NEWS RELEASE

bridgebio pharma shares positive long-term data from an ongoing phase 2 study, which support the potential use of glycosylated alpha-dystroglycan (αdg) levels as a surrogate endpoint in limb-girdle muscular dystrophy type 2i/r9 (lgmd2i/r9)

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- Early assessment of increased glycosylated αDG levels at 3 months predicted subsequent ambulatory improvements at 9 months, supporting the use of glycosylated αDG levels as a potential surrogate endpoint in LGDM2I/R9

- Long-term data from the ongoing Phase 2 study of BBP-418 in patients with LGMD2I/R9 at Month 21 demonstrate a well-tolerated safety profile, and stabilization in 10 meter walk test (10MWT), 100-m timed test (100mTT), and the North Star Assessment (NSAD) scores

- A sustained decrease of ≥80% in creatine kinase (CK), a marker of muscle breakdown, was observed with BBP-418 treatment at 21 months

- The Phase 3 study continues to enroll in the U.S. and BridgeBio believes there is potential to pursue Accelerated Approval for BBP-418 based on recent interactions with the U.S. Food and Drug Administration (FDA) on the use of glycosylated αDG levels as a surrogate endpoint

PALO ALTO, Calif., Oct. 09, 2023 (GLOBE NEWSWIRE) -- BridgeBio Pharma, Inc. (Nasdaq: BBIO) (“BridgeBio” or the “Company”), a commercial-stage biopharmaceutical company focused on genetic diseases and cancers, shared new

long-term data from its Phase 2 trial in patients with limb-girdle muscular dystrophy type 2I/R9 (LGMD2I/R9) at the Annual Congress of World Muscle Society (WMS) in Charleston, South Carolina. The new long-term data remains consistent with earlier data from the Phase 2 study showing a well-tolerated safety profile and encouraging preliminary efficacy. Additionally, early changes in glycosylated αDG levels at 3 months appear to be associated with ambulatory improvements at 9 months, providing support for the possible use of glycosylated αDG levels as a surrogate endpoint in the ongoing Phase 3 study for accelerated approval.

BridgeBio presented 21-month results from its ongoing Phase 2 trial, including:

- Large (≥80%), sustained reduction in creatine kinase observed over an extended (up to 21-months) treatment period
- Stabilization in NSAD scores and ambulatory measures observed over 21-months of BBP-418 treatment
- BBP-418 continues to be well-tolerated with longer-term treatment
- No treatment-related SAEs or dose limiting toxicities observed with 21-months of BBP-418 dosing

“There is a serious unmet need for those with LGMD2I/R9. The consistent improvement observed in biomarkers as well as stabilization in ambulatory measures are encouraging and help predict the clinical benefit for patients. The data for BBP-418 give me hope that similar results will be carried through in the currently enrolling Phase 3 trial, FORTIFY, for patients with LGMD2I/R9,” said Amy Harper, M.D., professor in the department of neurology at Virginia Commonwealth University (VCU) and primary investigator of the Phase 2 clinical trial in LGMD2I/R9.

BridgeBio recently announced the first patient dosed in FORTIFY, its Phase 3 registrational study in patients with LGMD2I/R9 and is continuing to enroll throughout the U.S. with clinical trial sites planned for Europe and Australia. The Phase 3 FORTIFY registrational study is a randomized, double-blind, placebo-controlled study evaluating the safety and efficacy of BBP-418. FORTIFY has a planned interim analysis at 12 months focused on assessing glycosylated αDG as a surrogate endpoint to potentially support an accelerated approval. The NSAD and secondary endpoints will be evaluated at 36 months, and results are expected to provide confirmatory clinical data.

“To see that our therapy is potentially providing clinical improvements to people who otherwise could be non-ambulatory is deeply heartening for us,” said Douglas Sproule, M.D., M.Sc., chief medical officer of ML Bio Solutions, a BridgeBio affiliate that is focused on developing BBP-418 for LGMD2I/R9. “LGMD2I/R9 is a severely progressive disease and, inevitably, the weakening of the muscles causes many affected people to become fully dependent on a caregiver. Current treatments for LGMD2I/R9 are purely supportive. BBP-418 has the potential to be the first disease-modifying orally administered therapy available for patients.”

More information about FORTIFY, the ongoing Phase 3 clinical trial of BBP-418 (NCT05775848) can be found **here** on the ClinicalTrials.gov website.

About Limb-girdle Muscular Dystrophy Type 2I/R9 (LGMD2I/R9)

LGMD2I/R9 is a monogenic autosomal recessive disease caused by partial loss of function mutations in the fukutin-related protein (FKRP) gene, and FKRP mutations impair glycosylation of αDG, a protein associated with stabilizing muscle cells. Clinical manifestations typically present as a skeletal myopathy affecting the lower and then upper limbs, which is commonly later accompanied by respiratory muscle and cardiac muscle involvement. Patients who harbor a L276I homozygous genotype typically develop disease manifestations during late childhood with progression to loss of independent ambulation (25%), assisted ventilation (10%), and cardiomyopathy (30%) in adulthood. Cardiomyopathy is progressive, with an annual loss of 0.4% of left ventricular ejection fraction (LVEF). Patients with other (non-L276I homozygous) genotypes have an earlier childhood onset with a more severe clinical course, rapid loss of mobility by 20 years of age, more frequent cardiac involvement (60%), and eventual respiratory failure by 30 years of age in nearly all cases.

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 BridgeBio makes 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. BridgeBio intends 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. These forward-looking statements, including statements relating to the clinical and therapeutic potential of BridgeBio's programs and product candidates, including BBP-418 for the treatment of LGMD2I, the potential benefits of BBP-418, including that the early assessment of increased glycosylated αDG levels at 3 months predicted subsequent ambulatory improvements at 9 months, supporting the use of glycosylated αDG levels as a potential surrogate endpoint in LGDM2I/R9 and that BBP-418 continues to be well-tolerated with longer-term treatment , the progress of BridgeBio's ongoing and planned clinical trials of BBP-418, including the continued enrollment of the Phase 3 study in the U.S. with clinical trial sites planned for Europe, UK, and Australia, the potential and the opportunity to pursue Accelerated Approval Pathway for BBP-418 in LGMD2I/R9 in the U.S. based

on recent interactions with the FDA on the use of glycosylated αDG levels as a surrogate endpoint, the statement regarding the potential benefit of FORTIFY trial for patients with LGMD2I/R9 and of BBP-418 in the quotes of Dr. Harper and Dr. Sproule, the expectation that the results of the evaluation of secondary endpoints at 36 months will provide confirmatory clinical data, and the timing and success of BridgeBio's clinical trials and development pipeline, among others, reflect BridgeBio's current views about its plans, intentions, expectations, strategies and prospects, which are based on the information currently available to BridgeBio and on assumptions BridgeBio has made. Although BridgeBio believes that its plans, intentions, expectations, strategies and prospects as reflected in or suggested by those forward-looking statements are reasonable, BridgeBio 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 continue and complete its ongoing and planned clinical trials of BBP-418 for the treatment of LGMD2I, initial and ongoing data from clinical trials not being indicative of final data, the design and success of BridgeBio's ongoing and planned clinical trials, the FDA or other regulatory agencies not agreeing with BridgeBio's regulatory approval strategies, components of our filings, such as clinical trial designs, conduct and methodologies, or the sufficiency of data submitted, as well as those risks set forth in the Risk Factors section of BridgeBio's Annual Report on Form 10-K for the year ended December 31, 2022, and BridgeBio's other SEC filings. Moreover, BridgeBio operates in a very competitive and rapidly changing environment in which new risks emerge from time to time. These forward-looking statements are based upon the current expectations and beliefs of BridgeBio's management as of the date of this press release and are subject to certain risks and uncertainties that could cause actual results to differ materially from those described in the forward-looking statements. 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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