BioMarin and La Jolla Pharmaceutical Sign Worldwide (Excluding Asia Pacific) Development and Commercialization Agreement for Riquent

Riquent is Currently Being Evaluated in a Phase 3 Clinical Study for Lupus Nephritis, A High Unmet Need and Orphan Disease
BioMarin Conference call to be held on January 6 at 5:00 PM EST and La Jolla Conference call to be held on January 7 at 8:00 AM EST.

PRNewswire-FirstCall
NOVATO, Calif. and SAN DIEGO

BioMarin Pharmaceutical Inc. and La Jolla Pharmaceutical Company announced today that they have entered into an agreement to develop and commercialize Riquent(R), La Jolla's investigational drug for lupus nephritis, in the United States, Europe and all other territories of the world, excluding the Asia Pacific region. Following a successful Phase 3 trial, the parties will share equally in all losses and profits. In the United States, BioMarin and La Jolla will jointly commercialize Riquent. In Europe and other territories outside of Asia, BioMarin will be responsible for all commercialization activities.

Jean-Jacques Bienaime, Chief Executive Officer of BioMarin commented, "We are very pleased to become La Jolla Pharmaceutical's partner for Riquent. The development history of Riquent has been long and challenging, but we feel the current study addresses the shortcomings of prior studies and provides the best possible opportunity to demonstrate that Riquent reduces the frequency of renal flares in lupus nephritis patients. The ASPEN Phase 3 study is the largest clinical study ever conducted in lupus nephritis and there is increasing evidence that Riquent targets one of the most important underlying causes of kidney disease in lupus patients, antibodies to double stranded DNA.

Mr. Bienaime continued, "This product opportunity also represents an exceptionally good strategic fit for BioMarin. Lupus nephritis is a serious and potentially fatal orphan disease treated by specialists, primarily nephrologists and rheumatologists, and there are no products specifically approved to treat lupus renal disease. Importantly, if the ASPEN study is successful, Riquent provides BioMarin the opportunity to launch a product in the 2010 and 2011 timeframe in regions where we can leverage existing commercial operations."

"BioMarin is a proven leader in successfully bringing value-added therapies to patients suffering from orphan diseases and we are very pleased to have BioMarin as a partner to develop and commercialize Riquent in the U.S., Europe, the Middle East and Latin America," said Deirdre Y. Gillespie, M.D., President and CEO of La Jolla Pharmaceutical Company. "This is an outstanding partnership for us as it not only provides significant near term funding towards the completion of the ASPEN trial but also facilitates La Jolla's plans to build a U.S. commercial infrastructure going forward. 2009 is a pivotal year for La Jolla and we are pleased to start the year with this positive announcement. We look forward to the first interim analysis of the ASPEN trial data which is expected to occur later in this quarter," continued Dr. Gillespie.

Overview of Deal Terms

Under the terms of the agreement, BioMarin will receive a co-exclusive license to develop and commercialize Riquent and La Jolla could receive up to $289 million in cash through milestones and equity purchases by BioMarin. Specific payments include: $15 million upfront, up to approximately $92.5 million related to clinical milestones, $55 million for regulatory milestones, and up to approximately $126 million for achieving specified annual net sales milestones beginning at $250 million in sales.

At each of the two interim efficacy analyses (the first expected in the first quarter of 2009 and the second expected in mid-2009) or when the ASPEN study comes to a successful completion, BioMarin may exercise its option to fully participate and share all losses and profits on a 50:50 basis. Prior to BioMarin's decision to participate fully, La Jolla will fund 100% of all costs. La Jolla expects consideration from the deal to significantly cover the remaining costs of the Phase 3 ASPEN study. The collaboration also provides La Jolla an ability to participate equally in sales and marketing responsibilities in the United States to facilitate building its commercial infrastructure. La Jolla will maintain primary manufacturing responsibility, and work collaboratively with BioMarin to maximize supply chain and process efficiencies.

Upfront Payment

Upon signing, BioMarin will pay La Jolla a total upfront payment of $15 million, $7.5 million in cash and $7.5

million for the purchase of 3,391,035 preferred shares at a price per share of $2.21171. The preferred shares are initially convertible at a rate of three shares of common stock for every one preferred share. This is equivalent to a common stock purchase price of $0.73724, a 20% premium to the average closing price for La Jolla's common stock over the previous 20 trading days.

Clinical Milestones

Depending on the outcomes (non-futile or achievement of p-value, p<0.001) of two predefined interim efficacy analyses in the Phase 3 ASPEN study as well as the complete Phase 3 clinical results, BioMarin will pay La Jolla up to an additional $47.5 to $92.5 million in clinical milestone and full participation payments prior to approval. BioMarin may apply up to $20.0 million of these pre-approval clinical milestones toward additional purchases of La Jolla preferred stock. If the first interim efficacy analysis results in a non-futile determination by the Data Monitoring Board, BioMarin will pay a milestone of $15 million to maintain its license option. If the second interim efficacy analysis results in a non-futile determination by the Data Monitoring Board, BioMarin will pay a milestone of $22.5 million to continue its license option, $5 million of which may be used to purchase additional equity.

Riquent is being evaluated in the international Phase 3 ASPEN trial designed to demonstrate that Riquent treatment delays the time to renal flare and reduces proteinuria in patients with lupus renal disease. The Riquent Phase 3 program is the subject of a special protocol assessment and has fast track designation from the Food and Drug Administration and Orphan Drug designation in the United States and Europe. The first and the second interim efficacy analysis are expected to occur in the first quarter of 2009 and mid- 2009, respectively. The final efficacy analysis is expected to occur in the second half of 2009. Assuming a positive outcome of the ASPEN Phase 3 trial, a New Drug Application is expected to be submitted in the first half of 2010. If approved, Riquent would be the first new drug approved specifically for lupus in more than 45 years.

BioMarin Conference Call Information

BioMarin will hold a conference call today, January 6, 2009, at 5:00 p.m. ET to discuss this announcement. This event can be accessed on the investor section of the BioMarin website at http://www.bmrn.com/.

Date: January 6, 2009
Time: 5:00 p.m. ET
U.S. and Canada Toll-Free Dial in #: 866.700.7477
International Dial in #: 617.213.8840
Participant Code: 66691013
Replay Toll-Free Dial in #: 888-286-8010
Replay International Dial in #: 617-801-6888
Replay Code: 82654807

La Jolla Conference Call information

La Jolla will hold a conference call tomorrow morning, January 7, 2009, at 8:00 a.m. ET to discuss this announcement. This event can be accessed on the La Jolla website at http://www.ljpc.com/. A replay of the conference call will be available later in the day of the call on La Jolla's Web site http://www.ljpc.com/ and will be archived for several weeks. In addition, a replay of the conference call can be accessed by dialing 888-286-8010 (US) or 617-801-6888 (international). The passcode for the replay is 20790213.

About Lupus Nephritis

Lupus nephritis is a life-threatening, antibody-mediated disease and is characterized by periods of extreme, acute inflammation or renal flares which often require treatment with high-dose corticosteroids, immunosuppressive agents and hospitalization. Over time, lupus nephritis can lead to deterioration of kidney function and end-stage kidney disease, requiring long-term renal dialysis or kidney transplantation, and often results in morbidity and mortality.

About Riquent

Riquent is being developed to specifically treat lupus renal disease by preventing or delaying renal flares, a leading cause of sickness and death in lupus patients. It is also being studied to assess whether Riquent treatment improves proteinuria, as was observed in previous clinical trials. Proteinuria is an indicator of abnormal renal function. Riquent has been well tolerated in all 14 clinical trials, with no overall difference in the adverse event profiles for Riquent-treated patients compared with placebo-treated patients. Riquent specifically reduces circulating levels of anti-dsDNA antibodies and is also designed to specifically suppress the B cells that make these antibodies. Decreases in these antibodies are believed to be associated with a decreased risk of renal flare. Although clinical benefit has not yet been proven, Riquent treatment has significantly reduced these antibody levels in all clinical trials in which they were measured.

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 pre-clinical 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.

About La Jolla Pharmaceutical Company

La Jolla Pharmaceutical Company is dedicated to improving and preserving human life by developing innovative pharmaceutical products. The Company's leading product in development is Riquent(R), which is designed to treat lupus renal disease by preventing or delaying renal flares. Lupus renal disease is a leading cause of sickness and death in patients with lupus. The Company has also developed potential small molecule drug candidates to treat various other autoimmune and inflammatory conditions. The Company's common stock is traded on The NASDAQ Global Market under the symbol LJPC. More information about the Company is available on its Web site: http://www.ljpc.com/.

This press release contains forward-looking statements, which involve significant risks, assumptions and uncertainties, and a number of factors, both foreseen and unforeseen, that could cause actual results to differ materially from BioMarin and La Jolla's current expectations. Forward-looking statements include those that express a plan, belief, expectation, estimation, anticipation, intent, contingency, future development or similar expression. There can be no assurance that actual results will be consistent the expectations reflected in BioMarin and La Jolla's forward looking statements. For example, there can be no assurance that: the ASPEN trial will be successful or that Riquent will be approved for marketing; the ASPEN trial will proceed on schedule as currently planned; La Jolla will receive any contingent payments under the license agreement with BioMarin or even if Riquent is approved, that it will be successfully marketed. These or other risks are discussed under the caption "Risk Factors" in BioMarin and La Jolla's most recent Annual Reports on Form 10-K and subsequent Quarterly Reports on Form 10-Q, as filed with the SEC. Accordingly, you should not rely upon forward-looking statements as predictions of future events. We expressly disclaim any intent to update forward-looking statements.

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