BioMarin Announces Results From Phase 2 Clinical Study of 6R-BH4 in Poorly Controlled Hypertension

No Significant Difference Observed Between 6R-BH4 and Placebo

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NOVATO, Calif.

BioMarin Pharmaceutical Inc. (Nasdaq and SWX: BMRN) today announced results from its Phase 2a placebo-controlled double-blind study of 6R-BH4 in patients with poorly controlled hypertension. Results demonstrate that there was no statistically significant or clinically meaningful effect of 6R-BH4 on any efficacy or safety parameter measured, relative to placebo.

Jean-Jacques Bienaime, Chief Executive Officer of BioMarin stated, "We are surprised and disappointed by these results, especially considering the numerous encouraging pre-clinical and clinical studies of 6R-BH4 in diseases with endothelial dysfunction. We plan to analyze the data in detail to better understand the background therapy and other characteristics of patients in the study. We have no immediate plans to change the course of ongoing or planned clinical studies of 6R-BH4."

Mr. Bienaime continued, "We remain on track to file our new drug application for Phenoptin in PKU next quarter. While 6R-BH4 is the active ingredient in Phenoptin, this development has no effect on our program for Phenoptin in PKU. 6R-BH4 works by an entirely different mechanism of action and metabolic pathway in PKU, and the safety data in the hypertension study did not change the safety profile of the drug."

Study Design

The 8-week multi-center, randomized, double-blind, placebo-controlled study enrolled 116 patients with poorly controlled systemic hypertension, approximately half with type 2 diabetes. Among other eligibility criteria, to participate in the study, patients had elevated blood pressure while on at least two different medications for hypertension. Study patients received oral doses of 5 mg/kg of 6R-BH4 or placebo twice daily for an eight-week period.

Primary Efficacy Endpoint Results

Patients receiving placebo experienced a 6.4 mm Hg drop in systolic blood pressure compared to a drop of 4.8 mm Hg for patients receiving 6R-BH4. The difference was not statistically significant.

Conference Call Information

BioMarin will hold a conference call today, February 20, 2007, at 11:00 a.m. ET to discuss the results of the Phase 2 study of 6R-BH4 in poorly controlled hypertension and fourth quarter and year-end 2006 financial results. This event can be accessed on the investor section of the BioMarin website at http://www.bmrn.com/.

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Date: February 20, 2007
Time: 11:00 a.m. ET
U.S. and Canada Toll-Free Dial in #: 800.901.5217
International Dial in #: 617.786.2964
Participant Code: 35915200
Replay Toll-Free Dial in #: 888.286.8010
Replay International Dial in #: 617.801.6888
Replay Code: 94899251

About 6R-BH4
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6R-BH4, commonly known as BH4 or tetrahydrobiopterin, 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 and produces multiple positive effects, such as relaxing smooth muscle, reducing blood pressure, controlling inflammation and reducing platelet aggregation. Researchers have demonstrated that a deficiency of BH4 can disrupt NO synthesis, resulting in a loss of normal endothelial NO production. This loss of endothelial NO production, commonly referred to as endothelial dysfunction, has been associated with many cardiovascular diseases, including hypertension, diabetic vascular disease, peripheral arterial disease, coronary arterial disease and pulmonary hypertension, and has been shown to be a strong predictor of cardiovascular adverse events in a number of clinical studies.

6R-BH4 is the same enzyme cofactor currently being evaluated in BioMarin's Phenoptin(TM) (sapropterin dihydrochloride) for phenylketonuria (PKU). In March 2006, BioMarin and Merck Serono (a division of Merck KGaA, Darmstadt, Germany), BioMarin's corporate partner for the Phenoptin and 6R-BH4 programs, announced positive results from the Phase 3 clinical study of Phenoptin for PKU. All primary and secondary endpoints of the study were met. The type and incidence of adverse events was similar in the Phenoptin and placebo groups. Phenoptin was well tolerated and investigators reported that no serious adverse event occurred.

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 peripheral arterial disease. 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 Statements

This press release contains forward-looking statements about the business prospects of BioMarin Pharmaceutical Inc., including, without limitation, statements about: the final results of the Phase 2 clinical study of 6R-BH4 in poorly controlled hypertension; the timing of BioMarin's clinical trials of Phenoptin and 6R-BH4 for other indications; the continued clinical development of 6R-BH4; expectation regarding regulatory filings for Phenoptin; and actions by regulatory authorities, including actions related to 6R-BH4. 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 final analysis of the data from the Phase 2 clinical study of 6R-BH4 in poorly controlled hypertension; results and timing of current and planned preclinical studies and clinical trials; actions related to Phenoptin; the content and timing of decisions by the U.S. Food and Drug Administration, the European Commission and other regulatory authorities concerning each of the described product candidates; 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, as amended, and the factors contained in BioMarin's reports on Form 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.

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