IMMEDIATE RELEASE

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aTyr Pharma and its Hong Kong Subsidiary, Pangu BioPharma, Announce Positive Data from a Phase 1b/2a Clinical Trial Demonstrating Consistent Dose Response of ATYR1923 in Pulmonary Sarcoidosis

Pangu's basic and translational research in tRNA synthetase biology, conducted in collaboration with the Hong Kong University of Science and Technology, contributed to successful clinical proof-of-concept findings.

aTyr Pharma Management to host conference call and webcast today, September 13th at 8:30am ET/8:30pm HKT

HONG KONG – September 13, 2021 – aTyr Pharma, Inc. (Nasdaq: LIFE) ("aTyr" or "The Company"), a clinical stage biotherapeutics company engaged in the discovery and development of innovative medicines based on novel biological pathways, and the company's Hong Kong subsidiary, Pangu BioPharma Limited (Pangu), which supports basic and translational research in tRNA synthetase biology in collaboration with the Hong Kong University of Science and Technology (HKUST), today announced positive results from the Company's Phase 1b/2a double-blind, placebo-controlled clinical trial of its lead therapeutic candidate, ATYR1923, in 37 patients with pulmonary sarcoidosis, a major form of interstitial lung disease (ILD). ATYR1923 was safe and well-tolerated at all doses with no drug-related serious adverse events or signal of immunogenicity. Additionally, the study demonstrated consistent dose response for ATYR1923 on key efficacy endpoints and improvements compared to placebo, including measures of steroid reduction, lung function, sarcoidosis symptom measures and inflammatory biomarkers.

Researchers from Pangu and HKUST were instrumental in discovering a splice variant of histidyl-tRNA synthetase (HARS) that liberates a smaller, extracellular signaling domain from the full-length tRNA synthetase shown to modulate the immune system. ATYR1923 is a fusion protein comprised of this domain fused to the FC region of a human antibody.

"We are very pleased to be a part of this groundbreaking work for ATYR1923, which represents the first clinical proof-of-concept for a tRNA synthetase derived therapy. The consistent dose response and clinically meaningful benefit observed across key efficacy endpoints is quite notable," said Paul Schimmel, Ph.D., Professor of Molecular Medicine at The Scripps Research Institute and Founder of aTyr and Pangu. "The formative basic research leading to the discovery of the active extracellular signaling domain of HARS conducted by Pangu and HKUST laid the foundation for the development of ATYR1923. We look forward to the next steps for the ATYR1923 clinical program and the potential of this novel therapeutic to improve patient outcomes in patients with pulmonary sarcoidosis."

Based on the results of the study, the Company plans to meet with the U.S. Food and Drug Administration to present these data and plans for subsequent clinical development and path to registration for ATYR1923 for pulmonary sarcoidosis and expects to initiate a registrational trial next year.

Phase 1b/2a Clinical Trial in Patients with Pulmonary Sarcoidosis

The Phase 1b/2a study was a randomized, double-blind, placebo-controlled, multiple-ascending dose clinical trial in 37 patients with pulmonary sarcoidosis. The trial consisted of three cohorts testing doses of 1.0 mg/kg, 3.0 mg/kg and 5.0 mg/kg of ATYR1923 or placebo, dosed intravenously every month for six months. The primary objective of the study was to evaluate the safety, tolerability, immunogenicity and pharmacokinetic profile of multiple doses of ATYR1923 compared to placebo. Secondary objectives included the potential steroid-sparing effects of ATYR1923, in addition to other exploratory assessments of efficacy.

About Pulmonary Sarcoidosis and Other ILD

Pulmonary sarcoidosis is an inflammatory disease characterized by the formulation of granulomas, clumps of inflammatory cells, in one or more organs of the body. Approximately 200,000 Americans live with pulmonary sarcoidosis and the prognosis ranges from benign and self-limiting to chronic, debilitating disease, permanent loss of lung function and death. Current treatment options include corticosteroids and other immunosuppressive therapies, which have limited efficacy and are associated with serious side-effects that many patients cannot tolerate long-term.

Pulmonary sarcoidosis is a major form of ILD, which is an umbrella term used for a large group of diseases that cause scarring (fibrosis) of the lung. The scarring causes stiffness in the lungs which makes it difficult to breathe and get oxygen to the bloodstream. Lung damage from ILD is often irreversible and gets worse over time. Other major forms of ILD include connective-tissue disease related ILD (e.g., scleroderma-related ILD), chronic hypersensitivity pneumonitis and idiopathic pulmonary fibrosis (IPF).

Conference Call and Webcast

aTyr Pharma will host a conference call and webcast to discuss the results today, September 13th, at 8:30am ET/8:30pm HKT. Interested parties may access the call by dialing toll-free 844-358-9116 from the US, or 209-905-5951 internationally and using conference ID 1957829. Links to a live webcast and replay may be accessed on the aTyr website events page at: http://investors.atyrpharma.com/events-and-webcasts. A replay will be available for at least 90 days following the event.

About ATYR1923

aTyr is developing ATYR1923 as a potential therapeutic for patients with severe inflammatory lung diseases. ATYR1923, a fusion protein comprised of the immuno-modulatory domain of histidyl-tRNA synthetase fused to the FC region of a human antibody, is a selective modulator of neuropilin-2 that downregulates innate and adaptive immune response in inflammatory disease states. aTyr's lead indication for ATYR1923 is pulmonary sarcoidosis, a major form of interstitial lung disease. Clinical proof-of-concept for ATYR1923 was recently established in a Phase 1b/2a multiple-ascending dose, placebo-controlled study of ATYR1923 in patients with pulmonary sarcoidosis, which demonstrated safety and a consistent dose

response and trends of benefit of ATYR1923 compared to placebo on key efficacy endpoints, including steroid reduction, lung function, clinical symptoms and inflammatory biomarkers.

About aTyr

aTyr is a biotherapeutics company engaged in the discovery and development of innovative medicines based on novel biological pathways. aTyr's research and development efforts are concentrated on a newly discovered area of biology, the extracellular functionality and signaling pathways of tRNA synthetases. aTyr has built a global intellectual property estate directed to a potential pipeline of protein compositions derived from 20 tRNA synthetase genes and their extracellular targets. aTyr's primary focus is ATYR1923, a clinical-stage product candidate which binds to the neuropilin-2 receptor and is designed to down-regulate immune engagement in inflammatory lung diseases. For more information, please visit http://www.atyrpharma.com.

About the Hong Kong University of Science and Technology

The Hong Kong University of Science and Technology (HKUST) (www.ust.hk) is a world-class research university that focuses on science, technology and business as well as humanities and social science. HKUST offers an international campus, and a holistic and interdisciplinary pedagogy to nurture well-rounded graduates with global vision, a strong entrepreneurial spirit and innovative thinking. HKUST attained the highest proportion of internationally excellent research work in the Research Assessment Exercise 2014 of Hong Kong's University Grants Committee, and is ranked as the world's best young university in Times Higher Education's Young University Rankings 2018. Its graduates were ranked 26th worldwide and among the best from universities from Asia in Global Employability University Survey 2020.

Forward-Looking Statements

This press release contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995. Forward-looking statements 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 such safe harbor provisions for forward-looking statements and are making this statement for purposes of complying with those safe harbor provisions. These forward-looking statements include statements regarding potential therapeutic benefits and applications of ATYR1923; timelines and plans with respect to certain development activities (such as the timing of additional clinical trials and planned interactions with regulatory authorities); and certain development goals. These forward-looking statements also 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 these forward-looking statements, are reasonable, we can give no assurance that the plans, intentions, expectations or strategies will be attained or achieved. All forward-looking statements are based on estimates and assumptions by our management that, although we believe to be reasonable, are inherently uncertain. Furthermore, actual results may differ materially from those described in these forward-looking statements and will be affected by a variety of risks and factors that are beyond our control including, without limitation, uncertainty regarding the COVID-19 pandemic, risks associated with the discovery, development and regulation of our product candidates, the risk that we or our partners may cease or delay preclinical or clinical development activities for any of our existing or future

product candidates for a variety of reasons (including difficulties or delays in patient enrollment in planned clinical trials), the possibility that existing collaborations could be terminated early, and the risk that we may not be able to raise the additional funding required for our business and product development plans, as well as those risks set forth in our most recent Annual Report on Form 10-K, Quarterly Reports on Form 10-Q and in our other SEC filings. Except as required by law, we assume no obligation to update publicly any forward-looking statements, whether as a result of new information, future events or otherwise.