Aeglea BioTherapeutics and Immedica Announce Commercialization Agreement for Pegzilarginase for the Treatment of Arginase 1 Deficiency in Europe and Middle East

Aeglea receives $21.5 million upfront with up to approximately $130 million in milestone payments and royalties in the mid-twenties Immedica obtains exclusive commercialization rights in Europe and several Middle Eastern countries

Austin, Texas, and Stockholm, March 22, 2021 – Aeglea BioTherapeutics, Inc. (NASDAQ:AGLE), a clinical-stage biotechnology company developing a new generation of human enzyme therapeutics as innovative solutions for rare metabolic diseases, and Immedica Pharma AB (Immedica) today announced the license and supply agreement for pegzilarginase, a novel, recombinant human arginase 1 enzyme that has been shown to lower toxic levels of the amino acid arginine in patients with Arginase 1 Deficiency (ARG1-D). People living with ARG1-D, a rare, progressive disease characterized by high levels of arginine, may experience severe spasticity-related mobility limitations, seizures, developmental delay, intellectual disability and early mortality.

"With a proven track record of bringing important products to the rare disease community in Europe and the Middle East, Immedica represents an ideal partner as we plan for the potential approval and commercialization of pegzilarginase in key markets beyond the U.S.," said Anthony G. Quinn, M.B. Ch.B., Ph.D., president and chief executive officer of Aeglea. "There is an urgent need to deliver new therapies for Arginase 1 Deficiency due to its progressive and devastating nature. We are pleased to have a partner who believes in the potential of this program and shares our excitement for bringing pegzilarginase to market to further our mission of improving the lives of families affected by Arginase 1 Deficiency."

"We are excited to enter this partnership with Aeglea and expand our growing portfolio to include pegzilarginase, which fits strategically very well within our existing rare disease and urea cycle disorder portfolio and commercial infrastructure," says Anders Edvell, CEO of Immedica. "This agreement strengthens our position as a partner of choice for rare disease collaborations."

Under the terms of the agreement, Immedica will make an upfront payment of $21.5 million to Aeglea. Additionally, Aeglea will be eligible to receive up to approximately $130 million in regulatory and commercial milestones as well as mid-twenties percentage royalties on net sales. Immedica receives commercialization rights in Europe and several Middle East countries. Aeglea will continue to be responsible for certain clinical development activities and the manufacturing of pegzilarginase, and retains commercialization rights in the U.S. and rest of the world.

Pegzilarginase is currently being investigated as a treatment for ARG1-D in the PEACE study, a pivotal Phase 3 clinical trial with topline data expected in the fourth quarter of 2021. In Phase 1/2 and ongoing open-label extension studies, pegzilarginase has been shown to lower toxic levels of arginine in patients with ARG1-D. After a 56 week treatment period, all 13 patients

achieved plasma arginine levels within the target range (<200μM) and 11 patients (85%) were considered clinical responders based on improvements in mobility assessments. Pegzilarginase was also shown to have a favorable safety profile with the most common treatment-related serious adverse events being hypersensitivity and hyperammonemia, both of which were infrequent, expected and managed with standard of care. Most treatment-related adverse events were mild and decreased in frequency over time.

About Pegzilarginase in Arginase 1 Deficiency

Pegzilarginase is a novel recombinant human enzyme, which has been shown to rapidly and sustainably lower levels of the amino acid arginine in plasma. Aeglea is developing pegzilarginase for the treatment of patients with Arginase 1 Deficiency (ARG1-D), a rare debilitating and progressive disease characterized by the accumulation of arginine. ARG1-D presents in early childhood and patients experience spasticity, seizures, developmental delay, intellectual disability and early mortality. Aeglea's Phase 1/2 and Phase 2 open-label extension (OLE) data for pegzilarginase in patients with ARG1-D demonstrated clinical improvements and sustained lowering of plasma arginine. The Company's ongoing single, global pivotal Phase 3 PEACE trial is designed to assess the effects of treatment with pegzilarginase versus placebo over 24 weeks with a primary endpoint of plasma arginine reduction. Pegzilarginase has received multiple regulatory designations, including Rare Pediatric Disease, Breakthrough, Fast Track and Orphan Drug Designations from the U.S. Food and Drug Administration as well as Orphan Drug Designation from the European Medicines Agency.

About Aeglea BioTherapeutics

Aeglea BioTherapeutics is a clinical-stage biotechnology company redefining the potential of human enzyme therapeutics to benefit people with rare and devastating metabolic diseases with limited treatment options. Aeglea's lead product candidate, pegzilarginase, is in a pivotal Phase 3 trial for the treatment of Arginase 1 Deficiency and has received both Rare Pediatric Disease and Breakthrough Therapy Designation. In the second quarter of 2020, the Company initiated a Phase 1/2 clinical trial of AGLE-177 for the treatment of Homocystinuria. AGLE-177 has also been granted Rare Pediatric Disease Designation. Aeglea has an active discovery platform focused on engineering small changes in human enzymes to have a big impact on the lives of patients and their families. For more information, please visit http://aeglea.com.

About Immedica

Immedica is a fast-growing private European niche pharma group. Its headquarters is based in Stockholm, Sweden, and it has pan-European and Middle East commercial coverage. In addition, Immedica provides some of its products to other parts of the world via a network of regional distributors.

Immedica provides significant know-how and experience from commercialization of niche/specialty care products across Europe and the Middle East, and the company's management team has an outstanding track record of operating niche pharma products internationally. Immedica has capabilities to provide optimal access of specialty care medicines to patients with significant medical needs, including key areas such as regulatory affairs, pharmacovigilance, medical affairs, pricing & reimbursement, and product distribution.

Aeglea Safe Harbor / Forward Looking Statements

This press release contains "forward-looking" statements within the meaning of the safe harbor provisions of the U.S. Private Securities Litigation Reform Act of 1995. Forward-looking statements can be identified by words such as: "anticipate," "intend," "plan," "goal," "seek," "believe," "project," "estimate," "expect," "strategy," "future," "likely," "may," "should," "will" and similar references to future periods. These statements are subject to numerous risks and uncertainties that could cause actual results to differ materially from what we expect. Examples

of forward-looking statements include, among others, statements we make regarding our ability to obtain regulatory approval for, and commercialize, pegzilarginase, recognize milestone and royalty payments from our agreement with Immedica, cash forecasts, the timing and success of our clinical trials and related data, the timing and expectations for regulatory submissions and approvals, timing and results of meetings with regulators, the timing of announcements and updates relating to our clinical trials and related data, our ability to enroll patients into our clinical trials, the expected impact of the COVID-19 pandemic on our operations and clinical trials, success in our collaborations, the potential addressable markets of the our product candidates and the potential therapeutic benefits and economic value of our lead product candidate or other product candidates. Further information on potential risk factors that could affect our business and its financial results are detailed in our most recent Quarterly Report on Form 10-Q for the quarter ended September 30, 2020 filed with the Securities and Exchange Commission (SEC), and other reports as filed with the SEC. We undertake no obligation to publicly update any forward-looking statement, whether written or oral, that may be made from time to time, whether as a result of new information, future developments or otherwise.

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