June 1, 2017

Eleven Biotherapeutics Announces Data and Safety Monitoring Board (DSMB) Recommendation to Continue Phase 3 Registration Trial with Vicinium™ in Non-Muscle Invasive Bladder Cancer Based on Review of Safety and Efficacy Data

Phase 3 Trial Enrollment Exceeds 50%

Company Expects to Complete Patient Enrollment in 2H2017 and to Report Topline 3-Month Data in 2Q2018

CAMBRIDGE, Mass.--(BUSINESS WIRE)-- Eleven Biotherapeutics, Inc. (NASDAQ:EBIO), a late-stage clinical oncology company advancing a broad pipeline of novel product candidates based on its Targeted Protein Therapeutics (TPTs) platform, today announced that its Phase 3 registration trial of Vicinium in non-muscle invasive bladder cancer (NMIBC) has exceeded 50% enrollment and that the independent Data and Safety Monitoring Board (DSMB) for the trial has recommended that the trial continue as planned. The DSMB reviewed available data to assess the risk/benefit to patients on drug and recommended that the trial continue without modification.

"We are very pleased that the DSMB recommended we continue enrolling our Phase 3 trial after their review of available safety and efficacy data," said Stephen Hurly, President and Chief Executive Officer of Eleven Biotherapeutics. "Patients with Bacillus Calmette-Guérin (BCG) unresponsive NMIBC have limited therapeutic options and frequently require cystectomies to prevent disease progression. Bladder removal, however, is a serious and life-altering surgery associated with significant morbidity and mortality. Vicinium may offer patients a positive non-surgical risk/benefit profile versus the standard of care. We look forward to advancing our trial as we continue to gain important information about the activity of Vicinium in patients with NMIBC."

"Urologists are looking for new treatment options for their patients with NMIBC once they stop responding to BCG. Their patients want alternatives to cystectomy. However, the NMIBC treatment landscape has not seen meaningful advances in forty years," commented Arthur DeCillis, Chief Medical Officer of Eleven Biotherapeutics. "With this positive step behind us, we look forward to continuing our Phase 3 registration trial and to reporting topline 3-month data in the second quarter of next year."

Vicinium is a single protein anti-epithelial cell adhesion molecule (anti-EpCAM) antibody fragment fused with *Pseudomonas* Exotoxin A (ETA) that is designed to specifically target and deliver a potent anti-cancer payload directly into tumor cells. The ongoing Phase 3 registration trial is a single-arm study evaluating Vicinium in patients with high-grade NMIBC, who have previously received two courses of BCG and whose disease is now BCG-unresponsive. Eleven Biotherapeutics plans to enroll 134 patients, at over 70 centers in the United States and Canada. The trial's primary endpoint is the complete response rate in patients with carcinoma-in-situ (CIS). Secondary endpoints include time to disease recurrence and event free survival. The Company expects to complete patient enrollment in the second half of 2017 and to report 3-month data in the second quarter of 2018.

About Eleven Biotherapeutics

Eleven Biotherapeutics, Inc. is a late-stage clinical oncology company advancing a broad pipeline of novel product candidates based upon the Company's TPT platform. The Company's TPTs incorporate a tumor-targeting antibody fragment and a protein cytotoxic payload into a single protein molecule in order to achieve focused tumor cell killing. The Company believes its TPT approach offers significant advantages in treating cancer over existing ADC technologies. The Company believes its TPTs provide effective tumor targeting with broader cancer cell-killing properties than are achievable with small molecule payloads that require tumor cell proliferation and face multi-drug resistance mechanisms. Additionally, the Company believes that its TPT's cancer cell-killing properties promote an anti-tumor immune response that will potentially combine well with immuno-oncology drugs such as checkpoint inhibitors. For more information please refer to the Company's website at www.elevenbio.com.

Cautionary Note on Forward-Looking Statements

Any statements in this press release about future expectations, plans and prospects for the Company, the Company's strategy, future operations, and other statements containing the words "anticipate," "believe," "estimate," "expect," "intend," "may," "plan," "predict," "project," "target," "potential," "will," "would," "could," "should," "continue," and similar expressions, constitute forward-looking statements within the meaning of The Private Securities Litigation Reform Act of 1995. Actual results may differ materially from those indicated by such forward-looking statements as a result of various important

factors, including: the uncertainties inherent in the initiation and conduct of clinical trials, our ability to successfully develop our product candidates and complete our planned clinical programs, our ability to obtain marketing approvals for our product candidates, expectations regarding our ongoing clinical trials, availability and timing of data from clinical trials, whether interim results from a clinical trial will be predictive of the final results of the trial or results of early clinical studies will be indicative of the results of future studies, the adequacy of any clinical models, expectations regarding regulatory approvals and other factors discussed in the "Risk Factors" section of the Company's Annual Report on Form 10-K, Quarterly Reports on Form 10-Q and other reports filed with the Securities and Exchange Commission. In addition, the forward-looking statements included in this press release represent the Company's views as of the date hereof. The Company anticipates that subsequent events and developments will cause the Company's views to change. However, while the Company may elect to update these forward-looking statements at some point in the future, the Company specifically disclaims any obligation to do so. These forward-looking statements should not be relied upon as representing the Company's views as of any date subsequent to the date hereof.

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