
BRINGING
THE
PROMISE
TO
MARKET

A MESSAGE FROM THE CEO

Less than 10 years after opening our doors, we are approaching the anticipated launch of our third product. Our proven ability to deliver products to market has transformed BioMarin from a product development-focused company to a fully integrated commercial company with growing product revenues and an exciting product pipeline. As important as our commercial success, is the fact that our products are bringing the promise of new therapeutics to patients around the world suffering from rare genetic diseases.

Building A Meaningful Revenue Base

The efficiency and speed of our research, development and manufacturing efforts is at the heart of our ability to quickly bring products to market. We have successfully advanced each of our two flagship drugs, Aldurazyme® (laronidase) and Naglazyme® (galsulfase), from research to commercialization in about five years. Revenues for both products increased substantially in 2006, as we further expanded our reach to patients and secured regulatory approvals worldwide. This sales growth, coupled with substantial improvements to our capital structure, has significantly improved the company's financial profile.

2006 marked important progress in the commercialization efforts of Aldurazyme. By the end of the year, worldwide sales increased 26 percent to $96.3 million and as a result, BioMarin's share of the profit in BioMarin/Genzyme LLC increased 63.6% over fiscal 2005.

In 2006, Naglazyme, our first independently launched therapy for MPS VI, was introduced in Europe after launching in the U.S. in mid-2005. When considered as a stand-alone business, this product became profitable in its first year on the market, and with $46.5 million in sales, we have clearly established our ability to independently launch products for rare diseases. This success now positions BioMarin as a commercial partner of choice for marketing highly-specialized pharmaceutical products addressing the needs of patients with rare diseases.

Expanding Product Profile

Growing revenues generated by Aldurazyme and Naglazyme are helping us advance additional compounds in our product pipeline that target both orphan genetic diseases and other conditions such as cardiovascular disease. This will present substantially larger market opportunities to augment our current enzyme replacement franchise.

Kuvan™ (sapropterin dihydrochloride), a small molecule oral therapeutic for the treatment of phenylketonuria (PKU), has demonstrated strong safety and efficacy in Phase 3 clinical trials and has been granted Fast Track status by the FDA. This product, co-developed with Merck Serono, will be the first approved drug treatment available for PKU, a metabolic disease that affects more than 50,000 people in the developed world. Pending priority review designation and a favorable regulatory review, U.S. approval for Kuvan is expected in late 2007. It will be the third drug we have successfully advanced from research to commercialization in less than five years. The same enzyme cofactor found in Kuvan, 6R-BH4, has also been found to play a key protective role in the cardiovascular system. To leverage this asset in markets beyond PKU, we are conducting several proof-of-concept studies for a variety of cardiovascular indications.

Finally, 2006 marked significant progress in our preclinical development of Phenylase™, an enzyme substitution therapy for the treatment of severe forms of PKU. We are currently performing additional preclinical studies with the goal of filing an IND by the end of 2007 and initiating clinical studies in early 2008.

Looking Forward

We are proud of our track record of bringing innovative, high-value biopharmaceuticals to market. By leveraging clear-cut development strategy and efficient, adaptive clinical development models we have achieved regulatory approvals far faster than the industry average and the results of our progress are now becoming more tangible in the form of improved financial performance. 2006 was a year of growth and progress in our efforts to provide increased value to both investors and patients.

I would like to thank all of our employees and partners for their hard work and commitment to excellence. We appreciate your continued support of the company and look forward to keeping you informed of our progress throughout the year.

Sincerely,

Jean-Jacques Bienaimé
Chief Executive Officer

BIOMARIN®

2006 Milestones

Commercial Success

BioMarin has transformed from a product development-focused company to a fully integrated commercial company with rapidly growing product revenues and an exciting product pipeline.

Global Expansion

In 2006, BioMarin further established its commercial presence in the United States and Europe and is now expanding into Latin America.

Improved Financial Profile

2006 was marked by significant increases in product sales, a decrease in net loss, the conversion of debt to common stock, and an increasing cash balance. This has enabled BioMarin to increase funding of growth opportunities.

Proven Business Model

After less than four years in development, BioMarin's third product, Kuvan, is expected to be approved in the U.S. by the end of 2007, further demonstrating the company's ability to recognize unique opportunities and develop them quickly.

Expanding Market Opportunities

BioMarin is now evaluating 6R-BH4 for a variety of cardiovascular indications. If successful, these programs could significantly expand the company's global market potential.

Fulfilling Unmet Needs

BioMarin is bringing the promise of new therapeutics to thousands of patients worldwide who might otherwise go untreated. In less than 10 years, the company has developed and commercialized two breakthrough drugs for rare genetic diseases and is well positioned to address many others with its exciting product pipeline.

EXPERIENCED MANAGEMENT

Jean-Jacques Bienaimé
Chief Executive Officer

Jeffrey Cooper
Senior Vice President,
Chief Financial Officer

Emil Kakkis, M.D., Ph.D.
Senior Vice President,
Chief Medical Officer

Stephen Aselage
Senior Vice President,
Global Commercial Operations

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

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

G. Eric Davis
Vice President,
General Counsel & Secretary

Mark Wood
Vice President,
Human Resources

Phenylase Core Team
Top row, left to right: James Dickow (Director, Cell Culture), Gia DePillis, Ph.D. (Director, Regulatory Affairs), Laurie Tsuruda, Ph.D., D.A.B.T. (Associate Director, Pharmacology/ Toxicology), Marlyn Morimoto, MS (Sr. Financial Analyst), Paul Fitzpatrick, Ph.D. (Principal Scientist, Research and Development), Mubarack Muthalif, Ph.D., MBA (Senior Program Manager, Product Development/ Phenylase Core Team Leader) and Julie Wilson, Ph.D. (Senior Product Manager).

In the Laboratory
Second row, left to right: Carroll Henschell (Study Monitor, Pharmacology/Toxicology) and Sean Bell, Ph.D. (Scientist, Research & Development).

Kuvan Core Team
Third row, left to right: Mark Henderson, Ph.D. (Associate Director, Quality Assurance), Alejandro Dorenbaum. M.D. (Sr. Medical Director), Benjamin Dewees, RAC (Senior Manager, Regulatory Affairs), Julie Schraeder (Director, Finance), Dong Wei, Ph.D. (Senior Program Manager, Product Development), Dan Oppenheimer, Ph.D. (Senior Director, Program Management / Kuvan Program Leader), Sandra Shpilberg, MBA (Director of Marketing), V. Miles Rios, Jr., BA, MA (Associate Director, Clinical Operations).

Representatives From European Sales Team
Bottom row, left to right: Philippe Carteron de Balmont (Country Manager, Switzerland, France, & BeNeLux), Guy Eggleton (Director of Operations, BioMarin Europe Ltd.), David Boothe, BSc (Director of Marketing, BioMarin Europe) and Felis Iglesias (Country Manager, Spain, Portugal).

Preclinical | Phase I

ON THE MARKET

NAGLAZYME® *for mucopolysaccharidosis VI (MPS VI)*

MPS VI is a rare, inherited metabolic disease caused by a deficiency in arylsulfatase B, an enzyme involved in the carbohydrate recycling throughout the body. Over time, the disease progresses, causing severe disabilities such as impaired cardiac and pulmonary function, delayed physical development, skeletal and joint deformities, reduced endurance and impaired vision and hearing. The majority of untreated patients die from disease-related complications between childhood and early adulthood.

Naglazyme is the first drug approved to treat MPS VI in the United States and Europe. It is an enzyme replacement therapy, using a recombinant version of arylsulfatase B to replace or supplement low to non-existent levels of the natural enzyme in the body.

ALDURAZYME® *for mucopolysaccharidosis I (MPS I)*

Like MPS VI, MPS I is a rare, inherited metabolic disease caused by a deficiency of alpha-L-iduronidase, an enzyme involved in the carbohydrate recycling throughout the body. Symptoms can include impaired cardiac and pulmonary function, delayed physical development, skeletal and joint deformities, reduced endurance, and delayed mental function. Untreated patients can die from complications before adulthood.

Aldurazyme is the first drug approved to treat MPS I in the United States and Europe, and was recently approved for use in Japan. It is an enzyme replacement therapy, using a recombinant version of alpha-L-iduronidase to replace or supplement low to non-existent levels of the natural enzyme in the body.

PRE-NDA

KUVAN™ *for PKU*

Phenylketonuria (PKU) is a rare, inherited metabolic disease resulting from a deficiency of phenylalanine hydroxylase, the enzyme responsible for converting phenylalanine (Phe) to tyrosine. Sustained elevated blood Phe levels can result in serious and irreversible neurological damage. In all countries where modern medical technology is available, all newborns are screened for PKU. There are approximately 50,000 PKU patients under the age of 40 in the developed world.

Kuvan (sapropterin dihydrochloride), an oral small molecule therapeutic, is the synthetic form of 6R-BH4 (tetrahydrobiopterin), a naturally-occurring enzyme cofactor that works in conjunction with phenylalanine hydroxylase to metabolize Phe.

IN DEVELOPMENT

6R-BH4 *for cardiovascular disease and sickle cell disease*

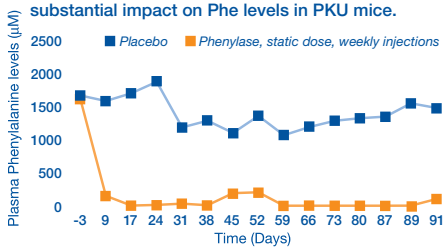
6R-BH4 (tetrahydrobiopterin), more commonly known as BH4, is a naturally-occurring enzyme cofactor that is required for numerous biochemical and physiologic processes, including the synthesis of nitric oxide (NO). NO has been shown to play a key protective role throughout the cardiovascular system.

A deficiency of BH4 can disrupt the production of endothelial NO, leading to endothelial dysfunction, which has been associated with many cardiovascular diseases impacting millions of people worldwide.

PHENYLASE™ *for PKU*

BioMarin expects to file an IND application for Phenylase in late 2007. Phenylase is an investigational enzyme substitution therapy designed to treat severe PKU, specifically in patients who are non-BH4 responsive. The active ingredient, phenylalanine ammonia lyase, is designed to break down Phe that builds up due to lack of or diminished activity of the enzyme phenylalanine hydroxylase.

Study shows weekly treatment of Phenylase has substantial impact on Phe levels in PKU mice.



Phase II

Phase III

Phase 1 clinical trials of Naglazyme were completed in 2001, demonstrating product safety and early indications of efficacy.

A Phase 2 open-label study of Naglazyme was conducted in the U.S. and Australia in 2002 evaluating the safety and efficacy of the drug in subjects ranging in age from six to 22. Long-term results showed patients improved their walk distances over baseline and stair-climbing ability. Functional improvements were also observed.

Conducted at six international sites, the Phase 3 trial of Naglazyme was completed in early 2004. The trial enrolled patients ranging in age from five to 29 years old and ran for 24 consecutive weeks. Patients in this study also showed statistically significant improvement in measurements of endurance, including walk distances and stair climbing.

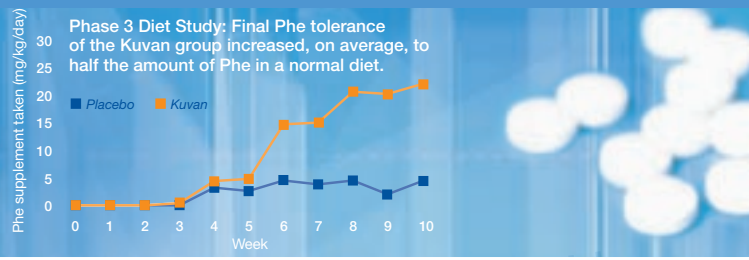
In 1998, BioMarin and Genzyme formed BioMarin/Genzyme LLC to develop and commercialize Aldurazyme worldwide. Early trials demonstrated safety and promising clinical results and in late 2001, a pivotal Phase 3 trial demonstrated a statistically significant increase in patients' pulmonary capacity, as well as increased endurance.

An extension study followed 45 patients who continued the treatment for an additional three years in which they demonstrated further improvement or stabilization in pulmonary function and endurance.

BioMarin's strategic partner, Merck Serono, is supporting the development of Kuvan and has rights to market the product outside of the U.S. and Japan. BioMarin retains exclusive rights to market Kuvan in the United States.

Data from Phase 3 clinical trials demonstrated the safety and tolerability of Kuvan, as well as its ability to increase Phe tolerance and reduce blood Phe levels in PKU patients.

Approval for Kuvan is expected in late 2007 in the U.S. and in 2008 in Europe.



BH4 has also shown promise as a treatment for diseases where endothelial dysfunction plays a larger role. To identify potential indications, BioMarin is conducting small, proof-of-concept studies for a number of cardiovascular diseases.

In the second quarter of 2007, the company expects to initiate a Phase 2 clinical study in sickle cell disease (SCD), an inherited blood disorder that affects red blood cells. There are an estimated 70,000 – 100,000 SCD patients in the U.S.

In the second quarter of 2007, an investigator-sponsored Phase 1 trial of BH4 in pulmonary arterial hypertension (PAH) is expected to be initiated. PAH is a chronic, life-threatening disease afflicting 100,000 to 200,000 people worldwide.

BioMarin has transformed from a product development-oriented company to a fully integrated commercial company with growing product revenues and an exciting product pipeline.

BioMarin products are bringing the promise of new therapeutics to patients around the world suffering from rare genetic diseases.

BioMarin is now a commercial partner of choice for marketing highly-specialized pharmaceutical products that target both orphan diseases and more common conditions such as cardiovascular disease.

By leveraging clear-cut development strategy and efficient, adaptive clinical development models, BioMarin has achieved regulatory approvals far faster than the industry average.

Expected milestones in 2007–08 include the anticipated launch of Kuvan, the initiation of clinical studies of Phenylase and preclinical studies of 6R-BH4 for a variety of cardiovascular indications.

BLA / NDA / MAA

Strong safety and efficacy data generated in the Naglazyme clinical trials formed the basis of BioMarin's U.S. and European regulatory submissions in late 2004. In all, 56 patients participated in the trials in seven different countries.

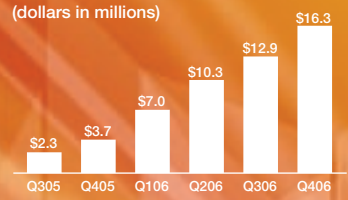
As Naglazyme moved into Phase 3 clinical trials, the company expanded its manufacturing facility to meet growing needs and product demand. The product received FDA approval in May 2005 and EMEA approval in January 2006, making it the first approved drug treatment for MPS VI.

With FDA and EC EMEA approval granted in 2003, Aldurazyme became the first commercially available treatment for MPS I, and provided physicians with the ability to change the course of the disease and provide better outcomes for patients worldwide.

On The Market

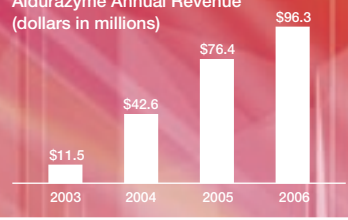
Naglazyme was launched in the U.S. in June 2005 and in the EU in February 2006. In 2007 and beyond, growth will be driven by factors such as new patient identification, geographic expansion and increasing dosage requirements.

Naglazyme Revenue by Quarters
(dollars in millions)

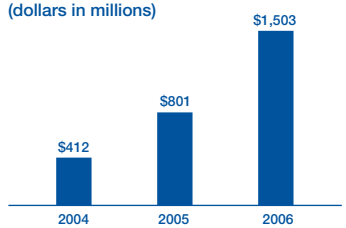


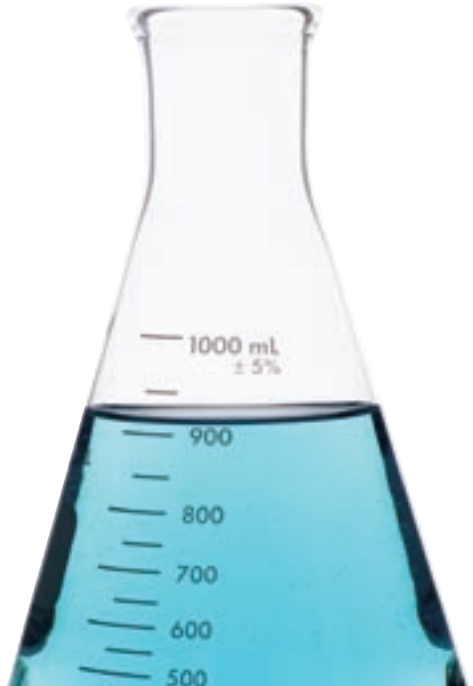
The product has been on the market since 2003 and has generated increased sales worldwide through 2006.

Aldurazyme Annual Revenue
(dollars in millions)



BioMarin's market capitalization has more than tripled over the last two years.
(dollars in millions)





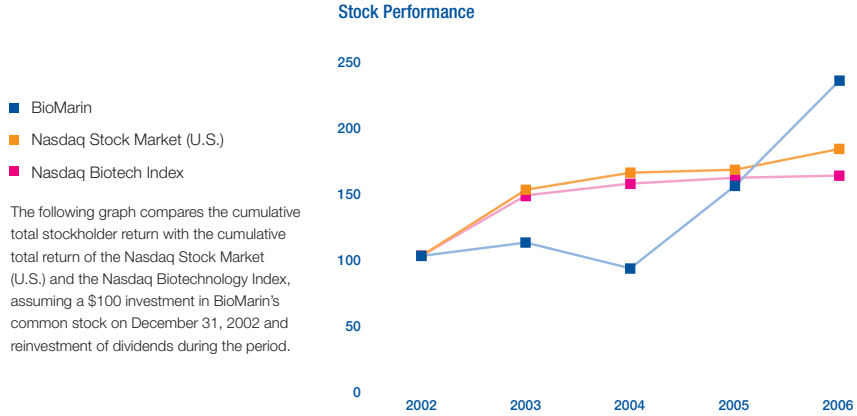
BioMarin's two flagship drugs, Aldurazyme® (galsulfase) for MPS I, and Naglazyme® (laronidase) for MPS VI, successfully advanced from research to commercialization status in about five years.

Naglazyme, BioMarin's first independently launched therapy, became profitable in its first year on the market. With full worldwide rights, the company is now enjoying commercial success in the United States and Europe and is expanding into Latin America.

BIOMARIN®

BioMarin develops and commercializes innovative biopharmaceuticals for serious diseases and medical conditions. The company's portfolio comprises two approved products and multiple clinical and preclinical product candidates.

Stock Performance



The following graph compares the cumulative total stockholder return with the cumulative total return of the Nasdaq Stock Market (U.S.) and the Nasdaq Biotechnology Index, assuming a $100 investment in BioMarin's common stock on December 31, 2002 and reinvestment of dividends during the period.

Corporate Headquarters

BioMarin Pharmaceutical Inc.
105 Digital Drive
Novato, CA 94949
Tel: 415-506-6700
Fax: 415-382-7889
E-mail: ir@bmrn.com
www.bmrn.com

Stock Listing

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

Independent Accountants

KPMG LLP
San Francisco, CA

Transfer Agent

Mellon Investor Services LLC
480 Washington Boulevard
Jersey City, NJ 07310
Tel: 800-522-6645 (Domestic)
201-680-6578 (International)

Executives

Jean-Jacques Bienaimé
Chief Executive Officer

Jeffrey Cooper
Senior Vice President
Chief Financial Officer

Emil Kakkis, M.D., Ph.D.
Senior Vice President,
Chief Medical Officer

Stephen Aselage
Senior Vice President
Global Commercial Operations

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

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

Jeff Ajer
Vice President
Sales & Marketing Operations

William Aliski
Vice President &
General Manager
European Operations

G. Eric Davis
Vice President
General Counsel & Secretary

Steven Jungles
Vice President
Supply Chain

Daniel Maher
Vice President
Product Development

R. Andrew Ramelmeier, Ph.D.
Vice President
Manufacturing and
Process Development

Victoria Sluzky, Ph.D.
Vice President
Quality & Analytical Chemistry

Amy Waterhouse
Vice President
Regulatory & Government Affairs

Mark Wood
Vice President
Human Resources

Board of Directors

Jean-Jacques Bienaimé
Chief Executive Officer
BioMarin Pharmaceutical Inc.

Joseph Klein, III
Managing Director
Gauss Capital Advisors, LLC

Pierre Lapalme
Former President &
Chief Executive Officer
North America Ethypharm, Inc.

Michael Grey
President &
Chief Executive Officer
SGX Pharmaceuticals, Inc.

Alan Lewis, Ph.D.
President &
Chief Executive Officer
Novocell, Inc.

Elaine Heron, Ph.D.
Chairman &
Chief Executive Officer
Labcyte Inc.

Randy Meier
Executive Vice President,
Eye Care Business and
Operations, and
Chief Financial Officer
Advanced Medical Optics, Inc.

Forward-Looking Statement: *This Annual Report contains 'forward-looking statements' as defined under securities laws. These statements can generally be identified by the use of terminology such as 'believes', 'expects', 'anticipates', 'plans', 'intends', 'may', 'will', 'projects', 'continues', 'estimates', 'potential', 'opportunity', and so on. The company's 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 current clinical trials, the company's ability to obtain regulatory approval for product candidates, its ability to successfully market products. and other factors discussed in the enclosed Form 10-K and the section entitled 'Risk Factors' therein.*

One should not place undue influence on these forward-looking statements that 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 the company may issue in the future. BioMarin Pharmaceutical Inc. does 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.

BioMarin Pharmaceutical Inc.
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Fax: 415.382.7889