NEWS RELEASE

bridgebio completes enrollment of fortify, phase 3 registrational study of bbp-418 in limb-girdle muscular dystrophy type 2i/r9 (lgmd2i/r9)

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- Study exceeded target enrollment, with an expected topline data readout from the interim analysis expected in 2025

- BridgeBio believes there is an opportunity to pursue Accelerated Approval in the U.S. for BBP-418 in LGMD2I/R9 based on a potential biomarker surrogate endpoint of glycosylated alpha-dystroglycan (αDG) at time of the interim analysis

- If successful, BBP-418 could be the first approved therapy for individuals living with LGMD2I/R9 in the U.S.

- Enrollment completion announced on 10th annual LGMD Awareness Day, a collaborative yearly effort on September 30th to globally raise awareness of individuals living with LGMD

PALO ALTO, Calif., Sept. 30, 2024 (GLOBE NEWSWIRE) -- BridgeBio Pharma, Inc. (Nasdaq: BBIO) (“BridgeBio” or the “Company”), a commercial-stage biopharmaceutical company focused on genetic diseases, announced, on LGMD Awareness Day, the completion of enrollment of FORTIFY, the Company’s Phase 3 registrational study of BBP-418 in individuals with LGMD2I/R9.

FORTIFY is a randomized, double-blind, placebo-controlled Phase 3 study evaluating the safety and efficacy of BBP-418, an investigational oral therapy in development for the treatment of individuals living with LGMD2I/R9. The study includes a planned interim analysis at 12 months focused on assessing glycosylated αDG as a surrogate

endpoint to potentially support Accelerated Approval. The primary endpoint, to be evaluated at 36 months, is the North Star Assessment (NSAD) for limb-girdle type muscular dystrophies and is designed to provide confirmatory clinical data supporting the efficacy of BBP-418. More information about the Phase 3 clinical trial of BBP-418 (NCT05775848) can be found **here** on clinicaltrials.gov. A topline data readout from the interim analysis is expected in 2025.

“Living with a progressive muscle wasting condition like LGMD2I/R9 means that individuals are continually losing the ability to perform daily activities independently and relying on friends and family to support them as they experience a continued decline in strength and health. For patients with LGMD2I/R9, there are currently no approved treatment options, but the promise seen in the rapid enrollment of this clinical trial provides hope for patients and their families that this may change in the future,” said Kelly Brazzo, CEO of CureLGMD2i Foundation.

“Completing rapid enrollment of FORTIFY is an important milestone and underscores the need for a therapeutic option for patients,” said Douglas Sproule, M.D., M.Sc., Chief Medical Officer of ML Bio Solutions, a BridgeBio company developing BBP-418 for LGMD2I/R9. “Based on multiple encouraging discussions with the FDA, we believe there is an opportunity to pursue Accelerated Approval in the U.S. for BBP-418 in LGMD2I/R9 based on a potential surrogate endpoint biomarker of glycosylated αDG at time of the interim analysis. If successful, BBP-418 could be the first approved disease-modifying therapy for individuals living with LGMD2I/R9 in the U.S. We’re immensely grateful to the patients, their families, and the trial site investigators participating in our study and look forward to sharing pivotal data with the community.”

BBP-418 has previously received Orphan Drug, Fast Track, and Rare Pediatric Disease Designations from the FDA and Orphan Drug Designation from the European Medicines Agency (EMA). Consistent with the Rare Pediatric Designation from the FDA, if BBP-418 is approved, BridgeBio may qualify for a Priority Review Voucher.

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 alpha-dystroglycan (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. Individuals who harbor a homozygous L276I 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). Individuals with other FKRP genotypes typically 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. 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, Twitter** and **Facebook**.

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," "continues," "estimates," "expects," "hopes," "intends," "may," "plans," "projects," "remains," "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 and market potential of BridgeBio's programs and product candidates, including BBP-418 for the treatment of LGMD2I/R9, the potential benefits of BBP-418, including its potential to address unmet need for patients with LGMD2I/R9, the potential and the opportunity to pursue Accelerated Approval Pathway for BBP-418 in LGMD2I/R9 in the U.S., the potential that BridgeBio may qualify for a Priority Review Voucher based on receipt of the Rare Pediatric Designation from the FDA, the expected timeline of announcing the topline data from the interim analysis of FORTIFY in individuals with LGMD2I/R9 in 2025, the statements regarding the potential benefit of our clinical trial or of our product candidate in the quotes of Dr. Sproule and Kelly Brazzo, and the progress, timeline and success of BridgeBio's ongoing and planned clinical trials of BBP-418, 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/R9, initial and ongoing data from its clinical trials not being indicative of final data, the design and success of ongoing and planned clinical trials, difficulties with enrollment in our clinical trials, adverse events that may be encountered in our 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, the ability of BBP-418 to retain Orphan Drug, Fast Track, and Rare Pediatric Disease Designations from the FDA and Orphan Drug Designation from the European Medicines Agency and potential adverse impacts due to global health emergencies, including delays in regulatory review, manufacturing and supply chain interruptions, adverse effects on healthcare systems and disruption of the global economy, the impacts of current macroeconomic and geopolitical events, including changing conditions from hostilities in Ukraine and in Israel and the Gaza Strip, increasing rates of inflation and rising interest rates, on our business operations and expectations as well as those risks set forth in the Risk Factors section of BridgeBio's most recent Annual Report on Form 10-K, and BridgeBio's other filings with the U.S. Securities and Exchange Commission. 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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