



NEWS RELEASE

bridgebio pharma and affiliate phoenix tissue repair announce first patient dosing in phase 2 trial of protein replacement therapy for the treatment of recessive dystrophic epidermolysis bullosa (rdeb)

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BOSTON, Oct. 27, 2020 (GLOBE NEWSWIRE) -- BridgeBio Pharma, Inc. (Nasdaq: BBIO) and affiliate Phoenix Tissue Repair today announced that the first patient has been dosed in a Phase 2 study of BBP-589 (also known as PTR-01), an intravenously-administered recombinant collagen 7 (rC7) protein replacement therapy, in patients with recessive dystrophic epidermolysis bullosa (RDEB). RDEB is a rare genetic disorder caused by mutations in the gene encoding collagen type VII (C7).

RDEB is a devastating disease and is one of the most severe forms of epidermolysis bullosa, characterized by severe and painful skin blistering, as well as extreme fragility and scarring of mucous membranes throughout the body. There is currently no cure or effective treatment available for patients suffering from this disease.

"BBP-589 is a potentially disease-modifying rC7 replacement therapy that we hope may improve the skin manifestations as well as the systemic symptoms these patients endure. The results observed in the Phase 1 study give us confidence in our approach, and we are excited to continue advancing this therapeutic candidate through the clinic," said Sanuj K. Ravindran, M.D., executive chairman of Phoenix Tissue Repair.

The Phase 2, open label study is designed to examine the effect of BBP-589 on wound healing and various systemic endpoints and to evaluate the long-term safety and tolerability of the drug candidate. The study plans to enroll six patients who will receive doses of BBP-589 intravenously in two regimens over 18 weeks, followed by 12 weeks of

observation after completion of dosing to assess the durability of wound healing and other efficacy endpoints. To learn more about the BBP-589 Phase 2 clinical trial, please visit www.clinicaltrials.gov.

About Dystrophic Epidermolysis Bullosa (DEB)

DEB is a rare genetic disorder symptomatic from birth that is caused by mutations in the gene for a protein called collagen type VII (C7). The C7 protein is essential for the formation of anchoring fibrils, structures which connect the epidermis and dermis—the uppermost two layers of the skin. Patients with the recessive form of DEB (RDEB) tend to have particularly severe symptoms due to severe insufficiency of functional C7. Symptoms include extreme skin and mucosal fragility that present as recurrent, painful blistering and scarring of the skin, as well as ulcerations of the mouth, tongue and dental caries. In addition to the cutaneous and oral symptoms, severe forms are associated with erosions and scarring of mucous membranes of the eye, esophagus, genitals and anus. Joint contractures, mutilating deformities of hands and feet, malnutrition, growth retardation, recurrent infections and a significantly increased risk for squamous cell carcinoma are also common. There are currently no approved disease-modifying therapies for any form of DEB, and the standard of care focuses on wound and pain management.

About Phoenix Tissue Repair and BBP-589

Phoenix Tissue Repair is a Boston-based company that is an affiliate of BridgeBio Pharma and is focused on advancing a novel systemic treatment for recessive dystrophic epidermolysis bullosa (RDEB). BBP-589 is an investigational protein replacement therapy which uses a recombinant collagen type VII (rC7) for the treatment of RDEB. BBP-589 is designed to be systemically available through intravenous delivery. Phoenix Tissue Repair acquired worldwide rights to BBP-589 in 2017. Preclinical studies of BBP-589 have demonstrated C7 staining in basement membranes with de novo anchoring fibril formation and improved survival in models of RDEB.

BBP-589 has been granted Orphan Drug Designation by the U.S. Food and Drug Administration (FDA) and the European Medicines Agency.

About BridgeBio Pharma, Inc.

BridgeBio is a team of experienced drug discoverers, developers and innovators working to create life-altering medicines that target well-characterized genetic diseases at their source. BridgeBio was founded in 2015 to identify and advance transformative medicines to treat patients who suffer from Mendelian diseases, which are diseases that arise from defects in a single gene, and cancers with clear genetic drivers. BridgeBio's pipeline of over 20 development programs includes product candidates ranging from early discovery to late-stage development. For more information, visit bridgebio.com.

Forward-Looking Statements

This press release contains forward-looking statements, including statements relating to expectations, plans, and prospects regarding Phoenix Tissue Repair's clinical development plan, clinical trial results, timing and completion

of clinical trials, and ability to take advantage of expedited FDA review for BBP-589. Statements in this press release that are not statements of historical fact are considered forward-looking statements within the meaning of Section 27A of 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", "could", "estimates," "expects," "intends," "may," "plans," "potential", "predicts", "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 are neither forecasts, promises nor guarantees, and are based on the current beliefs of BridgeBio's management and Phoenix Tissue Repair's management as well as assumptions made by and information currently available to BridgeBio and Phoenix Tissue Repair. Such statements reflect the current views of BridgeBio and Phoenix Tissue Repair with respect to future events and are subject to known and unknown risks, uncertainties and assumptions, including, but not limited to, Phoenix Tissue Repair's ability to advance BBP-589 in clinical development in accordance with its plans, the results from any clinical trials and nonclinical studies of BBP-589, the results from prior clinical trials and nonclinical studies of BBP-589 not being indicative of the results from future clinical trials and nonclinical studies of BBP-589, and the nature of Phoenix Tissue Repair's interactions with regulatory authorities. Moreover, BridgeBio and Phoenix Tissue Repair operate in a very competitive and rapidly changing environment in which new risks emerge from time to time. Although BridgeBio and Phoenix Tissue Repair believe that BridgeBio's and Phoenix Tissue Repair's plans, intentions, expectations, strategies and prospects as reflected in or suggested by these forward-looking statements are reasonable, neither BridgeBio nor Phoenix Tissue Repair can give any 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, without limitation, those risks and uncertainties described under the heading "Risk Factors" in BridgeBio's most recent Quarterly Report on Form 10-Q and Annual Report on Form 10-K filed with the SEC and in subsequent filings made by BridgeBio with the SEC, which are available on the SEC's website at www.sec.gov. Moreover, BridgeBio and Phoenix Tissue Repair operate in very competitive and rapidly changing environments 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 and Phoenix Tissue Repair'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. We anticipate that subsequent events and developments will cause our views to change. Except as required by law, each of BridgeBio and Phoenix Tissue Repair disclaims any intention or responsibility for updating or revising any forward-looking statements contained in this press release in the event of new information, future developments or otherwise. You should, therefore, not rely on these forward-looking statements as representing our views as of any date subsequent to the date of this press release.

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