Aeglea BioTherapeutics Announces Sale of Pegzilarginase to Immedica Pharma

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Global rights to pegzilarginase in development for Arginase 1 Deficiency sold to Immedica Pharma for $15 million upfront cash proceeds and up to $100 million of contingent milestone payments

Marketing Authorisation Application for pegzilarginase is under review by the European Medicines Agency

WALTHAM, Mass., July 27, 2023 /PRNewswire/ -- Aeglea BioTherapeutics, Inc. ("Aeglea") (NASDAQ:AGLE), a biotechnology company advancing a pipeline of antibody therapeutics with best-in-class potential to transform the treatment of inflammatory bowel disease ("IBD"), today announced that it has entered into an agreement to sell the global rights to pegzilarginase, an investigational treatment for the rare metabolic disease Arginase 1 Deficiency ("ARG1-D"), to Immedica Pharma AB ("Immedica") for $15 million in upfront cash proceeds and up to $100 million in contingent milestone payments. The sale of pegzilarginase to Immedica supersedes the previous license agreement between Aeglea and Immedica.

"We are thrilled that Immedica will be continuing our efforts with pegzilarginase and consolidating development of the potential therapy for the treatment of ARG1-D," said Jonathan Alspaugh, President and Chief Financial Officer of Aeglea. "Immedica has made substantial progress in working towards a European market approval. We believe it is ultimately in the best interest of the ARG1-D community that Immedica will seek to continue the dialogue with the FDA to discuss a path forward for pegzilarginase in the United States while advancing the program globally."

The milestone payments are contingent on formal reimbursement decisions by national authorities in key European markets and pegzilarginase approval by the U.S. Food and Drug Administration ("FDA"), among other events. The upfront payment and contingent milestone payments if paid, net of expenses and adjustments, will be distributed to holders of Aeglea's Contingent Value Rights ("CVR") pursuant to the CVR agreement resulting from Aeglea's acquisition of Spyre Therapeutics, Inc. ("Spyre").

About Pegzilarginase in ARG1-D
Pegzilarginase is a novel recombinant human enzyme engineered to degrade the amino acid arginine and has been shown to rapidly and sustainably lower levels of the amino acid arginine in plasma. Pegzilarginase has been in development for the treatment of people with 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. The PEACE Phase 3 clinical trial met its primary endpoint with a 76.7% reduction in mean plasma arginine compared to placebo. Additionally, 90.5% of pegzilarginase treated patients achieved normal plasma arginine levels. Based on the results from PEACE and a previous Phase 1/2 clinical trial, a Marketing Authorisation Application was submitted to the European Medicines Agency by Immedica. In April 2022, Aeglea announced the submission of a Biologics License Application (BLA) to the FDA. In June 2022, Aeglea announced the receipt of a Refusal to File letter from the FDA for the BLA of pegzilarginase for the treatment of ARG1-D.

About Aeglea BioTherapeutics
In June 2023, Aeglea completed the asset acquisition of Spyre and shifted its disease focus to IBD. Aeglea is advancing a pipeline of antibody therapeutics with the potential to transform the treatment of IBD. The approaches combine novel antibody engineering, rational therapeutic combinations, and precision immunology approaches to maximize efficacy, safety, and convenience of treatments for IBD.

For more information, please visit http://aeglea.com.

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Forward-Looking Statements
Certain statements in this press release, other than purely historical information, may constitute "forward-looking statements" within the meaning of the federal securities laws, including for purposes of the safe harbor provisions under the United States Private Securities Litigation Reform Act of 1995, concerning Aeglea, the sale of pegzilarginase by Aeglea and other matters. These forward-looking statements include, but are not limited to, express or implied statements relating to Aeglea's management team's expectations, hopes, beliefs, intentions or strategies regarding the future including, without limitation, statements regarding potential receipt of contingent milestone payments and the distribution of such payments to holders of CVRs, and the continued development of pegzilarginase. In addition, any statements that refer to projections, forecasts or other characterizations of future events or circumstances, including any underlying assumptions, are forward-looking statements. The words "opportunity," "potential," "milestones," "pipeline," "can," "goal," "aim," "strategy," "target," "seek," "anticipate," "achieve," "believe," "contemplate," "continue," "could," "estimate," "expect," "intends," "may," "might," "plan," "possible," "predict," "project," "should," "will," "would" and similar expressions (including the negatives of these terms or variations of them) may identify forward-looking statements, but the absence of these words does not mean that a statement is not forward-looking. These forward-looking statements are based

on current expectations and beliefs concerning future developments and their potential effects. There can be no assurance that future developments affecting Aeglea will be those that have been anticipated. These forward-looking statements involve a number of risks, uncertainties (some of which are beyond Aeglea's control) or other assumptions that may cause actual results or performance to be materially different from those expressed or implied by these forward-looking statements. These risks and uncertainties include, but are not limited those uncertainties and factors described under the heading "Risk Factors," "Risk Factor Summary" and "Forward-Looking Statements" in Aeglea's most recent Annual Report on Form 10-K filed with the Securities and Exchange Commission (the "SEC") on March 2, 2023, the Quarterly Report on Form 10-Q filed with the SEC on May 11, 2023, as well as discussions of potential risks, uncertainties, and other important factors included in other filings by Aeglea from time to time. Should one or more of these risks or uncertainties materialize, or should any of Aeglea's assumptions prove incorrect, actual results may vary in material respects from those projected in these forward-looking statements. Nothing in this press release should be regarded as a representation by any person that the forward-looking statements set forth herein will be achieved or that any of the contemplated results of such forward-looking statements will be achieved. You should not place undue reliance on forward-looking statements in this press release, which speak only as of the date they are made and are qualified in their entirety by reference to the cautionary statements herein. Aeglea does not undertake or accept any duty to release publicly any updates or revisions to any forward-looking statements. This press release does not purport to summarize all of the conditions, risks and other attributes of an investment in Aeglea.

SOURCE Aeglea BioTherapeutics, Inc.