UNITED STATES
SECURITIES AND EXCHANGE COMMISSION
Washington, D.C. 20549

FORM 8-K

CURRENT REPORT
Pursuant to Section 13 or 15(d)
of the Securities Exchange Act of 1934

Date of Report (Date of earliest event reported): March 7, 2019

AEGLEA BIOTHERAPEUTICS, INC.
(Exact name of registrant as specified in its charter)

Delaware	001-37722	46-4312787
(State or other jurisdiction of incorporation)	(Commission File Number)	(IRS Employer Identification No.)

901 S. MoPac Expressway Barton Oaks Plaza One Suite 250 Austin, TX	78746
(Address of principal executive offices)	(Zip Code)

(512) 942-2935
(Registrant's telephone number, including area code)

(Former name or former address, if changed since last report)

Check the appropriate box below if the Form 8-K filing is intended to simultaneously satisfy the filing obligation of the registrant under any of the following provisions:

☐ Written communications pursuant to Rule 425 under the Securities Act (17 CFR 230.425)

☐ Soliciting material pursuant to Rule 14a-12 under the Exchange Act (17 CFR 240.14a-12)

☐ Pre-commencement communications pursuant to Rule 14d-2(b) under the Exchange Act (17 CFR 240.14d-2(b))

☐ Pre-commencement communications pursuant to Rule 13e-4(c) under the Exchange Act (17 CFR 240.13e-4(c))

Indicate by check mark whether the registrant is an emerging growth company as defined in Rule 405 of the Securities Act of 1933 (§230.405 of this chapter) or Rule 12b-2 of the Securities Exchange Act of 1934 (§240.12b-2 of this chapter).

Emerging growth company ☒

If an emerging growth company, indicate by check mark if the registrant has elected not to use the extended transition period for complying with any new or revised financial accounting standards provided pursuant to Section 13(a) of the Exchange Act. ☒

Item 2.02 Results of Operations and Financial Condition.

On March 7, 2019, Aeglea BioTherapeutics, Inc. issued a press release announcing its financial results for the quarter and year ended December 31, 2018. A copy of the press release is attached as Exhibit 99.1 to this report.

The information in this Item 2.02, including Exhibit 99.1 to this report, shall not be deemed to be "filed" for purposes of Section 18 of the Securities Exchange Act of 1934, as amended (the "Exchange Act"), or otherwise subject to the liabilities of that section or Sections 11 and 12(a)(2) of the Securities Act of 1933, as amended (the "Securities Act"). The information contained in this Item 2.02 and in the accompanying Exhibit 99.1 shall not be incorporated by reference into any other filing under the Exchange Act or under the Securities Act, except as shall be expressly set forth by specific reference in such filing.

Item 9.01 Financial Statements and Exhibits.

(d) Exhibits

Exhibit Number	Description
99.1	Press release issued by Aeglea BioTherapeutics, Inc. regarding its financial results for the quarter and year ended December 31, 2018, dated March 7, 2019.

SIGNATURE

Pursuant to the requirements of the Securities Exchange Act of 1934, the registrant has duly caused this report to be signed on its behalf by the undersigned hereunto duly authorized.

AEGLEA BIOTHERAPEUTICS, INC.

Date: March 7, 2019

By: /s/ Charles N. York II
Charles N. York II
Chief Financial Officer

Exhibit 99.1

Aeglea BioTherapeutics Reports Fourth Quarter and Full Year 2018 Financial Results and Corporate Highlights

On Track to Dose First Patient in Pivotal Phase 3 Trial of Pegzilarginase for ARG1-D in Q2 of 2019

Initiated IND-Enabling Studies for New Pipeline Programs

Progress with Pegzilarginase Cancer Indication-Phase 2 Combination Study Initiated

Gross Proceeds of $69 Million from February 2019 Public Offering Extends Cash Runway Through Q1 of 2021

Company to Host Conference Call and Webcast Today at 4:30 p.m. ET

Austin, Texas, March 7, 2019 - 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 reported its fourth quarter and full year 2018 financial results and provided corporate highlights.

"2018 was a very successful year for the company," said Anthony G. Quinn, M.B. Ch. B, Ph.D., president and chief executive officer of Aeglea. "We made substantial progress with our clinical programs for pegzilarginase in Arginase 1 Deficiency (ARG1-D) and oncology, leveraged our unique drug hunting capabilities to generate new pipeline programs for cystinuria and homocystinuria and strengthened Aeglea's balance sheet. As we look to 2019, the Company is well positioned to advance our clinical and research programs, as well as to leverage our human enzyme design capabilities to address other diseases with significant unmet medical need."

Program Highlights

Pegzilarginase in Arginase 1 Deficiency

- Completed dosing of Phase 1/2 trial in February 2019, with 14 out of 16 enrolled patients completing eight weeks of repeat dosing. Clinical data from this trial, which informed the design of the Phase 3 PEACE (Pegzilarginase Effect on Arginase 1 Deficiency Clinical Endpoints) trial protocol design, included the demonstration that pegzilarginase is effective in reducing plasma arginine levels with accompanying improvements in important disease-related abnormalities. Insights from standardized clinical assessments and feedback from physicians and caregivers also established the value of mobility and adaptive behavior assessments in capturing the clinical benefits of pegzilarginase. Additionally, pegzilarginase appeared safe and well tolerated. Aeglea expects to report Phase 1/2 data at an oral presentation at the Society for Inherited Metabolic Disorders (SIMD) annual meeting being held April 6-9 in Seattle, Washington.

- In December 2018, the Company announced the design of its single, global pivotal Phase 3 PEACE trial based on input from the U.S. Food and Drug Administration (FDA) and the European Medicine Agency (EMA). Pegzilarginase is believed to be the first-ever investigative therapy for patients with ARG1-D that addresses the high arginine levels that are the key drivers of this devastating disease. PEACE is a global, randomized, double-blind, placebo-controlled trial designed to assess the effects of treatment with pegzilarginase versus placebo over 24 weeks with a primary endpoint of plasma arginine reduction. Secondary endpoints will include mobility and adaptive behavior as assessments of clinically meaningful effects, in addition to safety and pharmacokinetics. Aeglea expects to dose the first patient in the PEACE trial in the second quarter of 2019 and anticipates topline data will be available in the first quarter of 2021.

- Pediatric review voucher (PRV) eligible given receipt of rare pediatric disease designation from FDA for treatment of ARG1-D.

Pegzilarginase in Cancer

- Completed the Phase 1b safety evaluation and dose selection for pegzilarginase in combination with KEYTRUDA® (pembrolizumab), Merck's anti-PD-1 therapy. This trial is designed to assess the safety and efficacy of pegzilarginase with KEYTRUDA in patients with extensive disease small cell lung cancer (SCLC) who have relapsed or progressive disease after platinum-based chemotherapy.

    - Initiated enrollment in the open label Phase 2 study of pegzilarginase in combination with KEYTRUDA in December 2018. This trial is designed to assess the safety and efficacy of pegzilarginase with KEYTRUDA in

patients with extensive disease SCLC, with top-line data anticipated in the first half of 2020.

- 16 patients were enrolled across three cohorts in Phase 1b, with selection of the Phase 2 dose of 0.27 mg/kg/week of pegzilarginase in combination with KEYTRUDA. Of nine patients in the Phase 1b trial treated with 0.27 mg/kg/week, three patients had stable disease (SD) at 9 weeks and one partial response (PR) was observed. Three patients remain on treatment. The safety profile was consistent with pegzilarginase monotherapy observations.

- Completed enrollment in the single agent Phase 1 cohort expansions investigating pegzilarginase in patients with heavily pre-treated cutaneous melanoma, uveal melanoma and SCLC. In October 2018, the Company presented interim clinical data demonstrating monotherapy anti-tumor activity with pegzilarginase in the advanced melanoma cohorts. For SCLC, no objective responses and no new safety findings were observed in the 13 heavily-pretreated SCLC patients in the single-agent expansion arm, which completed enrollment in December 2018.

AEB4104 for Homocystinuria

- Initiated IND-enabling activities of AEB4104 for homocystinuria, a severe, progressive, rare genetic disease with significant unmet medical need. In homocystinuria disease models, AEB4104 lowered plasma homocysteine and homocystine levels with improved survival and correction of disease-related abnormalities. Aeglea is progressing manufacturing, pharmacology and other IND enabling activities to support preclinical toxicology studies and advancement towards clinical trials. The Company anticipates filing an IND or CTA in the first quarter of 2020.

AEB5100 for Cystinuria

- Initiated IND-enabling activities for AEB5100, a modified human enzyme approach with a unique specificity for degrading cystine, for the treatment of patients with cystinuria. In a cystinuria disease model, AEB5100 lowered blood levels of cystine, decreased the amount of cystine in the urine and reduced both cystine crystal and kidney stone formation. Aeglea is progressing manufacturing, pharmacology and other IND enabling activities to support preclinical toxicology studies and advancement towards clinical trials. The Company anticipates filing an IND or CTA in the second half of 2020.

Corporate Highlights

- In February 2019, the Company closed an underwritten public offering, which resulted in total gross proceeds of $69 million, of 4.6 million shares of its common stock at a price to the public of $8.00 per share, including 1.1 million shares pursuant to the underwriters' exercise in full of their option to purchase additional shares of common stock. In addition, and in lieu of common stock, Aeglea sold to certain investors pre-funded warrants to purchase up to an aggregate 4.0 million shares of common stock at a purchase price of $7.9999 per warrant, which represents the per share public offering price for the common stock less the $0.0001 per share exercise price for each such pre-funded warrant.
- Russell Cox, who has served as an Aeglea director since June 2015, assumed the position of chairman of the board in 2019. Armen Shanafelt, Ph.D., who served as chairman since February 2014, will continue as a director.
- Leslie Sloan, Ph.D., was appointed chief operating officer, a new position within Aeglea. Dr. Sloan was previously senior vice president of operations and will continue to report to the CEO. Dr. Sloan will be responsible for a number of key areas at the company, including operational management of drug development, regulatory affairs, manufacturing, commercial readiness and program management.

Upcoming Events

- Aeglea will provide an oral presentation titled "Sustained Reductions in Plasma Arginine Following Pegzilarginase Administration in Patients with Arginase-1 Deficiency are Accompanied by Improvements in Mobility and Adaptive Behavior" on Sunday, April 7th from 4:00-4:15 p.m. PT at the Society for Inherited Metabolic Disorders (SIMD) annual meeting being held April 6-9 in Seattle, Washington.
- Aeglea will present at the following conferences, with details regarding the date and time of the presentations and webcasts announced prior to the events.

  - H.C. Wainwright & Co., Global Life Sciences Conference to be held April 7- 9th in London, England
  - 18th Annual Needham Healthcare Conference to be held April 9-10th in New York, NY

Fourth Quarter and Full Year 2018 Financial Results

As of December 31, 2018, Aeglea had available cash, cash equivalents and marketable securities of $74.5 million, which excludes approximately $69.0 million in gross proceeds from a public offering to new and existing shareholders in February 2019. Based on Aeglea's current operating plan, and taking into account the $69.0 million raised in February 2019, management believes it has sufficient capital resources to fund anticipated operations through the first quarter of 2021.

Grant revenues were the result of a $19.8 million research grant received from the Cancer Prevention and Research Institute of Texas (CPRIT). The grant contract concluded in May 2018 with the full $19.8 million in grant proceeds collected and recognized as revenue over the life of the award. Aeglea did not recognize any grant revenues in the fourth quarter of 2018, compared with $1.5 million in the fourth quarter of 2017. Grant revenues of $3.9 million were recognized in the year ended December 31, 2018, compared with $5.2 million in the year ended December 31, 2017.

Research and development expenses totaled $11.8 million for the fourth quarter of 2018, compared with $5.8 million for the fourth quarter of 2017. The increase was primarily due to additional clinical development activity of Aeglea's lead product candidate, pegzilarginase, and a ramp-up in manufacturing activities for pegzilarginase. Aeglea completed and exceeded enrollment in its Phase 1/2 clinical trial in patients with Arginase 1 Deficiency, completed enrollment of three single-agent cohort expansions in a Phase 1 clinical trial for advanced solid tumor patients, and completed enrollment of a Phase 1 combination trial in patients with small cell lung cancer.

Research and development expenses totaled $36.7 million for the year ended December 31, 2018, compared with $22.8 million for the year ended December 31, 2017. The increase was primarily associated with advancing the clinical development of pegzilarginase and expanding our internal clinical development capabilities and research laboratory. Aeglea completed and exceeded enrollment in its Phase 1/2 clinical trial for pegzilarginase in patients with Arginase 1 Deficiency. Additionally, the Company initiated and completed enrollment of three single-agent cohort expansions for the Phase 1 trial in patients with advanced solid tumors and completed enrollment of its Phase 1 combination trial in patients with small cell lung cancer.

General and administrative expenses totaled $3.5 million for the fourth quarter of 2018, compared with $2.3 million for the fourth quarter of 2017. This increase was primarily due to additional employee headcount, compensation, and consulting costs.

General and administrative expenses totaled $12.6 million for the year ended December 31, 2018, compared with $10.1 million in the year ended December 31, 2017. This increase was primarily due to additional employee headcount and compensation to support Company growth.

Net loss totaled $14.9 million and $6.5 million for the fourth quarter of 2018 and 2017, respectively, with non-cash stock compensation expense of $1.4 million and $0.7 million for the fourth quarter of 2018 and 2017, respectively. Net loss totaled $44.3 million and $27.2 million for the years ended December 31, 2018 and 2017, respectively, with non-cash stock compensation expense of $4.3 million and $2.5 million for the years ended December 31, 2018 and 2017, respectively.

Conference Call & Webcast Details

Aeglea will hold a conference call on Thursday, March 7, 2019 at 4:30 p.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 March 14, 2019 by dialing +1 (877) 660-6853 within the United States or +1 (201) 612-7415 internationally. The conference ID is 13688006.

To access the live and archived webcast of the presentation, please visit the Presentation & 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 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 interim Phase 1/2 data demonstrated clinical improvements and rapid and sustained lowering of plasma arginine in Arginase 1 Deficiency patients.

About AEB4104 in Homocystinuria

AEB4104 is a novel recombinant human enzyme that degrades the amino acid homocysteine and its related homocystine dimer. Aeglea is developing AEB4104 for the treatment of patients with cystathionine beta synthase (CBS) deficiency, also known as Classical Homocystinuria. Homocysteine accumulation plays a key role in multiple progressive and serious disease-related complications, including thromboembolic vascular events; skeletal abnormalities including severe osteoporosis; developmental delay and intellectual disability; lens dislocation and severe myopia. AEB4104 is intended to lower the abnormally high blood levels of homocysteine to the normal range in patients with homocystinuria. Preclinical data demonstrated that AEB4104 improved important disease-related abnormalities and survival in a mouse model of homocystinuria.

About AEB5100 in Cystinuria

AEB5100 is a novel recombinant human enzyme that degrades cystine and the parent amino acid cysteine. Aeglea is developing AEB5100 for the treatment of patients with cystinuria, a rare genetic disease characterized by frequent and recurrent kidney stone formation from an early age. Affected individuals have severe symptoms, frequent hospital admissions, a requirement for multiple procedural interventions and an increased risk of both hypertension and chronic kidney disease. Cystinuria occurs due to genetic

mutations in amino acid transporters that lead to increased amounts of cystine in the urine. The high urine cystine concentrations leads to kidney stones formation. Preclinical data demonstrated that AEB5100 lowered blood levels of cystine and cysteine, decreased the amount of cystine in the urine and reduced kidney stone formation in a mouse model of cystinuria.

About Pegzilarginase in Cancer

Pegzilarginase is an enhanced human arginase that enzymatically degrades the amino acid arginine. In some cancers, tumor cells are unable to produce 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. Aeglea 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 our cash forecasts, the timing and success of our clinical trials and related data, 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 Annual Report on Form 10-K for the year ended December 31, 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.

KEYTRUDA® is a registered trademark of Merck Sharp & Dohme Corp., a subsidiary of Merck & Co., Inc., Kenilworth, NJ, USA.

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Financials

Aeglea BioTherapeutics, Inc.
Consolidated Balance Sheets

(In thousands, except share and per share amounts)

	December 31,	
	2018	2017
ASSETS		
CURRENT ASSETS		
Cash and cash equivalents	$ 22,461	$ 12,817
Marketable securities	52,052	37,482
Accounts receivable - grant	—	3,078
Prepaid expenses and other current assets	2,158	1,614
Total current assets	76,671	54,991
Property and equipment, net	1,018	854
Other non-current assets	50	232
TOTAL ASSETS	$ 77,739	$ 56,077
LIABILITIES AND STOCKHOLDERS' EQUITY		
CURRENT LIABILITIES		
Accounts payable	$ 663	$ 389
Deferred revenue	—	20
Accrued and other current liabilities	9,576	5,220
Total current liabilities	10,239	5,629
Other non-current liabilities	72	111
TOTAL LIABILITIES	10,311	5,740
STOCKHOLDERS' EQUITY		
Preferred stock, $0.0001 par value; 10,000,000 shares authorized as of December 31, 2018 and December 31, 2017; no shares issued and outstanding as of December 31, 2018 and December 31, 2017	—	—
Common stock, $0.0001 par value; 500,000,000 shares authorized as of December 31, 2018 and December 31, 2017; 24,140,097 shares and 16,670,188 shares issued and outstanding as of December 31, 2018 and December 31, 2017, respectively	2	2
Additional paid-in capital	184,314	122,950
Accumulated other comprehensive loss	(27)	(102)
Accumulated deficit	(116,861)	(72,513)
TOTAL STOCKHOLDERS' EQUITY	67,428	50,337
TOTAL LIABILITIES AND STOCKHOLDERS' EQUITY	$ 77,739	$ 56,077

Aeglea BioTherapeutics, Inc.
Consolidated Statements of Operations

(In thousands, except share and per share amounts)

	Year Ended December 31,		
	2018	**2017**	**2016**
Revenues:			
Grant	$ 3,888	$ 5,205	$ 4,628
Operating expenses:			
Research and development	36,719	22,815	18,143
General and administrative	12,632	10,066	8,391
Total operating expenses	49,351	32,881	26,534
Loss from operations	(45,463)	(27,676)	(21,906)
Other income (expense):			
Interest income	1,172	482	244
Other expense	(57)	(42)	(36)
Total other income	1,115	440	208
Net loss	$ (44,348)	$ (27,236)	$ (21,698)
Net loss per share, basic and diluted	$ (2.13)	$ (1.80)	$ (2.22)
Weighted-average common shares outstanding, basic and diluted	20,822,560	15,128,192	9,791,728