NEWS RELEASE

bridgebio pharma announces positive interim results from a phase 2 trial of infigratinib in achondroplasia demonstrating an increase in annualized height velocity of 1.52 cm/year in children 5 years of age and older, and adds 5th cohort to trial

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- At the highest dose level evaluated to date (Cohort 4, 0.128 mg/kg once daily), the mean change from baseline in annualized height velocity (AHV) was +1.52 cm/yr (p=0.02, n=11) and the responder rate was 64% in children 5 years and older[1]

- Infigratinib was well-tolerated with no serious adverse events (SAE) and no discontinuations due to adverse events (AE) including in Cohort 5 (dose: 0.25 mg/kg once daily) participants dosed to date, with a median duration of follow-up of 48.1 weeks; only a limited number of AEs were assessed as related to study drug and all were Grade 1, the lowest level

- Given infigratinib's profile to date, and after discussions with regulators, BridgeBio has begun dosing children in Cohort 5 (dose: 0.25 mg/kg once daily)

- Infigratinib is the only known oral product candidate currently under clinical investigation for achondroplasia, with granted and filed patent applications expiring as late as 2041

PALO ALTO, Calif., July 26, 2022 (GLOBE NEWSWIRE) -- BridgeBio Pharma, Inc. (Nasdaq: BBIO) (BridgeBio), a commercial-stage biopharmaceutical company focused on genetic diseases and cancers, today announced positive

interim results from PROPEL 2, a Phase 2 trial of the investigational therapy infigratinib in children with achondroplasia. Achondroplasia (ACH) is the most common cause of disproportionate short stature. Achondroplasia impacts health and can lead to medical complications such as obstructive sleep apnea, middle ear dysfunction, kyphosis, and spinal stenosis. Achondroplasia affects approximately 55,000 people in the United States (US) and Europe, including up to 13,000 children and adolescents with open growth plates. The condition is uniformly caused by an activating mutation in fibroblast growth factor receptor 3 (FGFR3). Infigratinib is an oral small molecule that inhibits FGFR3, therefore designed to target this well-described disease at its source.

At the highest dose level evaluated to date (0.128 mg/kg once daily), mean increase in annualized height velocity (AHV) was 1.52 cm/yr over baseline (p=0.02, n=11) for all follow-up data available at time of data cut in children 5 years of age and older. In this group, 64% were responders (defined with a strict criterion of an increase ≥25% in AHV from baseline) and the average percent change from baseline in AHV was 60%.

Earlier cohorts in PROPEL 2 did not achieve the target efficacious exposure as suggested by our preclinical data and no dose response was observed. An increase in AHV over baseline of 0.22 cm/yr (p=0.54, n=38) was observed across these earlier combined cohorts for children 5 years of age and older.

PROPEL 2 enrolled children as young as 3 years of age in the study. Consistent with other trials in the younger age population, both AHV and height Z-scores were analyzed to help control for greater variability in growth seen by age and sex in children 3 to <5 years old. In this age group, the average increase in ACH height Z-score at 6 months was 0.21 standard deviation score (SDS) and an increase in AHV of 0.61 cm/yr over baseline (n=5) was observed. Across Cohort 4, the median duration of follow-up was 26.9 weeks at time of data cut.

“These interim data demonstrate compelling initial proof-of-concept for an oral FGFR inhibitor in children with achondroplasia. We are encouraged by what we have seen to date, including the overall safety profile and promising initial efficacy data of infigratinib. We look forward to completing enrollment in Cohort 5 with the goal of presenting the full study results next year,” said Professor Ravi Savarirayan, M.D., Ph.D., clinical geneticist and group leader of skeletal therapies research at the Murdoch Children’s Research Institute in Australia, the lead investigator for PROPEL 2.

Median follow-up across the entire study is 48.1 weeks across 62 participants included in the safety population at time of data cut. No treatment-related SAEs have been reported to date in any cohort. 90.3% of participants experienced at least one treatment-emergent adverse event (TEAE), the majority of which were Grade 1, unrelated to study drug, and consistent with a pediatric achondroplasia population. Only 9.7% of participants had a TEAE related to study drug, all of which were Grade 1, and included dyspepsia, decreased appetite, flatulence, hypercholesterolemia, hyperphosphatemia, and vitamin D decrease.

A single case of mild hyperphosphatemia (Grade 1) led to a dose reduction in a participant in Cohort 3 (dose: 0.064 mg/kg once daily), the only dose adjustment made to date in the study. Phosphorus levels for this participant only exceeded upper limit of normal by <10% and returned to and remain normal at the reduced dose (0.032 mg/kg once daily). No other cases of hyperphosphatemia have been observed in any other cohort (including Cohort 5) to date, and no trends in phosphorous by dose have been observed. No other AEs required dose modifications or interruptions.

No children have discontinued treatment as the result of an adverse event. No bone-related AEs were observed to date.

“I am encouraged by this interim efficacy and safety data and thankful, as ever, to be partnered with the healthcare providers, children, and families who are making this study possible. These data, combined with our positive proof-of-concept data in LGMD2i and ADH1 earlier this year, highlight BridgeBio’s ability to efficiently prosecute high value programs in large areas of unmet need. We look forward to exploring the potential of infigratinib in achondroplasia and related skeletal dysplasias in the near future,” said Neil Kumar, Ph.D., founder and CEO of BridgeBio.

BridgeBio, after discussions with regulatory agencies, has begun enrolling Cohort 5. Cohort 5 participants are receiving approximately twice the dose of Cohort 4 (0.25 mg/kg once daily vs 0.128 mg/kg once daily in Cohort 4). At the conclusion of the ongoing trial, BridgeBio intends to present full data at a medical conference in the first half of 2023. Additionally, BridgeBio expects to evaluate development of infigratinib in other FGFR-driven skeletal dysplasias, which affect more than 50,000 people in the US and Europe, building on this positive interim data from PROPEL 2 as well as preclinical data in hypochondroplasia presented at the Endocrine Society (ENDO) 2022 Annual Conference earlier this year.

Infigratinib is not approved in any country for the treatment of children or adults with achondroplasia. For more information on infigratinib, please see the following **link.**

References

[1]Response defined as ≥25% increase in AHV from baseline

About BridgeBio Pharma, Inc.

BridgeBio Pharma (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 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 BridgeBio's Phase 2 trial of infigratinib, an investigational therapy for the treatment of children with achondroplasia, including BridgeBio's ability to complete enrollment and dosing in Cohort 5, expectations, plans and prospects regarding BridgeBio's regulatory approval process for infigratinib, cuts of current data being indicative of future cuts of follow-up data, interim results being predictive of final results, interim efficacy and safety data being predictive of final efficacy and safety data, the potential for infigratinib to be the first oral product for the treatment of achondroplasia, BridgeBio's ability to secure intellectual property protection for infigratinib and the duration of such protection, the potential patient population that infigratinib, if approved, could address in the United States and Europe, BridgeBio's ability to efficiently prosecute high value programs in large areas of unmet need, BridgeBio's ability to develop infigratinib in other FGFR-driven skeletal dysplasias, and the success of BridgeBio's continuing discussions with regulatory agencies regarding dosing and trial design, 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, BridgeBio's ability to complete the Phase 2 trial of infigratinib, including its ability to complete enrollment and dosing in Cohort 5, the success of cuts of current data not being indicative of future cuts of follow-up data, interim results not being predictive of final results, interim efficacy and safety data not being predictive of final efficacy and safety data, the potential for infigratinib to be the first oral product for the treatment of achondroplasia, BridgeBio's ability to obtain intellectual property protection for infigratinib and the duration of any such protection, BridgeBio's ability to advance infigratinib in clinical development according to its plans and discussions with regulatory agencies, and potential adverse impacts due to the global COVID-19 pandemic such as delays in regulatory review, manufacturing and clinical trials, supply chain interruptions, adverse effects on healthcare systems and disruption of the global economy; as well as those set forth in the Risk Factors

section of BridgeBio's most recent Annual Report on Form 10-K filed with the U.S. Securities and Exchange Commission (SEC) and in subsequent SEC filings, which are available on the SEC's website at **www.sec.gov**. Moreover, BridgeBio 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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