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For Immediate Release:

BioMarin Receives Marketing Approval for Naglazyme in European Union

2006 Revenue Guidance for Naglazyme and Aldurazyme Provided

Conference Call to be Held Today at 5:00 p.m. ET (23:00 CET)

Novato, Calif, January 30, 2006 – BioMarin Pharmaceutical Inc. (Nasdaq and SWX: BMRN) announced today that the European Commission has granted marketing authorization for Naglazyme™ (galsulfase), the first specific treatment approved in the European Union for patients with the genetic disease mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome). As the first drug ever approved for MPS VI, Naglazyme has been granted orphan drug status in the European Union, which confers 10 years of market exclusivity.

Naglazyme has been approved in the 25 member states of the European Union, Iceland and Norway for long-term enzyme replacement therapy in patients with a confirmed diagnosis of MPS VI. All clinical post-authorization commitments requested by the Committee for Medicinal Products for Human Use (CHMP) will be fulfilled through a voluntary surveillance program that will monitor patients on commercial therapy. No additional clinical trials are required. BioMarin will launch Naglazyme in the European Union on a country-by-country basis.

"I have observed the dramatic improvement enzyme replacement therapy with Naglazyme can have on patients afflicted with MPS VI and am pleased that the therapy will soon be available to individuals outside of clinical studies," stated Ed Wraith, M.D., Consultant Pediatrician at the Royal Manchester Children's Hospital located in Manchester, United Kingdom, and a principal investigator for Naglazyme clinical trials. "Naglazyme holds the potential to positively impact the lives of MPS VI patients and their caregivers."

"We are excited that BioMarin has succeeded in bringing the first enzyme replacement therapy for MPS VI to market, first in the United States and now in the European Union," stated Jean-Jacques Bienaimé, Chief Executive Officer of BioMarin. "With European commercial operations in place, we are ready to launch Naglazyme. Additionally, we are now positioned to partner with companies looking to bring other products for rare diseases to the European marketplace." Mr. Bienaimé continued, "We are pleased with the growing sales and profitability of Aldurazyme, and with Naglazyme now approved in Europe and the United States, we expect combined worldwide sales of Aldurazyme by our joint venture and Naglazyme by us for 2006 to be in the range of \$118 million to \$132 million."

2006 Naglazyme and Aldurazyme Revenue Guidance

- BioMarin estimates sales of Naglazyme for 2006 to be in a range of \$28 million to \$32 million.

- BioMarin and Genzyme estimate sales of Aldurazyme® (laronidase) for 2006 to be in a range of \$90 million to \$100 million.

Conference Call and Webcast Scheduled for Today, January 30

BioMarin will host a conference call and webcast today, January 30 at 5:00 p.m. ET to discuss today's announcement and to provide an update on its investigational product candidates. This event can be accessed on the investor section of the BioMarin website at www.BMRN.com.

Date: January 30, 2006
Time: 5:00 p.m. ET (23:00 CET)
U.S. & Canada Toll-free Dial in #: 866.831.6291
International Dial in #: 617.213.8860
Participant Code: 95593593
Replay Toll-free Dial in #: 888.286.8010
Replay International Dial in #: 617.801.6888
Replay Code: 91000661

About MPS VI

MPS VI (also known as Maroteaux-Lamy syndrome) is a debilitating, life-threatening genetic disease caused by a deficiency of the enzyme *N*-acetylgalactosamine 4-sulfatase. This enzyme deficiency leads to the accumulation of certain complex carbohydrates, glycosaminoglycans (GAGs), in the lysosomes, giving rise to progressive cellular, tissue and organ system dysfunction. The majority of individuals with MPS VI die from disease-related complications between childhood and early adulthood. Additional information can be found at www.mpsvi.com.

About Naglazyme

Naglazyme is the first and only enzyme replacement therapy indicated for the treatment of MPS VI. As the first drug approved for MPS VI, regulatory agencies in both the United States and European Union have granted Naglazyme orphan drug status, which confers seven years and 10 years of market exclusivity, respectively. Additional information can be found at www.naglazyme.com.

Naglazyme is indicated for patients with MPS VI. Naglazyme has been shown to improve walking and stair-climbing capacity.

The most common adverse events observed in clinical trials in Naglazyme-treated patients were headache, fever, arthralgia, vomiting, upper respiratory infections, abdominal pain, diarrhea, ear pain, cough, and otitis media. Severe reactions included angioneurotic edema, hypotension, dyspnea, bronchospasm, respiratory distress, apnea, and urticaria. The most common symptoms of infusion reactions included fever, chills/rigors, headache, rash, and mild to moderate urticaria. Nausea, vomiting, elevated blood pressure, retrosternal pain, abdominal pain, malaise, and joint pain were also reported. No patients discontinued Naglazyme infusions for adverse events and all patients that completed the double-blind portion of the trial continue to receive weekly infusions of Naglazyme. Nearly all patients developed antibodies as a result of treatment, but the level of the immune response did not correlate with the severity of adverse events or impact the improvements experienced in endurance. Because antihistamine use may increase the risk of apneic episodes, evaluation of airway patency should be considered prior to the initiation of treatment. Consideration to delay Naglazyme infusion should be given when treating patients who present with an acute febrile or respiratory illness.

About BioMarin

BioMarin develops and commercializes innovative biopharmaceuticals for serious diseases and medical conditions. The company's product portfolio is comprised of three approved products and multiple clinical and preclinical product candidates. Approved products include Naglazyme™

(galsulfase) for mucopolysaccharidosis VI (MPS VI), a product wholly developed and commercialized by BioMarin, Aldurazyme[®] (laronidase) for mucopolysaccharidosis I (MPS I), and Orapred[®] (prednisolone sodium phosphate oral solution) for inflammatory conditions. Investigational product candidates include Phenoptin[™] (sapropterin dihydrochloride), a Phase 3 product candidate for the treatment of phenylketonuria (PKU). For additional information, please visit www.BMRN.com. Information on BioMarin's website is not incorporated by reference into this press release.

Forward-Looking Statement

This press release contains forward-looking statements about the business prospects of BioMarin Pharmaceutical Inc., including, without limitation, statements about: financial projections, including revenue expectations for Aldurazyme and Naglazyme; the development and commercialization of Naglazyme and Phenoptin; and actions by regulatory and governmental authorities. These forward-looking statements are predictions and involve risks and uncertainties such that actual results may differ materially from these statements. These risks and uncertainties include, among others: possible delays in launching Naglazyme in the E.U. and slow market penetration in the U.S. and E.U.; actions by governmental authorities related to the reimbursement of Naglazyme; the content and timing of decisions by the FDA and European Commission and other regulatory authorities concerning Naglazyme and Phenoptin; issues or complications associated with post-marketing commitments; the results of current and future clinical trials of Phenoptin; and those factors detailed in BioMarin's filings with the Securities and Exchange Commission, including, without limitation, the factors contained under the caption "Factors That May Affect Future Results" in BioMarin's 2004 Annual Report on Form 10-K and the factors contained in BioMarin's reports on Forms 10-Q and 8-K. Stockholders are urged not to place undue reliance on forward-looking statements, which speak only as of the date hereof. BioMarin is under no obligation, and expressly disclaims any obligation, to update or alter any forward-looking statement, whether as a result of new information, future events or otherwise.

Aldurazyme[®] is a registered trademark of BioMarin/Genzyme LLC.

Orapred[®] is a registered trademark of Medicis Pediatrics, Inc. and is used under license.

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