ArmaGen Announces Strategic Licensing and Collaboration Agreement with Shire to Develop AGT-182 for Treatment of Hunter Syndrome

ArmaGen to Receive up to Approximately $225 Million Including Initial Upfront Payment of $15 Million in Cash and Equity, an Additional Equity Investment, R&D Funding, Development and Sales Milestones, in Addition to Future Royalties

Calabasas, Calif., July 23, 2014 – ArmaGen, a privately held biotechnology company focused on developing revolutionary therapies to treat severe neurological disorders, announced today that it has entered into a worldwide licensing and collaboration agreement with Shire plc to develop AGT-182, an investigational enzyme replacement therapy (ERT) for potential treatment of both the central nervous system (CNS) and somatic (body-related) manifestations of Hunter syndrome. Also known as MPS II, Hunter syndrome is a rare, severe, progressive and life-limiting lysosomal storage disorder.

Under the agreement, Shire receives worldwide commercialization rights for AGT-182. ArmaGen will receive payments up to approximately $225 million, including an initial upfront payment of $15 million in cash and equity, an additional equity investment, R&D funding, development and sales milestones, in addition to future royalties up to double digits. The collaboration between ArmaGen and Shire on AGT-182 will be managed by a joint steering committee, with representatives from both companies. ArmaGen will be responsible for conducting a Phase 1/2 study of AGT-182 and expects to initiate the trial before the end of 2014. Shire will be responsible for further clinical development, including Phase 3 trials, registration and commercialization of AGT-182 worldwide.

“The agreement with Shire validates the clinical potential of ArmaGen’s lead therapy and its ability to cross the blood-brain barrier to treat the progressive and devastating neurological complications of Hunter syndrome,” said James Callaway, Ph.D., Chief Executive Officer of ArmaGen. “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 our Phase 1/2 clinical trial of AGT-182 in collaboration with Shire and, in parallel, advancing ArmaGen’s proprietary pipeline of innovative therapies.”

“Our agreement with ArmaGen strengthens our long-standing commitment to the Hunter syndrome community to bring forward novel therapies that have the potential to dramatically redefine the treatment paradigm and address the most critical unmet needs,” said Philip J. Vickers, Ph.D., Global Head of Research and Development at Shire. “AGT-182 has the potential to be an important new therapy to our existing portfolio of Hunter syndrome programs. We plan

to apply our proven ability to develop therapies for rare genetic diseases to progress AGT-182 as a potential treatment that offers hope to patients with Hunter syndrome and their families."

About Hunter Syndrome

Hunter syndrome, also known as mucopolysaccharidosis type II, or MPS II, is a lysosomal storage disorder caused by inadequate activity of the enzyme iduronate-2-sulfatase (IDS), which is needed to break down complex sugars produced by the body. The buildup of these complex sugars, known as mucopolysaccharides, interferes with functioning of certain cells and organs, leading to serious complications including developmental delays and mental impairment. Symptoms of Hunter syndrome include growth delay, joint stiffness and coarsening of facial features. In severe cases, patients experience respiratory and cardiac problems, enlargement of the liver and spleen, and neurological deficits that can lead to premature death. Hunter syndrome primarily affects males and is almost always severe, progressive and life-limiting. Available treatments for Hunter syndrome are not expected to cross the blood-brain barrier (BBB) in clinically relevant amounts and therefore do not address the progressive neurological complications of the disease.

About AGT-182

AGT-182 is a novel, investigational ERT that has received orphan drug designation from both the U.S. Food and Drug Administration (FDA) and the European Medicines Agency (EMA). Using ArmaGen's proprietary technology, AGT-182 is designed to take advantage of the body's natural system for transporting products across the blood-brain barrier (BBB) by binding to 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 is attracted to a receptor on the BBB. The IDS enzyme is designed to travel through the BBB attached to that antibody.

About ArmaGen

ArmaGen is a privately held biotechnology company focused on developing revolutionary therapies for severe neurological disorders. The company is developing a robust pipeline of innovative therapies for the treatment of neurological complications of lysosomal storage disorders such as Hunter syndrome, Hurler syndrome, metachromatic leukodystrophy and Sanfilippo A syndrome, as well as central nervous system diseases such as Alzheimer's and Parkinson's. ArmaGen's pipeline is based on decades of scientific leadership in engineering therapies to cross the BBB and a dominant intellectual property portfolio. The company is advancing its pipeline through licensing and collaboration agreements, in-house development programs, and future partnering opportunities. For more information, visit www.armagen.com.

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