NEWS RELEASE

NS Pharma

April 18, 2025

FDA Grants Orphan Drug Designation to NS-229 for the Treatment of Eosinophilic Granulomatosis with Polyangiitis

PARAMUS, NJ: April 18, 2025 – NS Pharma, Inc. (NS Pharma) a subsidiary of Nippon Shinyaku Co., Ltd. (Nippon Shinyaku), announced today that the U.S Food & Drug Administration (FDA) has granted Orphan Drug Designation to NS-229, which is being developed for the treatment of the rare disease eosinophilic granulomatosis with polyangiitis (EGPA). NS-229 is being investigated as a selective Janus kinase 1 (JAK1) inhibitor to help regulate immune cell function and prevent the immune system from causing tissue damage.

FDA Orphan Drug Designation status is granted for treatments of rare diseases affecting fewer than 200,000 people in the U.S. This designation provides NS Pharma with a seven-year market exclusivity period, in support of the company's continued development and evaluation of this therapy.

About EGPA

EGPA is a rare autoimmune disease causing inflammation in the small-to-medium-sized blood vessels which can cause tissue and organ damage to the lungs, sinuses, peripheral nerves, skin, and kidneys. EGPA is generally preceded by symptoms of bronchial asthma and allergic rhinitis. The cause is unknown. It is estimated that EGPA affects between 5,600 and 14,500 people in the U.S.*

"There are several factors associated with the inflammatory response in EGPA that could be regulated by JAK1," explained NS Pharma Vice President, Research & Development, Takeshi Seita. "Our therapy has been designed to target this specific enzyme."

A Phase 2 global study of NS-229 is being conducted by Nippon Shinyaku and NS Pharma.

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About NS Pharma, Inc.

NS Pharma, Inc., is a wholly owned subsidiary of Nippon Shinyaku Co., Ltd. NS Pharma is a registered trademark of the Nippon Shinyaku group of companies. For more information, please visit nspharma.com.

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**Estimated prevalences 1.7 / 100,000[1)] and 4.4 / 100,000[2)] were multiplied by a 2023 U.S. population estimate of around 330 million and rounded to nearest hundred.*

1) Bell, CF., Lau, M., Shen, Q. Clinical and Economic Characteristics of Patients Diagnosed with Eosinophilic Granulomatosis with Polyangiitis (EGPA, formerly Churg-Strauss Syndrome) in the United States [abstract]. Arthritis Rheumatol. 2018; 70 (suppl 9).

2) Berti A, Cornec D, Crowson CS, Specks U, Matteson EL. The Epidemiology of Antineutrophil Cytoplasmic Autoantibody-Associated Vasculitis in Olmsted County, Minnesota: A Twenty-Year US Population-Based Study. Arthritis Rheumatol. 2017;69:2338-2350.