Aeglea BioTherapeutics Doses First Patient in Pivotal Phase 3 PEACE Trial of Pegzilarginase in Arginase 1 Deficiency

Topline Data from PEACE Anticipated in First Quarter of 2021

New Data from ARG1-D Phase 1/2 Extension Study Expected in September 2019

AUSTIN, Texas, June 03, 2019 (GLOBE NEWSWIRE) -- Aeglea BioTherapeutics, Inc. (NASDAQ: AGLE), a clinical-stage biotechnology company that engineers next-generation human enzymes to provide solutions for diseases with unmet medical need, today announced the dosing of the first patient in the Company's Phase 3 PEACE (Pegzilarginase Effect on Arginase 1 Deficiency Clinical Endpoints) clinical trial. The pivotal trial is intended to further evaluate the efficacy and safety of pegzilarginase for the treatment of Arginase 1 Deficiency (ARG1-D), a progressive disease presenting in early childhood that results in severe complications and early mortality. The Company expects to report topline data from the PEACE trial in the first quarter of 2021. Aeglea anticipates reporting new data in September 2019 from all patients who have each received at least 20 doses in the Company's ongoing Phase 1/2 extension study.

"Aeglea is reminded daily of the challenges patients with ARG1-D face due to the limitations of current disease management," said Anthony G. Quinn, M.B. Ch.B, Ph.D., president and chief executive officer of Aeglea. "The dosing of our first patient in the pivotal Phase 3 PEACE trial is a major step in establishing the clinical effectiveness of pegzilarginase. Aeglea is committed to working with physicians, patients, caregivers, and the ARG1-D community as we advance this potentially transformative therapy for patients with this devastating and progressive disease."

"We are excited to be involved in the PEACE trial as it provides us the opportunity to build on our experience and data from the Phase 1/2 trial, where we observed excellent control of arginine levels and stability or improvement in mobility and adaptive behavior," said Markey McNutt, M.D., Ph.D. an assistant professor with the Eugene McDermott Center for Human Growth and Development at UT Southwestern.

PEACE is a global, randomized, double-blind trial designed to assess the effects of treatment with pegzilarginase versus placebo over 24 weeks with a primary endpoint of statistically significant plasma arginine reduction from baseline. The primary endpoint assesses the effectiveness of pegzilarginase in lowering plasma arginine levels given the evidence that plasma arginine control has the potential to improve the clinical status and to slow disease progression in patients with ARG1-D. Secondary endpoints will include mobility and adaptive behavior as assessments of clinically meaningful effects, in addition to safety and pharmacokinetics. Upon completion of the 24-week treatment period, patients may qualify to participate in a long-term extension study of pegzilarginase.

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 rare debilitating disease presenting in childhood with persistent hyperargininemia, severe progressive neurological abnormalities and early mortality. Pegzilarginase is intended for use as an enzyme replacement therapy in patients to reduce elevated blood arginine levels. Aeglea's Phase 1/2 data for pegzilarginase in patients with Arginase 1 Deficiency

demonstrated clinical improvements and sustained lowering of plasma arginine. Aeglea is currently recruiting patients for its single, global pivotal Phase 3 PEACE trial designed to assess the effects of treatment with pegzilarginase versus placebo over 24 weeks with a primary endpoint of plasma arginine reduction.

About Aeglea BioTherapeutics

Aeglea is a clinical-stage biotechnology company that engineers next-generation human enzymes with enhanced properties and novel activity to provide solutions for diseases with unmet medical need. Aeglea is developing pegzilarginase, its lead investigational therapy, for the treatment of Arginase 1 Deficiency and in combination with an immune checkpoint inhibitor for small cell lung cancer. Aeglea has two pipeline programs in IND-enabling studies for Homocystinuria and Cystinuria and an active discovery pipeline. 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 our cash forecasts, the timing and success of our clinical trials and related data, the timing and expectations for regulatory submissions and approvals, the timing of announcements and updates relating to our clinical trials and related data, our ability to enroll patients into our clinical trials, success in our collaborations 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 March 31, 2019 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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