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 RECEIVES APPROVAL IN CHINA TO INITIATE IVOSIDENIB PHASE I BRIDGING REGISTRATIONAL STUDY FOR THE TREATMENT OF IDH1 MUTANT RELAPSED OR REFRACTORY AML

CStone Pharmaceuticals (the "**Company**" or "**CStone**") announces that it has received approval from China National Medical Products Administration ("**NMPA**") to initiate a Phase I bridging registrational study of ivosidenib (TIBSOVO) in China for the treatment of relapsed or refractory ("**R/R**") acute myeloid leukemia ("**AML**") with an isocitrate dehydrogenase-1 ("**IDH1**") mutation. This stand-alone trial is designed to evaluate the efficacy, safety and pharmacokinetics of ivosidenib in patients in China with IDH1 mutant R/R AML.

AML is the most common type of acute leukemia in adults and is characterized by rapid disease progression. There are approximately 20,000 new cases of AML in the United States each year, with a five-year survival rate of approximately 27%, as compared to at least 30,000 new cases in China annually and a five-year survival rate of below 20%. The majority of AML patients develop tolerance to treatments or eventually relapse, leading to R/R AML which has a poor prognosis. With an increasing life expectancy and aging population in China, the incidence of AML may rise significantly in the country. Interruption to the differentiation of hematopoietic stem cells due to IDH1 mutation is associated with around 6% to 10% of all AML cases. Among the currently approved treatments for AML in China, there is no effective therapy for this patient population.

Discovered and developed by CStone's partner, Agios Pharmaceuticals, Inc. (NASDAQ: AGIO), ivosidenib was approved by the U.S. Food and Drug Administration ("**FDA**") in July 2018 for the treatment of adult R/R AML patients with an IDH1 mutation as detected by an FDA-approved test.

"Ivosidenib is the first and only approved precision therapy in the United States that targets IDH1 mutation in R/R AML and the only asset in CStone's pipeline that has obtained a marketing authorization. This approval for the registrational study in China marks a big milestone for us," commented Dr. Frank Ningjun Jiang, executive director and chief executive officer of CStone. "Through precision therapies, patients can potentially benefit from treatments targeting the specific genetic mutations that drive the cancer. CStone remains committed to advancing precision therapies and bringing more targeted treatment options to patients."

"Ivosidenib was first approved as a treatment for IDH1 mutant R/R AML. The FDA recently approved a supplemental New Drug Application for ivosidenib to include newly diagnosed AML patients with an IDH1 mutation who are more than 75 years old or who have comorbidities that preclude the use of intensive induction chemotherapy. In March 2019, the FDA granted Breakthrough Therapy designation for ivosidenib in combination with azacitidine for the treatment of the same newly diagnosed AML patients. These milestones demonstrate ivosidenib's promising clinical utility. We are poised to rapidly roll out this trial for ivosidenib as a monotherapy for R/R AML in China. We hope the study will generate favorable clinical data, paving the way for the drug to quickly enter the China market," noted Dr. Jason Jianxin Yang, CStone's chief medical officer.

About TIBSOVO (ivosidenib)

In the United States, TIBSOVO is indicated for the treatment of AML with a susceptible IDH1 mutation as detected by an FDA-approved test 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.

IMPORTANT SAFETY INFORMATION

WARNING: DIFFERENTIATION SYNDROME

Patients treated with TIBSOVO have experienced symptoms of differentiation syndrome, which can be fatal if not treated effectively. Symptoms may include fever, dyspnea, hypoxia, pulmonary infiltrates, pleural or pericardial effusions, rapid weight gain or peripheral edema, hypotension, and hepatic, renal or multi-organ dysfunction. If differentiation syndrome is suspected, initiate corticosteroid therapy and hemodynamic monitoring until the relief of symptoms.

WARNINGS AND PRECAUTIONS

Differentiation Syndrome: See Boxed WARNING. In the clinical trial, 25% (7/28) of patients with newly diagnosed AML and 19% (34/179) of patients with relapsed or refractory AML treated with TIBSOVO experienced differentiation syndrome. Differentiation syndrome is associated with rapid proliferation and differentiation of myeloid cells and may be life-threatening or fatal if not treated effectively. Symptoms of differentiation syndrome in patients treated with TIBSOVO included non-infectious leukocytosis, peripheral edema, pyrexia, dyspnea, pleural effusion, hypotension, hypoxia, pulmonary edema, pneumonitis, pericardial effusion, rash, fluid overload, tumor lysis syndrome, and creatinine increased. Of the 7 patients with newly

diagnosed AML who experienced differentiation syndrome, 6 (86%) patients recovered, and of the 34 patients with relapsed or refractory AML who experienced differentiation syndrome, 27 (79%) patients recovered, after treatment or after dose interruption of TIBSOVO. Differentiation syndrome occurred as early as 1 day and up to 3 months after TIBSOVO initiation and has been observed with or without concomitant leukocytosis.

If differentiation syndrome is suspected, initiate dexamethasone 10 mg IV every 12 hours (or an equivalent dose of an alternative oral or IV corticosteroid) and hemodynamic monitoring until improvement. If concomitant non-infectious leukocytosis is observed, initiate treatment with hydroxyurea or leukapheresis, as clinically indicated. Taper corticosteroids and hydroxyurea after resolution of symptoms and administer corticosteroids for a minimum of 3 days. Symptoms of differentiation syndrome may recur with premature discontinuation of corticosteroid and/or hydroxyurea treatment. If severe signs and/or symptoms persist for more than 48 hours after initiation of corticosteroids, interrupt TIBSOVO until signs and symptoms are no longer severe.

QTc Interval Prolongation: Patients treated with TIBSOVO can develop QT ("**QTc**") prolongation and ventricular arrhythmias. One patient developed ventricular fibrillation attributed to TIBSOVO. Concomitant use of TIBSOVO with drugs known to prolong the QTc interval (e.g., anti-arrhythmic medicines, fluoroquinolones, triazole anti-fungals and 5-HT_3 receptor antagonists) and CYP3A4 inhibitors may increase the risk of QTc interval prolongation. Conduct monitoring of electrocardiograms and electrolytes. In patients with congenital long QTc syndrome, congestive heart failure, or electrolyte abnormalities, or in those who are taking medications known to prolong the QTc interval, more frequent monitoring may be necessary.

Interrupt TIBSOVO if QTc increases to greater than 480 msec and less than 500 msec. Interrupt and reduce TIBSOVO if QTc increases to greater than 500 msec. Permanently discontinue TIBSOVO in patients who develop QTc interval prolongation with signs or symptoms of life-threatening arrhythmia.

Guillain-Barré Syndrome: Guillain-Barré syndrome occurred in less than 1% (2/258) of patients treated with TIBSOVO in the clinical study. Monitor patients taking TIBSOVO for onset of new signs or symptoms of motor and/or sensory neuropathy such as unilateral or bilateral weakness, sensory alterations, paresthesias, or difficulty breathing. Permanently discontinue TIBSOVO in patients who are diagnosed with Guillain-Barré syndrome.

ADVERSE REACTIONS

- The most common adverse reactions including laboratory abnormalities (no less than 20%) were hemoglobin decreased (60%), fatigue (43%), arthralgia (39%), calcium decreased (39%), sodium decreased (39%), leukocytosis (38%), diarrhea (37%), magnesium decreased (36%), edema (34%), nausea (33%), dyspnea (32%), uric acid increased (32%), potassium decreased (32%), alkaline phosphatase increased (30%), mucositis (28%), aspartate aminotransferase increased (27%), phosphatase decreased (25%), electrocardiogram QT prolonged (24%), rash (24%), creatinine increased (24%), cough (23%), decreased appetite (22%), myalgia (21%), constipation (20%) and pyrexia (20%).

- **In patients with newly diagnosed AML**, the most frequently reported Grade 3 and above adverse reactions (no less than 5%) were fatigue (14%), differentiation syndrome (11%), electrocardiogram QT prolonged (11%), diarrhea (7%), nausea (7%), and leukocytosis (7%). Serious adverse reactions (no

less than 5%) were differentiation syndrome (18%), electrocardiogram QT prolonged (7%) and fatigue (7%). There was one case of posterior reversible encephalopathy syndrome.

- **In patients with R/R AML,** the most frequently reported Grade 3 and above adverse reactions (no less than 5%) were differentiation syndrome (13%), electrocardiogram QT prolonged (10%), dyspnea (9%), leukocytosis (8%), and tumor lysis syndrome (6%). Serious adverse reactions (no less than 5%) were differentiation syndrome (10%), leukocytosis (10%) and electrocardiogram QT prolonged (7%). There was one case of progressive multifocal leukoencephalopathy.

DRUG INTERACTIONS

Strong or Moderate CYP3A4 Inhibitors: Reduce TIBSOVO dose with strong CYP3A4 inhibitors. Monitor patients for increased risk of QTc interval prolongation.

Strong CYP3A4 Inducers: Avoid concomitant use with TIBSOVO.

Sensitive CYP3A4 Substrates: Avoid concomitant use with TIBSOVO.

QTc Prolonging Drugs: Avoid concomitant use with TIBSOVO. If co-administration is unavoidable, monitor patients for increased risk of QTc interval prolongation.

LACTATION

Because many drugs are excreted in human milk and because of the potential for adverse reactions in breastfed children, we advise women not to breastfeed during treatment with TIBSOVO and for at least 1 month after the last dose.

Please see full Prescribing Information, including Boxed WARNING.

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 22, 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.