



NEWS RELEASE

bridgebio pharma reports first quarter 2022 financial results and business update

2022-05-05

–Completed sale to Sentyln Therapeutics of BridgeBio’s NULIBRY™ (Fosdenopterin) for Injection

–Updated strategic collaboration with Helsinn Group to develop, manufacture and commercialize infigratinib in oncology indications in the U.S.; BridgeBio is eligible to receive regulatory and commercial milestone payments as well as tiered royalties on adjusted net sales from Helsinn

–Released positive data from Phase 2 study of BBP-418 in patients with limb-girdle muscular dystrophy type 2i (LGMD2i), which demonstrated a 43% increase in the ratio of glycosylated alpha-dystroglycan (aDG) to total aDG from baseline measured across all three dosing cohorts, signifying the oral therapy has the potential to address both the root cause of LGMD2i and drive functional improvements for patients

–Reported data from ongoing Phase 2 open-label extension (OLE) study of acoramidis (AG10) in patients with symptomatic transthyretin (TTR) amyloid cardiomyopathy (ATTR-CM) which demonstrated acoramidis continued to be well-tolerated and potently stabilize TTR, deepening BridgeBio’s conviction in its Month 30 readout given the stability or improvement with respect to key cardiac biomarkers

–Ended quarter with \$633.5 million in cash, cash equivalents and marketable securities, providing financial runway into 2024

PALO ALTO, Calif., May 05, 2022 (GLOBE NEWSWIRE) -- BridgeBio Pharma, Inc. (Nasdaq: BBIO) (BridgeBio or the

Company), a commercial-stage biopharmaceutical company focused on genetic diseases and cancers, today reported its financial results for the first quarter ended March 31, 2022 and provided an update on the Company's operations.

"Building on our positive Phase 2 data in LGMD2i, we are excited for a summer of ongoing focused execution where we have the opportunity to deliver meaningful data in two additional large conditions with great unmet need: ADH1 and achondroplasia," said Neil Kumar, Ph.D., founder and CEO of BridgeBio.

BridgeBio's key programs:

- Acoramidis (AG10) – Transthyretin (TTR) stabilizer for transthyretin amyloid cardiomyopathy (ATTR-CM): The Company released updated data from its ongoing Phase 2 open-label extension (OLE) study of acoramidis in patients with ATTR-CM, which demonstrated that acoramidis continued to be well-tolerated and potently stabilize TTR. NT-proBNP, a biomarker of cardiac failure and independent predictor of mortality in ATTR-CM patients, was stable or improving throughout the study and serum TTR levels were sustainably increased from baseline at Month 30. Topline data from the Month 30 primary endpoint of the ongoing Phase 3 trial of acoramidis in ATTR-CM (ATTRibute-CM), a hierarchical composite including all-cause mortality and cardiovascular hospitalizations, are expected in mid-2023.
- Encaleret – Calcium-sensing receptor (CaSR) inhibitor for autosomal dominant hypocalcemia type 1 (ADH1): Data from BridgeBio's Phase 2b study of encaleret in ADH1 to be presented at the Endocrine Society's 2022 ENDO conference in Atlanta from June 11-14, 2022. The planned presentation will include the profile of encaleret on clinically relevant mineral homeostasis parameters over 24 weeks of outpatient evaluation. Achieving simultaneous blood and urine calcium normalization is a challenge for patients with ADH1 due to the limitations of the current standard of care. If approved, encaleret could be the first therapy indicated for the treatment of ADH1, a condition caused by gain of function variants of the CASR gene estimated to be carried by 12,000 individuals in the United States alone. The Company intends to initiate a Phase 3 registrational trial of encaleret in patients with ADH1 in 2022 and anticipates Phase 3 topline data in 2023. Issued and filed patents are expected to provide market protection for encaleret in ADH1 to 2041.
- Low-dose infigratinib – FGFR1-3 inhibitor for achondroplasia and hypochondroplasia: Initial data from the ongoing Phase 2 dose-escalation and expansion study is anticipated in mid-2022. The Company also plans to share multiple presentations at the Endocrine Society's 2022 ENDO conference in Atlanta from June 11-14, 2022, including preclinical data for low-dose infigratinib in a mouse model of hypochondroplasia (an FGFR3-driven skeletal dysplasia), new analyses of natural history data in achondroplasia, and the results of a qualitative research study conducted to identify concepts and quality-of-life questionnaires relevant for the

achondroplasia community. Achondroplasia is the most common form of genetic short stature and one of the most common genetic diseases, with a prevalence of over 55,000 cases in the United States and European Union. Low-dose infigratinib is the only known product candidate in clinical development for achondroplasia that is designed to target the disease at its genetic source and the only orally administered product candidate in clinical development for achondroplasia.

- BBP-631 – AAV5 gene therapy candidate for congenital adrenal hyperplasia (CAH): Dosed first patient in Phase 1/2 gene therapy trial in January 2022. Initial Phase 1/2 data readout anticipated in second half of 2022. CAH is one of the most prevalent genetic diseases potentially addressable with adeno-associated virus (AAV) gene therapy, with more than 75,000 cases estimated in the United States and European Union. The disease is caused by deleterious mutations in the gene encoding an enzyme called 21-hydroxylase, leading to a lack of endogenous cortisol production. BBP-631 is designed to provide a functional copy of the 21-hydroxylase-encoding gene (CYP21A2) and potentially address many aspects of the disease course.
- BBP-418 – Glycosylation substrate for limb-girdle muscular dystrophy type 2i (LGMD2i): Announced positive Phase 2 data at the Muscular Dystrophy Association 2022 Annual Meeting on March 14, 2022. BridgeBio plans to engage with regulatory health bodies in 2022 to discuss potential paths to approval and subsequently intends to initiate a Phase 3 clinical trial. If successful, BBP-418 could be the first approved therapy for patients with LGMD2i. BridgeBio believes its initial Phase 2 results indicate the potential for BBP-418 to increase glycosylation of α DG and drive functional improvements for patients, as well as reduce CK, a key marker of muscle breakdown. Furthermore, the 90- and 180-day data show improvements on walk tests from baseline, which the Company believes suggests a potential impact on clinical function and on the rate of disease progression.
- RAS cancer portfolio: BridgeBio expects to select a RAS development candidate in 2022. It previously announced the discovery of its next-generation KRAS G12C dual inhibitors, the first-known compounds that directly bind and inhibit KRAS in both its active (GTP bound) and inactive (GDP bound) conformations, and PI3ka:RAS breakers, small molecules that block RAS driven PI3Ka activation – a novel approach with the potential to inhibit oncogenic PI3Ka signaling without adverse effects on glucose metabolism. RAS is one of the most well-known oncogenic drivers with approximately 30% of all cancers being driven by RAS mutations, including large proportions of lung, colorectal and pancreatic tumors.

Recent corporate updates

- Sale to Sentyln Therapeutics, Inc., of NULIBRY (Fosdenopterin) for Injection: Sentyln has acquired global rights to NULIBRY and will be responsible for the ongoing development and commercialization of NULIBRY in the

United States and developing, manufacturing and commercializing fosdenopterin globally. BridgeBio will share development responsibilities for fosdenopterin through approval of the marketing authorization application already under accelerated assessment with the European Medicines Agency and through approval of its regulatory submission with the Israeli Ministry of Health. Sentyln will provide cash payments upon the achievement of certain regulatory milestones. BridgeBio will be eligible to receive commercial milestone payments as well as tiered royalties on adjusted net sales of NULIBRY. NULIBRY is approved by the U.S. Food and Drug Administration (FDA) to reduce the risk of mortality in patients with molybdenum cofactor deficiency Type A, an ultra-rare, life-threatening pediatric genetic disorder.

- Updated strategic collaboration with Helsinn Group to develop, manufacture and commercialize infigratinib in oncology indications in the U.S.: Under the terms of the amended and restated agreement, Helsinn will gain an exclusive license to commercialize infigratinib in the United States and will be responsible for developing, manufacturing and commercializing infigratinib in oncology indications worldwide except for achondroplasia or any other skeletal dysplasias, and except in mainland China, Hong Kong and Macau. BridgeBio will be eligible to receive regulatory and commercial milestone payments as well as tiered royalties on adjusted net sales from Helsinn. BridgeBio will retain all rights to develop, manufacture and commercialize infigratinib in skeletal dysplasia, including achondroplasia. In 2021, Helsinn and BridgeBio obtained accelerated approval for TRUSELTIQ™ (infigratinib) from the FDA for the treatment of adults with previously treated, unresectable locally advanced or metastatic cholangiocarcinoma with a fibroblast growth factor receptor 2 fusion or other rearrangement as detected by an FDA-approved test.
- Business development – Due to the need to conserve capital and prioritize focused execution, BridgeBio is engaged in partnering and out-licensing discussions for the following programs:
 - BBP-589 – Recombinant collagen 7 for dystrophic epidermolysis bullosa
 - Completed Phase 2 clinical trial evaluating safety and tolerability of BBP-589, as well as clinical proof-of-concept, and initiated a Phase 2 extension study
 - BBP-681 – Topical PI3KA inhibitor for venous and lymphatic malformations
 - Phase 1/2 clinical trial underway
 - BBP-561 – KLK 5/7 inhibitor for Netherton syndrome
 - Investigational New Drug application (IND) enabling studies underway
 - BBP-815 – AAV gene therapy for nonsyndromic hearing loss
 - Preclinical development
 - BBP-818 – AAV gene therapy for classic galactosemia

- IND-enabling studies underway
- BBP-472 – PI3KB inhibitor for PTEN autism
 - Preclinical development

First Quarter 2022 Financial Results:

Cash, Cash Equivalents and Marketable Securities

Cash, cash equivalents and marketable securities, excluding restricted cash, totaled \$633.5 million as of March 31, 2022, compared to \$787.5 million as of December 31, 2021. The net decrease of \$154.0 million pertains primarily to payments for operating costs and expenses of \$160.6 million, which includes \$18.8 million in payments for debt-related interests, timing of payments of accrued compensation and benefits of \$16.9 million and \$3.9 million in payments related to the restructuring initiative that the Company implemented during the current quarter as described below. During the current quarter, the Company received an upfront payment of \$10.0 million upon closing of the asset purchase agreement between its affiliate, Origin Biosciences, and Sentyln Therapeutics.

“We made significant progress in reducing our expenditures in the first quarter, highlighted by a combined reduction of over \$100 million in 2022-2023 operating expenses through the Helsinn and Sentyln deals, but we are not yet done. Our priorities are focusing resources towards select internal programs while finding value-creating partnerships for others that we believe allow science to advance seamlessly towards patients. Our expectation is that operating expenses and cash burn will meaningfully decline in the second quarter as restructuring charges from the beginning of the year decline and anticipated additional business development activity allows us to decrease further from that baseline, establishing a reduced run rate for the rest of the year. We currently expect to have runway up to and beyond the readout of our ATTR catalyst in mid-2023 without requiring equity dilution. We reiterate our guidance of runway into 2024,” said Brian Stephenson, Ph.D., CFA, BridgeBio’s Chief Financial Officer.

BridgeBio is engaged in partnering and out-licensing discussions for a select number of programs, which are listed on the **Company’s website**.

Operating Costs and Expenses

Operating costs and expenses increased by \$7.4 million to \$175.4 million for the first quarter of 2022 as compared to \$168.0 million for the same period in the prior year. During the current quarter, the Company incurred \$22.7 million in restructuring, impairment and related charges, as part of its restructuring initiative. In January 2022, the Company committed to a restructuring initiative designed to drive operational changes in its business processes, efficiencies and cost savings to advance corporate strategy and development programs. The restructuring initiative

included, among other components, consolidation and rationalization of facilities, reprioritization of development programs and the reduction of our workforce. The Company has substantially completed the activities and has incurred most of the costs related to the restructuring initiative during the current quarter. The Company estimates to incur total charges in the range of approximately \$23.0 million to \$25.0 million for the fiscal year 2022, consisting primarily of impairments and write-offs of long-lived assets, severance and employee-related costs, and exit and other related costs. The estimate of the range of costs is subject to certain assumptions, such as the Company's ability to sublease certain office spaces. Actual results may differ from those estimates or assumptions. The nonrecurring charges related to the restructuring initiative were partially offset by a \$15.3 million decrease in the Company's normal operating expenses compared to the same period in the prior year, primarily due to cost savings and impacts from such restructuring initiative.

The Company's research and development expenses have not been significantly impacted by the global COVID-19 pandemic for the periods presented. While BridgeBio experienced some delays in certain of its clinical enrollment and trial commencement activities, it continues to adapt in this unprecedented time to enable alternative site, telehealth and home visits, at-home drug delivery, as well as mitigation strategies with its contract manufacturing organizations. The longer-term impact, if any, of COVID-19 on BridgeBio's operating costs and expenses is currently unknown.

BRIDGEBIO PHARMA, INC.
Condensed Consolidated Statements of Operations
(in thousands, except shares and per share amounts)

	Three Months Ended March 31,	
	2022	2021
Revenue	\$ 1,694	\$ 462
Operating costs and expenses ⁽¹⁾ :		
Research, development and others	108,997	122,559
Selling, general and administrative	43,713	45,407
Restructuring, impairment and related charges	22,662	—
Total operating costs and expenses	<u>175,372</u>	<u>167,966</u>
Loss from operations	(173,678)	(167,504)
Other income (expense), net:		
Interest income	267	394
Interest expense	(20,344)	(9,738)
Other income, net	(7,575)	5,766
Total other income (expense), net	<u>(27,652)</u>	<u>(3,578)</u>
Net loss	(201,330)	(171,082)
Net loss attributable to redeemable convertible noncontrolling interests and noncontrolling interests	4,933	8,003
Net loss attributable to common stockholders of BridgeBio	<u>\$ (196,397)</u>	<u>\$ (163,079)</u>
Net loss per share, basic and diluted	<u>\$ (1.35)</u>	<u>\$ (1.18)</u>
Weighted-average shares used in computing net loss per share, basic and diluted	<u>145,882,149</u>	<u>138,627,729</u>

(1) Amounts include stock-based compensation expense as follows:

	Three Months Ended March 31,	
	2022	2021
Research, development and others	\$ 8,557	\$ 22,449
Selling, general and administrative	14,552	12,447
Restructuring, impairment and related charges	1,172	—
Total stock-based compensation expense	\$ 24,281	\$ 34,896

BRIDGEBIO PHARMA, INC.
Condensed Consolidated Balance Sheets
(In thousands)

	March 31, 2022	December 31, 2021
Assets	(Unaudited)	(1)
Cash and cash equivalents and marketable securities	\$ 633,454	\$ 787,515
Investment in equity securities	37,772	49,148
Receivable from licensing and collaboration agreements	10,983	19,749
Prepaid expenses and other current assets	34,021	32,446
Property and equipment, net	17,182	30,066
Operating lease right-of-use assets	13,936	15,907
Intangible assets, net	30,476	44,934
Other assets	35,325	33,027
Total assets	\$ 813,149	\$ 1,012,792
Liabilities, Redeemable Convertible Noncontrolling Interests and Stockholders' Deficit		
Accounts payable	\$ 10,107	\$ 11,884
Accrued liabilities	88,993	118,247
Operating lease liabilities	19,857	22,366
2029 Notes	733,581	733,119
2027 Notes	540,355	539,934
Term loans	434,114	430,752
Other long-term liabilities	26,829	22,069
Redeemable convertible noncontrolling interests	336	1,423
Total BridgeBio stockholders' deficit	(1,040,996)	(870,414)
Noncontrolling interests	(27)	3,412
Total liabilities, redeemable convertible noncontrolling interests and stockholders' deficit	\$ 813,149	\$ 1,012,792

(1) The condensed consolidated financial statements as of and for the year ended December 31, 2021 are derived from the audited consolidated

financial statements as of that date.

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 and follow us on LinkedIn and Twitter.

BridgeBio Pharma, Inc. Forward-Looking Statements

This press release contains forward-looking statements. Statements 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 our programs and product candidates, including the availability and success of complete data from our ongoing Phase 2 OLE of acoramidis in patients with symptomatic ATTR-CM, availability and success of topline results from Part B of our Phase 3 ATTRIBUTE-CM trial of acoramidis, the availability and success of additional data from our ongoing Phase 2b study of encaleret for ADH1, the timing and success of additional trials of encaleret for ADH1, the availability and success of initial data from our ongoing Phase 2 study of low-dose infigratinib for achondroplasia and our ongoing Phase 1/2 study of BBP-631 for CAH, the timing and success of regulatory discussions regarding potential paths to approval for BBP-418, the ability of BBP-418 to be the first approved therapy for patients with LGMD2i, the timing and success of a Phase 3 trial of BBP-418 in patients with LGMD2i, the timing of our selection of a RAS development candidate, the success of our asset purchase agreement with Sentynl Therapeutics, including our ability to achieve future milestone and royalty payments from Sentynl Therapeutics and the timing of these events, the success of our updated strategic collaboration with Helsinn Group, including our ability to achieve future milestone and royalty payments from Helsinn and the timing of these events, the timing and success of partnering and out-licensing discussions for certain programs in our pipeline, including BBP-589, BBP-681, BBP-561, BBP-815, BBP-818 and BBP-472, the success of our reduction in operating expenses and our expectations for our operating expenses

and cash burn for the second quarter, the success of our restructuring initiative and its savings being realized, as well as our anticipated cash runway, reflect our current views about our plans, intentions, expectations and strategies, which are based on the information currently available to us and on assumptions we have made. Although we believe that our plans, intentions, expectations and strategies 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, initial and ongoing data from our preclinical studies and clinical trials not being indicative of final data, the potential size of the target patient populations our product candidates are designed to treat not being as large as anticipated, the design and success of ongoing and planned clinical trials, future regulatory filings, approvals and/or sales, despite having ongoing and future interactions with the FDA or other regulatory agencies to discuss potential paths to registration for our product candidates, the FDA or such 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, the continuing success of our collaborations, the Company's ability to unlock additional funding under our credit facility, potential volatility in our share price, potential adverse impacts due to the global COVID-19 pandemic such as delays in regulatory review, manufacturing and supply chain interruptions, adverse effects on healthcare systems and disruption of the global economy, as well as those risks set forth in the Risk Factors section of our Annual Report on Form 10-K for the year ended December 31, 2021 and our other filings with the U.S. Securities and Exchange Commission. Moreover, we operate 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.

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