Aeglea BioTherapeutics Announces Positive Interim Clinical Data and Completion of Enrollment for Ongoing Phase 1/2 Trial of Pegzilarginase in Patients with Arginase 1 Deficiency

Exceeded Enrollment Target with 15 Patients in Phase 1/2 Trial

Additional Interim Data to be Presented at ASHG Conference in October 2018

Company Plans to Announce Pivotal Trial Design in Q4 2018 and Initiate Pivotal Trial in 1H 2019

Company to Host Clinical Update Conference Call Today at 8:30 a.m. ET

AUSTIN, Texas, Sept. 04, 2018 (GLOBE NEWSWIRE) -- Aeglea BioTherapeutics, Inc.(NASDAQ: AGLE), a clinical-stage biotechnology company that designs and develops innovative human enzyme therapeutics for patients with rare genetic diseases and cancer, today announced completion of enrollment and additional clinical data at the 2018 Society for the Study of Inborn Errors of Metabolism (SSIEM) Annual Symposium from its ongoing Phase 1/2 trial of pegzilarginase, its lead investigational therapy, in patients with the rare genetic disease Arginase 1 Deficiency (ARG1-D).

“I am thrilled that we exceeded our recruitment target in this Phase 1/2 trial of pegzilarginase by five patients, as enrollment completion is an important milestone for patients with ARG1-D,” said Anthony G. Quinn, M.B Ch.B, Ph.D., president and chief executive officer of Aeglea. “We are pleased to confirm our previous guidance of announcing the design of the first pivotal clinical trial for patients with ARG1-D in the fourth quarter of 2018 and initiating the pivotal trial in the first half of 2019.”

“Given the challenges of lowering arginine levels with current approaches, it is very encouraging to see the marked reductions in plasma arginine in our patient following treatment with pegzilarginase,” said George Diaz, M.D., Ph.D, professor and chief, Division of Medical Genetics, Icahn School of Medicine at Mount Sinai, and co-author on the presentation. “In addition, it is very exciting that we are seeing evidence of an impact on important disease manifestations with better walking, improved posture, and enhanced alertness.”

Highlights of the SSIEM presentation, entitled "Improvements in Arginase 1 Deficiency Related Disease Manifestations Following Plasma Arginine Reductions with Pegzilarginase," include the following:

- Administration of pegzilarginase resulted in marked reductions in plasma arginine and related guanidino compounds (GCs);
- Clinical improvements in one or more instruments of neuromotor function in all three patients completing eight weeks of repeat dose administration;

- Pegzilarginase was generally safe and well tolerated; most treatment-related adverse events (AEs) were mild and all were resolved;

- No marked or sustained increase in ADA titers in patients exposed to pegzilarginase and initial evidence of rapid tolerization in patients with low titer ADAs.

The Company plans to present new interim clinical data for the Phase 1/2 trial at the 2018 American Society of Human Genetics (ASHG) Conference in October.

Conference Call & Webcast Details

Aeglea will hold a clinical update conference call today, Tuesday, September 4, 2018 at 8:30 a.m. ET. To access the live conference call via phone, please dial 1-877-709-8155 (toll free) within the United States, or 1-201-689-8881 internationally. A replay of the call will be available through September 11, 2018 by dialing 1-877-660-6853 within the United States or 1-201-612-7415 internationally. The conference ID is 13678293.

To access the live and archived webcast of the presentation, please visit the Presentations & Events section of the Aeglea BioTherapeutics investor relations website. Please connect to the website at least 15 minutes prior to the presentation to allow for any software download that may be necessary.

About Pegzilarginase in Arginase 1 Deficiency

Pegzilarginase is an enhanced human arginase that enzymatically depletes the amino acid arginine. Aeglea is developing pegzilarginase for the treatment of patients with Arginase 1 Deficiency, a debilitating urea cycle disorder caused by deficiency of a key arginine metabolizing enzyme that leads to severe and progressive hyperargininemia-related neurological abnormalities, hyperammonemia and early mortality. Pegzilarginase is intended for use as an enzyme replacement therapy in patients to reduce elevated blood arginine levels. The Company's interim Phase 1/2 data demonstrated clinical improvements and rapid and sustained lowering of plasma arginine in Arginase 1 Deficiency patients.

About Aeglea BioTherapeutics

Aeglea is a clinical-stage biotechnology company that designs and develops innovative human enzyme therapeutics for patients with rare genetic diseases and cancer. The Company is developing pegzilarginase, its lead investigational therapy, for the treatment of Arginase 1 Deficiency, as monotherapy in arginine-dependent cancers and in combination with an immune checkpoint inhibitor for small cell lung cancer. In addition, Aeglea has an active research pipeline of other human enzyme-based approaches in both therapeutic areas. For more information, please visit http://aegleabio.com.

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 the timing and success of our clinical trials and related data, the timing of announcements and updates relating to our clinical trials, trial designs and related data, 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 June 30, 2018 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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