*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'S PARTNER AGIOS ANNOUNCES THE PHASE 3 CLARIDHY TRIAL OF TIBSOVO® (IVOSIDENIB) ACHIEVED ITS PRIMARY RESULT

The partner of CStone Pharmaceuticals (the "**Company**" or "**CStone**"), Agios Pharmaceuticals, Inc. (NASDAQ: AGIO) ("**Agios**"), announces that the global Phase 3 ClarIDHy trial of TIBSOVO® (ivosidenib) in previously treated cholangiocarcinoma patients with an isocitrate dehydrogenase 1 ("**IDH1**") mutation met its primary result. Treatment with TIBSOVO® demonstrated a statistically significant improvement in progression-free survival ("**PFS**") by independent radiology review compared with patients who received placebo. The safety profile observed in the study was consistent with previously published data.

A full analysis of the ClarIDHy trial will be submitted for presentation at the European Society for Medical Oncology Congress taking place in Barcelona, Spain from September 27 to October 1, 2019. Agios plans to submit a supplemental new drug application for TIBSOVO® in previously treated IDH1 mutant cholangiocarcinoma by the end of 2019.

Cholangiocarcinoma is a very aggressive tumor for which surgical resection is the primary treatment option. Many patients are undiagnosed until the mid-to-late stages when they have missed the time window for surgery. At present, there is no standard second- and third-line treatment for patients with advanced cholangiocarcinoma. Patients typically suffer a poor prognosis and short-term survival.

China is one of the countries with the highest rate of incidence of cholangiocarcinoma which may be associated with a variety of high-risk factors including hepatitis B and clonorchis sinensis infection, diseases that are endemic in China.

In June 2018, the Company entered into an exclusive collaboration and license agreement with Agios to develop and commercialize TIBSOVO® in Greater China region.

**ClarIDHy Phase 3 Trial**

The ClarIDHy trial is a global, randomized Phase 3 clinical trial in previously treated IDH1 mutant cholangiocarcinoma patients who have documented disease progression following one or two systemic therapies in the advanced setting. As of the January 31, 2019 data cut-off, the participation of 185 patients was randomized.

- Patients were randomized in 2:1 ratio to receive either single-agent TIBSOVO® 500 mg once daily or placebo with crossover to TIBSOVO® permitted at the time of documented radiographic progression per RECIST 1.1.

- The primary result of the trial is PFS as evaluated by independent radiology review with secondary result including investigator evaluated PFS, safety and tolerability, overall response rate, overall survival, duration of response, PK/PD and quality of life assessments.

- The study was designed with 96% power to detect a hazard ratio of 0.5 for PFS (TIBSOVO® versus placebo), with a one-sided alpha of 0.025.

- Thermo Fisher Scientific (NYSE: TMO) is providing next-generation sequencing to detect IDH1 mutations for all tumor samples as inclusion criteria for enrollment in the study and will develop and commercialize the validated companion diagnostic.

TIBSOVO® has not been approved in any country for the treatment of patients with advanced cholangiocarcinoma to date.

**About TIBSOVO® (ivosidenib)**

TIBSOVO® (ivosidenib) is indicated in the United States for the treatment of acute myeloid leukemia ("**AML**") with a susceptible IDH1 mutation as detected by the test approved by U.S. Food and Drug Administration in:

- adult patients with newly-diagnosed AML who are more than 75 years old or who have comorbidities that preclude use of intensive induction chemotherapy; and

- adult patients with relapsed or refractory AML.

For more information about TIBSOVO® (ivosidenib), please visit: www.tibsovo.com.

**About CStone**

CStone is a biopharmaceutical company focused on developing and commercializing innovative immuno-oncology and precision medicine 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 with a strategic emphasis on immuno-oncology combination therapies. Currently, 4 late-stage 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 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, May 23, 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.*