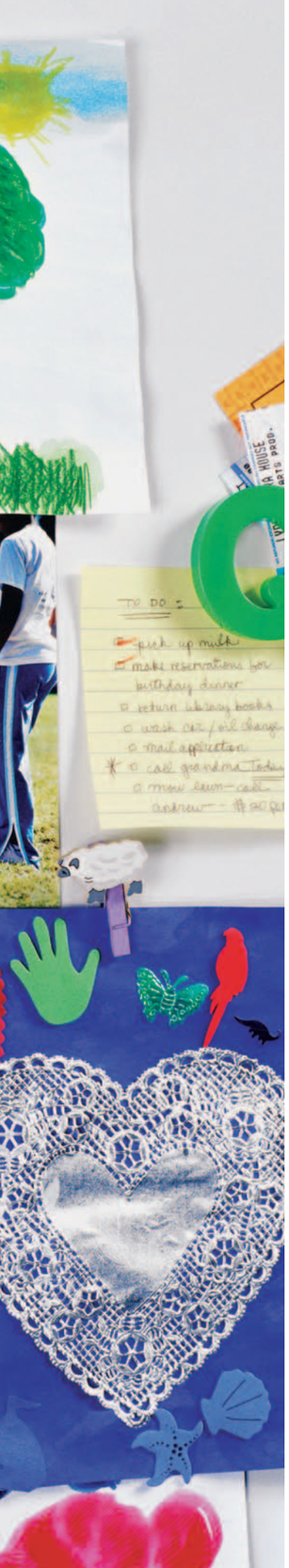
Annual Report 2004

BIOMARIN™



BioMarin

BioMarin develops and commercializes innovative biopharmaceuticals for serious diseases and medical conditions. Since the founding of the company in 1997, BioMarin has transformed into a fully integrated biopharmaceutical company with expertise to advance products from the clinic into the market.

Matching Proven Science With Proven Needs BioMarin applies its scientific know-how and clinical and regulatory expertise to rapidly and efficiently develop therapies that address the critical needs of individuals seeking new treatment options. The company has made significant progress on this front and today has two approved products on the market and multiple opportunities in its development pipeline.

Aldurazyme® (laronidase) for **MPS I** (mucopolysaccharidosis I) received marketing approval in the United States and European Union in the second quarter of 2003, making it the company's first marketed product. Today, Aldurazyme continues to be the only drug therapy for this progressive and life-threatening disease.

In May 2004, BioMarin added **Orapred**® (prednisolone sodium phosphate oral solution) for asthma to its product portfolio. The patent-protected, taste-masking technology of Orapred helps cover the bitter taste of prednisolone, making it easier for children to take without experiencing a natural gag reflex induced by other liquid formulations of prednisolone.

Looking Forward BioMarin is working to bring additional products to market, including **rhASB** (galsulfase) for **MPS VI** (mucopolysaccharidosis VI) and **Phenoptin**™ (sapropterin hydrochloride) for **PKU** (phenylketonuria)—two product candidates which, if approved, could become the first drug therapies for the treatment of these inherited metabolic diseases.

Opportunities to Improve Patient Lives

Products on the Market

Aldurazyme® (laronidase) for MPS I First drug therapy for the treatment of MPS I	**Orapred**® (prednisolone sodium phosphate oral solution) for asthma Follow-on formulations in development

Pipeline

	Preclinical testing	Phase 1	Phase 2	Phase 3	BLA/NDA MAA
rhASB for MPS VI					
Phenoptin™ for PKU					
Vibrilase™ for Burns					
Phenylase™ for Severe PKU					

MPS I and MPS VI are both progressive and life-threatening lysosomal storage disorders caused by a deficiency of specific enzymes. Individuals with MPS I and MPS VI commonly experience debilitating symptoms, including impaired cardiac and pulmonary function, delayed physical development, skeletal and joint deformities, impaired vision and hearing, and reduced endurance. MPS I and MPS VI frequently result in death between childhood and early adulthood.

PKU is one of the most commonly inherited metabolic diseases. It is caused by a deficiency of phenylalanine hydroxylase (PAH), an enzyme needed to metabolize phenylalanine (Phe). Phe is an amino acid found in most protein-containing foods. If not metabolized, it accumulates to abnormally high levels in the blood and other tissues. Sustained high Phe levels can cause serious neurological complications, including mental retardation, mental illness, loss of IQ, seizures and tremors, and cognitive problems.

How do I feel about PKU? It is a lot harder than some people think. My mom cooks me goodies, but they will never match up to real bread or pizza.

Madeleine, Age 12, PKU

Creating Possibilities

Imagine opening a refrigerator to find only a few foods you could eat without jeopardizing your health. Many individuals living with PKU experience this daily. To manage the disease and maintain non-toxic blood Phe levels, individuals must adhere to a severely restricted diet, one consisting of foods that are low in Phe and supplemented with medical foods, many of which are unpalatable and expensive. In October 2000, a Consensus Panel convened by the National Institutes of Health concluded that individuals need to adhere to this diet throughout life and that failure to do so can result in significant decline of mental and behavioral performance. Despite this conclusion, however, this diet is too difficult for many to comply with; approximately 80 percent of individuals with PKU stray from it during adolescence and adulthood.

BioMarin is seeking to help individuals with PKU better manage their disease. To do this, the company is currently evaluating two investigational approaches for treating the full spectrum of PKU: Phenoptin, an oral small molecule therapeutic for mild to moderate forms of the disease, and Phenylase™(phenylalanine ammonia lyase), an injectable enzyme substitution therapy for more severe forms. If proven to be safe and effective, these investigational therapies could provide individuals with PKU tools to better manage their disease, a task which is currently difficult to do given the presence of Phe in most commonly consumed foods.*

- Bread: 146 mg of Phe (slice)
- Corn on the Cob: 256 mg of Phe (ear)
- Cake: 328 mg of Phe (slice)
- Steak: 945 mg of Phe (85 g)

*Approximate Phe levels. Individuals with classical PKU typically consume a Phe-restricted diet containing approximately 300 mg to 600 mg of Phe per day. An individual's allowable daily Phe intake varies on several factors, including their size and severity of the disease.

When I graduate from college, I want to design clothing for people who are small like me—we want to be fashionable too!

Kendra, College Freshman, MPS VI

Moving Forward

With each milestone we reach, we also are helping others reach important milestones of their own—milestones that reflect they are growing up and moving forward with their lives.

In 2004, BioMarin reached significant milestones throughout its product portfolio.

By the close of the year, over 350 individuals worldwide were receiving **Aldurazyme** for MPS I, the first specific drug therapy for this disease. BioMarin expects this number to increase as more individuals are identified and as marketing authorization is received in additional countries.

The company also took steps towards expanding the market of **Orapred** for asthma. While the current liquid formulation is used primarily for treating young children, follow-on formulations, including an oral disintegrating tablet, could be used to treat older children and adults.

BioMarin submitted marketing applications in the United States and European Union for **rhASB**, the company's investigational treatment for MPS VI. The U.S. Food and Drug Administration (FDA) granted the application six-month review status, setting the stage for a decision by mid-2005. If approved, BioMarin plans to commercialize rhASB within the United States and to seek a partner for ex-U.S. commercialization. Regulatory agencies in both the United States and European Union have granted rhASB for MPS VI orphan drug designation. There is currently no specific drug therapy for the treatment of MPS VI.

The company also advanced clinical development of **Phenoptin** for PKU, moving from the initiation of the development program into a Phase 3 clinical trial in just over one year. BioMarin estimates that approximately 30 to 50 percent of the 50,000 diagnosed individuals living in the developed world could potentially benefit from this therapy. **Phenylase**, the company's preclinical-stage enzyme substitution therapy, is being evaluated for treatment of those with more severe forms of PKU, likely those who do not respond to Phenoptin. The U.S. FDA has granted orphan drug designation to both Phenoptin and Phenylase for the treatment of PKU. The European Medicines Evaluation Agency has also granted Phenoptin orphan drug designation.

I am inspired to do my best knowing that what I do could positively impact others and help them accomplish their own dreams and aspirations.

Cheri Piscia-Nichols, Clinical Research Associate, has worked at BioMarin for 7 years

To Our Stockholders

The year of 2004 was marked by significant progress and important changes in our business.

Throughout the year, we continued to meet critical milestones. After announcing positive data from our Phase 3 trial of **rhASB** for MPS VI, we then successfully prepared and filed marketing authorization applications in both the United States and European Union. Today, we continue to work closely with regulatory agencies and hope to bring rhASB to market in the United States in the second half of 2005, and to European countries soon thereafter, pending regulatory approvals. If approved, rhASB could become our second commercialized product to be developed and manufactured by BioMarin and our first in-house product to be marketed by our U.S.-based sales force, which we added to our business in May 2004.

We also made significant progress in our PKU program, moving it from a pilot study at the beginning of the year, into a Phase 2 trial by the close of the year, and into a Phase 3 trial in early 2005. Additionally, the partnership we formed this year with Daiichi Suntory Pharma Co., Ltd., further strengthens our intellectual property position surrounding the manufacturing of **Phenoptin** and helps ensure an adequate supply of the investigational drug for clinical trials.

From a commercial perspective, we seek to fully leverage our commercial infrastructure, which includes a sales force that calls on over 17,000 pediatricians nationwide. With this strategic business asset in place, we now have the capability to market approved products in the United States without the need for an external partner. This positions us to capture greater value from products emerging from our pipeline as well as the opportunity to enter into co-promotion agreements with companies seeking to bring products to market.

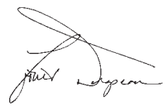
With regard to our marketed products, in 2004, we were pleased to see the first quarterly profit from **Aldurazyme** for MPS I, which we developed in partnership with Genzyme Corporation. We expect to see continued revenue growth of Aldurazyme in 2005 as more individuals with MPS I are identified and individuals currently in extended clinical studies are transitioned to commercial therapy.

Sara Isbell, Research Associate II, Cellular Genetics

We recognize the substantial challenge we face with **Orapred**, a U.S. FDA-approved liquid oral steroid that we added to our portfolio in May 2004. The recent introduction of generic competition has created a highly competitive environment. We are committed to doing everything possible to ensure Orapred positively impacts our bottom line. We believe that Orapred and its proprietary taste-masking technology continues to offer advantages over other liquid formulations of prednisolone and that follow-on formulations, including an oral disintegrating tablet ('Orapred ODT'), provide us with the opportunity to further expand our target market.

We have come a long way since our founding in 1997, thanks to our dedicated employees and leadership team, the pioneering spirit of individuals involved in our clinical trials, and the support of our stockholders.

On behalf of BioMarin and our Board of Directors, I express my thanks to all and I look forward to sharing our progress with you throughout the year.



Louis Drapeau, Acting Chief Exective Officer

Stock Listing

BioMarin Pharmaceutical Inc. is listed on the Nasdaq National Market and the SWX Swiss Exchange under the symbol BMRN.

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Forward-Looking Statement

This Annual Report contains 'forward-looking statements' as defined under securities laws. Many of these statements can be identified by the use of terminology such as 'believes,' 'expects,' 'anticipates,' 'plans,' 'intends,' 'may,' 'will,' 'projects,' 'continues,' 'estimates,' 'potential,' 'opportunity,' and so on. Our actual results or experience could differ significantly from the forward-looking statement. Factors that could cause or contribute to these differences include the results of our current clinical trials, our ability to successfully market our products, if we are able to obtain regulatory approval and the other factors discussed in the enclosed Form 10-K and the section entitled 'Factors That May Affect Future Results' therein.

You should not place undue influence on these forward-looking statements which speak only as of the date that they were made. These cautionary statements should be considered in connection with any written or oral forward-looking statements that we may issue in the future. We do not undertake any obligation to release publicly any revisions to these forward-looking statements after completion of the distribution of this Annual Report to reflect later events or circumstances or to reflect the occurrence of unanticipated events.

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MATCHING PROVEN SCIENCE WITH PROVEN NEEDS