NEWS RELEASE

# bridgebio pharma presents updated results from phase 2 open-label extension study of acoramidis in transthyretin amyloid cardiomyopathy (attr-cm)

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- NT-proBNP, a biomarker of cardiac failure and independent predictor of mortality in ATTR-CM patients, was stable or improving throughout the study. At Month 30, median change from baseline in NT-proBNP was -437 pg/mL with 68% of participants observing NT-proBNP levels below their baseline

- Serum TTR levels were sustainably increased from baseline, with mean concentration rising from 21.55 mg/dL at baseline to 30.06 mg/dL at Month 30 (+41%)

- Acoramidis remained generally well-tolerated with no safety signals of clinical concern identified

- Topline data from ongoing Phase 3 trial of acoramidis in ATTR-CM (ATTRibute-CM) are expected in mid-2023

PALO ALTO, Calif., April 03, 2022 (GLOBE NEWSWIRE) -- BridgeBio Pharma, Inc. (Nasdaq: BBIO) (“BridgeBio” or the “Company”), a commercial-stage biopharmaceutical company focused on genetic diseases and cancers, today announced updated data from its ongoing Phase 2 open-label extension (OLE) study of acoramidis (AG10) in patients with symptomatic transthyretin (TTR) amyloid cardiomyopathy (ATTR-CM). The results were featured in an oral presentation at the American College of Cardiology (ACC) Annual Scientific Session & Expo, taking place in Washington, D.C. on April 2 – 4, 2022.

An interim analysis of the ongoing Phase 2 OLE study was completed based on available data through August 31, 2021. This corresponds to a median of 38 months since Phase 2 enrollment in the first half of 2018 and 35 months

of continuous acoramidis treatment in the OLE. Acoramidis was generally well-tolerated and resulted in sustained, near-complete TTR stabilization as measured by established ex vivo assays and increased serum TTR levels. Median N-terminal Pro-brain natriuretic peptide (NT-ProBNP) was stable or improving in trial participants throughout the OLE. In ATTR-CM patients, NT-ProBNP concentrations are strongly correlated with mortality and typically increase progressively in untreated patients.[1] The Phase 2 OLE data continue to suggest long-term tolerability of acoramidis in ATTR-CM patients and a stabilization of disease progression in treated participants.

“Over approximately three years in this study, acoramidis continued to be well tolerated and potently stabilize TTR. In patients with advanced symptomatic disease that would be expected to decline rapidly, participants remained remarkably stable or improved with respect to key cardiac biomarkers,” said Ahmad Masri, M.D., MS, director of the Cardiac Amyloidosis Program at Oregon Health & Science University. “These results provide additional optimism for the results of the ongoing Phase 3 study of acoramidis expected next year.”

The ongoing OLE study enrolled 47 participants who had completed the 28-day randomized, placebo-controlled Phase 2 study of acoramidis in ATTR-CM patients with New York Heart Association (NYHA) class II or III symptoms. Participants received 800 mg of acoramidis hydrochloride twice daily during the OLE. An interim analysis of the ongoing Phase 2 OLE study was completed based on available data through August 31, 2021. The data demonstrated:

- 31 of 47 participants remained in the OLE study; of the 16 discontinuations, adverse events (AEs) with an outcome of death, cardiac transplant or transition to hospice were reported for 11 participants
- Acoramidis remained generally well-tolerated with a pattern of AEs consistent with underlying disease, progression of disease, concurrent illnesses, and age of participants. No safety signals of clinical concern were identified
- Acoramidis demonstrated near-complete TTR stabilization. Serum TTR levels were sustainably increased from baseline, with mean concentration rising from 21.55 mg/dL at baseline to 30.06 mg/dL at Month 30 (+41%). Near-complete stabilization was verified using established ex-vivo assays with mean stabilization of 102.5 ± 8.9% at Month 30
- Median NT-proBNP were stable or improving in study participants. At Month 30, median change from baseline in NT-proBNP was -437 pg/mL (interquartile range: -950, 316). 68% of participants with available samples at Month 30 (15/22) had NT-proBNP levels below their baseline, suggesting an improvement in their heart failure severity

BridgeBio’s Phase 3 study investigating acoramidis in ATTR-CM (ATTRibute-CM) is ongoing with Month 30 topline data expected in mid-2023. In the Month 12 readout, no benefit of acoramidis relative to placebo was observed on the six-minute walk test, but improvements in the Kansas City Cardiomyopathy Questionnaire Overall Score, NT-proBNP, and serum TTR level were observed. The Company remains optimistic in the Month 30 primary endpoint, a

hierarchical composite including all-cause mortality and cardiovascular hospitalizations.

“The Phase 2 OLE data deepen our conviction in the Month 30 readout given the stability or improvement of NT-proBNP change from baseline in patients with an otherwise rapidly progressive disease,” said Neil Kumar, Ph.D., founder and CEO of BridgeBio. “We are committed to the ATTR community and hope to provide a new treatment option for ATTR-CM patients.”

About Acoramidis

Acoramidis (AG10) is an investigational, orally-administered small molecule designed to potently stabilize tetrameric transthyretin, or TTR, thereby halting at its outset the series of molecular events that give rise to TTR amyloidosis, or ATTR. Acoramidis is currently being evaluated in Phase 2 and Phase 3 studies in patients with ATTR. Acoramidis was designed to mimic a naturally-occurring variant of the TTR gene (T119M) that is considered a “rescue mutation” because it has been shown to prevent or minimize ATTR in individuals carrying pathogenic, or disease-causing, mutations in the TTR gene. For patients with ATTR, TTR stabilization offers the chance to both preserve the protective benefits of TTR and address the root cause of disease.

About Transthyretin Amyloidosis (ATTR)

Likely affecting more than 400,000 patients globally, ATTR is an underdiagnosed and life-threatening disease with limited treatment options that can devastate the heart and nervous system. When the transthyretin (TTR) becomes unstable due to inherited variants or aging, it can accumulate as amyloid fibrils in various organs in the body, causing ATTR. TTR amyloid deposits predominantly in the heart and/or peripheral nerves, causing cardiomyopathy (ATTR-CM) and/or polyneuropathy (ATTR-PN). ATTR often dramatically impairs the quality of life, functional independence and life expectancy of patients, as well as impacting caregivers due to the progressive nature of the disease. If left untreated life expectancy from diagnosis is approximately four years.

References

[1]Lane, T. et al. Circulation. 2019;140:16–26.

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 over 30 development programs ranges from early science to advanced clinical trials and its commercial organization is focused on delivering the company’s first two approved therapies. 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 prospects of success for Part B results from the Phase 3 ATTRibute-CM Study, the market opportunity for AG10, and the timing, prospects of success and clinical trial results of our ongoing Phase 2 OLE study of AG10 in patients with symptomatic ATTR-CM, reflect our current views about our plans, intentions, expectations, strategies and prospects, and are based on the information currently available to us and on assumptions we have made and are not forecasts, promises nor guarantees. Although we believe that our plans, intentions, expectations, strategies and prospects as reflected in or suggested by these 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, the success of our product candidates to treat genetically driven diseases and cancers with clear genetic drivers, our anticipated cash runway and our being fully funded through the completion of the ATTRibute-CM study and our ability to access additional funding upon achievement of portfolio milestones, as well as those risks set forth in the Risk Factors section of our most recent Annual Report on Form 10-K and BridgeBio Pharma's other SEC filings. Moreover, we operate 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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