

July 29, 2024

Nobelpharma Co., Ltd.

**Launch of “Sargmalin® for inhalation 250 µg”
for Autoimmune Pulmonary Alveolar Proteinosis**

Nobelpharma Co., Ltd. (Headquarters: Chuo-ku, Tokyo, Managing Director & CEO: Jin Shiomura) announced today that “Sargmalin® for inhalation 250 µg” for the indication of autoimmune pulmonary alveolar proteinosis (this “Product”) has been launched.

We are deeply grateful for all of the efforts of the principal investigator, Dr. Koh Nakata, Niigata University Medical and Dental Hospital Advanced Clinical Research Center Specially Appointed Professor, and many physicians participated in the investigator initiated studies, as well as for the cooperation of the patients in the clinical studies and the assistance from Japan Agency for Medical Research and Development (AMED) and National Institute of Biomedical Innovation, Health and Nutrition, owing to which the Product was approved.

This Product is a “sargramostim (genetically modified)” based inhalation with granulocyte-macrophage colony-stimulating factor (GM-CSF), one of the cytokine having a promoting effect for proliferation and differentiation of myeloid precursor, produced with fermentum. This Product has been in the US market since 1991 as an injectable drug for neutrophil recovery following induction chemotherapy and myeloid cell recovery following peripheral stem-cell transplantation in patients with acute myeloid leukemia on the grounds that it increases and functionally activates neutrophils, eosinophils and monocytes.

This Product is the inhalation medicine for the indication of autoimmune pulmonary alveolar proteinosis, which has shown the efficacy and safety for the first time in the world.

Autoimmune pulmonary alveolar proteinosis is developed as the hyperproduced anti-GM-CSF autoantibody blocks mature alveolar macrophage from decomposing waste products including alveolar surfactant. Only established treatments are segmental lung lavage and whole lung lavage. The post-operative rapid symptomatic improvement

can be expected with these treatments, removing accumulated waste products by injecting saline into lungs under general anesthesia, while there exist clinical challenges of being highly invasive and requiring hospitalization as well as the therapeutic opportunities limited to facilities with experienced specialists.

This Product, as inhaled, acts directly on alveolar macrophage to accelerate its maturation and promotes decomposition of the waste products of alveolar surfactant by such matured macrophage, leading to improvement in lung functions.

The inhalation therapy with this Product is less invasive compared to segmental and whole lung lavage and makes home treatment possible. We are confident this will become a new option of “drug treatment” for the clinical practice as well as a rewarding drug that contributes to improve the satisfaction of treatment for autoimmune pulmonary alveolar proteinosis.

In Japan, autoimmune pulmonary alveolar proteinosis has become a designated intractable disease since 2015 and the number of patients is expected to be 730 to 770. We are gratified the launch of this Product assists the treatment of autoimmune pulmonary alveolar proteinosis.

Nobelpharma is committed to continue contributing to society by providing critical but neglected pharmaceuticals and medical devices.

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