



## **Amicus Therapeutics and Shire plc Enter Into \$440 Million ex-US Licensing Agreement to Develop and Commercialize Amigal(TM), Plicera(TM) and At2220**

### **Amicus to Host Conference Call at 8:00 A.M. Eastern Time**

CRANBURY, N.J., Nov 08, 2007 /PRNewswire-FirstCall via COMTEX News Network/ -- Amicus Therapeutics, Inc. (Nasdaq: FOLD) announced today that it has entered into a strategic collaboration with Shire Human Genetic Therapies, Inc., a subsidiary of Shire plc (LSE: SHP, Nasdaq: SHPGY, TSX: SHQ), to jointly develop Amicus' three lead pharmacological chaperone compounds for lysosomal storage disorders. Shire will receive rights to commercialize these products outside of the United States. Amicus will retain all rights to commercialize these products in the United States.

The collaboration includes Amigal(TM) (migalastat hydrochloride) currently in Phase 2 clinical trials for the treatment of Fabry disease, Plicera(TM) (isofagomine tartrate) currently in Phase 2 clinical trials for the treatment of Gaucher disease, and AT2220 (deoxynojirimycin), which the company is currently studying in Phase 1 clinical trials for the treatment of Pompe disease.

Under the terms of the deal, Amicus will receive an initial, non-refundable licensing payment of US\$ 50 million. Joint development costs toward global approval of the three compounds will be shared 50/50 going forward, and Amicus is eligible to receive an additional US\$ 150 million if certain clinical and regulatory milestones are met for the three programs through approvals. Amicus is also eligible to receive up to US\$ 240 million in sales-based milestones, as well as tiered double-digit royalties. Not including royalties and cost sharing, the deal is valued at up to US\$ 440 million.

John F. Crowley, Amicus' President & CEO said:

"We are immensely pleased to enter into this partnership with Shire, which leverages both companies' unique experience and expertise in developing therapies for lysosomal storage disorders. The combination of Amicus' strong science foundation in pharmacological chaperones and Shire's proven track record in drug development and commercialization will greatly enhance our efforts to bring these novel therapies to patients."

Matthew Emmens, Shire's CEO said:

"Amicus' pharmacological chaperone products have the potential to be an excellent addition to our current enzyme replacement therapy business. This technology should provide significant benefit to patients with these serious genetic diseases."

Amicus will lead worldwide development operations through the end of Phase 2 clinical trials. The companies will share responsibility for Phase 3 clinical trial execution. This will leverage Shire's significant ex-US regulatory and clinical experience, as well as its commercial infrastructure.

Sylvie Gregoire, President of Shire Human Genetic Therapies added:

"We are excited about this opportunity for Shire to expand its therapeutic platform beyond enzyme replacement therapies for lysosomal storage disorders. We look forward to working closely with Amicus on the development of these new therapies."

John F. Crowley also noted that, "this partnership is another step in Amicus' evolution as a biopharmaceutical company and it provides a significant validation of our platform technology for the treatment of lysosomal storage disorders. The partnership will also enhance our ability to rapidly advance our chaperone technologies to other diseases of misfolded or unstable proteins. It is a huge step forward for us."

### **Conference Call**

Amicus will host a conference call at 8 a.m. Eastern Time today to discuss the Amicus-Shire agreement. To listen to the conference call, please dial: 800-829-9048 from the United States and Canada or 913-312-9312 (International). A playback of the call will be available beginning today at 11:00 a.m. Eastern Time through November 18, and may be accessed by dialing: 888-203-1112 from the United States and Canada or 719-457-0820 (International). The reservation number for the replay is

## About Fabry Disease

Fabry disease is a lysosomal storage disorder caused by inherited genetic mutations in the GLA gene, which result in deficient activity of the enzyme alpha-galactosidase A (alpha-GAL). Deficient alpha-GAL activity leads to lysosomal accumulation of globotriaosylceramide (GL-3), which is believed to cause the various symptoms of Fabry disease, including pain, kidney failure and increased risk of heart attack and stroke. Fabry disease is estimated to affect approximately 5,000 to 10,000 people in the developed world, but recent evidence suggests that the disease may be significantly under diagnosed. The U.S. Food and Drug Administration's Office of Orphan Products Development has granted orphan designation for Amigal in the United States, and the European Commission has designated Amigal as an orphan medicinal product in the European Union.

## About Gaucher Disease

Gaucher disease, the most commonly diagnosed lysosomal storage disorder, is caused by inherited genetic mutations in the GBA gene, which result in deficient activity of the enzyme acid beta-glucosidase, also known as glucocerebrosidase (GCase). Deficient GCase activity leads to lysosomal accumulation of glucocerebroside inside certain cells, which is believed to cause the various symptoms of Gaucher disease, including an enlarged liver and spleen, abnormally low levels of red blood cells and platelets and skeletal complications. In some cases there is significant impairment of the central nervous system. Gaucher disease affects an estimated 8,000 to 10,000 people worldwide. The U.S. Food and Drug Administration's Office of Orphan Products Development has granted orphan drug designation for the active ingredient in Plicera in the United States and the European Commission has designated Plicera as an orphan medicinal product in the European Union.

## About Pompe Disease

Pompe disease affects an estimated 5,000 to 10,000 patients worldwide and is clinically heterogeneous in the age of onset, the extent of organ involvement, and the rate of progression. The early onset form of the disease is the most severe, progresses most rapidly, and is characterized by musculoskeletal, pulmonary, gastrointestinal, and cardiac symptoms that usually lead to death from cardio-respiratory failure between 1 and 2 years of age. The late onset form of the disease begins between childhood and adulthood and has a slower rate of progression that is characterized by musculoskeletal and pulmonary symptoms that usually lead to progressive weakness and respiratory insufficiency. The U.S. Food and Drug Administration's Office of Orphan Products Development has granted orphan drug designation for the active ingredient in AT2220 in the United States.

## About Amicus Therapeutics

Amicus Therapeutics is a biopharmaceutical company developing novel, oral therapeutics known as pharmacological chaperones for the treatment of a range of human genetic diseases. Pharmacological chaperone technology involves the use of small molecules that selectively bind to and stabilize proteins in cells, leading to improved protein folding and trafficking, and increased activity. Amicus is initially targeting lysosomal storage disorders, which are severe, chronic genetic diseases with unmet medical needs. Amicus has two product candidates in Phase 2 clinical trials, Amigal(TM) for the treatment of Fabry disease and Plicera(TM) for the treatment of Gaucher disease. The Company is also conducting Phase 1 clinical trials of AT2220 for the treatment of Pompe disease.

## Forward-Looking Statements

Amicus cautions you that statements included in this press release that are not a description of historical facts are "forward-looking statements" within the meaning of Section 21E of the Private Securities Litigation Reform Act of 1995. 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 potential progress 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 respective Phase 2 clinical trials for Amigal(TM) and Plicera(TM), and the Phase 1 clinical trial for AT2220 may not proceed in the timeframes or in the manner Amicus expects or at all. Further, the results of earlier clinical trials may not be predictive of future results; Amicus and its licensors may not be able to obtain, maintain and successfully enforce adequate patent and other intellectual property protection of its product candidates; and other risks detailed in the public filings of Amicus with the Securities and Exchange Commission. 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

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