NEWS RELEASE

bridgebio pharma reports month 12 topline results from phase 3 attribute-cm study

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- ATTRibute-CM did not meet its primary endpoint at Month 12. Mean observed six-minute walk distance (6MWD) decline for the acoramidis and placebo arms were 9 meters and 7 meters, respectively. Both declines are similar to healthy elderly adults and less than prior untreated ATTR-CM cohorts

- The company observed improvements at Month 12 on the Kansas City Cardiomyopathy Questionnaire Overall Score (KCCQ-OS, nominal $p < 0.05$), a quality-of-life measurement, N-terminal pro BNP (NT-proBNP, median +0.6% vs. +24.3%, nominal $p < 0.05$), a cardiac biomarker, and serum TTR concentration (mean +38.5% vs. -0.7%, nominal $p < 0.01$), a measure of TTR stabilization

- Acoramidis was generally well-tolerated with no safety signals of clinical concern identified. 27% fewer treatment emergent adverse events (AEs) leading to death occurred in participants receiving acoramidis than in participants receiving placebo (4.5% vs. 6.2%)

- The ATTRibute-CM independent data monitoring committee recommends continuing the study based on unblinded data reviews

- BridgeBio is fully funded through the completion of ATTRibute-CM and expects to realize at least four other clinical-stage pipeline catalysts beyond acoramidis in 2022

- BridgeBio to host investor call on December 27, 2021 at 8:00 AM ET

PALO ALTO, Calif., Dec. 27, 2021 (GLOBE NEWSWIRE) -- BridgeBio Pharma, Inc. (Nasdaq: BBIO) today announced topline results from Month 12 (Part A) of ATTRibute-CM, an ongoing global Phase 3 study investigating acoramidis for the treatment of symptomatic transthyretin (TTR) amyloid cardiomyopathy (ATTR-CM). The mean observed decline in 6MWD at Month 12 in participants receiving acoramidis or placebo with baseline eGFR ≥ 30 mL/min/1.73m^2 were 9 meters and 7 meters, respectively. Decline observed in both arms of ATTRibute-CM was similar to expected functional decline in healthy elderly adults.[1] The declines were also substantially less than the >40 meter annual declines observed in previous untreated arms reviewed by the company.[2] The decline in the ATTRibute-CM placebo group was more than 70% lower than the decline observed in the ATTR-ACT treatment group.[2]

The ATTRibute-CM independent data monitoring committee recommends continuing the study based on unblinded data reviews. Despite the unexpected performance of the six-minute walk test, the trial's steering committee co-chairs and the Company agree that there is potential for acoramidis to demonstrate benefit on the Month 30 endpoint which includes all-cause mortality and cardiovascular hospitalizations.

"This result is disappointing and baffling. I am, along with many others, searching for answers regarding the 6MWD," said Neil Kumar, Ph.D., founder and CEO of BridgeBio. "The results do not appear to be due to a baseline imbalance. The hypotheses we are currently evaluating include context bias, training bias, and an evolution in diagnosis and standard of care. The drug does appear to be pharmacologically active and well-tolerated, and we observed improvement on quality of life with promising trends on adverse events leading to death. The drug seems to be doing what we are asking of it. If we observe enough clinical outcome events at Month 30, I am still hopeful that we will demonstrate the benefit of acoramidis treatment."

"Although these results were not what we hoped, the most important moment in this trial will be the Part B readout at 30 months, where we will see the effects of acoramidis on all-cause mortality and cardiovascular hospitalizations. From what I've seen so far, I remain enthusiastic about getting to that endpoint," said Daniel Judge, M.D., professor in the Division of Cardiology at the Medical University of South Carolina, and co-chair of the ATTRibute-CM Steering Committee.

ATTRibute-CM enrolled 632 participants with symptomatic ATTR-CM, associated with either wild-type or variant TTR, with New York Heart Association (NYHA) Class I-III symptoms. The study is designed as a two-part study with Part A (Month 12) comparing change from baseline in 6MWD and Part B (Month 30) utilizing a hierarchical comparison including all-cause mortality and cardiovascular hospitalizations.

ATTRibute-CM enrolled a similar patient population as ATTR-ACT, excepting a smaller proportion of U.S. participants and TTR variant carriers. Participants were randomized 2:1 between treatment (acoramidis 800 mg) and placebo twice daily. Based on data available after 12 months of treatment, the Company observed:

- In the primary analysis, change from baseline in 6MWD was not improved in the acoramidis arm relative to the placebo arm (p = 0.76)
    - Key differences in NYHA class, geographic distribution, and TTR variant status compared to the ATTR-ACT population do not appear to have affected the primary outcome of ATTRibute-CM
    - The only participant sub-population the company has reviewed to date that exhibited substantial placebo decline in 6MWD by Month 12 was the variant population. In that population, observed decline in placebo and acoramidis was -40 meters and -2 meters, respectively
- Acoramidis improved Kansas City Cardiomyopathy Questionnaire Overall Summary Score relative to placebo (nominal $p < 0.05$, mixed model repeated measures without imputation)
- Acoramidis improved NT-proBNP relative to placebo. Median percent change from baseline at Month 12 in acoramidis-treated and placebo-treated participants were +0.6% and +24.3%, respectively (nominal $p < 0.05$ based on absolute changes from baseline between groups)
- Acoramidis increased serum TTR levels relative to placebo. Mean percent change from baseline at Month 12 in acoramidis-treated and placebo-treated participants were +38.5% and -0.7%, respectively (nominal $p < 0.01$ based on absolute changes from baseline between groups)
- Acoramidis was generally well-tolerated with no safety signals of clinical concern identified. To protect the integrity of Part B, the Sponsor's access to unblinded adverse event data for Part A excludes AEs leading to a cardiovascular hospitalization (as determined by investigators) excepting events with the outcome of death
    - Adverse events occurred in 85.3% of placebo-treated participants and 91.9% of acoramidis-treated participants
    - Serious adverse events occurred in 23.2% of placebo-treated participants and 20.2% of acoramidis-treated participants
    - Adverse events with outcome of death occurred in 6.2% of placebo-treated participants and 4.5% of acoramidis-treated participants

BridgeBio is well-capitalized through the completion of ATTRibute-CM and remains on track to deliver on additional catalysts in 2022 and 2023. "With the financing announced in November 2021, we currently have approximately $800 million in cash, cash equivalents and marketable securities with access to up to $300 million on achieving portfolio milestones through year-end 2022. We have ongoing clinical trials in multiple genetic diseases, including achondroplasia, autosomal dominant hypocalcemia type 1, limb-girdle muscle dystrophy type 2i and dystrophic epidermolysis bullosa, and we believe we are well-positioned to deliver in 2022," said Brian Stephenson, Ph.D., chief financial officer of BridgeBio.

[1] Enright, P.L. et al. Chest 2003.

[2] Maurer, M.S. et al. NEJM 2018.; Lane, T. et al., Circulation 2019.

Webcast Information

BridgeBio will host a conference call and simultaneous webcast to share updates on the Phase 3 Part A data for acoramidis on December 27th at 8:00 AM ET. To access this call, dial (800) 379-2666 (U.S.) or (409) 937-8964 (International) with Conference ID: 2895217. A link to the webcast may be accessed from the event calendar page of BridgeBio's website at **https://investor.bridgebio.com/**. A replay of the conference call and webcast will be archived on the Company's website and will be available for at least 30 days following the event.

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.

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.

About BridgeBio Pharma, Inc.

BridgeBio Pharma, Inc. (BridgeBio) is a 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 our clinical trial results for Part A of the Phase 3 ATTRibute-CM Study, the prospects of success for Part B results from the Phase 3 ATTRibute-CM Study, the market opportunity for AG10, 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, 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 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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