Aeglea BioTherapeutics Announces Positive Interim Clinical Data for Pegzilarginase in Advanced Melanoma Patients at the European Society for Medical Oncology 2018 Congress

Pegzilarginase Monotherapy Demonstrates Anti-Tumor Activity

AUSTIN, Texas, Oct. 22, 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 that it presented positive clinical data for pegzilarginase in melanoma patients at the European Society for Medical Oncology (ESMO) 2018 Congressin Munich, Germany. The poster, titled “Initial cohort expansion results of sustained arginine depletion with Pegzilarginase in melanoma patients in a phase 1 advanced solid tumor trial,” was presented on October 21. Clinical data from the Company’s ongoing Phase 1 clinical trial investigating pegzilarginase as a single agent includes expansion cohorts for cutaneous melanoma and uveal melanoma.

“This ongoing clinical study of pegzilarginase demonstrated single agent anti-tumor activity in what is a difficult-to-treat population of heavily pre-treated melanoma patients,” said James Wooldridge, M.D., chief medical officer of Aeglea. “These findings are in line with expectations from our single agent preclinical studies. Given the significant synergies we observed in preclinical studies with immune checkpoint inhibitors, we look forward to data readouts from the Phase 1/2 combination clinical trial.”

Data highlights from ESMO 2018:

- Pegzilarginase demonstrated single agent anti-tumor activity in patients with advanced melanoma

  --Of the 28 patients included in the two cohorts, there was one confirmed partial response (PR) and eight patients with stable disease (SD). Six patients remained on treatment at the time of the data cutoff.
  --Anti-tumor activity appeared greater in patients with tumors that lack ASS1 (argininosuccinate synthetase 1) expression, which is consistent with preclinical studies that suggest tumors lacking ASS1 expression are dependent on extracellular arginine for survival.

- Pegzilarginase rapidly and sustainably depleted plasma arginine with a manageable safety profile, treatment related adverse events were grade three or lower

About Pegzilarginase in Cancer

Pegzilarginase is an enhanced human arginase that enzymatically degrades the amino acid arginine. In some cancers, tumor cells stop producing specific amino acids and must acquire them from the blood, making the tumor cells susceptible to starvation through depletion of those amino acids. Aeglea is developing pegzilarginase to exploit vulnerabilities in some cancers that lead to an increased

dependency on extracellular arginine. Pegzilarginase targets these arginine dependent cancers by depleting blood arginine levels to below the normal range. Preclinical data demonstrated that the resulting arginine starvation inhibits proliferation, induces cell death, increases turnover of cell components and promotes anti-tumor immune responses. The Company's Phase 1 data in advanced solid tumors demonstrated that pegzilarginase was well tolerated at doses that produced marked and sustained reductions in blood arginine levels below the normal range.

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. 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, the potential therapeutic benefits and economic value of our lead product candidate or other product. 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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