NEWS RELEASE

NIPPON SHINYAKU CO., LTD.

November 25, 2020

Nippon Shinyaku Submitted Additional Application of Uptravi® for "Chronic Thromboembolic Pulmonary Hypertension (CTEPH)"

KYOTO, Japan, November 25, 2020 – Nippon Shinyaku Co., LTD. (Nippon Shinyaku; Headquarters, Kyoto; President, Shigenobu Maekawa) announced that it submitted an additional application for chronic thromboembolic pulmonary hypertension (CTEPH) of Uptravi® Tablets 0.2mg and 0.4mg (Generic name: Selexipag), which is Nippon Shinyaku's original product for the treatment of pulmonary arterial hypertension (PAH), to the Ministry of Health, Labour and Welfare (MHLW).

CTEPH is a disease in which thrombus form inside the pulmonary vessels (pulmonary embolism), and this causes increasing pressure on the pulmonary arteries and decreasing blood flow in the lungs and heart. Symptoms such as feeling hard to breathe when moving the body or getting tired quickly develop. If the disease progresses, symptoms such as swollen feet and breathing hard even after a little movement may develop due to poor heart function. The number of patients in Japan is approximately 3,800, and it is designated as an intractable disease.

Treatment consists of surgery to remove the thrombus, catheters to widen the blood vessels, or orally available medicine that opens the pulmonary arteries.

Uptravi® was designated as an orphan drug by MHLW for the indication of CTEPH in June 2016.

In Japan, we have conducted development jointly with Janssen Pharmaceutical K.K., one of the Janssen Pharmaceutical Companies of Johnson & Johnson (Janssen; HQs, Tokyo; President, Chris Hourigan).

For CTEPH patients in Japan who need an additional treatment option, we hope to add this indication for Uptravi® as soon as possible.

<About Uptravi®>

Uptravi® is an orally available agonist which bind in selective and long-acting manner on prostacyclin IP receptors. Uptravi® shows vasodilatory effect by acting selectively on IP receptor, and improves pulmonary hemodynamics. In Japan, it was launched in November 2016 for the treatment of PAH. Overseas, Janssen, a global partner, has received approval and markets Uptravi® in over 55 countries worldwide for the treatment of PAH. In Japan, we are promoting the provision and collection of drug information on Uptravi® in cooperation with Janssen.

<About prostacyclin receptor (IP receptor) agonists>

Binding of PGI_2 to prostacyclin (prostaglandin I_2/PGI_2) receptors in the body causes dilation of peripheral blood vessels and inhibition of platelet aggregation. PGI_2 receptor agonists, such as Uptravi®, are drugs that enhance these effects.

<About Orphan Drugs>

This is a medical product designated by the MHLW after reviewing on the fact that the number of patients in Japan is less than 50,000 and that it is particularly necessary for medical care.

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