



NEWS RELEASE

bridgebio pharma to present preliminary findings on its novel bioassay measuring glycosylated alpha-dystroglycan (α dg) in patients with limb-girdle muscular dystrophy type 2i (lgmd2i)

2023-03-17

- BridgeBio will also share 15-month Phase 2 data and review the Phase 3 clinical trial design of BBP-418, a potential therapeutic for patients with LGMD2I, with initiation of its Phase 3 study expected in mid-2023
 - Preliminary findings and study results will be presented in an oral presentation and posters at the Muscular Dystrophy Association (MDA) 2023 Annual Meeting
- Additionally, 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 findings on the bioassay, the updated Phase 2 study results and the preliminary Phase 3 study design

PALO ALTO, Calif., March 17, 2023 (GLOBE NEWSWIRE) -- BridgeBio Pharma, Inc. (Nasdaq: BBIO) ("BridgeBio" or the "Company"), a commercial-stage biopharmaceutical company focused on genetic diseases and cancers, announced today that preliminary data on the Company's muscle tissue-based bioassay measuring glycosylated α DG in LGMD2I patients will be featured in an oral presentation and posters at the MDA 2023 Annual Meeting, taking place in Dallas, Texas on March 19 - 22, 2023. 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.

BridgeBio was selected to share an oral presentation and posters on its ongoing Phase 2 trial with 15-month results, including the preliminary design of its pivotal Phase 3 study in patients with LGMD2I, which is expected to launch in mid-2023.

Oral Presentation and Poster Details:

Preliminary Results from MLB-01-003: An Open Label Phase 2 Study of BBP-418 in Patients with Limb-girdle Muscular Dystrophy Type 2I

Date/ Time: Monday, March 20 at 2:30 – 3:00 pm CT

Presenter: 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

Location: Coronado A; Hilton Anatole, Dallas, TX

Session: Ultra-Rare Track

Longitudinal Measurement Using a Novel Western Blot Assay of Glycosylation of Alpha- dystroglycan in Patients with Limb-girdle Muscular Dystrophy Type 2I/R9 FKR-related: Preliminary Results from MLB-01-001

Date/ Time: Monday, March 20 and Tuesday, March 21 at 6:00 – 8:00 pm CT

Location: Trinity Exhibit Hall; Hilton Anatole, Dallas, TX

Poster Number: #140

Preliminary Results from MLB-01-003: An Open Label Phase 2 Study of BBP-418 in Patients with Limb-girdle Muscular Dystrophy Type 2I

Date/ Time: Monday, March 20 and Tuesday, March 21, at 6:00 – 8:00 pm CT

Location: Trinity Exhibit Hall; Hilton Anatole, Dallas, TX

Poster Number: #139

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 FKR gene, and FKR mutations impair glycosylation of α DG, 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](https://www.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. These forward-looking statements, including statements relating to the clinical and therapeutic potential of the Company's programs and product candidates, including our clinical development program for BBP-418 for the treatment of LGMD2I, the availability of data from our Phase 2 clinical trial of BBP-418 and timing of this event, the progress of the Company's ongoing and planned clinical trials of BBP-418 for the treatment of LGMD2I, including the expectation 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, and the timing and success of BridgeBio's clinical trials and development pipeline, 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, our ability to

continue and complete our ongoing and planned clinical trials of BBP-418 for the treatment of LGMD2I, initial and ongoing data from our clinical trials not being indicative of final data, the design and success of ongoing and planned clinical trials, the U.S. Food and Drug Administration (FDA) or other regulatory agencies not agreeing with our regulatory approval strategies, components of our filings, such as clinical trial designs, conduct and methodologies, or the sufficiency of data submitted, our ability to advance our clinical trials and development pipeline, 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 our 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.

BridgeBio Media Contact:

Vikram Bali

contact@bridgebio.com

(650)-789-8220