The forward-looking statements made in this announcement relate only to the events or information as of the date on which the statements are made in this announcement. Except as required by law, we undertake no obligation to update or revise publicly any forward-looking statements, whether as a result of new information, future events or otherwise, after the date on which the statements are made or to reflect the occurrence of unanticipated events. You should read this announcement completely and with the understanding that our actual future results or performance may be materially different from what we expect. In this announcement, statements of, or references to, our intentions or those of any of our directors and/or our Company are made as of the date of this announcement. Any of these intentions may alter in light of future development.

CStone Pharmaceuticals

基石藥業

(Incorporated in the Cayman Islands with limited liability)

(**Stock Code: 2616**)

VOLUNTARY ANNOUNCEMENT

CSTONE ANNOUNCES FIRST PATIENT IN CHINA DOSED WITH AVAPRITINIB IN GLOBAL PHASE III CLINICAL TRIAL IN ADVANCED GIST

CStone Pharmaceuticals (the "**Company**" or "**CStone**") announces that the first patient has been dosed in China with avapritinib in the ongoing global Phase III VOYAGER clinical trial. This study is designed to evaluate the safety and efficacy of avapritinib as a third-line or fourth-line treatment for patients with gastrointestinal stromal tumors ("**GIST**"), in comparison with that of regorafenib, the current standard of care treatment for GIST. To be eligible, patients must have been previously treated with imatinib and one or two additional tyrosine kinase inhibitors. The trial's primary efficacy endpoint is progression-free survival (PFS).

Avapritinib, an orally available, potent and highly selective inhibitor of KIT and PDGFRA, was discovered by CStone's partner Blueprint Medicines Corporation (NASDAQ: BPMC) ("**Blueprint**"). Approximately 90% of GIST cases are associated with mutations of KIT and PDGFRA tyrosine kinases, leading to dysregulation of cell growth. Previously published preclinical results have shown that avapritinib can potently treat GIST associated with KIT and PDGFRA mutations.

Clinical data from the ongoing Phase I NAVIGATOR study presented in June 2019 demonstrated encouraging anti-tumor activity and favorable tolerability in patients with PDGFRA Exon 18 mutant and fourth-line GIST, two populations with no effective therapies. Blueprint has recently submitted a New Drug Application ("**NDA**") to U.S. Food and Drug Administration ("**FDA**") for this indication.

As of the data cut-off date of November 16, 2018 (the "**data cut-off date**"):

- In 43 evaluable patients with PDGFRA Exon 18 mutant GIST (including 38 patients with PDGFRα D842V-driven GIST), the objective response rate ("**ORR**") was 86% and the median duration of response ("**DOR**") was not reached;

- In 111 evaluable patients with fourth-line GIST, the ORR was 22% and the median DOR was 10.2 months; and

- avapritinib had a favorable safety profile, with most adverse events determined by investigators to be Grade 1 or 2 as of the data cut-off date.

Dr. Frank Ningjun Jiang, chairman, executive director and chief executive officer of CStone, commented: "Development of precision therapy in oncology is one of CStone's core strategies. GIST is a rare disease, and avapritinib has demonstrated its efficacy in treating GIST patients with tumor mutations resistant to currently available therapies. Our partner Blueprint has submitted the NDA for avapritinib to the FDA, and the agent is expected to be CStone's second product that gets approved in the United States. As we continue to make progress with the VOYAGER trial in China, we hope that the clinical data can soon support the approval of avapritinib in the country, and ultimately allow the product to benefit patients with advanced GIST who now lack effective treatments."

Dr. Jason Jianxin Yang, CStone's chief medical officer, noted: "In clinical trials in PDGFRA Exon 18 mutant and fourth-line GIST patients, avapritinib has demonstrated improved ORR and DOR compared to currently approved second-line and third-line treatments. We are pleased that the first Chinese patient has been enrolled and dosed in this Phase III trial of avapritinib as a third-line agent for advanced GIST. We will do our best to have more Chinese clinical trial centers participate in this important global clinical study."

About GIST

GIST is a sarcoma, or tumor of bone or connective tissue, of the gastrointestinal ("**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 age 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.

Most GIST cases are caused by a spectrum of clinically relevant mutations that force the KIT or PDGFRA protein kinases into an increasingly active state. Because currently available therapies primarily bind to the inactive protein conformations, certain primary and secondary mutations typically lead to treatment resistance and disease progression.

In unresectable or metastatic GIST, clinical benefits from existing treatments can vary by mutation type. Mutational testing is critical to tailor therapy to the underlying disease driver and is recommended in expert guidelines. Currently, there are no approved therapies for patients with KIT-driven GIST whose disease progresses beyond imatinib, sunitinib and regorafenib. In patients with metastatic PDGFRα D842V-driven GIST, progression occurs in a median of approximately three to four months with available therapy.

About Avapritinib

Avapritinib is an investigational and oral precision therapy that selectively and potently inhibits KIT and PDGFRA mutant kinases. It is a type 1 inhibitor designed to target the active kinase conformation; all

oncogenic kinases signal via this conformation. 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.

Blueprint is initially developing avapritinib for the treatment of advanced GIST, advanced systemic mastocytosis ("**SM**"), and indolent and smoldering SM. The FDA has granted Breakthrough Therapy Designation to avapritinib for two indications: one for the treatment of unresectable or metastatic GIST harboring the PDGFRα D842V mutation and one for the treatment of advanced SM, including the subtypes of aggressive SM, SM with an associated hematologic neoplasm and mast cell leukemia.

Blueprint has an exclusive collaboration and license agreement with CStone for the development and commercialization of avapritinib and certain other drug candidates in mainland China, Hong Kong, Macau and Taiwan. Blueprint retains development and commercial rights for avapritinib in the rest of the world.

About CStone

CStone is a biopharmaceutical company focused on developing and commercializing innovative immuno-oncology and molecularly-targeted drugs to address unmet medical needs for cancer patients in China and worldwide. Since the Company's inception in 2015, CStone has assembled a world-class management team that has a full spectrum of complementary skillsets from preclinical research to clinical development and commercialization. With combination therapies as a core strategy, the Company has built a rich oncology pipeline of 15 oncology drug candidates. Currently five late-stage drug candidates are at or near 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 and differentiated 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 9, 2019

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.