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Amicus Therapeutics and GlaxoSmithKline Expand Fabry Disease Collaboration

Companies Jointly to Develop Proprietary Enzyme Replacement Therapy (ERT) for Fabry Disease Co-Formulated With Chaperone Migalastat HCl

Fabry Products to be Commercialized by Amicus in U.S. and by GSK ex-U.S.

GSK to Increase Ownership in Amicus to 19.9% with \$18.6 Million Investment in Common Stock Priced at \$6.30 Per Share

CRANBURY, N.J. and LONDON, July 17, 2012 (GLOBE NEWSWIRE) -- Amicus Therapeutics (Nasdaq:FOLD) and Glaxo Group Limited (GSK) today announced an expansion of their collaboration to develop and commercialize the investigational pharmacological chaperone migalastat HCl for Fabry disease.

The expanded alliance comprises three components:

- Co-development of all current and future formulations of migalastat HCl for Fabry disease, including a co-formulation of migalastat HCl with GSK/JCR Pharmaceutical Co., Ltd's investigational enzyme replacement therapy (ERT) for Fabry disease;
- Commercialization arrangements for all future Fabry products. Amicus will have commercial rights to all Fabry products in the United States and GSK will commercialize all products in the rest of world; and
- Increased GSK ownership in Amicus with an \$18.6 million investment in common stock priced at \$6.30 per share, bringing GSK's total ownership stake in Amicus to 19.9%.

"We have strengthened our relationship with Amicus through the expanded Fabry collaboration and additional equity investment in the Company," said Marc Dunoyer, Global Head of GSK Rare Diseases and a member of the GSK Corporate Executive Team. "Amicus has a very successful track record as our development partner, and long-standing relationships with the Fabry community. We look forward to their leadership in the U.S. commercialization of now several potential medicines for patients with Fabry disease. This is an important step in our strategic vision, allowing us to undertake and fund an enlarged scientific program with a view to turning molecules into medicines for rare diseases faster and more effectively than ever before."

The global Fabry collaboration combines Amicus' U.S. presence, pharmacological chaperone development expertise, and established relationships in the rare and orphan disease community with GSK's global rare disease unit and worldwide regulatory, commercial, and manufacturing capabilities. Amicus and GSK are now committed to the parallel development of three different uses of migalastat HCl for Fabry disease:

- **Migalastat HCl monotherapy in Phase 3:** Phase 3 global registration studies ([Study 011](#) and [Study 012](#)) are currently underway in patients with genetic mutations that are amenable to chaperone monotherapy. Results from Study 011 are anticipated in the third quarter of 2012 to support a New Drug Application (NDA) submission to the U.S. Food and Drug Administration (FDA). If approved, Amicus will be responsible for the U.S. commercial launch.
- **Migalastat HCl co-administered with ERT in Phase 2:** A Phase 2 study of migalastat HCl co-administered with ERT for Fabry disease ([Study 013](#)) is currently ongoing. In January 2012, Amicus announced positive preliminary results from Study 013.
- **Migalastat HCl co-formulated with a proprietary preclinical ERT:** Amicus and GSK, in collaboration with Japan-based JCR, are developing migalastat HCl co-formulated with a proprietary recombinant human alpha-Gal A enzyme (JR-051). This ERT was developed by JCR and licensed to GSK for all markets outside Japan. Preclinical studies conducted by Amicus, GSK and JCR suggest that this co-formulated chaperone-ERT product may provide greater alpha-Gal A enzyme uptake into tissue and markedly reduced levels of GL-3 in Fabry disease-relevant tissues compared to recombinant enzyme alone. Amicus and GSK believe that this co-formulated chaperone-ERT product for Fabry disease has the potential to enter clinical studies in 2013. Further details of this program and preclinical results will be presented on today's conference call and webcast.

John F. Crowley, Chairman and Chief Executive Officer of Amicus said, "GSK has added significant value to the Fabry program through its global scale and capabilities as well as the dedicated focus of GSK Rare Diseases. Through our expanded

agreement, GSK is increasing its investment in the Fabry development program and Amicus is transforming into a commercial-stage biopharmaceutical company within the U.S. Amicus is leveraging its chaperone-ERT platform to advance migalastat HCl in multiple potential uses for patients with Fabry disease."

Expanded Amicus-GSK Collaboration for Fabry Disease: Key Highlights

- Amicus will commercialize all formulations of migalastat HCl in the U.S., while GSK will commercialize in the rest of the world.
- Amicus and GSK will continue to share research and development costs for all formulations of migalastat HCl, with Amicus funding 25% and GSK funding 75% of these costs for monotherapy and co-administration during the remainder of 2012. Amicus and GSK will be responsible for 40% and 60% of these costs, respectively, for co-formulation immediately and for all formulations in 2013 and beyond.
- GSK will make an \$18.6 million equity investment in Amicus, bringing GSK's total ownership stake in Amicus to 19.9%. GSK will purchase 2,949,581 shares of common stock at \$6.30 per share, a 7% premium over the 15-day average closing sale price of Amicus's common stock as reported by Nasdaq.
- Amicus will receive a \$3.5 million cash payment from GSK this quarter to reflect Amicus' achievement of a clinical development milestone during the second quarter 2012.
- GSK will be eligible to receive U.S. regulatory approval and product launch milestones totaling \$20 million for migalastat HCl monotherapy and chaperone-ERT co-administration.
- GSK will be eligible to receive additional regulatory and time-based milestone payments totaling up to \$35 million within 7 years following the launch of a co-formulated chaperone-ERT product. Amicus will also be responsible for certain additional pass-through milestone payments and single-digit royalties on the net U.S. sales of the co-formulated chaperone-ERT product that GSK must pay to a Third Party.

Conference Call and Audio-Visual Webcast

Amicus Therapeutics will host a conference call and audio-visual webcast today, July 17, 2012 at 5:00 P.M. ET to discuss the expanded agreement with GSK and provide additional details surrounding the new chaperone-ERT co-formulation. Interested participants and investors may access the live conference call by dialing 877-303-5859 (U.S./Canada) or 678-224-7784 (international).

An audio-visual webcast can also be accessed via the Investors section of the Amicus Therapeutics corporate web site at <http://www.amicusrx.com>, and will be archived for 30 days. Web participants are encouraged to go to the Web site 15 minutes prior to the start of the call to register, download and install any necessary software.

A telephonic replay of the call will be available for seven days beginning at 8 p.m. ET today. Access numbers for this replay are 855-859-2056 (U.S./Canada) and 404-537-3406 (international); participant code 11288561.

About Fabry Disease

Fabry disease is an inherited lysosomal storage disease that is currently 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 central nervous system, heart, kidneys, 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 disorders 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 human genetic diseases. Amicus' late-stage programs for lysosomal storage disorders include migalastat HCl monotherapy in Phase 3 for Fabry disease; migalastat HCl co-administered with enzyme replacement therapy (ERT) in Phase 2 for Fabry disease; and AT2220 co-administered with ERT in Phase 2 for Pompe 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, the projected cash position for the Company, and business development and other transactional opportunities. 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. 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, 2011. 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 'Financial review & risk section' in the company's Annual Report 2011 included as exhibit 15.2 to the company's Annual Report on Form 20-F for 2011.

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