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): August 10, 2020

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.)**

805 Las Cimas Parkway
Suite 100
Austin, TX **78746**
(Address of principal executive offices) **(Zip Code)**

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

N/A

(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))

Securities registered pursuant to Section 12(b) of the Act:

Title of each class	Trading symbol(s)	Name of each exchange on which registered
Common Stock, $0.0001 Par Value Per Share	AGLE	The Nasdaq Stock Market LLC (Nasdaq Global Market)

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 August 10, 2020, Aeglea BioTherapeutics, Inc. issued a press release announcing its financial results for the quarter ended June 30, 2020. 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 ended June 30, 2020, dated August 10, 2020.
104	Cover Page Interactive Data File (embedded within the Inline XBRL document).

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: August 10, 2020

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

Aeglea BioTherapeutics Reports Second Quarter 2020 Financial Results and Corporate Highlights

Pegzilarginase Showed Durable Clinical Response and Sustained Reduction in Plasma Arginine at 56 Week Analysis of Phase 1/2 Open-Label Extension Study

Initiated Phase 1/2 Clinical Trial of ACN00177 for the Treatment of Homocystinuria

Austin, Texas, August 10, 2020 - Aeglea BioTherapeutics, Inc. (NASDAQ:AGLE), a clinical-stage biotechnology company developing a new generation of human enzyme therapeutics as innovative solutions for rare and other high-burden diseases, today reported its second quarter 2020 financial results, and provided recent corporate and program highlights.

"Despite the operating challenges posed by the global pandemic, we continued to advance our pegzilarginase program in the first half of the year. The presentation of long-term data showing sustained lowering of arginine levels and durable clinical response with pegzilarginase treatment, as well as progress in our patient identification efforts, reinforce our belief in its potential as a life-changing therapy for those with Arginase 1 Deficiency and lay a strong foundation for the commercial launch of pegzilarginase," said Anthony Quinn, M.B Ch.B, Ph.D., president and chief executive officer of Aeglea. "Additionally, we initiated our Phase 1/2 clinical trial of ACN00177 for Homocystinuria in the second quarter. We are continuing our patient identification activities and look forward to dosing the first patient once the clinical sites are able to begin screening patients."

Recent Highlights and Updates

Pegzilarginase in Arginase 1 Deficiency

- In May, Aeglea presented results from its 56 week analysis from the Company's completed Phase 1/2 clinical trial and ongoing open-label extension study during a late-breaking oral presentation at the 6th Congress of the European Academy of Neurology. Key results include:
  - Treatment with pegzilarginase resulted in a significant reduction in plasma arginine from baseline with all 13 patients achieving plasma arginine levels within the target range (<200 µM).
  - 85% (11 of 13) of patients were clinical responders based on mobility improvements evaluated using three assessments: 6MWT (6 Minute Walk Test), GMFM (Gross Motor Function) Part D (standing) and Part E (walking, running, and jumping).
  - Pegzilarginase was shown to have a favorable safety profile with more than 750 doses administered.
- To date, Aeglea has identified more than 240 Arginase 1 Deficiency patients. The number of identified patients represents more than 50% and 30% of the estimated genetic prevalence patient population in the U.S. and key European markets (France, Germany, Spain, Italy and the United Kingdom), respectively.

ACN00177 in Homocystinuria

- Aeglea initiated its Phase 1/2 clinical trial for ACN00177, a novel engineered human enzyme therapy designed to treat Homocystinuria, a serious metabolic disorder characterized by elevated plasma homocysteine which leads to a wide range of life-altering complications and reduced life expectancy.

Corporate

- Eric Bradford, M.D., M.Sc., M.B.A. has been promoted to Chief Development Officer. Dr. Bradford will oversee the clinical programs for pegzilarginase and ACN00177 as well as shape the clinical development strategy for future programs from the Company's platform of novel human enzymes.
- Chief Medical Officer Ravi M. Rao, M.B BChir, Ph.D., will depart the company to pursue other opportunities. Dr. Rao will continue to support Aeglea in a medical advisor role through a transitional period.

"Ravi has been a valued and impactful member of the Aeglea team. While we are disappointed by his planned departure, we wish him the best as he returns to his roots in immunology research and development," said Dr. Quinn. "I look forward to working more closely with Eric as we continue to strengthen our capabilities and advance pegzilarginase towards potential approval and launch."

Upcoming Events

Aeglea will be attending the following virtual investor conferences in the coming quarter.

- Wells Fargo Securities Healthcare Conference, September 9-10
- H.C. Wainwright Healthcare Conference, September 13-15

- Cantor Fitzgerald Global Healthcare Conference, September 15-17

Further, Aeglea's leadership looks forward to participating in dialogue about the Company's enzyme therapeutics platform during the following industry events, with additional details to be announced.

- World Orphan Drug Congress USA 2020, August 24-26
- Child Neurology Society Annual Meeting-International Child Neurology Congress 2020, October 19-23

Second Quarter 2020 Financial Results

As of June 30, 2020, Aeglea had available cash, cash equivalents, marketable securities and restricted cash of $159.2 million. Based on Aeglea's current operating plans, management believes it has sufficient capital resources to fund anticipated operations through 2022.

Research and development expenses totaled $16.9 million for the second quarter of 2020 and $14.8 million for the second quarter of 2019. The increase was primarily associated with investing in manufacturing and pre-commercial activities for Aeglea's lead product candidate, pegzilarginase; ramp-up in manufacturing activities for ACN00177 in Homocystinuria; and personnel-related expenses offset by decreasing clinical development expenses as a result of completing a Phase 1/2 clinical trial in patients with Arginase 1 Deficiency and closing out cancer trials.

General and administrative expenses totaled $4.7 million for the second quarter of 2020 and $3.8 million for the second quarter of 2019. This increase was primarily due to additional employee headcount, ramping up commercial capabilities, and additional facilities to support company growth.

Net loss totaled $21.4 million and $18.0 million for the second quarter of 2020 and 2019, respectively, with non-cash stock compensation expense of $1.6 million and $1.2 million for the second quarter of 2020 and 2019, respectively.

About Pegzilarginase in Arginase 1 Deficiency

Pegzilarginase is an enhanced human arginase that enzymatically lowers levels of the amino acid arginine. Aeglea is developing pegzilarginase for the treatment of patients with Arginase 1 Deficiency (ARG1-D), a rare debilitating, progressive disease presenting in childhood with persistent hyperargininemia, spasticity, developmental delay, intellectual disability, seizures and early mortality. Pegzilarginase is intended for use as an enzyme therapy to reduce elevated blood arginine levels in patients with ARG1-D. Aeglea's Phase 1/2 and Phase 2 open-label extension data for pegzilarginase in patients with ARG1-D demonstrated clinical improvements and sustained lowering of plasma arginine. The Company's single, global pivotal Phase 3 PEACE trial is designed to assess the effects of treatment with pegzilarginase versus placebo over 24 weeks with a primary endpoint of plasma arginine reduction.

About ACN00177 in Homocystinuria

Aeglea is developing ACN00177 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, intellectual disability, lens dislocation and severe near-sightedness. ACN00177 has been designed as a novel recombinant human enzyme, which degrades the amino acid homocysteine and its related homocystine dimer. With this mechanism, ACN00177 is intended to lower the abnormally high blood levels of homocysteine in patients with Homocystinuria. Preclinical data demonstrated that ACN00177 improved important disease-related abnormalities and survival in a mouse model of Homocystinuria. The Company initiated a Phase 1/2 trial in the second quarter of 2020 and continues patient identification and administrative activities. The timing of first patient dosing in this Phase 1/2 trial will depend on determinations by individual sites as they adjust to impacts from COVID-19.

About Aeglea BioTherapeutics

Aeglea BioTherapeutics is a clinical-stage biotechnology company redefining the potential of human enzyme therapeutics to benefit people with rare and other high burden diseases. Aeglea's lead product candidate, pegzilarginase, is in a pivotal Phase 3 trial for the treatment of Arginase 1 Deficiency and has received both Rare Pediatric Disease and Breakthrough Therapy Designation. The Company initiated a Phase 1/2 clinical trial of ACN00177 for the treatment of Homocystinuria in the second quarter of 2020. Aeglea has an active discovery platform, with the most advanced program for Cystinuria. 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, timing and results of meetings with regulators, the timing of announcements and updates relating to our clinical trials and related data, our ability to enroll patients into our clinical trials, the expected impact of the COVID-19 pandemic on our operations and clinical trials, success in our collaborations, the potential addressable markets of the our product candidates 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, 2020 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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Financials

Aeglea BioTherapeutics, Inc.
Condensed Consolidated Balance Sheets
(Unaudited)

(In thousands, except share and per share amounts)

	June 30, 2020	December 31, 2019
ASSETS		
CURRENT ASSETS		
Cash and cash equivalents	$ 65,713	$ 19,253
Marketable securities	92,017	52,696
Prepaid expenses and other current assets	4,136	2,556
Total current assets	161,866	74,505
Restricted cash	1,500	1,500
Property and equipment, net	4,896	2,385
Operating lease right-of-use assets	4,557	4,726
Other non-current assets	92	67
TOTAL ASSETS	$ 172,911	$ 83,183
LIABILITIES AND STOCKHOLDERS' EQUITY		
CURRENT LIABILITIES		
Accounts payable	$ 4,151	$ 3,154
Operating lease liabilities	228	351
Accrued and other current liabilities	11,511	14,854
Total current liabilities	15,890	18,359
Non-current operating lease liabilities	4,695	4,712
Other non-current liabilities	68	31
TOTAL LIABILITIES	20,653	23,102
STOCKHOLDERS' EQUITY		
Preferred stock, $0.0001 par value; 10,000,000 shares authorized as of June 30, 2020 and December 31, 2019; no shares issued and outstanding as of June 30, 2020 and December 31, 2019	—	—
Common stock, $0.0001 par value; 500,000,000 shares authorized as of June 30, 2020 and December 31, 2019; 44,599,847 shares and 29,084,437 shares issued and outstanding as of June 30, 2020 and December 31, 2019, respectively	5	3
Additional paid-in capital	387,475	255,142
Accumulated other comprehensive income	39	51
Accumulated deficit	(235,261)	(195,115)
TOTAL STOCKHOLDERS' EQUITY	152,258	60,081
TOTAL LIABILITIES AND STOCKHOLDERS' EQUITY	$ 172,911	$ 83,183

Aeglea BioTherapeutics, Inc.
Condensed Consolidated Statements of Operations
(Unaudited)

(In thousands, except share and per share amounts)

	Three Months Ended June 30,		Six Months Ended June 30,	
	2020	2019	2020	2019
Operating expenses:				
Research and development	16,869	14,806	31,431	29,195
General and administrative	4,691	3,816	9,151	7,084
Total operating expenses	21,560	18,622	40,582	36,279
Loss from operations	(21,560)	(18,622)	(40,582)	(36,279)
Other income (expense):				
Interest income	161	619	461	1,126
Other expense, net	(19)	(16)	(25)	(33)
Total other income	142	603	436	1,093
Net loss	$ (21,418)	$ (18,019)	$ (40,146)	$ (35,186)
Net loss per share, basic and diluted	$ (0.40)	$ (0.55)	$ (0.93)	$ (1.14)
Weighted-average common shares outstanding, basic and diluted	52,941,603	32,840,357	43,019,670	30,936,623