NEWS RELEASE

bridgebio pharma shares preliminary findings on novel bioassay measuring glycosylated alpha-dystroglycan (αdg) in patients with limb-girdle muscular dystrophy type 2i (lgmd2i)

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- BridgeBio has developed a validated bioassay that directly measures glycosylated αDG, which is central to LGMD2I disease, and enables monitoring of responses to disease-modifying therapies in LGMD2I patients

- BridgeBio also shared 15-month results from its ongoing Phase 2 study, which showed a doubling of glycosylated αDG in LGMD2I patients treated with BBP-418

- A sustained decrease of ≥70% in creatine kinase (CK), a marker of muscle breakdown, was observed with BBP-418 treatment at 15 months

- Improvements in ambulatory and clinical function measures were observed after 15 months of treatment with BBP-418

- Based on the Phase 2 results, BridgeBio is embarking on a registration-enabling Phase 3 study with an initiation in mid-2023

PALO ALTO, Calif., March 21, 2023 (GLOBE NEWSWIRE) -- BridgeBio Pharma, Inc. (Nasdaq: BBIO) (“BridgeBio” or the “Company”), a commercial-stage biopharmaceutical company focused on genetic diseases and cancers, today announced the Company’s novel validated muscle tissue-based bioassay, which measures the amount of

glycosylated αDG in patients with LGMD2I, at the MDA 2023 Annual Meeting. Additionally, BridgeBio shared updated results at Month 15 for its Phase 2 clinical trial and the design for its pivotal Phase 3 study for patients with LGMD2I. BridgeBio will host an investor call with Jeffrey Rosenfeld, M.D., Ph.D., a specialist in neuromuscular medicine and professor of neurology at Loma Linda University School of Medicine on Tuesday, March 21 at 8:30 am ET to discuss the results shared at the meeting.

The novel bioassay was developed by BridgeBio to measure the amount of glycosylated αDG, and to evaluate the impact of BBP-418 treatment. When deficient, glycosylated αDG is the direct cause of muscle breakdown in LGMD2I. The bioassay allowed the Company to evaluate natural history and Phase 2 data to inform the Phase 3 study design. Highlights of the results include:

- Development of a novel, validated, quantitative method to measure glycosylated αDG in muscle
- Observation of ~90% reduction in glycosylated αDG in muscle biopsies from patients with LGMD2I compared to healthy individuals
- Higher glycosylated αDG in L276I/L276I homozygous patients (~11%) who are characterized by a milder clinical phenotype than other fukutin-related protein (FKRP) genotypes (~5%)
- Stable glycosylated αDG levels in untreated patients with LGMD2I over ~6-12 months

“The αDG bioassay offers a unique opportunity to directly measure the core causative step leading to muscle cell breakdown and muscle weakness in LGMD2I. There is a serious unmet need for patients living with this progressive and debilitating condition, and I am encouraged by the very promising results from this therapy, especially considering the relevance of the αDG bioassay. The profound effects on glycosylated-αDG observed in the Phase 2 trial, alongside the encouraging trends on clinical endpoints, including North Star Assessment for Dysferlinopathy (NSAD) and ambulatory measures has made me especially optimistic for a phase 3 trial,” said Jeffrey Rosenfeld, M.D., Ph.D.

Additionally, BridgeBio presented 15-month results from its ongoing Phase 2 trial and the design of its registration-enabling Phase 3 study. Phase 2 study results showed:

- Increased glycosylation of αDG was observed following treatment initiation with BBP-418, with an approximate doubling of glycosylated αDG at 3 months which was sustained over 15 months
- Greater than 70% reduction in CK sustained at 15 months
- Improvements from baseline in NSAD test scores and ambulatory measures at 15 months
- No treatment-related serious adverse events or dose limiting toxicities were observed with BBP-418

The planned Phase 3 registrational study in patients with LGMD2I **(NCT05775848)** is designed as a randomized, placebo-controlled, double-blind two-arm study comparing the effect of BBP-418 to standard of care. Key measures include glycosylated-αDG using the validated bioassay and changes in NSAD scores and ambulatory measures over

a 36-month period.

“LGMD2I is highly progressive and, inevitably, the severity of the disease causes people to become fully dependent on a caregiver, requiring a wheelchair or even ventilation assistance. Our novel bioassay enables us to assess whether our therapy is working and signifies, through our Phase 2 data, that the effect of BBP-418 treatment on restoration of glycosylation of αDG is sustained over time. At 15 months, we are continuing to see consistent improvements in clinical endpoints from the Phase 2 trial such as increased NSAD score, increased 10-meter walk test velocity, and reduced time to complete 100-meter timed test compared to baseline; as opposed to the declines observed in these endpoints in the natural history population. Based upon these results, we are embarking on a registration-enabling Phase 3 study in mid-2023, which we hope confirms a therapeutic benefit for patients,” 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.

BBP-418 is designed to supply supraphysiological levels of the ribitol substrate upstream of the mutant FKRP enzyme to help drive residual activity of the enzyme to glycosylate αDG, with the goal of stabilizing muscle cells during contraction, and potentially halting further muscle damage. If approved, BridgeBio believes BBP-418 could be the first approved orally administered therapy for the treatment of patients with LGMD2I.

Webcast Information

BridgeBio will host an investor call and simultaneous webcast to discuss the LGMD2I program updates shared at MDA on Tuesday, March 21 at 8:30 am ET. 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 Limb-girdle Muscular Dystrophy Type 2I (LGMD2I)

LGMD2I 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 homozygous genotype typically develop disease manifestations during late childhood with progression to loss of independent ambulation (25%), assisted ventilation (5%), and cardiomyopathy (10%) in adulthood. Cardiomyopathy is progressive, with an annual loss of 0.4% of left ventricular ejection fraction (LVEF). Patients with heterozygous 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 (25%), 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 and achieving a doubling of glycosylated αDG at 3 months sustained over 15 months and a decrease of greater than 70% in CK at 15 months, the progress of BridgeBio's ongoing and planned clinical trials of BBP-418, including the intent to initiate a Phase 3 clinical trial in mid-2023, the typical clinical manifestations of LGMD2I and progression to loss of independent ambulation, assisted ventilation and cardiomyopathy, the impact on patients with heterozygous genotypes, the potential benefits of BridgeBio's novel bioassay, including the ability to measure glycosolated αDG levels from muscle biopsy samples, the potential for BBP-418 to be the first approved orally administered therapy for the treatment of LGMD2I, 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 U.S. Food and Drug Administration (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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