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BIOMARIN PHARMACEUTICAL INC  
Form DEFA14A  
April 18, 2001

SCHEDULE 14A INFORMATION

PROXY STATEMENT PURSUANT TO SECTION 14(a) OF THE  
SECURITIES EXCHANGE ACT OF 1934

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

Check the appropriate box:  
 Preliminary Proxy Statement

Confidential, for Use of the Commission Only (as permitted by Rule 14a  
6(e)(2))  Definitive Proxy Statement  Definitive Additional Materials   
Soliciting Material Pursuant to Section 240.14a-11(c) or Section 240.14a 12

BioMarin Pharmaceutical Inc.

-----  
(Name of Registrant as Specified In Its Charter)

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

Payment of Filing Fee (Check the appropriate box):

No fee required.  
 Fee computed on table below per Exchange Act Rules 14a-6(i)(1) and 0-11.

(1) Title of each class of securities to which transaction applies:

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

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(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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(4) Proposed maximum aggregate value of transaction:

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(5) Total fee paid:

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-

Fee paid previously with preliminary materials.

Check box if any part of the fee is offset as provided by Exchange Act  
Rule 0-11(a)(2) and identify the filing for which the offsetting fee  
was paid previously. Identify the previous filing by registration  
statement number, or the Form or Schedule and the date of its filing.

(1) Amount Previously Paid:

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

(3) Filing Party:

(4) Date Filed:

BioMarin Pharmaceutical Inc.  
371 Bel Marin Keys Boulevard, Suite 210  
Novato, CA 94949

NOTICE OF ANNUAL MEETING OF STOCKHOLDERS  
TO BE HELD ON MAY 24, 2001

TO THE STOCKHOLDERS:

NOTICE IS HEREBY GIVEN that the 2001 Annual Meeting of Stockholders (the "Annual Meeting") of BioMarin Pharmaceutical Inc. ("BioMarin" or the "Company") will be held on Thursday, May 24, 2001 at 10:00 a.m., local time, at the Company's facility located at 46 Galli Drive, Novato, California 94949 for the following purposes:

1. To elect five directors of the Company;
2. To ratify the selection by the Board of Directors of Arthur Andersen LLP as the Company's independent auditors for the year ending December 31, 2001;
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 April 3, 2001 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, however, 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:

Raymond W. Anderson

Chief Operating Officer, Chief Financial Officer, Secretary  
and Vice President, Finance and Administration

Novato, California  
April 3, 2001

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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, CA 94949

PROXY STATEMENT FOR  
2001 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 for use at the Annual Meeting of Stockholders of the Company (the "Annual Meeting") to be held on Thursday, May 24, 2001, or at any adjournment of the Annual Meeting, for the purposes set forth herein and in the foregoing Notice of Annual Meeting of Stockholders. The Annual Meeting will be held at the Company's facility located at 46 Galli Drive, Novato, California 94949. The phone number is 415-884-6700. Copies of solicitation material and the Company's Annual Report including the financial statements and financial statement schedules will be furnished to brokerage houses, fiduciaries and custodians to forward to beneficial owners of common stock of the Company held in their names. The Annual Report to Stockholders for the fiscal year ended December 31, 2000, including financial statements, will be mailed to stockholders entitled to vote at the Annual Meeting concurrently with this Proxy Statement and accompanying Proxy on or about April 17, 2001.

## Record Date; Outstanding Securities

The voting securities of the Company entitled to vote at the Annual Meeting consist of shares of common stock. Only stockholders of record at the close of business on April 3, 2001 are entitled to notice of and to vote at the Annual Meeting. On April 3, 2001, there were 37,134,859 shares of common stock, par value \$0.001 per share, issued and outstanding. Each share of common stock is entitled to one vote. As of the date of record, no shares of the Company's Preferred Stock were outstanding.

## 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 filing with the Company (Attention: Raymond W. Anderson, Chief Financial Officer) a written revocation, or a duly executed Proxy bearing a later date, or by appearing at the Annual Meeting and electing to vote in person.

## Voting

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

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### 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 or by facsimile or by e-mail or by telegram.

### 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 thirty 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.

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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. Except in certain specific circumstances, 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 under Delaware law for approval of proposals presented to stockholders. An exception to this procedure relates to the election of directors. The five nominees receiving the highest number of votes "FOR" a director will be elected as directors. This number is called a plurality. In general, Delaware law also provides that a quorum consists of a majority of shares entitled to vote that are present or represented by Proxy at the Annual Meeting.

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 nominees for directors set forth herein; (ii) for the ratification of Arthur Andersen LLP as independent auditors of the Company for the fiscal year ending December 31, 2001; and (iii) at the discretion of the proxy holder, upon such other business as may properly come before the Annual Meeting or any adjournment thereof.

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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 procedures to be followed by the Inspector are consistent with the general statutory requirements in Delaware concerning voting of shares and determination of a quorum.

### Submission of Stockholder Proposals for 2002 Annual Meeting

Stockholders, who intend to submit a proposal for inclusion in the Company's Proxy materials for the 2002 Annual Meeting of Stockholders, must submit the proposal to the Company no later than December 20, 2001. Stockholders who intend to present a proposal at the 2002 Annual Meeting of Stockholders without inclusion of such proposal in the Company's proxy materials for the 2002 Annual Meeting are required to provide notice of such proposal to the Company no later than March 29, 2002. 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.

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### SECURITY OWNERSHIP OF CERTAIN BENEFICIAL OWNERS

The following table sets forth certain information with respect to the beneficial ownership of our common stock as of April 3, 2001 as to (i) each person, or group of affiliated persons, who is known by us to own beneficially more than 5% of our common stock; (ii) each of our directors; (iii) each of our executive officers named in the Summary Compensation Table set forth herein under the caption "Executive Compensation" (we refer to all these officers as the "Named Executive Officers"); (iv) all of our 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 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 ("SEC"), or based upon the actual knowledge of the Company.

| Name of Beneficial Owner<br>-----  | Number of<br>Shares | Number of<br>Shares | Beneficially<br>Owned (1)<br>----- | Subject<br>To Options<br>----- | Pe<br>C<br>--- |
|--|---------------------|---------------------|------------------------------------|--------------------------------|----------------|
| Glyko Biomedical Ltd.<br>Box 25, Commerce Court West<br>199 Bay Street<br>Toronto, Ontario<br>Canada M5L 1A9 |                     |                     | 11,367,617                         | 0                              |                |
| BB BioVentures, L.P. (2)<br>One Cambridge Center, 9th Floor<br>Cambridge, MA 02142                           |                     |                     | 2,725,787                          | 0                              |                |
| Genzyme Corporation<br>One Kendall Square<br>Cambridge, MA 02139   |                     |                     | 2,102,563                          | 0                              |                |
| Fredric D. Price (3)   |                     |                     | 122,836                            | 97,836                         |                |
| Grant W. Denison, Jr. (3)  |                     |                     | 1,346,939                          | 46,322                         |                |
| John C. Klock, M.D. (3)  |                     |                     | 815,513                            | 0                              |                |
| Christopher M. Starr, Ph.D. (3)  |                     |                     | 615,722                            | 253,105                        |                |
| Emil D. Kakkis, M.D., Ph.D. (3)  |                     |                     | 173,986                            | 171,185                        |                |
| Raymond W. Anderson (3)  |                     |                     | 200,237                            | 197,549                        |                |

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| Name of Beneficial Owner<br>-----   | Number of<br>Shares | Number of<br>Shares | Beneficially<br>Owned (1)<br>----- | Subject<br>To Options<br>----- | Pe<br>C<br>--- |
|---|---------------------|---------------------|------------------------------------|--------------------------------|----------------|
| John L. Jost, Ph.D. (3)   |                     |                     | 109,453                            | 107,260                        |                |
| Ansbert S. Gadicke, M.D., Ph.D. (3) (4)                                     |                     |                     | 2,821,412                          | 95,625                         |                |
| Erich Sager (3) (5)   |                     |                     | 11,525,892                         | 79,375                         |                |
| Gwynn R. Williams (3)   |                     |                     | 57,500                             | 57,500                         |                |
| All current executive officers and directors<br>as a group (13 persons) (6) |                     |                     | 18,004,156                         | 1,309,794                      |                |

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

(1) The "Number of Shares Beneficially Owned" column is based on 37,134,859 shares of common stock outstanding at April 3, 2001. Shares of common stock subject to options or warrants that are currently exercisable or exercisable within 60 days of April 3, 2001 (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

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outstanding for the purpose of computing the percentage ownership of any other person. The "Number of Shares Subject to Options" enumerates for each principal stockholder, director and Named Executive Officer and for all officers and directors in the aggregate, the shares of common stock subject to options exercisable within 60 days of April 3, 2001. These shares are included in the calculation of the "Number of Shares Beneficially Owned."

- (2) Includes shares held by MPM Asset Management 1998 Investors L.L.C. and MPM BioVentures Parallel Fund L.P., both of which are affiliated with BB BioVentures L.P.
- (3) The mailing address for such stockholder is c/o BioMarin Pharmaceutical Inc., 371 Bel Marin Keys Blvd., Suite 210, Novato, California 94949
- (4) Includes 2,725,787 shares currently held by BB BioVentures L.P., MPM Asset Management 1998 Investors L.L.C. and MPM BioVenture Parallel Fund L.P., of which Dr. Gadick is an affiliate. Dr. Gadick disclaims beneficial ownership in these shares except to the extent of his pecuniary interest.
- (5) Includes 11,367,617 shares held by Glyko Biomedical Ltd. of which Mr. Sager is an officer. Mr. Sager disclaims beneficial ownership of these shares, except to the extent of his pecuniary interest.
- (6) See footnotes 2, 4 and 5.

### PROPOSAL ONE: ELECTION OF DIRECTORS

The Company has a Board of Directors, currently consisting of five directors,

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which will be elected at the Annual Meeting. The Proxy holders may not vote the Proxies for a greater number of persons than the number of nominees named. Unless otherwise instructed, the Proxy holders will vote the Proxies received by them for the five 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 nominees who shall be designated by the present Board of Directors 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 of Directors' 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 or until that person's successor has been elected.

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

### NOMINEES FOR DIRECTOR

Set forth below is certain information regarding our directors:

| Name                                    | Age | Position with BioMarin                            | Dir |
|---|-----|---|-----|
| Fredric D. Price                        | 55  | Chief Executive Officer and Chairman of the Board | Oc  |
| Grant W. Denison, Jr.                   | 51  | Director  | Oc  |
| Ansbert S. Gadicke, M.D., Ph.D. (1) (2) | 43  | Director  | Dec |
| Erich Sager (1) (2)                     | 43  | Director  | Nov |
| Gwynn R. Williams (1) (2)               | 67  | Director  | Oc  |

(1) Member of the Compensation Committee.

(2) Member of the Audit Committee.

There is no family relationship between any director and any executive officer of the Company.

Fredric D. Price has served as Chairman and Chief Executive Officer since November 2000. From September 1994 until September 2000, Mr. Price was President, Chief Executive Officer and a member of the Board of Directors of AMBI Inc., formerly Applied Microbiology, Inc., (Nasdaq: AMBI), a biotechnology and nutrition company. From July 1991 to September 1994, he served as Vice President, Finance & Administration and Chief Financial Officer of Regeneron Pharmaceuticals, Inc. (Nasdaq: REGN), and prior to that was a strategy consultant for biopharmaceutical CEOs and investment groups. Mr. Price began his career at Pfizer Pharmaceuticals, where he simultaneously held line and staff positions as Vice President, reporting directly to a member of the board of Pfizer Inc. Mr. Price received an M.B.A. from the Wharton School of the University of Pennsylvania and a B.A. from Dartmouth College. He is also a member of the Advisory Board of equity4life AG, a health care investment company based in Zurich, Switzerland.

Grant W. Denison, Jr. has served as a director of the Company since its inception. Mr. Denison served as Chief Executive Officer of the Company from inception until his resignation in October 2000. Mr. Denison also served as Chairman of the Board of the Company from April 1997 until October 2000. From July 1993 to April 1997, Mr. Denison served as President, Consumer Products and Corporate Senior Vice President, Business Development at Searle, a pharmaceutical company. From July 1989 to June 1993, Mr. Denison served as Vice

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President Corporate Planning at Monsanto Company, a diversified life sciences company. From April 1986 to June 1989, Mr. Denison served as Vice President Corporate Planning and President, U.S. Operations at Searle, a pharmaceutical company. From 1985 to 1986, Mr. Denison served as Vice President, International Operations at Squibb Medical Systems, a medical devices company. From 1980 to 1985, Mr. Denison served as Vice President, Planning and Business Development at Pfizer, Inc., a pharmaceutical company. Mr. Denison serves as a director of Natestch Pharmaceutical Company Inc. and Genetronics. Mr. Denison received an A.B. in Mathematical Economics from Colgate University and an M.B.A. from Harvard Graduate School of Business Administration.

Ansbert S. Gadicke, M.D., Ph.D. has served as a director of the Company since December 1997. Since July 1992, Dr. Gadicke has served as the Chairman of the Board and Managing Director of MPM Capital, L.P., an investment company specializing in the healthcare industry. From 1989 to 1992, Dr. Gadicke was a consultant with Boston Consulting Group. Dr. Gadicke currently serves on the boards of DoubleTwist, Inc., Endeavor, LXN Corporation, MediGene AG, Novirio Pharmaceuticals Limited, Omrix Pharmaceuticals, Inc., Pharmasset Ltd., ViaCell, Inc. and Transform, Inc. Dr. Gadicke received a Ph.D. and an M.D. from J.W. Goethe Universitat, Frankfurt, Germany.

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Erich Sager has served as a director of the Company 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 BioNebraska, Inc., Dentalview, Inc., Kimsa Holding, LaMont Asset Management, SA and Sermont Asset Management, SA. 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 the Company since its inception. Mr. Williams founded AstroMed Limited and Astroscan Limited, UK manufacturers of scientific equipment, in March 1984, which entities, in December 1997, merged into Life Science Resources Ltd. Previously, Mr. Williams was a partner of Arthur Andersen & Co., a mathematician with General Motors Research, and a mathematician with British Steel. Mr. Williams was a founder of Glyko Biomedical Ltd. and its predecessor Glyko, Inc. Mr. Williams received a B.S. in Theoretical Physics from the University of Wales.

### 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 and a Compensation Committee whose functions are briefly described below. The Board has not established a Nominating Committee.

The Board of Directors of the Company held a total of seven meetings during the year ended December 31, 2000 and took action by Unanimous Written Consent on 11 occasions. No director participated in fewer than 75% of all such meetings and actions of the Board of Directors and the committees thereof, held during fiscal 2000, if any, upon which such director served.

Audit Committee. The Audit Committee provides oversight of the (a) financial reporting process, the system of internal controls and the audit process of the Company and (b) the Company's independent auditors. The Audit Committee also recommends to the Board of Directors the appointment of the Company's

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independent auditors. On June 15, 2000, the Board adopted a new written Audit Committee Charter (the "Audit Charter"), a copy of which is included with this Proxy. As required by the Audit Charter, Mr. Sager was appointed to the Audit Committee as the third member. As also required by the Audit Charter, each of the members of the Audit Committee is an independent director as defined by Rule 4200(a)(15) of the National Association of Securities Dealers. The members of the Audit Committee are Dr. Gadicke, Mr. Sager, and Mr. Williams. During the fiscal year 2000, the Audit Committee met on two (2) occasions. Additionally, prior to each financial press release, Mr. Williams, as a representative of the Audit Committee, discussed the information included in such release with our independent auditors.

Compensation Committee. The Compensation Committee, which consists of Dr. Gadicke, Mr. Sager and Mr. Williams, sets general compensation policy for the Company and has final approval power over compensation of executive officers. The Compensation Committee also has final approval power over guidelines and criteria for employees' bonuses and administers the Company's 1997 Stock Plan and 1998 Director Option Plan. The Compensation Committee met twice in 2000.  
Compensation Committee Interlocks and Insider Participation

None of the members of the Compensation Committee is currently or has 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 of Directors or compensation committee of any entity that has one or more executive officers serving as a member of the Company's Board of Directors or Compensation Committee.

The Board unanimously recommends voting "For" each of the foregoing nominees for director.

### PROPOSAL TWO: RATIFICATION OF APPOINTMENT OF INDEPENDENT AUDITORS

The Board of Directors has selected Arthur Andersen LLP, independent auditors, to audit the financial statements of the Company for the fiscal year ending December 31, 2001, and recommends that stockholders vote for ratification of such appointment. Although action by stockholders is not required by law, the Board of Directors has determined that it is desirable to request approval of this selection by the stockholders. Notwithstanding the selection, the Board of Directors, in its discretion, may direct the appointment of new independent auditors at any time during the year, if the Board of Directors feels that such a change would be in the best interest of the Company and its stockholders. In

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the event of a negative vote on ratification, the Board of Directors will reconsider its selection.

Arthur Andersen LLP has audited the Company's financial statements annually since inception on March 21, 1997. Representatives of Arthur Andersen LLP are expected to be present at the Annual Meeting with the opportunity to make a statement if they desire to do so and are expected to be available to respond to appropriate questions.

For the year ended December 31, 2000, Arthur Andersen LLP billed the Company the following amounts for the respective professional services:

|  |           |
|--|-----------|
| Audit Fees:  | \$125,000 |
| Financial Information Systems Design and<br>Implementation Fees: | \$ -      |
| All Other Fees:  | \$ 32,550 |

The Audit Committee has considered the nature and amount of these fees and believes that the provision of the services relating to Financial Information Systems and All Other Fees is compatible with maintaining Arthur Andersen LLP's independence.

The Board unanimously recommends voting "For" the ratification of the appointment of Arthur Andersen LLP as independent auditors of the Company for the fiscal year ending December 31, 2001.

## EXECUTIVE OFFICERS

In addition to Fredric D. Price the Company's Chief Executive Officer and Chairman of the Board, whose biography appears above, the following table sets forth as of April 3, 2001 information concerning the Company's other executive officers:

| Name                            | Age | Position with BioMarin  |
|---------------------------------|-----|---|
| Raymond W. Anderson             | 59  | Chief Operating Officer, Chief Financial Officer and Vice President, Finance and Administration |
| Christopher M. Starr, Ph.D.     | 48  | Vice President, Research and Development  |
| John L. Jost, Ph.D.             | 56  | Vice President, Manufacturing   |
| Robert A. Baffi, Ph.D.          | 46  | Vice President, Quality Assurance/Quality Control   |
| Emil D. Kakkis, M.D., Ph.D.     | 40  | Vice President, Business Development  |
| Stuart J. Swiedler, M.D., Ph.D. | 45  | Vice President, Scientific and Clinical Affairs   |
| Brian K. Brandley, Ph.D.        | 44  | Vice President of BioMarin and Managing Director, Glyko, Inc., a wholly-owned subsidiary        |

Raymond W. Anderson has served as Chief Financial Officer and Vice President, Finance and Administration since June 1998 and has served as Chief Operating Officer since April 2000. Mr. Anderson has served as Secretary since August 2000. Mr. Anderson served as the Vice President, Finance and Chief Financial Officer at Fusion Medical Technologies, Inc., a medical technology company developing drug delivery systems, from July 1997 to June 1998. Mr. Anderson served as the Vice President, Finance and Chief Financial Officer at Fidus Medical Technology, Inc., a medical technology company specializing in cardiac arrhythmias, from October 1996 to July 1997. Mr. Anderson served as a director of Recombinant Capital, a consulting firm, from July 1994 to October 1996. Mr. Anderson served as the Vice President, Finance and Chief Financial Officer of Glycomed Incorporated, a biotechnology company, from April 1989 to July 1994. Mr. Anderson was the Chief Financial Officer at Chiron Corporation, a biotechnology company, from 1985 to 1989. Mr. Anderson was a Controller and Director of Financial Planning and Analysis at Syntex Laboratories, a pharmaceutical company, from 1981 to 1985. Mr. Anderson served as a director of Glyko Biomedical Ltd. and its predecessor, Glyko, Inc., a carbohydrate analytical and diagnostic company, from October 1989 to June 2000. Mr. Anderson received a B.S. in Engineering from the United States Military Academy, a M.S. in Administration from George Washington University and a M.B.A. from the Harvard Graduate School of Business Administration.

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Christopher M. Starr, Ph.D. has served as Vice President, Research and Development since the Company's inception. From July 1991 to April 1998, Dr. Starr served as the Vice President, Research and Development for Glyko, Inc. Dr. Starr held the position as National Research Associate at the National Institutes of Health in Bethesda, Maryland from August 1986 to June 1991. Dr. Starr received a B.S. in Biology from Syracuse University and a Ph.D. in Biochemistry and Molecular Biology from the State University of New York, Health Science Center, Syracuse, NY.

John L. Jost, Ph.D. has served as Vice President, Manufacturing since June 1999. Dr. Jost devoted his time from November 1997 to June 1999 to personal affairs. Dr. Jost served in several positions at Genentech, Inc., a biotechnology company, from February 1983 to November 1997. From June 1992 to November 1997, he was Director of Manufacturing Sciences at Genentech. From 1971 to 1983, he served in various scientific positions in process development at The Upjohn Company, a pharmaceutical company, ending as a Senior Research Scientist. Dr. Jost received a B.S. and a Ph.D. in Chemical Engineering from the University of Minnesota.

Emil D. Kakkis, M.D., Ph.D. has served as Vice President of BioMarin since September 1998. From July 1994 to August 1998, Dr. Kakkis was a Physician Specialist at the Department of Pediatrics at Harbor-UCLA Medical Center. From July 1991 to June 1994, Dr. Kakkis completed a Fellowship in Genetics at the UCLA Intercampus Medical Genetics Training Program. Dr. Kakkis received a B.A. in Biology from Pomona College and received a Ph.D. in Biological Chemistry from UCLA. Dr. Kakkis received an M.D. from UCLA.

Stuart J. Swiedler, M.D., Ph.D. has served as Vice President of Scientific and Clinical Affairs since June 1998. From November 1997 to June 1998, Dr. Swiedler was as an independent biotechnology consultant. From February 1993 to November 1997, Dr. Swiedler served, in chronological order, with Glycomed Incorporated, a biotechnology company, as Assistant Vice President, Biology from February 1993 to July 1994, as Assistant Vice President, Research from July 1994 to May 1995, and as Vice President, Research from May 1995 to November 1997. Dr. Swiedler received a B.S. in Biology from the State University of New York at Albany. Dr. Swiedler received an M.D. and a Ph.D. in Biochemistry from the Johns Hopkins University School of Medicine.

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Brian K. Brandley, Ph.D. has served as Vice President of BioMarin since October 1998. Dr. Brandley has served as the Managing Director of Glyko, Inc. since April 1998. He was an Assistant Professor at Rush University from July 1995 to April 1998, and was the Senior Scientist, Head of Cell Biology at Glycomed Incorporated, a biotechnology company, from July 1988 to July 1995. Dr. Brandley received a B.S. and a M.S. in Biology from the University of Miami. Dr. Brandley received a Ph.D. in Biology from the University of Sydney, Australia.

Robert A. Baffi, Ph.D., has served as Vice President of Quality Assurance and Quality Control since May 2000. From 1986 to 2000, Dr. Baffi served in a number of progressively more responsible positions at Genentech, Inc., primarily in the functional area of quality control. Dr. Baffi's most recent role within the quality organization at Genentech was Director of Collaborations where he was responsible for providing coordination and support in all areas of quality assurance and control to over 14 Genentech collaborative partners. He was also responsible for assessments of the quality capabilities of potential collaborative partners. Prior to Genentech, Dr. Baffi worked for Cooper BioMedical as a research scientist and at Becton Dickinson Research Center as a

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post-doctoral fellow. Dr. Baffi received a Ph.D. in Biochemistry, a M.Phil. and a B.S. in Biochemistry from the City University of New York.

### EXECUTIVE COMPENSATION

#### Summary Compensation Table.

We are 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 executive officers, other than the Chief Executive Officer, who were serving as executive officers at the end of fiscal year 2000; and (c) up to two additional individuals for whom such disclosure would have been provided under clause (a) and (b) above but for the fact that the individual was not serving as an executive officer of the Company at the end of fiscal year 2000; 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 Chief Executive Officer; (b) Raymond W. Anderson, Christopher M. Starr, Ph.D., Emil D. Kakkis, M.D., Ph.D. and John L. Jost, Ph.D., the four most highly-compensated executive officers, other than the Chief Executive Officer, who were serving as executive officers at the end of fiscal year 2000 and whose salary and bonus exceeded \$100,000; and (c) Grant W. Denison, Jr. and John C. Klock, M.D. our former Chief Executive Officer and former President, respectively. We refer to all of these officers as the "Named Executive Officers."

The entries under the column "Other Annual Compensation" for Mr. Price, include the value of his restricted stock grant issued upon his employment commencement, a tax gross-up benefit relating to the compensation earned from the restricted stock grant and payments for temporary housing and for Mr. Denison, relate to payments for temporary housing. Each of these amounts paid to Mr. Price is described in more detail in the section captioned "Employment Agreements." The entries under the column "All Other Compensation" in the table represent the premiums paid for life insurance benefits and vested 401(k) matching for each Named Executive Officer.

## SUMMARY COMPENSATION TABLE

| NAME AND PRINCIPAL POSITION  | YEAR                 | ANNUAL COMPENSATION (1)       |                              |                                   | LONG-TERM<br>COMPENSATION                |
|--|----------------------|-------------------------------|------------------------------|-----------------------------------|--|
|  |                      | SALARY (\$)                   | BONUS (\$)                   | OTHER ANNUAL<br>COMPENSATION (\$) | AWARDS                                   |
|  |                      |                               |                              |                                   | SECURITIES<br>UNDERLYING<br>OPTIONS/SARS |
| Fredric D. Price (1)<br>Chairman and Chief Executive<br>Officer  | 2000<br>1999<br>1998 | 99,487<br>--<br>--            | 100,000<br>--<br>--          | 578,424<br>--<br>--               | 523                                      |
| Christopher M. Starr, Ph.D.<br>(2)<br>Vice President, Research and<br>Development  | 2000<br>1999<br>1998 | 190,000<br>150,000<br>139,560 | 75,000<br>150,000<br>87,550  | --<br>--<br>--                    | 75<br>94<br>200                          |
| Emil D. Kakkis, M.D., Ph.D.<br>(2)<br>Vice President, Business<br>Development  | 2000<br>1999<br>1998 | 229,327<br>225,000<br>75,000  | 50,000<br>--<br>50,000       | --<br>--<br>--                    | 50<br>64<br>200                          |
| Raymond W. Anderson (2)<br>Chief Operating Officer,<br>Chief Financial Officer,<br>Secretary and Vice President,<br>Finance and Administration | 2000<br>1999<br>1998 | 222,466<br>189,625<br>90,484  | 25,000<br>100,000<br>--      | --<br>--<br>--                    | 58<br>72<br>200                          |
| John L. Jost, Ph.D. (2)<br>Vice President, Manufacturing   | 2000<br>1999<br>1998 | 200,000<br>106,154<br>--      | 50,000<br>--<br>--           | --<br>--<br>--                    | 17<br>200                                |
| Grant W. Denison, Jr. (3)<br>Former Chief Executive Officer  | 2000<br>1999<br>1998 | 214,615<br>257,143<br>202,500 | 128,572<br>257,143<br>87,550 | --<br>32,805<br>--                | 32<br>154<br>400                         |
| John C. Klock, M.D. (4)<br>Former President  | 2000<br>1999<br>1998 | 195,769<br>250,000<br>222,450 | 125,000<br>250,000<br>87,550 | --<br>--<br>--                    | 32<br>121<br>300                         |

(1) Mr. Price was appointed as the Chief Executive Officer effective October

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31, 2000.

- (2) Options were earned for the fiscal year 2000 and granted on December 29, 2000. Long-term options vest 6/48ths at June 30, 2001 and 1/48ths per month thereafter.

Short-term options vest 25% per 2001 calendar quarter for options granted on December 29, 2000. Short-term options earned on June 30, 2000 for Mr. Denison, Dr. Klock and Dr. Starr in the amounts of 17,582, 17,807 and 20,959, respectively, vested 100% on June 30, 2000.

Short-term options include options to purchase 15,000 shares granted to each of Mr. Denison and Dr. Klock under the 1998 Director Option Plan upon termination of their respective employment with the Company.

- (3) Mr. Denison resigned as Chief Executive Officer and Chairman on October 31, 2000, but remains a member of the Board.
- (4) Dr. Klock resigned as President on July 31, 2000 and resigned as Director on November 1, 2000.

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Stock Option Grants Table. The following table sets forth certain information for each grant of options to purchase the Company's common stock during fiscal 2000 to each of the Named Executive Officers. Except as otherwise noted, all these options were granted under the 1997 Stock Plan and have a term of ten years subject to early termination in the event the officer's services to the Company cease.

Fiscal 2000 Stock Option Grants

| Name                  | Number of Securities Underlying Options Granted | Percent of Total Options Granted to Employees in 2000 (1) | Exercise Price Per Share (\$) (2) | Option Term Expiration Date | Potential Assumed Price |
|-----------------------|---|---|-----------------------------------|-----------------------------|-------------------------|
| Fredric D. Price      | 500,000   | 18%   | 12.50                             | 10/30/10                    |                         |
|                       | 23,288  |   | 9.50                              | 01/01/11                    |                         |
| Grant W. Denison, Jr. | 17,582  | 1%  | 17.00                             | 06/30/10                    |                         |

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|                             |            |    |       |          |
|-----------------------------|------------|----|-------|----------|
|                             | 15,000 (5) |    | 13.00 | 10/31/10 |
| John C. Klock, M.D. (4)     | 17,807     | 1% | 17.00 | 01/30/01 |
|                             | 15,000 (5) |    | 19.00 | 01/30/01 |
| Christopher M. Starr, Ph.D. | 20,959     | 3% | 17.00 | 06/30/10 |
|                             | 54,235     |    | 9.50  | 01/01/11 |
| Emil D. Kakkis, M.D., Ph.D. | 50,000     | 2% | 9.50  | 01/01/11 |
| Raymond W. Anderson         | 58,469     | 2% | 9.50  | 01/01/11 |
| John L. Jost, Ph.D.         | 17,365     | 1% | 9.50  | 01/01/11 |

- (1) Based on an aggregate of 2,923,835 shares subject to options granted by the Company during fiscal year 2000 to employees, consultants and the Named Executive Officers.
- (2) Options were granted at an exercise price equal to the fair market value of the common stock, as determined by the Board of Directors, on the date of grant or by the closing price on the Nasdaq National Market.
- (3) The 5% and 10% assumed annual rates of compounded stock price appreciation are mandated by rules of the Securities and Exchange Commission. The Company cannot assure any 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 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 the Company.
- (4) Dr. Klock's options terminated on January 30, 2001 due to his resignation as an officer and director of the Company. These options expired unexercised.
- (5) These options were granted under the 1998 Director Option Plan and have a term of ten years subject to early termination in the event the Director's services to the Company cease.

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Fiscal Year-End 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 at December 31, 2000.

| Name                        | Fiscal Year-End Option Value Table |               | Value of Unexercised In-the-Money Options at Year-End (1) |               |
|-----------------------------|------------------------------------|---------------|---|---------------|
|                             | Exercisable                        | Unexercisable | Exercisable   | Unexercisable |
| Fredric D. Price            | 27,778                             | 495,510       | \$ -  |               |
| Grant W. Denison, Jr.       | 89,518                             | 185,833       | \$ 293,278  |               |
| John C. Klock, M.D.         | 42,803                             | 81,563        | \$ 8,398  |               |
| Christopher M. Starr, Ph.D. | 209,731                            | 139,652       | \$ 853,956  |               |
| Emil D. Kakkis, M.D., Ph.D. | 144,710                            | 169,792       | \$ 695,904  |               |
| Raymond W. Anderson         | 175,032                            | 149,095       | \$ 859,194  |               |
| John L. Jost, Ph.D.         | 82,633                             | 156,427       | \$ -  |               |

(1) Based on closing price on December 31, 2000 of \$9.6875 per share less exercise price per share.

Employment Agreements

We are party to employment agreements with each executive officer on the terms enumerated on the chart below. Each of these employment agreements is terminable without cause by the Company upon six months prior written notice to the officer, or by the officer upon three months prior written notice to the Company. The Company is obligated to pay the officer's salary and benefits until this termination. Except in the case of Mr. Price, in the event that the officer is involuntarily terminated within one year of a change of control of the Company, he will receive (i) a severance payment equal to six months of his annual salary; (ii) a bonus equal to 50% of the annual bonus that he would otherwise be entitled to, and (iii) immediate vesting of 50% of the unvested portion of his outstanding options to purchase Company capital stock.

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| Name of Executive Officer   | 2000<br>Annual<br>Salary Rate (1) | Annual Bonus  | Initial Grant of<br>to Purchase<br>Equity Securities   |
|-----------------------------|-----------------------------------|---|--|
| Fredric D. Price            | \$400,000                         | Annual bonus, payable in cash based on performance. | Option to purchase up to 500,000 shares of our common stock at a price of \$12.50 per share.   |
| Christopher M. Starr, Ph.D. | \$230,000                         | Annual bonus, payable in cash or stock.             | 400,000 shares of our common stock at a price of \$1.00 per share.   |
| Brian K. Brandley, Ph.D.    | \$139,500                         | Annual bonus, payable in cash or stock.             | Option to purchase up to 150,000 shares of Glyko Biomedical Ltd.' common stock. Now exercisable for 65,415 shares of our common stock at a purchase price of \$5.27 per share. |
| Raymond W. Anderson         | \$230,000                         | Annual bonus, payable in cash or stock.             | Option to purchase up to 200,000 shares of our common stock at a purchase price of \$5.27 per share.   |

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|   |           |  | price of \$4.00 per sha  |
|---|-----------|--|--|
| Emil D. Kakkis, M.D., Ph.D.                   | \$225,000 | Eligible to receive a cash bonus based on achievement of milestones. | Option to purchase up 200,000 shares of our common stock at a purc price of \$4.00 per sha |
| John L. Jost, Ph.D.                           | \$200,000 | Annual bonus, payable in cash or stock.                              | Option to purchase up 200,000 shares of our common stock at a purc price of \$13.00 per sh |
| Stuart Swiedler, M.D., Ph.D.                  | \$210,000 | Annual bonus, payable in cash or stock.                              | Option to purchase up 150,000 shares of our common stock at a purc price of \$4.00 per sha |
| Robert A. Baffi, Ph.D.<br>(hired in May 2000) | \$200,000 | Annual bonus, payable in cash or stock.                              | Option to purchase up 200,000 shares of our common stock at a purc price of \$22.00 per sh |

(1) 2000 Annual Salary Rate reflected in the above table reflects 2000 annual salary rate as of December 31, 2000. Actual salaries paid in 2000, reflected in the table entitled "SUMMARY COMPENSATION TABLE" include mid-year salary adjustments.

Effective October 31, 2000, in connection with Mr. Price's employment, we entered into an employment agreement with him which provides for an initial

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payment to Mr. Price of \$357,624, an annual base salary of \$400,000 in the first year, \$450,000 in the second year, and \$500,000 in the third year, and a bonus of \$200,000 in the first year and a performance bonus of between 25% and 100% of his respective annual base salary for each of the second and third years. In addition, we granted Mr. Price 25,000 restricted shares of our common stock (which vest in 3 equal annual installments commencing on January 1, 2001), we granted Mr. Price an option to purchase 500,000 shares of our common stock, at a purchase price of \$12.50 per share (which vests monthly over 36 months, commencing October 30, 2000) and we further agreed to grant Mr. Price additional options on each anniversary of the agreement, in an amount to be determined by the Board. We also agreed to reimburse Mr. Price for all expenses incurred in relocating to the area with a tax gross-up adjustment and to extend him a \$1,500,000 interest-deferred loan for the purchase of a house.

The agreement has a three-year term, which will automatically renew for an additional three-year period unless either Mr. Price or we give the other notice of our intent not to renew the agreement. If we decide not to renew the agreement, we must pay Mr. Price an amount such that our net payment after deduction of all payroll taxes and all income taxes at the highest marginal rates applicable to him will equal the base salary and bonus we paid him in the third year of the agreement. Additionally, the expiration for any vested options will be one year from the termination of the agreement and all unvested options will remain unvested and unexercisable.

Either party can terminate the agreement on sixty days' notice. However, in the event there is a change in control which results in Mr. Price's actual or constructive termination, he is entitled to a severance payment equal to twice the aggregate of his annual base salary and bonus payable in the year in which termination occurs, forgiveness of all outstanding principal and interest on the interest-deferred loan, acceleration of the full unvested portion of his 25,000 share restricted stock grant and all stock options and an additional payment equal to Mr. Price's maximum total income tax liability applicable to the total severance package. If Mr. Price is terminated other than for cause, he is entitled to receive a severance payment equal to twice his applicable annual base salary and bonus if he is terminated in the first year of the agreement and equal to his applicable annual base salary and bonus if he is terminated in a subsequent year, forgiveness of all outstanding principal and interest on the interest deferred-loan and acceleration of the full unvested portion of his 25,000 share restricted stock grant and all stock options. Additionally, if he is terminated other than for cause prior to the second anniversary of the agreement, he is entitled to an additional payment equal the maximum income tax liability associated with forgiveness of the loan and such additional payment.

We provided a three-year loan to Dr. Starr for the purchase of his shares referenced in column four of the table above and to each of Mr. Denison and Dr. Klock for the purchase of 1,300,000 and 800,000 shares of the Company's common stock all of which were purchased pursuant to the Founders' Stock Purchase Agreement described below. Each loan bears interest at a rate of 6%. For each of them, respectively, if his employment is terminated by us for any reason, he has the right to sell any or all of these shares of common stock to us at a price per share equal to the lesser of the then-current per share market price of the shares or the original per share purchase price of \$1.00. Upon Dr. Klock's resignation as President, we repurchased 33,333 shares of common stock in consideration for reducing his note obligation by \$33,333 and Dr. Klock paid the remainder of his note including interest. The repurchase right was pursuant to provisions contained in the Founders' Stock Purchase Agreement. These repurchase rights have since lapsed with respect to all shares held by Mr. Denison and Dr. Starr.

Dr. Brandley's employment agreement was originally with Glyko, Inc. but was assigned to us in connection with our acquisition of Glyko, Inc.

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Through June 30, 2000, the founder's cash bonus for each of Mr. Denison, Dr. Klock and Dr. Starr was based on the difference between a minimum market capitalization and our quarterly market capitalization. The cash bonus is calculated as follows:

The Board of Directors established a minimum market capitalization of \$20.0 million for the first quarter of 1998. The minimum market capitalization increases by \$1.0 million per quarter until the end of the agreement in the second quarter of 2000. Our quarterly market capitalization is calculated at the end of each calendar quarter by multiplying the number of our common shares outstanding times the average closing price of our common stock for the last ten trading days of the quarter. If our common stock is not publicly traded the quarterly market capitalization is determined by multiplying the shares of our common stock outstanding by the price at which of our common stock was sold in the latest significant investment by an independent third-party investor. For each full \$5.0 million that the quarterly market capitalization exceeds the minimum market capitalization, the founders each receive a cash bonus of \$1,200 in the first calendar quarter and \$1,250 in the second quarter. Each founder's cash bonus is the sum of the two quarterly bonuses for 2000.

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Each founder's total cash bonus may not exceed 100% of base salary in any year. Additional amounts beyond the cash limit that may be earned in a year will be paid in stock options using the Black-Scholes option pricing model to calculate the value of the stock option based on period-end parameters.

There are no adjustments in the founders' annual base salaries that result from increases in market capitalization.

In December 1998, the Board approved a form of indemnification agreement to be entered into between us and each of our officers and directors. This indemnification agreement requires us, among other things, to indemnify officers and directors against liabilities that may arise by reason of their status or performance of their duties as 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 the Company and affiliates of the Company, see "Certain Relationships and Related Transactions".

### Section 162(m)

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

### Director Compensation

Directors do not receive cash compensation for their services as directors of the Company but are reimbursed for their reasonable expenses in attending meetings of the Board and while performing services for the Company. In July

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2000, under the Company's 1997 Stock Plan, Mr. Sager and Dr. Gadick were each issued an option to purchase an additional 50,000 shares of common stock of the Company at an exercise price set at the fair market value on the date of grant (closing price of common stock on the Nasdaq National Market), which was \$20.375, as consideration for services rendered by them to the Company in connection with the search for a new senior executive officer. In August 2000, under the Company's 1998 Director Option Plan, the Company issued to Dr. Klock an option to purchase 15,000 shares of common stock with an exercise price set at the fair market value (closing price of common stock on the Nasdaq National Market) on the date of grant, which was \$19.00, as consideration for his ongoing services to the Company as a director. In October 2000, under the 1998 Director Option Plan, the Company issued to Mr. Denison an option to purchase 15,000 shares of common stock with an exercise price set at the fair market value (closing price of common stock on the Nasdaq National Market) on the date of grant, which was \$13.00, as consideration for his ongoing services to the Company as a director. In November 2000, under the 1998 Director Option Plan, the Company issued to Mr. Sager and Mr. Williams options to purchase 15,000 shares of common stock with an exercise price set at the fair market value (closing price of common stock on the Nasdaq National Market) on the date of grant, which was \$9.6875 as consideration for their ongoing services to the Company as directors. In December 2000, under the 1998 Director Option Plan, the Company issued to Dr. Gadick an option to purchase 15,000 shares of common stock with an exercise price set at the fair market value (closing price of common stock on the Nasdaq National Market) on the date of grant, which was \$9.6875, as consideration for his ongoing services to the Company as a director.

### 1998 Director Option Plan

The 1998 Director Option Plan was adopted by the Board in December 1998. It was approved by the stockholders as of January 15, 1999. The plan provides for the grant of nonstatutory stock options to non-employee directors. A total of 200,000 shares of Company common stock have been reserved for issuance under the plan. The plan also provides for an annual increase in this number of shares equal to the lesser of: (1) 0.5% of the Company's outstanding capital stock, (2) 200,000 shares, or (3) a lesser amount determined by the Board.

In fiscal year 2000, options to purchase, in the aggregate, 75,000 shares were issued to directors.

The 1998 Director Option Plan provides that each non-employee director shall automatically be granted an option to purchase 20,000 shares of Company common stock on the date which that person first becomes a non-employee director. This option shall have a term of ten years. The shares subject to this initial option shall vest over one year. Each non-employee director shall thereafter also be automatically granted an option to purchase 15,000 shares of Company common stock on the first anniversary of the date of their respective initial appointments to the Board and each anniversary thereafter, provided that he or she retains the Board seat on his or her anniversary date. The shares subject to this annual option shall vest in full one year from the date of grant and shall have a term of ten years. These options shall 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 Company 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 the Company, each option 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 plan. However, if a non-employee director's status as a director of the Company or the successor corporation, as applicable, is terminated, other than upon a voluntary resignation by the non-employee director, the option shall

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immediately become fully vested and exercisable. If the successor corporation does not agree to assume or substitute the option, each option shall become 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 shall terminate.

Options granted under the plan must 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, but in no event later than the expiration of the option's ten-year term. No option granted under the 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 plan will terminate automatically ten years from the effective date of the plan.

The information contained below under the captions "Performance Graph," "Report of the Compensation Committee on Executive Compensation" and "Report of the Audit Committee" shall not be deemed to be "soliciting material" or to be "filed" with the Securities and Exchange Commission, nor shall such information be incorporated by reference into any future filing under the Securities Act of 1933, as amended, or the Securities Exchange Act of 1934, as amended, except to the extent that the Company specifically incorporates it by reference into such filing.

### PERFORMANCE GRAPH

The following graph compares the Company's 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, 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 the Company was publicly traded.

[OBJECT OMITTED]

REPORT OF THE COMPENSATION COMMITTEE ON EXECUTIVE COMPENSATION

The Compensation Committee is responsible for setting general compensation goals and operational guidelines for Company personnel, for setting all elements of the compensation of the executive officers of the Company, and for approving grants of stock options for the Company. In 2000, the Compensation Committee was composed of the three outside, non-management members of the Board of Directors.

Compensation Goals and Policies

The goal of the Company'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. The Company uses the following principles to help achieve that goal:

- (1.) The Company provides competitive compensation packages incorporating all compensation elements for executives and staff based upon internal Company policies and compensation packages at similarly situated pharmaceutical and biotechnology companies in the San Francisco Bay Area.
- (2.) The Company rewards executives and senior staff for outstanding performance by the individual and by the Company.
- (3.) The Company 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 our Employee Stock Purchase Plan.

Considerations for 2000 Compensation

Increases in base salary for 2000 were made effective January 2000 primarily based on the progress and achievements of the Company during 1999 and competitive conditions in the job marketplace for biotechnology expertise in the San Francisco Bay Area marketplace.

The Compensation Committee took particular note of 1999 achievements including a successful initial public offering, continued progress on the Company's lead programs: Aldurazyme for MPS-I, rhASB (then known as BM 102 or N-acetylgalactosamine-4 sulfatase) for MPS-VI, and Vibriolysin (then known as BM 202) for severe burn debridement. However, the Compensation Committee also noted

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that, in November 1999, the FDA had requested a confirmatory Phase III clinical trial for Aldurazyme which had the effect of significantly delaying the approval of that product and delaying the potential growth of stockholder value.

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 the Company. Except for the Company founders and others who were covered under separate employment agreements, the Committee also granted short-term incentive stock options as a reward for the Company's progress in comparison to plan in 1999. Grants under both programs were pro-rated for the portion of the year that the employee was in the service of the Company. Salary compensation for the staff below the rank of officer was generally increased by an average of 5%, which approximated the reported average salary increase in the biotechnology industry in the San Francisco Bay Area and which was also pro-rated for time in service during the year. Benefits were improved primarily in the Company matching payments under the 401(k) plan, a benefit feature that was weak in comparison to competitive peer companies. The Compensation Committee deemed that these compensation actions were appropriate for the progress made by the Company in 1999 and maintained a competitive balance with biotechnology companies of similar size and state of development in the region.

With respect to officer compensation, and based on an overall analysis of performance factors and in particular the delays resulting from an Aldurazyme Phase III clinical trial, the Compensation Committee determined that, in January, officers would not receive salary increases and would receive only 50% of normal long term compensation stock option grants (compared to the program algorithms used for all staff in 1999 and used for lower ranks in 2000). During the course of 2000, one officer was given a raise as a result of a promotion and one officer received an increase in base salary to a competitive level as a result of the expiration of his founders' employment agreement.

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### Chief Executive Officer Compensation

In the first half of 2000, the compensation of the three employee founders, including the then Chief Executive Officer, Mr. Grant W. Denison Jr., for 2000 was set by compensation and bonus formulas in their employment agreements, as amended, which were effective until June 30, 2000. The primary determinant of the founders' bonus for the first six months of 2000 was based on the difference between a minimum target market capitalization and the Company's market capitalization as measured quarterly.

Based on the application of the bonus formula for the first six months of 2000, the founders reached the limit for cash bonus, which was 100% of base salary. Additional bonus amounts beyond the cash limit were paid in stock options using

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the Black-Scholes option- pricing model based on the period-end market price and other market parameters.

As Chairman and Chief Executive Officer, Mr. Denison continued at his 1999 base salary from January 1, 2000 until October 31, 2000 when he resigned from both positions but remained as a member of the Board of Directors. No bonus was paid for the period from the end of his founders' employment agreement at June 30, 2000 until October 31, 2000.

As of October 31, 2000, Mr. Fredric D. Price was elected by the Board of Directors to the position of Chief Executive Officer and Chairman of the Board. The Compensation Committee negotiated a competitive compensation package appropriate to Mr. Price's demonstrated leadership capabilities and achievements. In negotiating this package, the Compensation Committee took into consideration the following factors:

- (1.) Mr. Price's six years of experience as the Chief Executive Officer of a biotechnology/nutrition company during which time he demonstrated a dedication to the successful commercialization of products and the vision to re-orient the business away from research programs which had challenges beyond the resources of that company.
- (2.) Mr. Price's three years of experience as the Chief Financial Officer of a biotechnology company. This period demonstrated the proven capability to develop and maintain strong, positive relationships with investors and analysts and to help secure a strong financial position for that company.
- (3.) Mr. Price's five years of experience as a strategy consultant to the biopharmaceutical industry and to investor groups during which time he developed a capacity for concise, insightful and realistic analysis and actionable recommendations and an understanding of the success factors and risks of biopharmaceutical development and commercialization.
- (4.) Mr. Price's record of advancement and achievement in the earlier phases of his career in progressively more responsible positions in both line and staff assignments at Pfizer Pharmaceuticals.
- (5.) Mr. Price's demonstrated problem solving and analytical capabilities while serving as a paid consultant to BioMarin in the one-month period prior to his election as Chief Executive Officer.

The Compensation Committee also considered Mr. Price's compensation in his previous position, competitive compensation levels for Chief Executive Officers in the biotechnology industry especially in the San Francisco Bay Area (as recorded in a well-regarded salary and compensation survey), and Mr. Price's qualifications in a favorable comparison to the qualifications of the slate of candidates developed for the Committee by an executive search firm.

Based on the above and additional considerations, the Compensation Committee signed a three-year employment agreement with Mr. Price that includes the following major compensation elements:

- (1.) A base annual salary in the first year of \$400,000, with increases to \$450,000 in the second year and to \$500,000 in the third year.
- (2.) A sign-on bonus of \$100,000 and annual cash bonuses based on the achievement of pre-set goals set by the Board of Directors. The cash bonus for the first year will be \$200,000 and the minimum cash bonus for each of the second and third years will be 25% of the base salary in those years. The maximum cash bonus in the second and third years will be 100% of base salary in those years.
- (3.) A stock grant of 25,000 restricted shares that will vest in three equal parts on January 1, 2001, January 1, 2002 and January 1, 2003. We also made a payment to Mr. Price of \$257,624 related to his tax liability resulting from this initial stock grant.
- (4.) A stock option to purchase 500,000 shares of the Company's common stock at the closing price on the Nasdaq National Market on October 30, 2000 which

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was \$12.50 per share. Subsequent option grants to purchase 125,000 shares will be granted on each anniversary date (October 31st) during the term of the agreement. The options vest monthly over a three- year period.

- (5.) A loan of up to \$1,500,000 with interest deferred for the purchase of a residence in connection with a relocation closer to the Company. Payment of relocation expenses including an appropriate gross-up to compensate for any tax liability incurred is also included.
- (6.) Significant severance and change of control benefits, including the following maximum benefits if he is actually or constructively terminated following a change of control or if he is terminated other than for cause: the payment of twice his annual base salary and bonus, full vesting of his restricted stock grant, forgiveness of his loan, and the payment of a gross- up bonus based upon Mr. Price's maximum tax liability related to the total severance package.

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- (7.) If we elect not to renew the employment agreement for an additional three-year term, we will pay Mr. Price an amount equal to his base salary and bonus paid in the third year of the agreement and a payment equal to his maximum tax liability related the total severance package.

The Compensation Committee believes that the above authorized compensation actions based upon Company 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 the Company and the creation of stockholder value.

Respectfully submitted on April 3, 2001 by the members of the Compensation Committee of the Board of Directors:

Erich Sager  
Ansbert S. Gadicke, M.D., Ph.D.  
Gwynn R. Williams

### REPORT OF THE AUDIT COMMITTEE

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

In this context, the Committee has met and held discussions with management and the independent auditors. Management represented to the Committee that the Company's consolidated financial statements were prepared in accordance with generally accepted accounting principles, and the Committee has reviewed and discussed the consolidated financials statements with management and the independent auditors. The 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 Committee has discussed with the independent auditors, the auditor's 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 Committee discussed with management and the independent auditors the overall scope and plans for the 2000 annual audit. The Committee meets 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 Committee

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recommended to the Board of Directors, 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, 2000, for filing with the U.S. Securities and Exchange Commission and the Swiss SWX New Market. The Committee and the Board also have recommended, subject to stockholder ratification, the selection of the Company's independent auditors for the year ending December 31, 2001.

Respectfully submitted on April 3, 2001 by the members of the Audit Committee of the Board of Directors:

Gwynn R. Williams  
Ansbert S. Gadicke, M.D., Ph.D.  
Erich Sager

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### CERTAIN RELATIONSHIPS AND RELATED TRANSACTIONS

Since January 1, 2000, 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 (1) compensation agreements and other arrangements, which are described elsewhere in this Proxy and (2) the transaction described below.

#### Transactions with Directors, Executive Officers and 5% Stockholders

Following Dr. Klock's resignation as our President and continuing until October 31, 2000, Mr. Sager performed certain consulting services for which we paid him a total of \$115,479. These services included assisting us in the executive search for and selection of a senior executive officer and, as Chairman of a Board management subcommittee, coordinating with and aiding the management of the Company in a wide variety of senior level tasks.

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During 2000, Glyko Biomedical Ltd. paid us \$7,700 per month in management fees for accounting and related governmental reporting services.

### 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 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 ("Founders' Shares") to the three founding officers of the Company, the Company has loaned \$1,300,000, \$800,000, and \$400,000 to Mr. Denison, Dr. Klock and Dr. Starr, respectively, to purchase common stock of the Company. The loans are evidenced by interest bearing promissory notes due on demand and are fully recourse. In August 2000, Dr. Klock paid-off the balance, including interest, of his note. In September, the Compensation Committee extended Mr. Denison's and Dr. Starr's notes until March 31, 2001.

The following table sets forth any indebtedness of directors or executive officers of the Company entered into in connection with the purchase of securities of the Company.

| Name of Borrower            | Lender  | Largest Amount of<br>Outstanding<br>Indebtedness (1) | Outstanding<br>Indebtedness<br>as of December 31,<br>2000 (1) |
|-----------------------------|---------|--|---|
| Grant W. Denison, Jr.       | Company | \$1,553,660  | \$1,553,660   |
| John C. Klock, M.D.         | Company | \$780,335  | --  |
| Christopher M. Starr, Ph.D. | Company | \$428,288  | \$409,875   |

(1) Includes accrued interest at 6% per annum.

### SECTION 16(a) BENEFICIAL OWNERSHIP REPORTING COMPLIANCE

Section 16(a) of the Securities Exchange Act of 1934, as amended (the "Exchange Act"), requires the Company's directors and officers and persons who own more than 10% of a registered class of the Company's equity securities to file reports of ownership and reports of changes in the ownership with the SEC and the National Association of Securities Dealers, Inc. 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 review of the copies of such reports furnished to us or written representations that no other reports were required, during the fiscal year ended December 31, 2000, all officers, directors, and greater than 10% stockholders complied with all Section 16(a) filing requirements.

The Company is aware that (i) the Change of Beneficial Ownership of Securities on Form 4 for Mr. Sager for the months of February, March and April 2000 which related to 14 transactions was filed in June 2000 and for the month of September 2000 which related to 3 transactions was filed in January 2001, for Dr. Brandley for the month of March 2000 which related to 2 transactions was filed in October 2000, for Dr. Starr for the month of May 2000 which related to 1 transaction was filed in July 2000, for Mr. Denison for the month of March 2000 which related to 2 transactions was filed in July 2000, for Dr. Klock for the month of May 2000

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which related to 1 transaction was filed in July 2000, for Dr. Swiedler for the month of May 2000 which related to 2 transactions was filed in July 2000, and (ii) the Initial Statement of Beneficial Ownership of Securities on Form 3 for Dr. Baffi for the month of May 2000 was filed in November 2000, were each filed late.

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### OTHER MATTERS

Except as otherwise indicated, information contained herein is given as of April 3, 2001. 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. However, if any other matters which are not now known to the Company's management should come properly before the Annual Meeting, the Proxy will be voted on such matters in accordance with the best judgment of the person voting it.

### 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 3rd day of April, 2001

Raymond W. Anderson

Chief Operating Officer, Chief Financial Officer, Secretary and  
Vice President, Finance and Administration

BioMarin Pharmaceutical Inc.

Audit Committee Charter

Introduction

BioMarin Pharmaceutical Inc.'s executive management is primarily responsible for the completeness and accuracy of its financial reporting and the adequacy of its internal financial and operating controls. Its Board of Directors has responsibility to oversee management's exercise of these responsibilities. The Board has determined that it is in the best interests of the Company to establish an Audit Committee whose authority and responsibilities are described by this Charter.

Organization

The full Board of Directors shall designate a committee of at least three independent directors to be an audit committee for the Company (the "Audit Committee" or "Committee"). The Audit Committee shall be composed of directors who are independent of the management of the Company and are free of any relationship that, in the opinion of the Board of Directors, would interfere with their exercise of independent judgement as a Committee member. Each member shall be financially literate and at least one member shall have accounting or related financial management expertise. Each of the foregoing qualifications shall be in compliance with all applicable rules promulgated by the Financial Accounting Standards Board ("FASB"), U.S. Securities and Exchange Commission ("SEC"), and the National Association of Securities Dealers ("NASD") or other

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regulatory body. Unless a chairperson is elected by the full Board, the members of the Committee may designate a chairperson by majority vote of the full Committee membership.

### Statement of Policy

The Audit Committee shall provide assistance to the Board of Directors in fulfilling their responsibilities to the stockholders, potential stockholders, and the investment community relating to corporate accounting, reporting practices, and the quality and integrity of the financial reports of Company. In so doing, it is the responsibility of the Audit Committee to maintain free and open means of communication between directors, the independent auditors, the internal auditors (when BioMarin is of sufficient size to make this function appropriate), and the financial management of the Company. The Audit Committee and the Board of Directors will review and assess the adequacy of this Charter annually.

### Responsibilities

In carrying out its responsibilities, the Audit Committee believes that its policies and procedures should remain flexible, in order to react to changing conditions and to ensure to the directors and stockholders that the corporate accounting and reporting practices of the Company are in accordance with all requirements and are of the highest quality.

In meeting its responsibilities:

1. The Committee will meet no less frequently than on a quarterly basis, prior to the release of earnings, and special meetings may be called when circumstances require and shall approve and maintain minutes for each such meeting. The Committee may ask members of management or others to attend the meeting and provide pertinent information as necessary.
2. The Committee, as representatives of the stockholders, has the ultimate authority to select, evaluate and, where appropriate, replace the independent auditors, or to nominate the independent auditors to be proposed for stockholder approval in the proxy statement. The Committee will review with management the performance, appointment and/or termination of the independent auditors and approve the compensation of the independent auditors. The Committee will recommend to the Board of Directors the independent auditors to be nominated to audit the financial statements of the Company and its divisions and subsidiaries and (if deemed appropriate) approve the discharge of the incumbent independent auditors.
3. The Committee will meet with the independent auditors and financial management of the Company to review the scope of the proposed audit for the current year and the audit procedures to be utilized, and at the conclusion of the audit, review the audit to include any comments and recommendations of the independent auditors.
4. The Committee will: review with the independent auditors, the Company's financial and accounting staff, and the internal auditor the adequacy and effectiveness of the internal accounting and financial controls of the Company including computerized information system controls and security; elicit any recommendations for the improvement of any internal control procedures or particular areas where new or more detailed controls or procedures are desirable; and review the adequacy of internal controls to expose any payments, transactions, or procedures that might be deemed illegal or otherwise improper.
5. The Committee will confirm and assure the independence of the independent auditors and the internal auditor and require that the independent auditors

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provide a formal written statement to the Committee setting forth all relationships between the independent auditors and the Company, consistent with the Independence Standards Board Standard No. 1. The Committee will discuss with the independent auditors any disclosed relationships or services which may impact the objectivity and independence of the independent auditors. The Committee will take, or recommend that the full Board of Directors take, appropriate action to ensure the independence of the independent auditors.

6. The Committee will inquire of management, the director of internal audit, and the independent auditors and others as deemed necessary about significant risks or exposures and assess the actions that management has taken to minimize these significant risks to the Company.
7. The Committee will review with management and the independent auditors at the completion of the annual audit:
  - (a) The Company's annual financial statements and related footnotes
  - (b) The independent auditor's audit of the financial statements and his or her report thereon.
  - (c) Any significant changes in the independent auditor's audit plan.
  - (d) Any serious difficulties or disputes with management encountered during the course of the audit.
  - (e) Other matters related to the conduct of the audit, which are to be communicated to the Committee under generally accepted auditing standards.
8. The Committee will review policies and procedures with respect to officers' expense reports and perquisites, including the use of Company assets, and consider the results of any review of these areas by the independent auditors or by the internal auditor.
9. The Committee will review legal and regulatory matters, including any litigation contingencies claims or assessments, that may have a material impact on the financial statements, related company compliance policies and programs, and reports received from regulators.
10. The Committee will meet with the independent auditors, the director of internal auditing, and management in separate executive sessions to discuss any matters that the Committee or these groups believe should be discussed privately with the Audit Committee.
11. The Committee will conduct or authorize investigations into any matters within the Committee's scope of responsibilities. The Audit Committee shall be empowered to retain independent counsel, accountants, or others to assist it in the conduct of its investigations.
12. The Committee will review accounting and financial succession planning and depth of staff within the Company.
13. The Committee will report Committee actions to the Board of Directors with such recommendations as the Committee may deem appropriate and will submit the minutes of all meetings to, or discuss the matters reviewed at each Committee meeting with, the Board of Directors.
14. The Committee will review with management and the independent auditors the Company's interim and year-end financial statements, including management's discussion and analysis, and audit findings (including any significant suggestions for improvements provided to management by the internal audit director, if any, and the independent auditors). Such review will include a discussion of significant adjustments recorded or adjustments passed and will conform with the requirements of SAS 61. Following such review, the Committee will recommend to the Board of Directors whether the audited financial statements should be included in the Company's annual report on

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Form 10-K and shall prepare a letter for the annual report that describes the Committee's composition and responsibilities and how they were discharged.

15. The Committee will review with the independent auditors or the director of internal auditing the results of their review of the Company's monitoring of compliance with the Company's code of conduct.
16. With respect to the internal audit function, the Committee will:
  - (a) Review and concur in the appointment, replacement, reassignment, or dismissal of the director of internal auditing.
  - (b) Consider, in consultation with the independent auditors and the director of internal auditing, the audit scope and plan of the internal auditors and the independent auditors.
  - (c) Review with the independent auditors and the director of internal audit, the coordination of audit effort to ensure completeness of coverage, reduction of redundant efforts, and the effective use of audit resources.
  - (d) Review significant internal audit's findings during the year and management's responses thereto.
  - (e) Inquire as to any difficulties encountered in the course of internal audits, including any restrictions on the scope of their work or access to required information.
  - (f) Review any changes in the scope of the audit plan.
  - (g) Review the internal auditing department budget and staffing.
  - (h) Approve the internal audit charter.
  - (i) Review internal audit's compliance with The IIA's Standards for the Professional Practice of Internal Auditing (Standards).
17. The Committee will perform other functions as assigned by law, any applicable regulatory body, including the NASD, the company's charter or by-laws, or the Board of Directors.

PROXY

BIOMARIN PHARMACEUTICAL INC.

2001 ANNUAL MEETING OF STOCKHOLDERS

MAY 24, 2001

THIS PROXY IS SOLICITED ON BEHALF OF THE BOARD OF DIRECTORS

The undersigned stockholder of BIOMARIN PHARMACEUTICAL INC., a Delaware corporation, hereby acknowledges receipt of the Notice of Annual Meeting of Stockholders and Proxy Statement, each dated April 3, 2001 and hereby appoints Fredric D. Price, Raymond W. Anderson, and Kim R. Tsuchimoto, 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 2001 Annual Meeting of Stockholders of BIOMARIN PHARMACEUTICAL INC. to be held on May 24, 2001 at 10am local time, at 46 Galli Drive, Novato, California 94949 and at any adjournment or adjournments thereof, and to vote all shares of common stock which the undersigned would be entitled to vote, if then and there personally present, on the matters set forth on the reverse side.

(Continued, and to be signed on the other side)

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1. ELECTION OF DIRECTORS:  
NOMINEES:

Fredric D. Price, Grant W. Denison, Jr.,  
Ansbert S. Gadicke, M.D., Ph.D., Erich Sager  
and Gwynn R. Williams

FOR [ ]

WITHHOLD FOR

INSTRUCTION: To withhold authority to vote for any individual nominee, write that nominee's name in the space provided below:

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2. Appointment of Arthur Andersen LLP as independent auditors of BIOMARIN PHARMACEUTICAL INC. for the fiscal year ending December 31, 2001

FOR [ ]

AGAINST [ ]

3. In their discretion, the proxies are authorized to vote upon such matters as may properly come before the Annual Meeting, or any adjournments thereof.

FOR [ ]

WITHHOLD FOR

THIS PROXY WILL BE VOTED AS DIRECTED OR, IF NO CONTRARY DIRECTION IS INDICATED, WILL BE VOTED FOR THE COMPANY'S NOMINEES FOR ELECTION TO THE BOARD OF DIRECTORS, THE APPOINTMENT OF ARTHUR ANDERSEN LLP OR, AS SAID PROXIES DEEM ADVISABLE, ON SUCH OTHER MATTERS AS MAY PROPERLY COME BEFORE THE ANNUAL MEETING, INCLUDING, AMONG OTHER THINGS, CONSIDERATION OF ANY MOTION MADE FOR ADJOURNMENT OF THE MEETING.

Signature(s) \_\_\_\_\_

Date: \_\_\_\_\_

This proxy should be marked, dated and signed by the stockholder(s) exactly as his or her name appears hereon, and returned promptly in the enclosed envelope. Persons signing in a fiduciary capacity should so indicate. If shares are held by joint tenants or as community property, both should sign.