January 22, 2007 AnGes MG, Inc.

# AnGes MG, Inc. Announces Marketing and Distribution Agreement with BioMarin Pharmaceutical Inc. for Naglazyme in Japan

AnGes MG, Inc. ("AnGes") announced today that they have entered into a Marketing and Distribution Agreement with BioMarin Pharmaceutical Inc. ("BioMarin") granting AnGes exclusive rights to market and distribute Naglazyme<sup>™</sup> (galsulfase) for patients with the genetic disease mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome) in Japan.

Naglazyme<sup>™</sup> (galsulfase) is a normal variant form of arylsulfatase B (IUPAC name N-acetylgalactosamin 4-sulfatase) that is lacking in mucopolysaccharidosis VI patients. It is produced by recombinant DNA technology at BioMarin. The U.S. Food and Drug Administration (FDA) approved the therapy in May 2005 and subsequently, the European Commission (EC) approved Naglazyme<sup>™</sup> in January 2006. Naglazyme<sup>™</sup> is a replacement therapy that is intended to provide exogenous enzyme and the clinical trials and usage experience thereafter have proved its efficacy for MPS VI.

Mucopolysaccharidosis VI is a very rare autosomal recessive metabolic disease. Due to the deficiency of arylsulfatase B, the normal breakdown of dermatan sulfate and chondroitin sulfate is blocked and subsequently, accumulate in the body. It is a progressive disease that manifests in reduced joint range of motion, bone deformities one year after birth, as well as hepatosplenomegaly, cloudy cornea, impaired hearing, cardiac abnormalities and valvular disease etc. However, since the application of the current treatment, 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.

Patient advocacy groups and medical societies have shown a strong interest in obtaining access to Naglazyme<sup>™</sup> for Japanese patients, as highlighted at the "Special Committee for Unapproved Products" held by Health, Labour and Welfare Ministry in Japan. AnGes will immediately be focused on securing approval and reimbursement for Naglazyme in order to provide treatment for patients with the severe genetic disease.

AnGes will pay upfront and will make milestone payments to BioMarin. However, those financial impacts are minimum.

## <Reference>

### **About BioMarin Pharmaceuticals**

Head quarter: 105 Digital Drive, Novato, CA 94949, U.S.A.
CEO: Jean-Jacques Bienaimé
Founded : 1997
Number of employees : Approx. 400
Business : Manufacture, develop and commercialize drugs.
Marketed Products :

Naglazyme® (galsulfase) for MPS VI

- Aldurazyme® (laronidase) for MPS I

### 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.