BioMarin Provides Update on Phenylase Product Development

Important Technical Hurdles Overcome - IND Filing Targeted for Late 2007

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BioMarin Pharmaceutical Inc. (Nasdaq and SWX: BMRN) announced today that investigators presented data from preclinical studies of Phenylase(TM) (phenylalalanine ammonia lyase) that suggest the company has successfully addressed key technical challenges associated with its development for the treatment of phenylketonuria (PKU). BioMarin plans to conduct additional preclinical studies of Phenylase, with the intention of filing an Investigational New Drug application (IND) with the U.S. Food and Drug Administration in late 2007.

"Data from preclinical studies of Phenylase suggest that we have successfully addressed important technical hurdles required for the development of a potentially safe and effective treatment for individuals with PKU," stated Emil Kakkis, M.D. Ph.D., Chief Medical Officer of BioMarin. "The data suggest that we have a molecule with the essential characteristics required for clinical development, and we expect to file an Investigational New Drug application in late 2007."

Preclinical data demonstrate that Phenylase administered once weekly via subcutaneous injection in a PAH(enu2) murine model of PKU resulted in a sustained decrease in blood phenylalanine (Phe) to normal levels for a 12-week period. Baseline blood Phe levels ranging from 1500uM to 2000uM decreased to less than 100uM after treatment with Phenylase. Researchers observed restoration of pigmentation and increase in weight in the Phenylase-treatment groups relative to placebo groups. Antibodies did not have an impact on observed efficacy in either group, nor were there signs of systemic allergic reaction or local injection site reactions. These data were presented on Sunday, September 10, 2006, at a meeting entitled, "Tetrahydrobiopterin and Alternative Treatment in PKU. Cardiovascular Disease, and Diabetes," held in conjunction with the International Congress on Inborn Errors of Metabolism taking place this week in Chiba, Japan. The studies were conducted in collaboration with researchers in the laboratory of Charles Scriver, M.D., at McGill University, located in Montreal, Quebec and the laboratory of Ray Stevens, Ph.D., at The Scripps Research Institute, located in La Jolla, California.

About Phenylase

Phenylase is an investigational enzyme substitution therapy currently being evaluated in preclinical studies for the treatment of severe PKU. The active ingredient in Phenylase, PEGylated phenylalanine ammonia lyase, is designed to catabolize the Phe that builds-up in the body due to lack of activity of the enzyme phenylalanine hydroxylase. BioMarin is seeking to develop Phenylase as a once-weekly, self-administered subcutaneous injection.

Phenylase is one of two investigational product candidates currently being evaluated by BioMarin for the treatment of PKU. The second, Phenoptin(TM) (sapropterin dihydrochloride), is a small-molecule oral therapeutic currently in Phase 3 clinical development. Phenylase and Phenoptin are both partnered with Serono S.A. as part of a strategic alliance established in May 2005.

About PKU

PKU, a genetic disorder affecting approximately 50,000 diagnosed patients in the developed world, is caused by a deficiency of the enzyme phenylalanine hydroxylase (PAH). PAH is required for the metabolism of phenylalanine (Phe), an essential amino acid found in most protein-containing foods. If the active enzyme is not present in sufficient quantities, Phe accumulates to abnormally high levels in the blood and brain, resulting in a variety of complications including severe mental retardation and brain damage, mental illness, seizures and tremors, and cognitive problems. As a result of global newborn screening efforts implemented in the 1960s and early 1970s, virtually all PKU patients in developed countries have been diagnosed at birth. The only treatment currently available for PKU patients is a highly restrictive and expensive medical food diet that most patients fail to adhere to the extent needed for achieving adequate control of blood Phe levels. To learn more about PKU, please visit www.PKU.com. Information on this website is not incorporated by reference into this press release.

About BioMarin

BioMarin develops and commercializes innovative biopharmaceuticals for serious diseases and medical conditions. The company's product portfolio is comprised of two approved products and multiple clinical and preclinical product candidates. Approved products include Naglazyme(R) (galsulfase) for mucopolysaccharidosis VI (MPS VI), a product wholly developed and commercialized by BioMarin, and Aldurazyme(R) (laronidase) for mucopolysaccharidosis I (MPS I), a product which BioMarin developed through a 50/50 joint venture with Genzyme Corporation. Investigational product candidates include Phenoptin(TM) (sapropterin dihydrochloride), a Phase 3 product

candidate for the treatment of phenylketonuria (PKU), and 6R-BH4 for cardiovascular indications, which is currently in Phase 2 clinical development for the treatment of poorly controlled hypertension. 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: the development of its product candidates Phenoptin and Phenylase for the treatment of PKU; and expectations regarding filings with regulatory agencies. 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: the results of preclinical and clinical trials related to Phenoptin and Phenylase; results and timing of current and planned clinical trials of Phenoptin for the treatment of PKU and; the content and timing of decisions by the U.S. Food and Drug Administration, the European Medicines Agency and other regulatory authorities; and those factors detailed in BioMarin's filings with the Securities and Exchange Commission, including, without limitation, the factors contained under the caption "Risk Factors" in BioMarin's 2005 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 statements.

NOTE: Naglazyme(R) is a registered trademark of BioMarin Pharmaceutical Inc.

Aldurazyme(R) is a registered trademark of BioMarin/Genzyme LLC.

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