

Amicus Therapeutics and GlaxoSmithKline Commence Second Phase 3 Study of Amigal™ for Fabry Disease

CRANBURY, NJ, US & LONDON, UK, September 12, 2011 – Amicus Therapeutics (Nasdaq: FOLD) and GlaxoSmithKline (GSK) today announced the first patient has commenced dosing in a Phase 3 global registration study ([Study 012](#)) to compare the safety and efficacy of Amigal™ (migalastat HCl) and enzyme replacement therapy (ERT) for the treatment of Fabry disease. The randomized, open-label, 18-month study will provide longer-term clinical data comparing migalastat HCl to ERT in patients with Fabry disease, a rare inherited lysosomal storage disorder.

Amicus and GSK are targeting up to 50 sites globally to enroll approximately 50 male and female Fabry patients who are currently receiving ERT treatment, and who have a genetic mutation that may be addressable with migalastat HCl. The primary outcome of efficacy will be renal function as measured by glomerular filtration rate (GFR).

John F. Crowley, Chairman and Chief Executive Officer of Amicus, stated, “In collaboration with GSK we are pleased to announce the dosing of the first patient in Study 012, the first Phase 3 pivotal study to compare Amigal to ERT. This is an important step in our clinical development plan and builds on the latest encouraging safety and renal function data from our ongoing Phase 2 extension study.”

Study 012 is the second of two Phase 3 studies intended to support the worldwide registration of migalastat HCl for Fabry disease. Amicus and GSK are also conducting a six-month, placebo-controlled Phase 3 study ([Study 011](#)) of migalastat HCl at 37 sites worldwide to support marketing applications for the U.S. Food and Drug Administration (FDA) and other regulatory agencies.

“We believe Amigal has the potential to provide an important treatment option in Fabry disease,” said Dr. Philippe Monteyne, Head of Development and Chief Medical Officer for GSK Rare Diseases. “We are delighted with the progress that the joint Amicus-GSK team has made to advance the clinical development program since we entered an alliance in October 2010.”

About Study 012 (Study AT1001-012)

[Study 012](#) is a randomized, open-label, 18-month Phase 3 study to compare the safety and efficacy of migalastat HCl (AT1001/GR181314A), and ERT in male and female patients with Fabry disease. The study was recently amended to randomize approximately 50 patients (30 to switch to migalastat HCl and 20 to remain on ERT). Eligible patients will have a genetic mutation that may be addressable with migalastat HCl.

Subjects who have been treated with either of the ERT preparations currently marketed, for at least 12 months, will be randomized on a 1.5:1 ratio to stop ERT and begin migalastat HCl treatment, or to continue receiving ERT treatment alone. Subjects in the migalastat HCl treatment arm will receive 150 mg of migalastat HCl every other day. Subjects in the ERT alone arm will continue on their current dose and regimen of ERT.

The primary outcome of efficacy will be renal function as measured by glomerular filtration rate (GFR) for the migalastat HCl and ERT groups at 18 months. The primary analysis will use descriptive statistics to compare the mean changes in GFR for each arm. Secondary outcomes of efficacy

include renal function as measured by 24-hour urine protein and other clinical outcomes. For more information please visit www.clinicaltrials.gov, study reference number NCT01218659.

About Migalastat HCl

Migalastat HCl is an orally-administered pharmacological chaperone in Phase 3 development for the treatment of Fabry disease. It has not been approved or licensed anywhere in the world. On October 29, 2010, Amicus announced a definitive agreement with Glaxo Group Limited (GSK) to develop and commercialize migalastat HCl. Under the terms of the agreement, GSK received an exclusive worldwide license to develop, manufacture and commercialize migalastat HCl.

In addition to the Phase 3 monotherapy studies (Study 011 and Study 012), Amicus and GSK are currently conducting a Phase 2 clinical study to evaluate migalastat HCl co-administered with ERT for the treatment of Fabry disease.

About Fabry Disease

Fabry disease is an inherited lysosomal storage disorder that is estimated to affect approximately 5,000 to 10,000 people worldwide. Fabry disease is caused by deficiency of an enzyme called alpha-galactosidase A (alpha-Gal A). The role of alpha-Gal A within the body is to break down a complex lipid called globotriaosylceramide (GL-3). Reduced or absent levels of alpha-Gal A activity leads to the accumulation of GL-3 in the affected tissues, including the kidneys, central nervous system, heart, and skin. This accumulation of GL-3 is believed to cause the various symptoms of Fabry disease, including pain, kidney failure, and increased risk of heart attack and stroke.

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 development for the treatment of Fabry disease.

About GlaxoSmithKline

GlaxoSmithKline - one of the world's leading research-based pharmaceutical and healthcare companies - is committed to improving the quality of human life by enabling people to do more, feel better and live longer. For further information please visit www.gsk.com

Amicus Forward-Looking Statements

This press release contains, and the accompanying conference call will contain, "forward-looking statements" within the meaning of the Private Securities Litigation Reform Act of 1995 relating to preclinical and clinical development of Amicus' candidate drug products, the timing and reporting of results from preclinical studies and clinical trials evaluating Amicus' candidate drug products, and the projected cash position for the Company, including achievement of development and commercialization milestone payments and sales royalties under our collaboration with GlaxoSmithKline. 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 preclinical studies and 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. With respect to statements regarding projections of the Company's cash position, actual results may differ based on market factors and the Company's ability to execute its operational and budget plans, including achievement of development and commercialization milestone payments and sales royalties under our collaboration with GlaxoSmithKline. 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.

GlaxoSmithKline cautionary statement regarding forward-looking statements

Under the safe harbor provisions of the U.S. Private Securities Litigation Reform Act of 1995, GSK cautions investors that any forward-looking statements or projections made by GSK, including those made in this announcement, are subject to risks and uncertainties that may cause actual results to differ materially from those projected. Factors that may affect GSK's operations are described under 'Risk Factors' in the 'Business Review' in the company's Annual Report on Form 20-F for 2010.

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