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CStone Pharmaceuticals

基石藥業

(Incorporated in the Cayman Islands with limited liability)

(**Stock Code: 2616**)

VOLUNTARY ANNOUNCEMENT

CSTONE'S PARTNER BLUEPRINT MEDICINES ANNOUNCED PART 1 RESULTS FROM PRIONEER TRIAL SHOWING BROAD ACTIVITY OF AVAPRITINIB ACROSS MEASURES OF MAST CELL BURDEN, CLINICAL OUTCOMES AND QUALITY OF LIFE IN INDOLENT SYSTEMIC MASTOCYTOSIS

The partner of CStone Pharmaceuticals (the "**Company**" or "**CStone**"), Blueprint Medicines Corporation (NASDAQ: BPMC) ("**Blueprint Medicines**"), announced on March 16, 2020 the updated results from the Phase 2 PIONEER trial of avapritinib in patients with indolent systemic mastocytosis ("**SM**"), demonstrating significant clinical improvements versus placebo. In Part 1 of the PIONEER trial, patients treated with avapritinib showed a statistically significant mean decline of approximately 30 percent in total symptom score ("**TSS**") in the 16th week, as measured by the *Indolent SM Symptom Assessment Form* ("**ISM-SAF**"), and reductions in symptom scores have deepened over time. In addition, patients treated with avapritinib achieved consistent improvements across objective measures of mast cell burden and patient-reported quality of life. Studies revealed that avapritinib was well-tolerated with no patients discontinuing treatment due to adverse events ("**AEs**"). Based on the data from Part 1 of the PIONEER trial, 25 mg once daily ("**QD**") has been selected as the recommended Part 2 dose ("**RP2D**"). Results from this data presentation will be available on an American Academy of Allergy, Asthma & Immunology ("**AAAAI**") virtual forum, which was established following the cancellation of the 2020 AAAAI Annual Meeting: https://education.aaaai.org/annual-meeting-abstracts/.

The key updates include:

- Avapritinib resulted in a statistically significant improvement in patient-reported outcomes, which is a primary measure of clinical benefit in PIONEER;
- Avapritinib was well-tolerated across all three doses, and no patient discontinuations took place due to AEs;
- 25 mg QD was selected as RP2D, based on consistent and clinically important improvements across multiple measures of efficacy and a well-tolerated safety profile.

SM is a rare disease driven by the KIT D816V mutation and characterized by uncontrolled mast cell proliferation and activation. The disorder can lead to debilitating symptoms and life-threatening complications. Avapritinib is a potent and highly selective inhibitor of D816V mutant KIT.

Blueprint Medicines plans to initiate patient screening for the registration-enabling Part 2 of the PIONEER trial in June 2020. Part 2 is designed to evaluate the efficacy of avapritinib at the RP2D versus placebo. Blueprint Medicines anticipates completing enrollment in Part 2 of the PIONEER trial by the end of 2020.

Blueprint Medicines has entered into an exclusive collaboration and license agreement with CStone Pharmaceuticals for the development and commercialization of avapritinib, pralsetinib and fisogatinib in mainland China, Hong Kong, Macau and Taiwan. Blueprint Medicines retains development and commercial rights for all three licensed products in the rest of the world.

Highlights from the Part 1 PIONEER Trial Data in Indolent SM

Part 1 of the PIONEER trial was designed to determine the RP2D by evaluating three doses of avapritinib (25 mg, 50 mg and 100 mg QD) versus placebo. Key eligibility criteria include adults with indolent SM confirmed by central pathology review and moderate-to-severe symptom burden despite best supportive care medicines. A total of 39 patients were enrolled in Part 1 across four concurrent cohorts, consisting of ten patients each in the three avapritinib dose cohorts and nine patients in the placebo cohort.

Patient-reported outcomes ("**PRO**") data were collected using the ISM-SAF, which was designed with input from disease experts, patients and regulatory authorities to support registration. The data cut-off date for all results are as of December 27, 2019.

Baseline Patient Characteristics

Patients had high symptom burden at baseline, with a mean ISM-SAF TSS of 53 on a scale of 0 to 110. Eight patients (21%) had an Eastern Cooperative Oncology Group Performance Status of 2, reflecting the inability to carry out any work activities. Patients received a median of four best supportive care medicines at baseline (range: two to nine). Median serum tryptase was 45 micrograms per liter (the upper limit of normal is 11.4 micrograms per liter). A high sensitivity polymerase chain reaction assay on peripheral blood detected the KIT D816V mutation in 37 patients (95%).

Clinical Activity

Avapritinib showed broad activity across measures of mast cell burden, the PRO clinical benefit measure and quality of life. The consistency of results observed across multiple measures of disease burden support the further evaluation of avapritinib in indolent SM. In the 16th week, patients had a statistically significant reduction in ISM-SAF TSS, with a mean improvement of approximately 30% across all avapritinib dose cohorts compared to approximately 3% in the placebo cohort (p=0.001). As of the data cut-off date, 37 patients (95%) have remained on study with a median follow-up of 18 weeks.

Results from the 25 mg QD dose cohort demonstrate important clinical activity, including meaningful declines in serum tryptase, bone marrow mast cells and KIT D816V allele burden. Treatment with avapritinib led to consistent reductions in the ISM-SAF TSS, gastrointestinal domain, skin domain and each individual symptom. Symptom improvements in patients treated at 25 mg QD continued to deepen over time.

Mean Percent Changes in ISM-SAF at 16 Weeks		
	Avapritinib, 25 mg QD	**Placebo**
TSS	-31%	-3%
Skin domain	-37%	+3%
Gastrointestinal domain	-25%	+6%
Neurological symptoms	-26%	-8%

Data from the Mastocytosis Quality of Life (MC-QoL) questionnaire, a PRO tool developed for mast cell disorders, show improvements in quality of life for patients receiving avapritinib and support the results observed with the ISM-SAF. Patients in the 25 mg QD dose cohort had a mean reduction of 34 percent in the total MC-QoL score and improvements in all four domains assessed (i.e. symptoms, social life functioning, emotions and skin). A 7% increase from baseline was observed in the placebo cohort.

Safety

The safety profile of avapritinib supports chronic dosing in indolent SM. All doses of avapritinib were well-tolerated and no patients discontinued treatment due to AEs. No patients treated with avapritinib in the 25 mg QD dose cohort had serious AEs, Grade 3 or higher AEs, or dose modifications. In the placebo cohort, two patients (22%) had Grade 3 AEs, one with seizure and one with diffuse cutaneous mastocytosis; these events also met criteria for serious AEs.

About the Clinical Development Program for Avapritinib

Avapritinib is an investigational, oral precision therapy that selectively and potently inhibits KIT and PDGFRA mutant kinases. Blueprint Medicines is pursuing a broad clinical development program for avapritinib for advanced, smoldering and indolent SM, as well as across multiple lines of GIST treatment.

Avapritinib is uniquely designed to selectively bind and inhibit D816V mutant KIT, the common driver of disease in approximately 95% of all SM patients. Preclinical studies have shown avapritinib potently inhibited

KIT D816V at sub-nanomolar potencies with minimal off-target activity. In addition, avapritinib has demonstrated broad inhibition of KIT and PDGFRA mutations associated with GIST, including potent activity against activation loop mutations that are associated with resistance to currently approved therapies.

About the Phase 2 PIONEER Trial

PIONEER is a randomized, double-blind, placebo-controlled and registration-enabling trial evaluating avapritinib in patients with indolent and smoldering SM. The trial includes three parts: dose-finding Part 1, registration-enabling Part 2 and long-term treatment Part 3. All patients who complete Parts 1 or 2 will have an opportunity to continue to receive treatment with avapritinib in Part 3. Key trial endpoints include the change in patient-reported disease symptoms as measured by the ISM-SAF TSS, quantitative measures of mast cell burden and safety. Part 1 has completed patient enrollment. Blueprint Medicines plans to initiate patient screening for Part 2 in June 2020 at sites in the United States, Canada and European Union.

About CStone

CStone is a biopharmaceutical company focused on developing and commercializing innovative immuno-oncology and precision medicines to address the unmet medical needs of cancer patients in China and worldwide. Established in 2015, CStone has assembled a world-class management team with extensive experience in innovative drug development, clinical research, and commercialization. The company has built an oncology-focused pipeline of 15 drug candidates with a strategic emphasis on immuno-oncology combination therapies. Currently, five late-stage candidates are at pivotal trials. With an experienced team, a rich pipeline, a robust clinical development-driven business model and substantial funding, CStone's vision is to become globally recognized as a leading Chinese biopharmaceutical company by bringing innovative oncology therapies to cancer patients worldwide.

For more information about CStone, please visit: www.cstonepharma.com.

By order of the Board
CStone Pharmaceuticals
Dr. Frank Ningjun Jiang
Chairman

Suzhou, People's Republic of China, March 19, 2020

As at the date of this announcement, the Board of Directors of the Company comprises Dr. Frank Ningjun Jiang as Chairman and Executive Director, Dr. Wei Li, Mr. Qun Zhao, Mr. Yanling Cao, Mr. Guobin Zhang and Dr. Lian Yong Chen as non-executive Directors, and Dr. Paul Herbert Chew, Mr. Ting Yuk Anthony Wu and Mr. Hongbin Sun as independent non-executive Directors.