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 DOSED IN THE PHASE I BRIDGING REGISTRATIONAL STUDY OF IVOSIDENIB

CStone Pharmaceuticals (the "**Company**" or "**CStone**") announced that the first patient has been dosed in the Phase I bridging registrational study of ivosidenib (TIBSOVO®) in China. This stand-alone trial is designed to validate the efficacy, safety, and pharmacokinetics of ivosidenib in patients with isocitrate dehydrogenase-1 ("**IDH1**") mutant relapsed or refractory ("**R/R**") acute myeloid leukemia ("**AML**").

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 patients with R/R AML with a susceptible IDH1 mutation as detected by an FDA-approved test. In May 2019, CStone submitted a new drug application for ivosidenib in Taiwan for the treatment of adult patients with IDH1 mutant R/R AML.

Current standard of care treatment for newly diagnosed AML patients mainly includes intensive induction chemotherapy (IC), followed by consolidation therapy such as allogeneic hematopoietic stem cell transplantation (Allo-HSCT) in order to attain durable remission. Approximately 35% to 40% of those treated patients achieve complete remission, while only about 25% achieve 3 years or longer survival. The majority of AML patients develop acquired resistance to treatment or eventually relapse, leading to R/R AML, which has a very poor prognosis in the absence of standard of care treatment options globally. With the emergence of DNA sequencing technology, the detection of genetic mutations has presented new opportunities and challenges in AML treatment. IDH1 mutations are associated with around 6% to 10% of all AML cases.

Dr. Frank Ningjun Jiang, chairman, executive director and chief executive officer of CStone, commented: "AML is the most common acute leukemia affecting adults with over 30,000 new cases estimated in China every year. AML is characterized by its rapid progression with a five-year survival rate below 20%. We are faced with the urgent need for clinical development, particularly for IDH1 mutant R/R AML patients, due to the lack of any effective treatment in China. We will rigorously press ahead with the clinical development of ivosidenib to achieve its regulatory approval in China which will allow more AML patients in Greater China to benefit from this precision therapy."

CStone's chief medical officer, Dr. Jason Yang, noted: "ivosidenib is a potent and highly selective IDH1 inhibitor, and the only targeted therapy currently approved by the FDA for IDH1 mutant AML. It is very encouraging that we have already initiated two registrational studies of ivosidenib in China, including the global Phase III AGILE study of ivosidenib in combination with azacitidine in adult patients with newly diagnosed IDH1 mutant AML who are not eligible for intensive chemotherapy. We will remain fully dedicated to the development of this drug to fulfill the treatment gap as quickly as possible and to better serve AML patients in the Greater China Region."

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. 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 symptom resolution.

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. 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. 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-HT3 receptor antagonists) and CYP3A4 inhibitors may increase the risk of QTc interval prolongation. Conduct monitoring of electrocardiograms (ECGs) 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 (PRES).

- **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 (PML).

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 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 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, November 20, 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.