AnGes MG, Inc.

AnGes MG, Inc. Receives Approval for Manufacturing and Marketing of Naglazyme

AnGes MG, Inc. (AnGes) announced that on March 28, the company has received approval of

Naglazyme (galsulfase) for patients with the genetic disease mucopolysaccharidosis VI (MPS VI;

Maroteaux-Lamy syndrome). Naglazyme is the first product for which AnGes has received

approval.

AnGes established a marketing and distribution agreement with BioMarin Pharmaceutical Inc. in

December 2006, through which AnGes obtained exclusive rights to market Naglazyme in Japan.

Naglazyme was developed to supplement the deficient enzyme in MPS VI by providing an

exogenous enzyme in a therapeutic approach known as enzyme replacement therapy (ERT).

Although hematopoietic stem cell transplant is an option for MPS VI patients, there are issues such

as finding appropriate donors and risks of transplantation; thus, there is a need for a safer and more

effective treatment. Naglazyme has been already sold in the US and Europe. In Japan, patient

advocacy groups and academic societies are strongly calling for the early use of Naglazyme.

AnGes plans to launch Naglazyme as soon as possible after NHI price listing.

Details of approval are as follows,

Brand name:

Naglazyme Intravenous Infusion 5mg

Generic name:

Galsulfase (Genetical Recombination)

Indication:

MPS VI

Drug dosage

1mg/kg of body weight administered once weekly as an intravenous infusion

## <Reference>

## About BioMarin Pharmaceutical Inc.

Headquarters: 105 Digital Drive, Novato, CA 94949, U.S.A.

CEO: Jean-Jacques Bienaimé

Founded: 1997

Number of employees: 520 (as of Jan. 2008)

Business: Manufacture, develop and commercialize drugs

Marketed Products:

Aldurazyme (laronidase) for MPS I

Naglazyme (galsulfase) for MPS VI

Kuvan (sapropterin dihydrochloride) for phenylketonuria (PKU)

## **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.