PHASE I CLINICAL STUDIES DEMONSTRATE SAFETY AND INITIAL "PROOF OF CONCEPT" FOR AMICUS' FABRY DISEASE DRUG

Lead Compound Based on Unique Pharmacological Chaperone Technology to Correct Misfolded Proteins Demonstrates Positive Biological Activity in Healthy Volunteers

Amicus Initiates Phase II Clinical Program

Cranbury, NJ, September 8, 2005 - Amicus Therapeutics, a biopharmaceutical company developing small molecule, orally-active pharmacological chaperones for the treatment of human genetic diseases, today announced positive results from Phase I clinical studies of its lead compound Amigal™ (migalastat hydrochloride) that is being developed using an entirely new approach for the treatment of Fabry disease. Based on these results in healthy volunteers, the company has commenced a Phase II clinical program to study Amigal in patients with Fabry disease. The Amicus executive team discussed the findings at the 42nd Annual Symposium of the Society for the Study of Inborn Errors of Metabolism held this week in Paris.

Amicus' technology represents a unique approach to treating the enzyme deficiency (alpha galactosidase A) that characterizes Fabry disease. Amigal acts as a pharmacological chaperone that restores natural function to the misfolded target protein - in this case the defective enzyme. In the Phase l studies, administration of Amigal to 16 healthy volunteers resulted in the following:

- Zero drug related adverse events
- High bioavailability in an oral formulation, and
- A favorable pharmacokinetic profile as measured by multiple parameters.

Notably, administration of Amigal also resulted in a statistically significant and dose-dependent increase in the activity of the target enzyme in each of the healthy volunteers who took part in the study,

"These exciting results are the first proof of concept for Amicus' unique pharmacological chaperone technology that offers the potential to dramatically improve treatment options for patients with genetic diseases," said Michel Bouvier, Ph.D., professor and director of biochemistry at the University of Montreal and a member of Amicus' Scientific Advisory Board. "The demonstration of increased enzyme activity in the healthy volunteers is intriguing. If these data are replicated, pharmacological chaperones may be applicable to therapeutic indications beyond the specific genetic disorders targeted today."

"This is a potentially groundbreaking advance that may transform the way we treat a wide range of genetic diseases," said Arthur Horwich, M.D., professor of genetics and pediatrics at Yale University and a member of Amicus' Scientific Advisory Board. "This unique technology represents a paradigm-shifting approach to the management of genetic diseases. If these data are confirmed in further clinical studies of Fabry patients, Amicus' pharmacological chaperone technology could allow us to move beyond the blunt approach of trying to replace missing proteins, and instead use an oral therapy to marshal the patients' own natural resources to correct defects in their innate protein production mechanism."

Based on these positive results, Amicus has initiated a Phase ll clinical program for Amigal in Fabry disease. The Phase II program includes investigational sites in the United States, Europe and Latin America and is designed to study the safety and preliminary efficacy of Amigal in Fabry patients. The first patient was enrolled August 31, 2005 and preliminary results are expected in early 2006. Amicus is also proceeding with several other programs, including a novel small molecule treatment for Gaucher disease, AT2101.

About Fabry Disease

Fabry disease is an inherited genetic disease caused by diminished or absent levels of a key enzyme called a-galactosidase A (a-GAL). In most Fabry patients, missense mutations alter the structure of a-GAL, which results in the degradation of the protein before it is allowed to leave the endoplasmic reticulum. As a result, a-GAL is unable to reach the lysosome - the area of the cell where the enzyme does its work. Reduced or absent levels of a-GAL activity leads to the accumulation of a complex lipid called globotriaosylceramide (GL-3) in the affected cells. The accumulation of GL-3, as well as the increased cell burden caused by the misfolding and accumulation of the a-GAL protein, leads to disease in the central nervous system, heart, kidneys and skin. Patients with Fabry disease experience pain, clouded vision, kidney failure and have an increased risk of heart attack and stroke. Fabry disease affects approximately 5,000 people worldwide but recent evidence suggests that the

disease may be significantly under-diagnosed. Pharmacological chaperones offer several potential advantages over competing approaches to the treatment of genetic disorders, including oral delivery and the ability to increase enzyme activity in hard to reach areas like the central nervous system.

About Amicus Therapeutics

Amicus Therapeutics is a biopharmaceutical company based in Cranbury, New Jersey, developing small molecule, orally-active 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 an active drug development program for Gaucher disease and is developing programs for a range of genetic diseases.