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): May 10, 2021

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 May 10, 2021, Aeglea BioTherapeutics, Inc. issued a press release announcing its financial results for the quarter ended March 31, 2021. 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 March 31, 2021, dated May 10, 2021.
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: May 10, 2021

By: /s/ Anthony Quinn
Anthony Quinn, M.B Ch.B, Ph.D
Chief Executive Officer

Aeglea BioTherapeutics Reports First Quarter 2021 Financial Results and Corporate Highlights

Completed Patient Randomization for Phase 3 PEACE Clinical Trial; Topline Data Expected in Q4

Entered into Ex-U.S. Commercialization Agreement for Pegzilarginase with Upfront and Milestone Payments Up to $151.5 Million Plus Royalty Percentage in the Mid-Twenties

Launched THINK ARGININE™, a Disease Education and Diagnostic Testing Initiative for Arginase 1 Deficiency

Austin, Texas, May 10, 2021 - Aeglea BioTherapeutics, Inc. (NASDAQ:AGLE), a clinical-stage biotechnology company developing a new generation of human enzyme therapeutics as innovative solutions for rare metabolic diseases, today reported its first quarter 2021 financial results, and provided recent corporate and program highlights.

"We've had a strong start to 2021 and I am excited to see the significant momentum we have built for our pegzilarginase program from a clinical perspective as well as strengthening our commercial foundation in preparation for a potential FDA approval and launch. In just the last two months, we completed patient randomization for PEACE, secured a commercialization partner for pegzilarginase in Europe and the Middle East and launched THINK ARGININE, an Arginase 1 Deficiency disease education and diagnostic testing initiative in the United States," said Anthony Quinn, M.B Ch.B, Ph.D., president and chief executive officer of Aeglea. "This substantial progress sets us up well for the rest of 2021. We expect to provide topline data from the PEACE study in the fourth quarter, continue progressing our commercial strategy for pegzilarginase and advance our AGLE-177 clinical program in Homocystinuria. These accomplishments are critical milestones as we move closer to our mission of bringing impactful therapies to patients with rare metabolic disorders who currently have limited treatment options."

Recent Highlights and Updates

Corporate

- In March, Aeglea announced it has entered into a license and supply agreement with Immedica Pharma AB granting Immedica exclusive commercialization rights in Europe and several Middle Eastern countries. Aeglea will receive an upfront payment of $21.5 million and is eligible for commercial and regulatory milestones of up to approximately $130 million and mid-twenties percentage royalties on net sales.

Pegzilarginase in Arginase 1 Deficiency

- In April, the Company completed patient screening and randomization for PEACE, its pivotal Phase 3 clinical trial. Trial enrollment of 32 patients exceeded the target of 30 patients, underscoring the high levels of interest seen from patients, caregivers and investigators. Topline data are expected in the fourth quarter of 2021.
- In May, we announced the launch of THINK ARGININE, a disease education initiative to improve the awareness and diagnosis of Arginase 1 Deficiency (ARG1-D). The initiative consists of a comprehensive healthcare provider education campaign and a no-charge diagnostic testing program for adults and children with suspected ARG1-D in the United States.
    - The no-charge diagnostic testing program includes amino acid testing working with Mayo Clinic Laboratories and genetic testing partnered with Invitae.

Upcoming Events

Aeglea will participate in the upcoming virtual conferences and events:

- ISPOR 2021, May 17-20
- Virtual Rare Disease Week on Capitol Hill 2021, July 14-22

First Quarter 2021 Financial Results

As of March 31, 2021, Aeglea had available cash, cash equivalents, marketable securities and restricted cash of $128.5 million, which excludes the upfront license receivable from Immedica for $21.5 million. Based on Aeglea's current operating plan, management believes it has sufficient capital resources to fund anticipated operations into 2023.

Research and development expenses totaled $11.9 million for the first quarter of 2021 and $14.6 million for the first quarter of 2020. The decrease was primarily associated with completing certain pre-commercial manufacturing activities for Aeglea's lead product

candidate, pegzilarginase.

General and administrative expenses totaled $6.4 million for the first quarter of 2021 and $4.5 million for the first quarter of 2020. This increase was primarily due to ramping-up the Company’s commercial capabilities and infrastructure.

Net loss totaled $18.2 million and $18.7 million for the first quarter of 2021 and 2020, respectively, with non-cash stock compensation expense of $1.8 million and $1.3 million for the first quarter of 2021 and 2020, respectively.

About Pegzilarginase in Arginase 1 Deficiency

Pegzilarginase is a novel recombinant human enzyme, which has been shown to rapidly and sustainably lower levels of the amino acid arginine in plasma. Aeglea is developing pegzilarginase for the treatment of patients 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. 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 ongoing 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. Pegzilarginase has received multiple regulatory designations, including Rare Pediatric Disease, Breakthrough, Fast Track and Orphan Drug Designations from the FDA as well as Orphan Drug Designation from the European Medicines Agency.

About AGLE-177 in Homocystinuria

AGLE-177 is a novel recombinant human enzyme, which degrades the amino acid homocysteine and its related homocystine dimer. Aeglea is developing AGLE-177 for the treatment of patients with cystathionine beta synthase (CBS) deficiency, also known as Classical Homocystinuria, a rare inherited disorder of methionine metabolism that results in elevated levels of homocysteine and homocystine. 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. Preclinical data demonstrated that AGLE-177 improved important disease-related abnormalities and survival in a mouse model of Homocystinuria. AGLE-177 has received U.S. Rare Pediatric Disease and Orphan Drug Designations as well as EU Orphan Drug Designation. Aeglea initiated a Phase 1/2 trial in 2020 and continues patient identification and administrative activities.

About Aeglea BioTherapeutics

Aeglea BioTherapeutics is a clinical-stage biotechnology company redefining the potential of human enzyme therapeutics to benefit people with rare metabolic diseases with limited treatment options. 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 AGLE-177 for the treatment of Homocystinuria in 2020. AGLE-177 has also been granted Rare Pediatric Disease Designation. Aeglea has an active discovery platform focused on engineering small changes in human enzymes to have a big impact on the lives of patients and their families. For more information, please visit http://aeglea.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 ability to obtain regulatory approval for, and commercialize, pegzilarginase, recognize milestone and royalty payments from our agreement with Immedica, 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 Annual Report on Form 10-Q for the quarter ended March 31, 2021 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)

	March 31, 2021	December 31, 2020
ASSETS		
CURRENT ASSETS		
Cash and cash equivalents	$ 80,231	$ 90,095
Marketable securities	46,429	56,178
Accounts receivable - license	21,500	—
Prepaid expenses and other current assets	3,252	3,516
Total current assets	151,412	149,789
Restricted cash	1,845	1,842
Property and equipment, net	5,518	5,642
Operating lease right-of-use assets	4,123	4,230
Other non-current assets	716	115
TOTAL ASSETS	$ 163,614	$ 161,618
LIABILITIES AND STOCKHOLDERS' EQUITY		
CURRENT LIABILITIES		
Accounts payable	$ 1,878	$ 2,254
Operating lease liabilities	315	319
Deferred revenue	19,226	—
Accrued and other current liabilities	10,951	13,870
Total current liabilities	32,370	16,443
Non-current operating lease liabilities	5,009	5,129
Deferred revenue, net of current portion	2,274	—
Other non-current liabilities	206	214
TOTAL LIABILITIES	39,859	21,786
STOCKHOLDERS' EQUITY		
Preferred stock, $0.0001 par value; 10,000,000 shares authorized as of March 31, 2021 and December 31, 2020; no shares issued and outstanding as of March 31, 2021 and December 31, 2020	—	—
Common stock, $0.0001 par value; 500,000,000 shares authorized as of March 31, 2021 and December 31, 2020; 49,019,901 shares and 47,959,086 shares issued and outstanding as of March 31, 2021 and December 31, 2020, respectively	5	5
Additional paid-in capital	417,951	415,824
Accumulated other comprehensive income	25	11
Accumulated deficit	(294,226)	(276,008)
TOTAL STOCKHOLDERS' EQUITY	123,755	139,832
TOTAL LIABILITIES AND STOCKHOLDERS' EQUITY	$ 163,614	$ 161,618

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

(In thousands, except share and per share amounts)

	Three Months Ended March 31,	
	2021	**2020**
Operating expenses:		
Research and development	11,855	14,562
General and administrative	6,354	4,460
Total operating expenses	18,209	19,022
Loss from operations	(18,209)	(19,022)
Other income (expense):		
Interest income	22	300
Other expense, net	(31)	(6)
Total other income (expense)	(9)	294
Net loss	$ (18,218)	$ (18,728)
Net loss per share, basic and diluted	$ (0.28)	$ (0.57)
Weighted-average common shares outstanding, basic and diluted	65,604,336	33,097,736