NEWS RELEASE

bridgebio pharma announces positive phase 1 data and phase 2/3 trial design for bbp-711, a potentially best-in-class go inhibitor for primary hyperoxaluria type 1 (ph1) and recurrent kidney stone formers

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– BBP-711 led to near complete inhibition of glycolate oxidase throughout the dosing period and greater than 10-fold increases in plasma glycolate, suggesting it has the potential to be both a best-in-class therapy and the first oral therapy for PH1 and recurrent kidney stone formers

– Based on the tolerability and potency of the oral therapy, BridgeBio has met with regulators and intends to initiate a Phase 2/3 pivotal study by the end of 2022

– At the end of 2022, BridgeBio also intends to launch a Phase 2 study of BBP-711 in adult recurrent kidney stone formers, which affects an estimated 1.5 million individuals in the United States and European Union

PALO ALTO, Calif., June 27, 2022 (GLOBE NEWSWIRE) -- BridgeBio Pharma, Inc. (Nasdaq: BBIO), a commercial-stage biopharmaceutical company focused on genetic diseases and cancers, today announced positive Phase 1 data for BBP-711 in healthy volunteers, supporting the development of the investigational therapy for patients with primary hyperoxaluria type 1 (PH1) and recurrent kidney stone formers. The data were shared in a **feature oral presentation** at European Society for Pediatric Nephrology (ESPN) 2022, taking place in Ljubljana, Slovenia.

BBP-711 is an orally-administered small molecule inhibitor of glycolate oxidase (GO) that is being developed to treat conditions of excess oxalate. Overproduction of oxalate in hyperoxaluria, including PH1 and recurrent kidney stone

formers with elevated oxalate, can lead to kidney stone formation, nephrocalcinosis and renal impairment. PH1 affects an estimated 5,000 patients in the United States and European Union, while recurrent stone formers are much more common, affecting an estimated 1.5 million individuals in the United States and European Union.

“PH1 is a severe genetic condition with a high degree of morbidity, including kidney stones, calcification in the kidneys and deposition of calcium oxalate in many vital organs. Despite major advances in the field of hyperoxaluria, PH1 remains a disease with high unmet medical need in many parts of the world,” said Scott Adler, M.D., chief medical officer and vice president of clinical development at Cantero Therapeutics, the BridgeBio affiliate developing BBP-711. “These data are encouraging for patients with PH1 as BBP-711 could become a novel oral therapy to prevent hyperoxaluria and its long-term consequences.”

BBP-711 was evaluated in a two-part, randomized, double-blinded, placebo-controlled, ascending dose Phase 1 study with 92 healthy volunteers. The Phase 1 data included:

- The pharmacokinetic (PK) parameters demonstrated that the drug was rapidly absorbed with a time to maximum concentration of ~2.5 hours and an elimination half-life of ~26 hours, supportive of once-daily dosing
- The results showed rapid and clinically meaningful pharmacodynamic (PD) increases in plasma glycolate of 10-15-fold above baseline levels, the highest response publicly reported to date of any GO-targeting agent tested in healthy volunteers
- PK-PD modeling of this data predicts that near-complete inhibition (maximal inhibition of > 95%) of GO can be sustained throughout the dosing period
- BBP-711 was well-tolerated with evaluated single doses 40 to 3,000 mgs and multiple doses of 75 to 1000 mgs. Treatment emergent adverse events were low in frequency (15-34%) and mild or moderate, with no changes in clinical laboratory measures, ECG, or vital signs observed

Based on the data, BridgeBio intends to initiate a Phase 2/3 study in PH1. The study is expected to consist of two parts: Part A will include a dose-finding period to identify a well-tolerated therapeutic dose for Part B; Part B will be a randomized, placebo-controlled trial. Key secondary endpoints include absolute change from baseline in 24-hour UOx excretion corrected for body surface area (BSA) and percentage of participants with 24-hour UOx below upper limit of normal (ULN).

“PH1 is a severe and progressive genetic disease. It is often undiagnosed or misdiagnosed in infancy, childhood, adolescence and adulthood and can lead to intensive dialysis or kidney and liver transplantation,” said Kim Hollander, executive director of the Oxalosis and Hyperoxaluria Foundation. “We are encouraged by the potential of an oral therapy for people living with this serious genetic condition. It provides hope to patients praying for a new treatment option, seeking to alleviate the burden of medical care management and achieve an improved quality of

life."

The Company also intends to launch a proof-of-concept Phase 2 study in adult recurrent kidney stone formers with elevated urinary oxalate excretion at the end of 2022, pending discussions with regulatory agencies. Additional studies are planned to address all patients with hyperoxaluria including patients less than six years of age and patients with impaired renal function.

About BridgeBio Pharma, Inc.
BridgeBio Pharma, Inc. (BridgeBio) is a commercial-stage biopharmaceutical company founded to discover, create, test and deliver transformative medicines to treat patients who suffer from genetic diseases and cancers with clear genetic drivers. BridgeBio's pipeline of development programs ranges from early science to advanced clinical trials. BridgeBio was founded in 2015 and its team of experienced drug discoverers, developers and innovators are committed to applying advances in genetic medicine to help patients as quickly as possible. For more information visit **bridgebio.com** and follow us on **LinkedIn** and **Twitter**.

BridgeBio Pharma, Inc. Forward-Looking Statements
This press release contains forward-looking statements. Statements we make in this press release may include statements that are not historical facts and are considered forward-looking within the meaning of Section 27A of the Securities Act of 1933, as amended (the "Securities Act"), and Section 21E of the Securities Exchange Act of 1934, as amended (the "Exchange Act"), which are usually identified by the use of words such as "anticipates," "believes," "estimates," "expects," "intends," "may," "plans," "projects," "seeks," "should," "will," and variations of such words or similar expressions. We intend these forward-looking statements to be covered by the safe harbor provisions for forward-looking statements contained in Section 27A of the Securities Act and Section 21E of the Exchange Act and are making this statement for purposes of complying with those safe harbor provisions. These forward-looking statements, including statements relating to the timing and success of BridgeBio's Phase 2/3 clinical trial of BBP-711, an investigational therapy for patients with primary hyperoxaluria type 1 (PH1) and recurrent kidney stone formers, expectations, plans and prospects regarding BridgeBio's regulatory approval process for BBP-711, the potential ability of BBP-711 to be a best-in-class therapy for PH1, recurrent kidney stone formers and other conditions caused by overproduction of oxalate, and the potential timing and success of a proof-of-concept Phase 2 study of BBP-711 in adult recurrent kidney stone formers with elevated urinary oxalate excretion, reflect our current views about our plans, intentions, expectations, strategies and prospects, which are based on the information currently available to us and on assumptions we have made. Although we believe that our plans, intentions, expectations, strategies and prospects as reflected in or suggested by those forward-looking statements are reasonable, we can give no assurance that the plans, intentions, expectations or strategies will be attained or achieved. Furthermore, actual results may differ materially from those described in the forward-looking statements and will be affected by a number of risks, uncertainties and assumptions, including, but not limited to, BridgeBio's

ability to initiate, continue and complete a Phase 2/3 clinical trial of BBP-711, an investigational therapy for patients with PH1 and recurrent kidney stone formers, past data from preclinical studies not being indicative of future data from clinical trials, BridgeBio's ability to advance BBP-711 in clinical development according to its plans, the potential ability of BBP-711 to be a best-in-class therapy for PH1, recurrent kidney stone formers and other conditions caused by overproduction of oxalate, and potential adverse impacts due to the global COVID-19 pandemic such as delays in regulatory review, manufacturing and clinical trials, supply chain interruptions, adverse effects on healthcare systems and disruption of the global economy; as well as those set forth in the Risk Factors section of BridgeBio's most recent Annual Report on Form 10-K filed with the U.S. Securities and Exchange Commission (SEC) and in subsequent SEC filings, which are available on the SEC's website at **www.sec.gov**. Moreover, BridgeBio operates in a very competitive and rapidly changing environment in which new risks emerge from time to time. Except as required by applicable law, we assume no obligation to update publicly any forward-looking statements, whether as a result of new information, future events or otherwise.

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