Press Release

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Shire Enters Strategic Licensing and Collaboration Agreement with ArmaGen

Strengthens Leadership Position in Treatments for Hunter syndrome and Commitment to MPS II Patient Community

Lexington, Massachusetts, US – July 23, 2014 - Shire plc (LSE: SHP, NASDAQ: SHPG), the global specialty biopharmaceutical company, and ArmaGen, a US privately held biotechnology company, today announced a worldwide licensing and collaboration agreement for AGT-182, an investigational enzyme replacement therapy (ERT) for the potential treatment of both the central nervous system (CNS) and somatic manifestations in patients with Hunter syndrome (MPS II). This collaboration strengthens Shire's rare disease pipeline of innovative therapies where there is high unmet need, and underscores the company's long standing commitment to the Hunter syndrome community.

Under the terms of the agreement, Shire will obtain worldwide commercialization rights for AGT-182 in exchange for payments of approximately $225 million to ArmaGen, including an initial upfront payment of $15 million in cash and equity, an additional equity investment, R&D funding, development milestones and sales milestones, in addition to royalty payments. As part of the agreement, ArmaGen will be responsible for conducting and completing the Phase I/II study which it expects to initiate before the end of 2014, after which point Shire will be responsible for further clinical development, including Phase III trials, and commercialization.

Dr. Philip J. Vickers, Global Head of Research and Development at Shire, said, "Our agreement with ArmaGen marks our continued promise to the Hunter syndrome community to bring novel therapies that have the potential to dramatically redefine the treatment paradigm and address the most critical unmet needs. AGT-182 has the potential to be an important new therapy to our portfolio of programs for the treatment of both the CNS and somatic manifestations of Hunter syndrome. We look forward to collaborating with ArmaGen and leveraging our ability to successfully develop medicines to treat this rare, life-threatening disease."

Shire researched, developed and commercialized the first treatment approved for Hunter syndrome. This agreement with ArmaGen expands Shire's commitment to finding treatments for Hunter syndrome, which also includes SHP-609, Shire's product currently being investigated to treat the CNS manifestations associated with Hunter syndrome.

James Callaway, Ph.D., Chief Executive Officer of ArmaGen said, "Shire is the ideal partner for AGT-182, based on the company's international reach and expertise in serving patients with Hunter syndrome. We look forward to beginning the Phase I/II clinical trial of AGT-182 in collaboration with Shire and leveraging their expertise with these patients."

About AGT-182

AGT-182, which has received orphan drug designation from both the U.S. Food and Drug Administration (FDA) and the European Medicines Agency (EMA), is designed to take advantage of the body's natural system for transporting products across the blood brain barrier (BBB) by using the same receptor that delivers insulin to the brain. AGT-182 is engineered by the fusion of the replacement IDS enzyme to an antibody that binds to a receptor on the BBB. The IDS enzyme is designed to cross the BBB attached to that antibody.

About Hunter syndrome (MPS II)

Hunter syndrome or Mucopolysaccharidosis II (MPS II) is a rare, life-threatening genetic disorder that results from the absence or insufficient levels of the lysosomal enzyme iduronate-2-sulfatase. Without this enzyme, cellular waste products called mucopolysaccharides, also known as glycosaminoglycans or GAGs accumulate in tissues and organs, which then begin to malfunction. Possible signs and symptoms include large head, enlarged abdomen, frequent ear infections, difficulty breathing, joint stiffness, and short stature. It is estimated that 2/3 of all MPS II patients will be affected with CNS disease; this translates into a prevalence of around 1,200 patients worldwide.

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NOTES TO EDITORS

Shire enables people with life-altering conditions to lead better lives.

Our strategy is to focus on developing and marketing innovative specialty medicines to meet significant unmet patient needs.

We focus on providing treatments in Neuroscience, Rare Diseases, Gastrointestinal and Internal Medicine and we are developing treatments for symptomatic conditions treated by specialist physicians in other targeted therapeutic areas, such as Ophthalmology.

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FORWARD - LOOKING STATEMENTS - "SAFE HARBOR" STATEMENT UNDER THE PRIVATE SECURITIES LITIGATION REFORM ACT OF 1995

Statements included in this announcement that are not historical facts are forward-looking statements. Forward-looking statements involve a number of risks and uncertainties and are subject to change at any time. In the event such risks or uncertainties materialize, Shire's results could be materially adversely affected. The risks and uncertainties include, but are not limited to, that:

- Shire's products may not be a commercial success;
- revenues from ADDERALL XR are subject to generic erosion and revenues from INTUNIV will become subject to generic competition starting in December 2014;
- the failure to obtain and maintain reimbursement, or an adequate level of reimbursement, by third-party payors in a timely manner for Shire's products may impact future revenues, financial condition and results of operations;
- Shire conducts its own manufacturing operations for certain of its Rare Diseases products and is reliant on third party contractors to manufacture other products and to provide goods and services. Some of Shire's products or ingredients are only available from a single approved source for manufacture. Any disruption to the supply chain for any of Shire's products may result in the Shire being unable to continue marketing or developing a product or may result in Shire being unable to do so on a commercially viable basis for some period of time.
- the development, approval and manufacturing of Shire's products is subject to extensive oversight by various regulatory agencies. Submission of an application for regulatory approval of any of our product candidates, such as our planned submission of a New Drug Application to the FDA for Lifitegrast as a treatment for the signs and symptoms of dry eye disease in adults, may be delayed for any number of reasons and, once submitted, may be subjected to lengthy review and ultimately rejected. Moreover, regulatory approvals or interventions associated with changes to manufacturing sites, ingredients or manufacturing processes could lead to significant delays, increase in operating costs, lost product sales, an interruption of research activities or the delay of new product launches;
- the actions of certain customers could affect Shire's ability to sell or market products profitably. Fluctuations in buying or distribution patterns by such customers can adversely impact Shire's revenues, financial conditions or results of operations;
- investigations or enforcement action by regulatory authorities or law enforcement agencies relating to Shire's activities in the highly regulated markets in which it operates may result in the distraction of senior management, significant legal costs and the payment of substantial compensation or fines;
- adverse outcomes in legal matters and other disputes, including Shire's ability to enforce and defend patents and other intellectual property rights required for its business, could have a material adverse effect on Shire's revenues, financial condition or results of operations;
- Shire faces intense competition for highly qualified personnel from other companies, academic institutions, government entities and other organizations. Shire is undergoing a corporate reorganization and the consequent uncertainty could adversely impact Shire's ability to attract and/or retain the highly skilled personnel needed for Shire to meet its strategic objectives;
- failure to achieve Shire's strategic objectives with respect to the acquisition of ViroPharma Incorporated may adversely affect Shire's financial condition and results of operations;

and other risks and uncertainties detailed from time to time in Shire's filings with the U.S. Securities and Exchange Commission, including its most recent Annual Report on Form 10-K.