Contacts:

Investors
Joshua A. Grass
Director, Business Development & Finance
BioMarin Pharmaceutical Inc.
415.506.6777

Media
Susan Ferris
Manager, Corporate Communications
BioMarin Pharmaceutical Inc.
415.506.6701

For Immediate Release:

BioMarin Announces U.S. Launch of Naglazyme for MPS VI

Novato, CA, June 21, 2005 – BioMarin Pharmaceutical Inc. (Nasdaq and SWX: BMRN) announced today that Naglazyme™ (galsulfase) is now available in the United States as the first specific therapy approved for the treatment of individuals with mucopolysaccharidosis VI (MPS VI), a rare and life-threatening genetic disease. Naglazyme was approved on May 31, 2005 by the U.S. Food and Drug Administration (FDA) and was shipped to distributors on Monday, June 20. The drug is being shipped to physicians this week. Naglazyme was developed by BioMarin and will be commercialized by the company's U.S.-based sales force. Naglazyme is indicated for patients with MPS VI. Naglazyme has been shown to improve walking and stair-climbing capacity.

"With the commercial launch of Naglazyme, individuals with MPS VI now have an approved treatment option that specifically addresses the underlying cause of this progressive and debilitating disease," stated Jean-Jacques Bienaimé, Chief Executive Officer and Director of BioMarin. "Naglazyme is the second product developed by BioMarin to receive FDA approval and the first such product that we are commercializing on our own. We have prepared extensively for this moment and are optimistic that our preparation will result in a robust product launch."

In a separate release issued today, BioMarin announced the appointment of Stephen Aselage to the position of Senior Vice President, Global Commercial Operations, effective July 1, 2005. BioMarin's commercial team includes four medical science liaisons who will be responsible for providing support to infusion centers and physicians who administer Naglazyme. The BioMarin sales force has mounted a disease awareness campaign targeted toward pediatricians and sub-specialists who are involved in the multi-disciplinary care of patients with MPS disorders to help drive new patient identification. Additionally, BioMarin has launched BioMarin Physician and Patient Support (BPPS), a free and confidential service established to assist patients and caregivers in their effort to receive insurance reimbursement for treatment with Naglazyme.

Naglazyme is administered once weekly via intravenous infusion and is dosed at 1 milligram of drug per kilogram of body weight. The annual cost of treatment will vary considerably according to each patient's weight.

ATU Program Under Way in France

In advance of product approval, BioMarin has received temporary use authorization (ATU) from regulatory authorities in France for named patient basis sales. Three patients who met certain selection criteria have begun to receive Naglazyme treatment. BioMarin expects to book revenue from these named patient basis sales and will continue to assist patients in Europe wherever possible prior to product approval in the European Union.

About Naglazyme

Naglazyme is an enzyme replacement therapy for the treatment of MPS VI. As the first therapy approved for MPS VI, the FDA has granted Naglazyme orphan drug status, which confers seven years of market exclusivity.

Data from the Phase 3 clinical trial and extension study indicate that Naglazyme was generally safe. 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. Over 95 percent of the infusion-related adverse events were considered mild or moderate and were easily managed. Infusion-related adverse events commonly included fever, chills/rigors, headache, rash, and mild to moderate urticaria. Severe reactions included angioneurotic edema, hypotension, dyspnea, bronchospasm, respiratory distress, apnea, and urticaria. 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 adverse events or impact the improvements experienced in endurance. 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. Naglazyme is available by prescription only. Full prescribing information is available at www.naglazyme.com.

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. Enzyme replacement therapy with Naglazyme addresses the underlying cause of the disease by providing individuals with MPS VI the enzyme they are naturally lacking. An estimated 1,100 individuals in the developed world have MPS VI. The majority of individuals with MPS VI die from disease-related complications between childhood and early adulthood. Additional information about MPS VI can be found at www.mpsvi.com.

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 product 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 severe asthma. Investigational product candidates include Phenoptin™ (sapropterin hydrochloride), a Phase 3 product candidate for the treatment of phenylketonuria (PKU). For additional information, please visit www.BMRN.com.

Information on BioMarin's websites, including www.BMRN.com, www.mpsvi.com and www.naglazyme.com, 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: the development and commercialization of Naglazyme; expectations related to post-marketing commitments for Naglazyme; and actions by regulatory 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: slow market penetration following launch; the content and timing of decisions by the European Commission and other regulatory authorities concerning Naglazyme; issues or complications associated with post-

marketing commitments; 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.

# # #