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Topline Results from Phase 3 Clinical Study of KRP-R120 for Interstitial Lung Disease

KYORIN Pharmaceutical Co., Ltd. announced today that aTyr Pharma Inc. (Head office: San Diego, CA, CEO and President: Sanjay S. Shukla, hereinafter, "aTyr") disclosed topline results for the global Phase 3 clinical study, EFZO-FIT™ study of KRP-R120 (Efzofitimod).

The study was a randomized, double-blind, placebo-controlled study to evaluate the efficacy and safety of intravenously administered KRP-R120 for the treatment of pulmonary sarcoidosis. The study was conducted in 10 countries and regions, including Japan. In the study, two doses of KRP-R120 (3 mg/kg and 5 mg/kg) were administered intravenously a total of 12 times every four weeks and compared to placebo.

As a result, the study did not meet primary endpoint for the change from baseline in mean daily oral corticosteroid (OCS) dose at week 48 (p=0.3313). The change from baseline in mean daily OCS dose reduced to an average of 2.79 mg for 5 mg/kg KRP-R120 vs 3.52 mg for placebo.

Meanwhile, the study's statistical analysis plan was designed on a hierarchical assessment basis. Therefore, since the primary endpoint was not met, all subsequent statistical testing is reported as nominal findings. The study demonstrated that complete steroid withdrawal was achieved 52.6% of patients treated with 5 mg/kg KRP-R120 vs 40.2% on placebo (p=0.0919). A change was observed from baseline in KSQ-lung score of 10.36 for 5 mg/kg KRP-R120 vs 6.19 for placebo (p=0.0479). Proportion of patients who achieved complete steroid withdrawal with KSQ-lung improvement was 29.5% of patients on 5 mg/kg KRP-R120 vs 14.4% in placebo (p=0.0199).

KRP-R120 was well-tolerated with a consistent safety profile observed previously in all studies conducted to date.

The EFZO-FIT topline results are planned to present at the upcoming European Respiratory Society Congress in September 30, 2025 in Amsterdam, Netherlands.

aTyr press release (September 15, 2025): "aTyr Pharma Announces Topline Results from Phase 3 EFZO-FIT™ Study of Efzofitimod in Pulmonary Sarcoidosis" (https://investors.atyrpharma.com/news-releases)

Based on the results, aTyr and KYORIN are moving forward with discussions on the future development plan for KRP-R120.

The costs for this study have already been factored into the consolidated earnings forecast for the fiscal year ending March 2026.

This document is for reference purposes only. In case of any discrepancy, the Japanese version shall prevail.

[Reference]

About aTyr

aTyr is a clinical stage biotechnology company leveraging evolutionary intelligence to translate tRNA synthetase biology into new therapies for fibrosis and inflammation. tRNA synthetases are ancient, essential proteins that have evolved novel domains that regulate diverse pathways extracellularly in humans. aTyr's discovery platform is focused on unlocking hidden therapeutic intervention points by uncovering signaling pathways driven by its proprietary library of domains derived from all 20 tRNA synthetases. aTyr's lead therapeutic candidate is efzofitimod, a first-in-class biologic immunomodulator in clinical development for the treatment of interstitial lung disease, a group of immune-mediated disorders that can cause inflammation and progressive fibrosis, or scarring, of the lungs. For more information, please visit www.atyrpharma.com.

About Sarcoidosis

Sarcoidosis is an inflammatory disease characterized by the formation of granulomas, clumps of inflammatory cells, in one or more organs in the body. Sarcoidosis affects people of all ages, but typically presents before the age of 50 years, with the incidence peaking at 20 to 39 years. The disorder usually begins in the lungs, skin or lymph nodes, but can affect almost any organ. Sarcoidosis in the lungs is called pulmonary sarcoidosis and affects 90% or more of patients with sarcoidosis have lung involvement. Pulmonary sarcoidosis is a major form of interstitial lung disease (ILD) a group of immune-mediated disorders which cause progressive fibrosis of the lung interstitium. Estimates of prevalence vary, however, current data indicate that approximately 200,000 Americans live with pulmonary sarcoidosis. The prognosis for patients with pulmonary sarcoidosis ranges from benign and self-limiting to chronic, debilitating disease and death.

About KRP-R120 (Efzofitimod)

KRP-R120 is 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 the innate and adaptive immune response in inflammatory disease states.

NRP2 (Neuropilin-2) function: NRP2 is a multifunctional single-spanning trans-membrane glycoprotein that plays a central role in lymphatic vessel development and modulates inflammatory reactions. NRP2 is expressed in various immune cells in inflammatory conditions where they regulate a myriad of functions, including migration, recruitment, phagocytosis and communication between different immune cells. aTyr confirms the contribution of NRP2 in pathogenesis of lung by detecting of the high expression of NPR2 in lung granulomas in patients with lung sarcoidosis.