NEWS RELEASE

bridgebio pharma receives fda fast track designation for investigational therapy for the treatment of limb-girdle muscular dystrophy type 2i (lgmd2i)

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- If successful, BridgeBio's drug could be the first approved therapy for patients with LGMD2i
- BridgeBio's investigational therapy for LGMD2i is one of more than 30 R&D programs in the company's diverse pipeline targeting genetic diseases and genetically-driven cancers
- 12 of BridgeBio's programs are in the clinic and two have received FDA approval

PALO ALTO, Calif., Sept. 15, 2021 /PRNewswire/ -- BridgeBio Pharma, Inc. (Nasdaq: BBIO), a commercial-stage biopharmaceutical company founded to discover, create, test and deliver meaningful medicines for patients with genetic diseases and cancers with clear genetic drivers, today announced that the United States Food and Drug Administration (FDA) granted Fast Track designation for the investigation of BBP-418 as a treatment option for Limb-girdle Muscular Dystrophy Type 2i (LGMD2i). The FDA grants development programs Fast Track designation to help drive the development and expedite its review process for drugs being investigated to treat serious conditions and fill unmet medical needs. The FDA utilizes this program to provide patients access to important new drugs as early as possible. This is the fifth Fast Track designation for an investigational therapy that BridgeBio has received this year.

BridgeBio's LGMD2i investigational therapy is one of the Company's 14 programs that are in the clinic or commercial setting for patients living with genetic diseases and genetically-driven cancers.

BridgeBio's first wave of programs are the now-approved drugs for Molybdenum Cofactor Deficiency (MoCD) Type A and previously-treated locally advanced or metastatic cholangiocarcinoma (CCA) harboring an FGFR2 fusion or rearrangement. The second wave of programs includes the Company's four major near-term catalysts for its product candidates for the treatment of transthyretin (TTR) amyloidosis (ATTR), achondroplasia, congenital adrenal hyperplasia (CAH) and autosomal dominant hypocalcemia type 1 (ADH1).

LGMD2i represents one of the leading programs in BridgeBio's ongoing third wave in development, which includes a variety of programs in the cancer and mendelian space already in the clinic.

With approximately 7,000 patients with potentially treatable mutations, LGMD2i is an inherited recessive muscular dystrophy caused by mutation of fukutin-related protein (FKRP). FKRP is a critical enzyme that adds a specific sugar molecule to a muscle cell structural protein called alpha-dystroglycan (αDG). Due to defective FKRP enzyme function, muscle cells of patients affected by LGMD2i lack a robust cushioning system that is provided by fully glycosylated αDG proteins. Pediatric and adult patients with LGMD2i most commonly present with upper and lower extremity ("limb") and thoracic ("girdle") dysfunction ("limb-girdle" pattern of weakness), and without treatment often develop additional severe clinical manifestations, including loss of independent ambulation, severe breathing issues which can require mechanical ventilation, cardiomyopathy and premature death.

"As of now, there are no approved treatment options for people born with Limb-girdle Muscular Dystrophy Type 2i. People living with this disease can lose their ability to perform routine daily activities, and ultimately may lose the ability to walk, need ventilatory support or face the risk of heart failure," said Douglas Sproule, M.D., M.Sc., chief medical officer of ML Bio Solutions, Inc., the BridgeBio company developing BBP-418. "We are grateful the FDA has granted our program Fast Track designation based on the potential of our investigational therapy to treat this very serious condition. We are hopeful the designation will allow us to address this unmet medical need by allowing us to potentially deliver our medicine to patients more quickly."

BBP-418 is being investigated as a treatment for LGMD2i. The investigational therapy is designed to overcome the enzymatic limitation of the defective FKRP enzyme by supplementing endogenous sugar molecules to glycosylate αDG and to improve muscle cell integrities, resulting in improved muscle strength and function for patients. Clinical trials to verify the safety and efficacy of BBP-418 are ongoing.

BBP-418 has received Orphan Drug Designation for the treatment of LGMD2i from the FDA and for LGMD from the European Medicines Agency. BridgeBio is currently advancing its Phase 2 clinical trial in subjects with a genetically confirmed diagnosis of LGMD2i. If the development program is successful, BBP-418 could be the first approved therapy for the treatment of patients with LGMD2i.

About Limb-girdle Muscular Dystrophy Type 2i

LGMD2i is a monogenic autosomal recessive disease caused by partial loss of function mutations in the FKRP gene, and these FKRP mutations impair glycosylation of α-DG, a protein associated with stabilizing muscle cells. LGMD2i is a disease that has pediatric symptomatic onset with most individuals developing manifestations of disease between 5 and 18 years of age. 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 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 success of ML Bio Solutions' clinical trials of BBP-418 for the treatment of LGMD2i, expectations, plans and prospects regarding ML Bio Solutions' regulatory approval process for BBP-418, the ability of BBP-418 to treat LGMD2i in humans, the potential for BBP-418 to be the first approved therapy for the treatment of LGMD2i 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, ML Bio Solutions' ability to continue and complete its clinical trials of BBP-418 for the treatment of LDMD2i, past data from preclinical studies not being indicative of future data from clinical trials, ML Bio Solutions' ability to advance BBP-418 in clinical development according to its plans, the ability of BBP-418 to be the first approved therapy for the treatment of patients with LGMD2i, BridgeBio's ability to advance its clinical trials and development pipeline, the success of BridgeBio's approved drugs, as well as those risks set forth in the Risk Factors section of BridgeBio Pharma's Annual Report on Form 10-K for the year ended December 31, 2020, and BridgeBio Pharma's other SEC filings. Moreover, ML BioSolutions operates 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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