



AMICUS THERAPEUTICS FILES INVESTIGATIONAL NEW DRUG APPLICATION FOR AT2101 FOR GAUCHER DISEASE

Phase I Clinical Trials Expected to Commence in June

Cranbury, NJ, APRIL 27, 2006 - Amicus Therapeutics, a biopharmaceutical company developing small molecule, orally-administered pharmacological chaperones for the treatment of human genetic diseases, today announced that it has submitted an investigational new drug application (IND) to the U.S. Food and Drug Administration (FDA) for AT2101 for the treatment of Gaucher disease. Pending FDA review of the IND, the company plans to begin Phase I clinical trials in June.

Gaucher disease is a lysosomal storage disorder that affects approximately 10,000 people in the developed world. Symptoms can be severe and debilitating, including an enlarged liver and spleen, abnormally low levels of red blood cells and platelets, and skeletal disease. In rare cases there can be significant impairment of the central nervous system. Gaucher disease is caused by a deficiency in the enzyme β -glucocerebrosidase (GCase) resulting from an inherited genetic mutation. In most cases, this mutation causes GCase to be misfolded and unable to perform its normal function.

AT2101, taken orally, is designed to act as a pharmacological chaperone that binds to and stabilizes the misfolded GCase enzyme, which allows it to be transported to the lysosome where it is active and performs its normal function. In pre-clinical tests, AT2101 has shown the ability to increase GCase activity in cells with a variety of different mutations that cause Gaucher disease, including the most common type of mutation, which accounts for more than 50% of the patient population.

"Our patient organization looks forward to working with Amicus to ensure that the potential of this therapy is evaluated in human clinical trials", said Rhonda Buyers, Executive Director, National Gaucher Foundation.

AT2101 for Gaucher disease is the second Amicus product to enter clinical trials. The company's lead compound, Amigal™ (migalastat hydrochloride), is a pharmacological chaperone in Phase II clinical trials for Fabry disease.

In January 2006, Amicus received orphan drug designation for AT2101 from the FDA's Office of Orphan Products Development.

About Amicus Therapeutics

Amicus Therapeutics is a biopharmaceutical company based in Cranbury, New Jersey, developing small molecule, orally-administered pharmacological chaperones for the treatment of human genetic diseases. Many of these diseases are the result of missense and other genetic errors that cause the misfolding and degradation or accumulation of a particular protein. Amicus' products act as pharmacological chaperones that selectively bind and "rescue" the misfolded target protein to restore its proper conformation and natural function, which in turn restores the function of the affected cells. Amicus' lead compound Amigal™ is in Phase II clinical trials for Fabry disease. The company has filed an IND for AT2101 for Gaucher disease, and is developing programs for a range of genetic diseases.