30/04
Emil D. Kakkis, M.D., Ph.D.	50,000(4)	1%	6.43	1/6/13	202,190	512,388	6/30/03
	75,000(6)	2%	7.76	12/31/13	366,017	927,558	6/30/04
Jeffrey I. Landau	40,000(4)	1%	6.43	1/6/13	161,752	409,911	6/30/03
	75,000(6)	2%	7.76	12/31/13	366,017	927,558	6/30/04
Stuart J. Swiedler, M.D., Ph.D.	30,000(4)	1%	6.43	1/6/13	121,314	307,433	6/30/03
	50,000(6)	1%	7.76	12/31/13	244,011	618,372	6/30/04

- (1) Based on an aggregate of 3,482,775 shares subject to options granted by BioMarin during fiscal year 2003 to employees, consultants and the Named Executive Officers.
- (2) Options were granted at an exercise price equal to the closing price of the BioMarin 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. BioMarin cannot assure any Named Executive Officer or any other holder of its securities that the actual stock price appreciation over the option term will be at the assumed 5% and 10% levels or at any other defined level. Unless the market price of its 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 or sales of appreciated stock. The values shown do not consider non-transferability or termination of the options upon termination of such employee's employment with BioMarin.
- (4) Options were granted at an exercise price of \$6.43 per share on January 6, 2003, representing bonuses for 2002 performance.
- (5) Options were granted on October 31, 2003, at an exercise price of \$7.00 per share, pursuant to Mr. Price's employment agreement executed in March 2003.
- (6) Options were granted at an exercise price of \$7.76 per share on December 31, 2003, representing bonuses for 2003 performance.

Fiscal 2003 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 2003.

Options Exercised During Fiscal Year 2003

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
Fredric D. Price		\$	713,913	609,375	\$ 87,083	\$ 459,167
Christopher M. Starr, Ph.D.	120,000	680,225	241,883	137,500	430,225	49,875
Emil D. Kakkis, M.D., Ph.D.	25,375	90,959	301,627	137,500	749,825	49,875
Jeffrey I. Landau			64,688	175,313	13,300	39,900
Stuart J. Swiedler, M.D., Ph.D.	23,072	105,592	191,019	91,875	404,775	29,925

(1) Based on closing price on December 31, 2003 of \$7.76 per share less exercise price per share.

Employment Agreements

BioMarin is party to employment agreements with the Named Executive Officers and each current executive officer on the terms enumerated on the chart below other than the Company's Chief Executive Officer, Mr. Price, whose agreement is described below. Each of these employment agreements is terminable without cause by BioMarin upon six months' prior written notice to the officer, or by the officer upon three months' prior written notice to BioMarin. BioMarin is obligated to pay the officer's salary and benefits until termination.

In January 2004, the Company instituted a new change of control policy that is applicable to each of the officers listed in the table below, other than Mr. Price. Under the new policy, in the event of a change of control, all options held by the officer will immediately vest. Additionally, for Dr. Starr, Dr. Kakkis, Mr. Landau and Dr. Baffi, in the event that any one of them is involuntarily terminated within one year of a change of control of BioMarin, such person will receive a severance payment equal to 12 months of his base salary and continuation of employer sponsored benefits for 12 months. Also, Mr. Drapeau and Dr. Swiedler, in the event that any one of them is involuntarily terminated within one year of a change of control of BioMarin, such person will receive a severance payment equal to 10 months of his base salary and continuation of employer sponsored benefits for 10 months.

Name of Executive Officer	2003 Salary Rate(1)	Annual Bonus	Initial Grant of Right to	
			Purchase Equity Securities(2)	Agreement Termination Date
Fredric D. Price	\$550,000	Annual bonus, payable in cash based on performance.	Options to purchase up to 500,000 shares of BioMarin's common stock at a purchase price of \$12.50 per share.	October 30, 2006
Christopher M. Starr, Ph.D.	\$275,500	Annual bonus, payable in cash and/or stock.	400,000 shares of BioMarin's common stock at a purchase price of \$1.00 per share.	None
Emil D. Kakkis, M.D., Ph.D.	\$270,250	Eligible to receive a cash bonus based on achievement of milestones and an annual bonus, payable in cash and/or stock.	Options to purchase up to 200,000 shares of BioMarin's common stock at a purchase price of \$4.00 per share.	None
Stuart J. Swiedler, M.D., Ph.D.	\$256,500	Annual bonus, payable in cash and/or stock.	Options to purchase up to 150,000 shares of BioMarin's common stock at a purchase price of \$4.00 per share.	None
Louis Drapeau, C.P.A.	\$250,000	Annual bonus, payable in cash and/or stock.	Options to purchase up to 125,000 and 25,000 shares of BioMarin's common stock at a purchase price of \$9.25 and \$4.24, respectively.	None
Jeffrey I. Landau	\$250,000	Annual bonus, payable in cash and/or stock.	Options to purchase up to 125,000 shares of BioMarin's common stock at a purchase price of \$9.25 per share.	None
Robert A. Baffi, Ph.D.	\$242,500	Annual bonus, payable in cash and/or stock.	Options to purchase up to 200,000 shares of BioMarin's common stock at a purchase price of \$22.00 per share.	None

- (1) 2003 Annual Salary Rate reflected in the above table reflects 2003 annual salary rate as of December 31, 2003. The table entitled Summary Compensation Table reflects actual salaries paid in 2003 and includes mid-year salary adjustments.
- (2) Options in this table were granted on commencement of employment for each individual.

On March 14, 2003, BioMarin and Mr. Price amended and restated, in its entirety, the former employment agreement between BioMarin and Mr. Price, dated October 31, 2000. The amended agreement provides for a continuation of the salary and bonus terms of the existing agreement until October 30, 2003 and thereafter provides for an annual base salary of \$550,000 for the year ending October 30, 2004, with the potential for increased amounts for the subsequent two years and a potential performance bonus of 0% to 125% of Mr. Price's base salary for each of the years ending October 30, 2004, 2005 and 2006. The amended agreement did not modify the terms of the interest-deferred loan made by BioMarin to Mr. Price in 2000 in the amount of \$860,000, which becomes due and payable to BioMarin on October 31, 2006.

Under the amended agreement, Mr. Price received a grant of 39,285 restricted shares (valued at \$275,000 based on a closing price of \$7 per share) of BioMarin's common stock on October 31, 2003 (to vest on October 31, 2004) and will receive, on October 31, 2004, a cash payment equal to his total income tax liability from receiving such restricted shares and from receiving such payment. Additionally, Mr. Price received an option to purchase an additional 500,000 shares of BioMarin's common stock on October 31, 2003, at a per share exercise price of \$7.00 (which option vests monthly over 36 months, from October 31, 2003). BioMarin further agreed to grant Mr. Price additional options to purchase between 50,000 and 200,000 shares of BioMarin common stock on each of

October 31, 2004 and October 31, 2005 (which will each vest monthly over 36 months, commencing on the date of grant).

Either party can terminate the amended agreement on sixty days' notice. In the event that there is a change in control which results in Mr. Price's actual or constructive termination, he is entitled to (i) a severance payment equal to twice the aggregate of his annual base salary and bonus payable in the year in which termination occurs, (ii) forgiveness of all outstanding principal and interest on the interest-deferred loan, (iii) acceleration of all his unvested stock options and the full unvested portion of his 39,285 share restricted stock grant (and a registration right with respect to such shares) and (iv) an additional payment equal to his income tax liability applicable to the severance package.

If Mr. Price is terminated other than for cause or he resigns with good reason, he is entitled to (i) a severance payment equal to his applicable annual base salary and bonus for the year in which termination occurs (plus an additional cash payment equal to the income tax liability associated with forgiveness of the loan and such additional payment), (ii) forgiveness of all outstanding principal and interest on the interest-deferred loan, (iii) if such termination occurs on or after October 31, 2003, acceleration of the full unvested portion of his 39,285 share restricted stock grant (and a registration right with respect to such shares) and (iv) the extension of the exercise period for all his vested stock options to one year following termination. Additionally, if he is terminated other than for cause between October 31, 2003 and October 30, 2005, he is entitled to an additional cash payment equal to the income tax liability associated with forgiveness of the loan and such additional payment.

If Mr. Price becomes disabled or dies between October 31, 2003 and October 30, 2006, he (or his estate) is entitled to (i) acceleration of the full unvested portion of his 39,285 share restricted stock grant, (ii) a payment equal to the aggregate base salary that would have been paid to him if he had continued to be employed through October 30, 2006, and (iii) the pro rata portion of his bonus earned for the year in which he became disabled or died.

The amended agreement has a term that expires on October 30, 2006 and will automatically renew for an additional three-year period unless either Mr. Price or BioMarin give the other notice of his/its intent not to renew the agreement. If BioMarin decides not to renew the agreement, BioMarin must pay Mr. Price an amount such that BioMarin's net payment after deduction of all income taxes at the highest marginal rates applicable to Mr. Price will equal the sum of (i) the base salary BioMarin paid him for the year ending October 30, 2006 and (ii) a percentage of such base salary equal to the same percentage of the bonus BioMarin paid him for the year ending October 30, 2005 compared to the base salary BioMarin paid him for the year ending October 30, 2005. Additionally, any vested options will be exercisable for one year from the termination of the agreement and all unvested options will remain unvested and unexercisable.

In December 1998, the Board approved a form of indemnification agreement to be entered into between BioMarin and each of its executive officers and Directors. This indemnification agreement requires BioMarin, 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)

BioMarin 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. BioMarin has adopted a policy that, where reasonably practicable, it will seek to qualify the variable compensation paid to its executive officers for an exemption from the deductibility limitations of Section 162(m).

Equity Compensation Plans

The following table provides certain information with respect to all of BioMarin's equity compensation plans in effect as of December 31, 2003.

Equity Compensation Plan Information

Plan Category	Number of Securities to be Issued Upon Exercise of Outstanding Options, Warrants and Rights(1)	Weighted Average Exercise Price of Outstanding Options, Warrants and Rights	Number of Securities Remaining Available for Future Issuance Under Equity Compensation Plans(2)
Equity compensation plans approved by stockholders	9,681,706	\$ 10.37	1,878,001
Equity compensation plans not approved by stockholders			
Total	9,681,706	\$ 10.37	1,878,001

- (1) Does not include any shares of BioMarin common stock issuable under its 1998 Employee Stock Purchase Plan. BioMarin issues shares under this plan once every six months based on employee elections in the preceding six months. Pursuant to the terms of this plan, the number of shares to be issued and the price per share is not determined until immediately before the date of issuance.
- (2) Includes options and shares of common stock issuable pursuant to BioMarin's 1997 Stock Plan, as amended, 1998 Director Option Plan and 1998 Employee Stock Purchase Plan. Pursuant to the terms of these plans, the number of shares of common stock available for future issuance under these plans increases on the first day of each fiscal year of BioMarin or, January 1 of each year. On the first day of each fiscal year of BioMarin, the common stock available for future issuance under the 1997 Stock Plan increases by the lesser of (i) 4% of the then outstanding capital stock of BioMarin, (ii) 2,000,000 shares, or (iii) a lower amount set by the Board. On the first day of each fiscal year of BioMarin, 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) 200,000 shares, (ii) 0.5% of the then outstanding capital stock of BioMarin, or (iii) a lower amount set by the Board.

Certain Relationships And Related Transactions

Since January 1, 2003, there has not been, nor is there currently proposed, any transaction or series of similar transactions to which the Company was or is to be a party in which the amount involved exceeds \$60,000 and in which any Director, executive officer, holder of more than 5% of the common stock of the Company or any member of the immediate family of any of the foregoing persons had or will have a direct or indirect material interest other than (i) compensation agreements and other arrangements, which are described elsewhere in this Proxy, 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 BioMarin. BioMarin 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. BioMarin paid REI approximately \$1.1 million and \$0.8 million in 2002 and 2003, respectively, primarily for research. BioMarin's joint venture with Genzyme is subject to a second agreement with REI that requires the joint venture to pay REI a royalty on sales of products covered by the license agreement through November 2019, of which Dr. Kakkis is entitled to certain portions, based on the sales level per the terms of the agreement. The license agreement was effective before Dr. Kakkis was a BioMarin officer. Pursuant to these agreements, Dr. Kakkis was entitled to approximately \$172,000 during 2003.

Indebtedness Of Directors And Executive Officers

Other than as described below, no Director or executive officer of the Company or associate of any Director or executive officer, is or at any time since January 1, 2003, the beginning of the most recently completed fiscal year, has been indebted to the Company.

Pursuant to the Stock Purchase Agreements under which the Company sold stock to founding officers of the Company, the Company has loaned \$400,000 to Dr. Starr to purchase common stock of the Company. The loans are evidenced by interest bearing promissory notes due on demand and are fully recourse. Dr. Starr repaid the loan in full in September 2003.

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 matures on October 31, 2006 and bears interest at the Federal mid-term rate (3.55% as of December 31, 2003). The balance of the note plus accrued interest at December 31, 2003 was \$964,112.

In March 2002, the Company entered into an employment agreement with Mr. Landau that entitles 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 is not completed. In each of June 2002 and June 2003, the Company loaned Mr. Landau \$36,000 which bear interest at 6.0% and 3.5% per annum, respectively, and are due upon termination of his employment with the Company. The balance of the notes plus accrued interest at December 31, 2003 was \$75,928. In the event the Company terminates Mr. Landau other than for cause, the Company has agreed to forgive these loans and to pay Mr. Landau a cash payment equal to the tax liability associated with the forgiveness of the loans and such cash payment.

The following table sets forth certain information related to any indebtedness of Directors or executive officers of the Company owed to the Company.

<u>Name of Borrower</u>	<u>Largest Amount of Outstanding Indebtedness (1)</u>	<u>Outstanding Indebtedness as of December 31, 2003</u>	<u>Maturity Date</u>	<u>Interest Rate</u>	<u>Security for Indebtedness</u>
Christopher M. Starr, Ph.D.	\$ 479,478	\$	(2)	6%	342,956 shares of BioMarin common stock
Fredric D. Price	\$ 964,112	\$ 964,112	October 31, 2006	3.55%(3)	Property purchased with proceeds of loan
Jeffrey I. Landau	\$ 75,928	\$ 75,928	(4)	(5)	None

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- (1) Including accrued interest as of December 31, 2003.
- (2) Under the terms of the loan, the loan is due upon Dr. Starr's termination of involvement with BioMarin.
- (3) Rate as of December 31, 2003.
- (4) Under the terms of the loans, the loans are due upon Mr. Landau's termination of employment with BioMarin.
- (5) The note issued in June 2002 bears interest at 6%; the note issued in June 2003 bears interest at 3.5%.

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 July 31, 1999 and reinvestment of dividends during the period. The period covered by the graph includes that portion of the fiscal year ended December 31, 1999 during which BioMarin was publicly traded.

	<u>BioMarin</u>	<u>Nasdaq Stock Market (U.S.)</u>	<u>Nasdaq Pharmaceutical Index</u>	<u>Nasdaq Biotech Index</u>
7/31/1999	100	100	100	100
12/31/1999	90	154	150	154
12/31/2000	75	93	187	189
12/31/2001	103	74	160	159
12/31/2002	54	51	103	87
12/31/2003	60	76	151	126

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 2003, the Compensation Committee was composed of three outside, independent 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 1998 Employee Stock Purchase Plan.

Considerations for 2003 Compensation

Increases in base salary for 2003 were made effective January 1, 2003, primarily based on the progress and achievements of BioMarin during 2002 and competitive conditions in the job marketplace for biotechnology expertise in the San Francisco Bay Area marketplace.

The Compensation Committee took particular note of 2002 achievements including the acquisitions of Synapse Technologies and Glyko Biomedical, the filing of the Biologics License Application in the United States and the Marketing Authorization Application in the European Union for Aldurazyme® and the initiation of the Phase 2 trial of Aryplase. Based on the Compensation Committee's judgment as to the value of these events and other less visible internal developments, the Compensation Committee awarded 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 4.8%, 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 2002 and maintained a competitive balance with biotechnology companies of similar size and state of development in the region.

With respect to officer salary, the Compensation Committee determined that, based on the factors described above, it was appropriate and elected to increase salaries by an average of 6.6%.

In February 2003, the Company paid the following performance bonuses to the Named Executive Officers (other than Mr. Price) for their service in 2002: Dr. Starr, \$60,000; Dr. Kakkis, \$60,000; Mr. Landau, \$42,500; and Dr. Swiedler, \$42,500. These bonuses were in recognition of the Company's achievements in 2002. Pursuant to Dr. Kakkis' employment agreement, Dr. Kakkis received a \$50,000 bonus in April 2003 upon the marketing approval of Aldurazyme.

In February 2004, the Company paid the following performance bonuses to the Named Executive Officers (other than Mr. Price) for their service in 2003: 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, 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 Aryplase, the raising of \$94 million in equity financings and the closing of a \$125 million convertible debt financing.

Chief Executive Officer Compensation

Effective October 31, 2000, Mr. Price was elected by the Board to the position of Chief Executive Officer and Chairman of the Board. At that time, the Compensation Committee negotiated a three-year employment agreement, which included a competitive compensation package appropriate to Mr. Price's demonstrated leadership capabilities and achievements. Mr. Price's base salary for 2003 was set by this employment agreement. Pursuant to the terms of his employment contract, Mr. Price was also entitled to a bonus of at least \$125,000 and an additional stock option grant to purchase 125,000 shares for his service in 2002. Based on the foregoing and the factors described below, in January 2003, the Company granted Mr. Price an option for the purchase of 125,000 shares of BioMarin common stock and a bonus of \$225,000.

The Compensation Committee negotiated an amended and restated employment agreement with Mr. Price, which was executed in March 2003 and is described in the Proxy. The Compensation Committee determined that the terms of the amended and restated employment agreement were fair and in the best interest of the Company due to the significant contributions Mr. Price has made to the Company in the last 30 months.

The terms of Mr. Price's compensation for 2003 were primarily determined prior to his service with BioMarin. However, the Compensation Committee believes that, during 2003, Mr. Price has amply demonstrated that his skills merit his remuneration. In particular, the Compensation Committee notes 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 Aryplase, 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. 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 March 15, 2004 by the members of the Compensation Committee of the Board of Directors:

Erich Sager, Chairman

Elaine Heron, Ph.D.

Vijay B. Samant

John Urquhart, M.D.

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 controls.

In this context, the Audit Committee has met and held discussions with management and the independent auditors. 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 financials statements with management and the independent auditors. The Audit Committee discussed with the independent auditors matters required to be discussed by Statement on Auditing Standards No. 61 (Communication With Audit Committees).

In addition, the Audit Committee has discussed with the independent auditors, the auditors' independence from the Company and its management, including the matters in the written disclosures required by the Independence Standards Board Standard No. 1 (Independent Discussions With Audit Committees).

The Audit Committee discussed with management and the independent auditors the overall scope and plans for the 2003 annual audit. The Audit Committee met with management and the independent auditors, with and without management present, to discuss the results of their examination, the evaluations of the Company's internal 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, 2003, for filing with the Securities and Exchange Commission and the Swiss SWX Exchange. The Audit Committee and the Board also have recommended the selection of KPMG LLP as the Company's independent auditors for the year ending December 31, 2004.

Respectfully submitted on March 15, 2004 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 Securities Exchange Act of 1934, as amended (the Exchange Act), requires the Company's Directors and executive officers and persons who own more than that 10% of a registered class of the Company's equity securities to file reports of ownership and reports of changes in the ownership with the SEC. Executive officers, Directors and greater than 10% stockholders are required by the SEC 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 us or written representation that no other reports were required, during the fiscal year ended December 31, 2003, all executive officers, Directors, and greater than 10% stockholders complied with all Section 16(a) filing requirements. On May 22, 2003 Mr. Sager filed a late Form 4 reporting the receipt of a stock option grant for the purchase of 20,000 shares on March 21, 2002.

OTHER MATTERS

Except as otherwise indicated, information contained herein is given as of March 15, 2004. The Company's management knows of no matters to come before the Annual Meeting other than the matters referred to in the Notice of Annual Meeting of Stockholders. The Company is not soliciting proxies for any matter that is not now known to the Company's management that should come properly before the Annual Meeting.

The matters to be considered at the Annual Meeting are of great importance to the stockholders of BioMarin. Accordingly, you are urged to read and carefully consider the information presented in this Proxy, and to complete, date, sign and promptly return the enclosed proxy card in the enclosed postage paid envelope.

CONTACT FOR QUESTIONS AND ASSISTANCE IN VOTING

Any stockholder who has a question about the Proxy or how to vote or revoke a proxy, or who wishes to obtain additional copies of this Proxy, should contact BioMarin's proxy solicitor at the address or telephone number listed below:

Morrow & Co., Inc.

445 Park Avenue 5th Floor

New York, NY 10022

1-800-607-0088

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

APPROVAL

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

DATED at Novato, California this 29th day of March, 2004

Louis Drapeau

Vice President, Finance, Chief Financial Officer and Secretary

BioMarin Pharmaceutical

Corporate Governance and Nominating Committee Charter

Purposes of the Committee

The purposes of the Corporate Governance and Nominating Committee (*Committee*) are to:

Oversee the composition of the Board to ensure that qualified individuals meeting the criteria of Nasdaq and regulations of the Securities and Exchange Commission (*SEC*) serve as members of the Board and its committees; and

Oversee the development and implementation of corporate governance principles, policies, codes of conduct and codes of ethics relating to the operation of the Board and its committees and the Company as a whole.

Membership of the Committee

Appointment and Removal

Members of the Committee shall be appointed by the Board and shall serve until a successor is duly elected or the member is removed or resigns.

A member of the Committee shall be automatically removed if (1) the member is no longer a Director of the Company, (2) is determined by the Board to no longer be *independent* as that term is defined by Nasdaq and, if applicable, the Sarbanes-Oxley Act, or (3) is ineligible because of other rules or requirements.

Committee members may otherwise be removed or replaced by vote of the Board.

Number and Qualifications

The Committee will be comprised of at least three directors. The members must be *independent* as that term is defined by Nasdaq and, if applicable, the Sarbanes-Oxley Act.

Committee Structure and Operations

The Board, taking into account the views of the Chairman of the Board, shall designate one member of the Committee as its chairperson.

The Committee shall meet when deemed necessary or desirable by the Committee or its chairperson, generally at least two times per year. The Committee may invite such members of management and other persons to its meetings as it may deem desirable or appropriate.

The Committee shall report regularly to the Board summarizing the Committee's actions and any significant issues considered by the Committee.

The Committee members shall annually submit to a performance evaluation of the Committee by the Board.

Committee Duties and Responsibilities

Identification, Nomination and Termination of Directors

In discharging its general purposes, the Committee shall have the following duties:

To identify individuals qualified to serve on the Board as vacancies arise and to seek out nominees to enhance the diversity and independence of the Board;

To recommend to the full Board for selection the Director nominees to be voted upon either by the Board (for service until the next annual meeting of stockholders) or at the next annual meeting of the stockholders of the Company (for service until the subsequent annual meeting of stockholders);

To recommend to the full Board the termination of the directorship of Directors in accordance with the Company's corporate governance principles or for other appropriate reasons;

To perform an annual evaluation of the Board, each committee of the Board and each member of the Board, based on procedures and guidelines established by the Committee;

To make recommendations to the full Board concerning the appropriate size and needs of the Board, including, without limitation, regarding committees of the Board to be maintained or created;

To annually review the compensation of members of the Board; and

To recommend to the Board appointees to committees of the Board and to recommend removal of committee members to the Board based on such criteria as the Committee shall from time to time establish.

Corporate Governance Principles and Code of Ethics

The Committee shall develop and recommend to the full Board a set of corporate governance principles applicable to the Board and the Company and, if deemed necessary by the Board, propose from time to time any amendments to such principles.

The Committee shall propose for consideration by the full Board a code of business conduct and ethics and, if deemed necessary by the Board, propose from time to time any amendments to such code.

The Committee shall ensure that, as part of each regularly scheduled meeting of the Board of Directors, the outside or non-management directors shall meet in executive session prior to the conclusion of such Board meeting. The Lead Director (as described below) shall chair such executive sessions. At the conclusion of the executive session, the management directors and other members of management, as the Committee deems appropriate, will be invited to join the executive session in order that they can be informed of all relevant issues.

Lead Director

The Committee shall nominate a Lead Director (the Lead) for appointment by the outside members of the full Board. The Lead will be an outside Director of the Board. The Lead will be responsible for the following:

Calling and conducting meetings of the outside Directors and for coordinating the activities of the outside Directors;

Presiding at all meetings of the Board at which the Chairman of the Board is not present, including executive sessions of the outside Directors;

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Serving as liaison between the Chairman of the Board and the outside Directors;

Approving information sent to the Board;

Approving meeting agendas for the Board;

Approving meeting schedules to assure that there is sufficient time for discussion of all agenda items; and

If requested by major stockholders, ensuring that the Lead is available for consultation and direct communication.

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Compliance

The Committee shall have the following additional duties:

To oversee and evaluate compliance by the Board and management of the Company with the Company's corporate governance principles and ethics standards and its code of conduct;

To appoint a Chief Compliance Officer with respect to such corporate governance principles and ethics standards and code of conduct, who will normally be the Chief Financial Officer of the Company; and

The Committee shall meet at least annually with management of the Company to discuss management compliance with the Company's corporate governance principles.

Training

The Committee shall coordinate the training of Directors as required by the SEC and the listing requirements of Nasdaq, as amended from time to time.

Other Duties:

The Committee has the following additional duties:

To recommend guidelines to the Board for corporate succession planning as it relates to the Company's Chief Executive Officer; and

To perform any other activities required by applicable law, rules or regulations, including the rules of the SEC and any stock exchange or market on which the Company's securities may be listed from time to time, and perform such other activities that are consistent with this Charter, the Company's Bylaws and governing instruments, as the Committee or the Board deem necessary or appropriate.

Committee Reports

The Committee shall produce the following reports and provide them to the Board:

An annual performance evaluation of the Committee, which evaluation shall compare the performance of the Committee with the requirements of this Charter. The performance evaluation shall also include a review of the adequacy of this Charter and shall recommend to the Board any revisions the Committee deems necessary or desirable, although the Board shall have the sole authority to amend this Charter. The performance evaluation shall be conducted in such manner as the Committee deems appropriate.

Delegation to Subcommittee

The Committee may, in its discretion, delegate all or a portion of its duties and responsibilities to a subcommittee of the Committee or to the Chairman of the Committee, whether or not such delegation is specifically contemplated under any plan or program.

Resources and Authority of the Committee

The Committee shall have the resources and authority appropriate to discharge its duties and responsibilities, including the authority to select, retain, terminate, and approve the fees and other retention terms of special counsel or other experts or consultants, as it deems appropriate, without seeking approval of the Board or management.

The Committee is authorized to have full access to all Company personnel, records and other informational sources.

The Committee is empowered to investigate any matter brought to its attention and to retain counsel, auditors or other experts as required.

FORM OF PROXY

BIOMARIN PHARMACEUTICAL INC.

ANNUAL MEETING OF STOCKHOLDERS

May 5, 2004

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 March 29, 2004, and hereby appoints Fredric D. Price and Louis Drapeau, or either 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 May 5, 2004 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 below.

1. Election of Directors. Nominees: Fredric D. Price, Franz L. Cristiani, Elaine J. Heron, Pierre Lapalme, Erich Sager, John Urquhart and Gwynn R. Williams.

“ FOR “ WITHHOLD FOR ALL

INSTRUCTION: To withhold authority to vote for any individual nominee, 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 auditors for the year ending December 31, 2004.

“ FOR “ AGAINST “ ABSTAIN

(PLEASE DATE AND SIGN ON REVERSE SIDE)

BioMarin's Board of Directors recommends a vote FOR each of Proposals 1 and 2. If not otherwise directed, this proxy will be voted FOR Proposals 1 and 2.

Dated this ____ day of _____, 2004

_____ Name of Stockholder

_____ Signature(s) of Stockholder

This proxy should be marked, dated and signed by the stockholders(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.