June 7, 2007
AnGes MG, Inc.

Announcement Regarding the Official Designation of Naglazyme as an Orphan Drug

The Ministry of Health, Labour and Welfare (MHLW), Japan, officially designated Naglazyme (galsulfase) for the treatment of mucopolysaccharidosis VI as an orphan drug on June 5, 2007.

Naglazyme is a drug for which AnGes MG, Inc. has been licensed by BioMarin Pharmaceutical Inc. (USA) regarding the rights for its development and distribution in Japan. This drug was developed as a means of enzyme replacement therapy, i.e., replenishment of the enzyme deficient in patients with mucopolysaccharidosis VI. At present, hematopoietic stem cell transplant is available as a means of treating mucopolysaccharidosis type VI. However, since hematopoietic stem cell transplant is frequently limited by the difficulty in finding an appropriate donor and the risks associated with the procedure, a new treatment that is safer and efficacious has been sought. In the USA and Europe, Naglazyme is already marketed. In Japan, patient advocacy groups and medical societies have shown a strong interest in obtaining access to Naglazyme for Japanese patients. AnGes MG is preparing to submit an application for approval of this drug to the MHLW, using the clinical data collected in the USA and Europe.

The orphan drug designation system is aimed at stimulating the research and development of drugs which are expected to be useful for the treatment of intractable but relatively rare diseases and which are keenly needed for the care of such conditions.

Requirements for orphan drug designation are specified in the Pharmaceutical Affairs Law, and include: (1) the number of target patients less than 50,000, (2) high needs from the viewpoint of healthcare, and so on. Drugs designated as orphan drugs are favorably treated by the government, including financial support to R & D, preferential examination for approval, extended period until re-examination, and so on.

<Reference >

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. The majority of individuals with MPS VI die from disease-related complications between childhood and early adulthood.

About Naglazyme

Naglazyme is the first and only enzyme replacement therapy indicated for the treatment of MPS VI. As the first drug approved for MPS VI, regulatory agencies in both the United States and European Union have granted Naglazyme orphan drug status, which confers seven years and 10 years of market exclusivity, respectively.

About BioMarin Pharmaceutical Inc.

Head Office: 105 Digital Drive, Novato, CA 94949, USA
CEO: Jean-Jacques Bienaimé
Founded: 1997
Number of Employees: 410 (as of Feb. 2007)
Business: Manufacture, develop and commercialize drugs.
Marketed Products:

Aldurazyme (laronidase) for MPS I
Naglazyme (galsulfase) for MPS VI