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 PUBLICATION IN *THE LANCET ONCOLOGY* SHOWING DURABLE CLINICAL BENEFITS OF AVAPRITINIB IN NAVIGATOR TRIAL PATIENTS WITH PDGFRA D842V MUTANT GIST

The partner of CStone Pharmaceuticals (the "**Company**" or "**CStone**"), Blueprint Medicines Corporation (NASDAQ: BPMC) ("**Blueprint Medicines**"), announced on June 29, 2020 that *The Lancet Oncology* published data from the NAVIGATOR clinical trial showing an 81% overall survival ("**OS**") rate at 24 months and a well-tolerated safety profile for avapritinib in patients with advanced PDGFRA D842V mutant gastrointestinal stromal tumor ("**GIST**"). The paper, titled "avapritinib in advanced PDGFRA D842V-mutant gastrointestinal stromal tumour (NAVIGATOR): a multicentre, open-label, phase 1 trial," was published online in *The Lancet Oncology* on June 29, 2020.

Blueprint Medicines has an exclusive collaboration and license agreement with CStone for the development and commercialization of avapritinib, fisogatinib and pralsetinib in the mainland of the People's Republic of China, Hong Kong Special Administrative Region, Macau Special Administrative Region and Taiwan. Blueprint Medicines retains development and commercial rights for all three licensed products in the rest of the world.

The key updates include:

- *The Lancet Oncology* paper reported efficacy and safety results from the NAVIGATOR trial, including all patients enrolled in the dose escalation part of the trial and the subset of patients with PDGFRA

D842V mutant GIST enrolled in the expansion part of the trial. The efficacy population comprised 56 patients with PDGFRA D842V mutant GIST. The safety population comprised 82 patients, including 26 patients with non-PDGFRA D842V mutant GIST enrolled in the dose escalation part of the trial. All results were as of November 16, 2018;

- In patients with PDGFRA D842V mutant GIST, the overall response rate was 88% (95% CI: 76-95%) with 9% of patients achieving a complete response. avapritinib demonstrated durable clinical benefit in this patient population with a 12-month duration of response rate of 70% (95% CI: 54-87%), a 12-month progression-free survival rate of 81% (95% CI: 69-93%) and a 24-month OS rate of 81% (95% CI: 67-94%);

- avapritinib was generally well-tolerated with most treatment-related adverse events reported as grade 1 or 2. The most common treatment-related adverse events were nausea, fatigue, diarrhea, periorbital edema, anemia, decreased appetite, vomiting and memory impairment. Cognitive effects occurred in 40% of patients, with the majority of events reported as grade 1.

About avapritinib

avapritinib is a kinase inhibitor approved by the Food and Drug Administration of the United States ("**USFDA**") under the brand name AYVAKIT™ for the treatment of adults with unresectable or metastatic GIST harboring a PDGFRA exon 18 mutation, including PDGFRA D842V mutations.

avapritinib is not approved for the treatment of any other indication, including systemic mastocytosis, in the U.S. by the USFDA or for any indication in any other jurisdiction by any other health authority.

Blueprint Medicines is developing avapritinib globally for the treatment of advanced, smoldering and indolent systemic mastocytosis. The USFDA granted a breakthrough therapy designation to avapritinib for the treatment of advanced systemic mastocytosis, including the subtypes of aggressive systemic mastocytosis, systemic mastocytosis with an associated hematologic neoplasm and mast cell leukemia.

About GIST

GIST is a sarcoma, or tumor of bone or connective tissue, of the GI tract. Tumors arise from cells in the wall of the GI tract and occur most often in the stomach or small intestine. Most patients are diagnosed between the ages of 50 to 80, and diagnosis is typically triggered by GI bleeding, incidental findings during surgery or imaging and, in rare cases, tumor rupture or GI obstruction.

About 5 to 6% of primary GIST cases are caused by a PDGFRA D842V mutation, the most common PDGFRA exon 18 mutation. Prior to the USFDA approval of avapritinib, there were no highly effective treatments for PDGFRA D842V mutant GIST. Published data have shown poor outcomes in patients with PDGFRA D842V mutant GIST treated with imatinib and other approved therapies, including a median OS of 15 months, a median progression-free survival of 3 months and an overall response rate of 0%.

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 at the end of 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, July 6, 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.