BioMarin Initiates Phase 1 Clinical Study of PEG-PAL in PKU

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BioMarin Pharmaceutical Inc. (Nasdaq and SWX: BMRN) today announced that the first patient has initiated treatment in the Phase 1 clinical study of PEG-PAL (PEGylated recombinant phenylalanine ammonia lyase) for the treatment of phenylketonuria (PKU). The study is expected to conclude enrollment in the fourth quarter of 2008.

"We are dedicated to serving the PKU community and hope to address the entire spectrum of PKU patients between PEG-PAL and Kuvan. We believe PEG-PAL holds tremendous potential to bring blood Phe down to normal levels and may help patients who either do not respond to Kuvan or who wish to reduce blood Phe levels beyond what is possible with Kuvan," said Jean-Jacques Bienaime, Chief Executive Officer of BioMarin. "Positive preclinical data shows sustained decreases in blood Phe levels in PKU mice without any notable safety concerns and we hope to see the same results in humans."

The Phase 1 clinical trial is an open-label, multi-center study to be conducted in approximately 35 PKU patients in a series of seven dose- escalating cohorts ranging from 0.001 to 1.0 mg/kg, with each cohort receiving a single dose, and a 6-week follow-up period.

The primary objective of the study is to assess the safety and tolerability of single, subcutaneous injections of PEG-PAL in subjects with PKU. The secondary objectives of the study are to evaluate the pharmacokinetics of single, subcutaneous injections of PEG-PAL administered at escalating doses and to evaluate the effect of PEG-PAL on blood Phe concentrations in subjects with PKU.

About PEG-PAL

PEG-PAL (PEGylated recombinant phenylalanine ammonia lyase) is an investigational enzyme substitution therapy for the treatment of PKU. Pharmacology studies conducted in the PKU mouse model demonstrated that weekly subcutaneous administrations of PEG-PAL resulted in a significant and stable decrease of plasma phenylalanine. BioMarin estimates that PEG-PAL could be a potential treatment option for a significant portion of the PKU population.

About BioMarin

BioMarin develops and commercializes innovative biopharmaceuticals for serious diseases and medical conditions. The company's product portfolio comprises three 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; Aldurazyme(R) (laronidase) for mucopolysaccharidosis I (MPS I), a product which BioMarin developed through a 50/50 joint venture with Genzyme Corporation; and Kuvan(R) (sapropterin dihydrochloride) Tablets, a product for the treatment of phenylketonuria (PKU), developed in partnership with Merck Serono, a division of Merck KGaA of Darmstadt, Germany. Other product candidates include 6R-BH4 for cardiovascular indications, which is currently in Phase 2 clinical development for the treatment of peripheral arterial disease and sickle cell disease, and PEG-PAL (PEGylated recombinant phenylalanine ammonia lyase), which is currently in Phase 1 clinical development for the treatment of PKU. For additional information, please visit http://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 candidate PEG-PAL, 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 current and planned clinical trials related to PEG-PAL; the content and timing of decisions by the U.S. Food and Drug Administration and other regulatory agencies, particularly with respect to PEG-PAL, 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 2007 Annual Report on Form 10-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.

BioMarin(R), Naglazyme(R) and Kuvan(R) are a registered trademarks of BioMarin Pharmaceutical Inc.

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

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