Amicus Therapeutics Announces First Patient in Phase 2 Study for Pompe Disease

AT2220 Co-Administered with Enzyme Replacement Therapy

Second Phase 2 Pharmacological Chaperone-ERT Co-Administration Study

CRANBURY, NJ, US, December 1, 2011 – Amicus Therapeutics (Nasdaq: FOLD), a biopharmaceutical company at the forefront of developing therapies for rare diseases, today announced the initial infusion of the first subject in an open-label Phase 2 drug-drug interaction study (Study 010) of AT2220 (duvoglustat HCl) co-administered with enzyme replacement therapy (ERT) in individuals with Pompe disease.

The purpose of Study 010 is to evaluate whether AT2220, an orally available, investigational pharmacological chaperone owned exclusively by Amicus, can be safely co-administered with the ERT alglucosidase alfa, the only approved therapy for Pompe disease. All subjects will be given a regularly scheduled ERT infusion. One hour prior to the initiation of the next ERT infusion, subjects will receive a single oral dose of AT2220. In Study 010, alglucosidase alfa will be measured in plasma and muscle tissue, with and without AT2220.

John F. Crowley, Chairman and Chief Executive Officer of Amicus Therapeutics said, “ERT is an important first generation treatment that has extended the lives of many individuals living with Pompe disease. We believe that chaperone-ERT co-administration has the potential to improve treatment outcomes for Pompe patients, and we are excited about commencing Study 010. Along with our work in Fabry, the co-administration approach may represent an important expansion of our technology platform into other lysosomal storage diseases where ERT is standard of care.”

In acid alpha-glucosidase (GAA) knock-out mouse models of Pompe disease, AT2220 co-administered with ERT increased ERT activity in plasma and uptake into key tissues, which corresponded with greater reductions in muscle glycogen, compared to ERT alone. Collectively these preclinical data highlight the potential for AT2220 to improve ERT in patients with Pompe disease.

“Results from Study 010, if positive, may form the basis for later stage studies that would allow us to evaluate the effect of AT2220 co-administered with ERT on glycogen reduction and ERT-related toxicity in patients with Pompe disease,” said Pol F. Boudes, Chief Medical Officer of Amicus.

Study Design

Study 010 is a Phase 2 open-label, multi-center study to evaluate the safety and pharmacokinetic (PK) effects of four increasing oral doses of AT2220 co-administered with ERT versus ERT alone in individuals with Pompe disease.

The study will enroll approximately 16 male or female subjects who have been on a stable dose and regimen of ERT for at least three months. All subjects will be given a regularly scheduled ERT infusion. One hour prior to the initiation of the next ERT infusion, subjects will receive a single oral dose of AT2220. Plasma enzyme activity and protein levels will be evaluated during each infusion. Muscle biopsies will be taken seven days after each infusion to measure tissue ERT activity with and without the chaperone, as well as the level of AT2220.

More information about Study 010, including patient eligibility, enrollment requirements and study location sites can be obtained by visiting www.clinicaltrials.gov: NCT1380743 or www.pompestudy.com, calling the patient hotline at 855-POMPE-33 (855-766-7333), or e-mailing inquiries to info@pompestudy.com.

About Amicus Therapeutics

Amicus Therapeutics (Nasdaq: FOLD) is a biopharmaceutical company at the forefront of developing therapies for rare diseases. The Company is developing orally-administered, small molecule drugs called pharmacological chaperones, a novel, first-in-class approach to treating a broad range of diseases including lysosomal storage disorders and diseases of neurodegeneration. Amicus' lead program Amigal™ (migalastat HCl) is in Phase 3 for the treatment of Fabry disease.

About AT2220 for Pompe Disease

AT2220 is an investigational, orally-administered pharmacological chaperone owned exclusively by Amicus. The Company is currently investigating AT2220 (duvoglostat HCl) co-administered with ERT in a Phase 2 study (Study 010) in individuals with Pompe disease.

Pompe disease is a lysosomal storage disease characterized by progressive skeletal muscle weakness and respiratory insufficiency. It is caused by a deficiency in GAA activity, which leads to accumulation of glycogen in tissues affected by the disease (primarily muscle). Pompe disease affects an estimated 5,000 to 10,000 individuals worldwide and is clinically heterogeneous in the age of onset, the extent of organ involvement, and the rate of progression.

Forward-Looking Statements

This press release contains “forward-looking statements” within the meaning of the Private Securities Litigation Reform Act of 1995 relating to clinical development of Amicus’ candidate drug products and the timing and reporting of results from clinical trials evaluating Amicus’ candidate drug products. Words such as, but not limited to, “look forward to,” “believe,” “expect,” “anticipate,” “estimate,” “intend,” “plan,” “targets,” “likely,” “will,” “would,” “should” and “could,” and similar expressions or words identify forward-looking statements. Such forward-looking statements are based upon current expectations that involve risks, changes in circumstances, assumptions and uncertainties. The inclusion of forward-looking statements should not be regarded as a representation by Amicus that any of its plans will be achieved. Any or all of the forward-looking statements in this press release may turn out to be wrong. They can be affected by inaccurate assumptions Amicus might make or by known or unknown risks and uncertainties. For example, with respect to statements regarding the goals, progress, timing and outcomes of

discussions with regulatory authorities and the potential goals, progress, timing and results of clinical trials, actual results may differ materially from those set forth in this release due to the risks and uncertainties inherent in the business of Amicus, including, without limitation: the potential that results of clinical or pre-clinical studies indicate that the product candidates are unsafe or ineffective; the potential that it may be difficult to enroll patients in our clinical trials; the potential that regulatory authorities may not grant or may delay approval for our product candidates; the potential that preclinical and clinical studies could be delayed because we identify serious side effects or other safety issues; the potential that we will need additional funding to complete all of our studies and, our dependence on third parties in the conduct of our clinical studies. Further, the results of earlier preclinical studies and/or clinical trials may not be predictive of future results. In addition, all forward looking statements are subject to other risks detailed in our Annual Report on Form 10-K for the year ended December 31, 2010. You are cautioned not to place undue reliance on these forward-looking statements, which speak only as of the date hereof. All forward-looking statements are qualified in their entirety by this cautionary statement, and Amicus undertakes no obligation to revise or update this news release to reflect events or circumstances after the date hereof. This caution is made under the safe harbor provisions of Section 21E of the Private Securities Litigation Reform Act of 1995.

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