NEWS RELEASE

NIPPON SHINYAKU CO., LTD.

May 20, 2026

Launch of Uptravi® Tablets 0.8 mg for the Treatment of Pulmonary Arterial Hypertension and Chronic Thromboembolic Pulmonary Hypertension in Japan

Kyoto, Japan, May 20, 2026 - Nippon Shinyaku Co., Ltd. (Nippon Shinyaku; Headquarters, Kyoto; President, Toru Nakai) announced today that it has launched Uptravi® Tablets 0.8 mg (generic name: selexipag) for the treatment for pulmonary arterial hypertension (PAH) and chronic thromboembolic pulmonary hypertension (CTEPH) in Japan.

Uptravi® Tablets are an orally administered, selective and sustained prostacyclin (IP) receptor agonist. By binding to IP receptors on vascular smooth muscle cells and increasing cyclic adenosine monophosphate (cAMP) production, the drug is believed to reduce pulmonary arterial pressure through vasodilatory effects and inhibition of vascular smooth muscle cell proliferation.
In Japan, Nippon Shinyaku currently markets Uptravi® Tablets at doses of 0.2 mg and 0.4 mg. Uptravi® Tablets are designed with a treatment regimen that allows for stepwise dose titration tailored to each patient's condition. With the addition of the higher-dose 0.8-mg tablet, the number of tablets required can be reduced, which is expected to improve patient adherence.

PAH is a disease with a poor prognosis characterized by abnormally high blood pressure in the pulmonary artery. It is classified into idiopathic PAH, heritable PAH and PAH associated with various diseases such as connective tissue disease and congenital heart disease. CTEPH is a disease in which thrombi 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.

Nippon Shinyaku focuses on rare and intractable diseases as a key area and continues to make further efforts to develop new treatments and provide product information. With the launch of Uptravi® Tablets 0.8 mg, we hope to contribute even more to patients suffering from PAH and CTEPH, their families, and healthcare providers.

Summary of Uptravi® tablets 0.8 mg

<table>
<tr><td>Brand name</td><td>Uptravi® tablets 0.8 mg</td></tr>
<tr><td>Generic name</td><td>Selexipag</td></tr>
<tr><td>Date of approval</td><td>February 13, 2026</td></tr>
<tr><td>Date of NHI reimbursement price listing</td><td>May 20, 2026</td></tr>
<tr><td>Date of launch</td><td>May 20, 2026</td></tr>
<tr><td>Number of approval</td><td>30800AMX00069000</td></tr>
<tr><td>Dosage Forms and Strengths</td><td>A round film-coated tablet containing 0.8 mg of selexipag per tablet</td></tr>
<tr><td>Indication</td><td>・Pulmonary arterial hypertension
・Chronic thromboembolic pulmonary hypertension for which surgical treatment is not indicated or which remained/recurred after surgical treatment</td></tr>
<tr><td>Dosage and administration</td><td>< Pulmonary arterial hypertension >
The usual dose for adults is started at 0.2 mg per dose, administered orally twice daily after meals. While confirming the tolerability, the maintenance dose is determined by increasing the 0.2 mg per dose to the maximum tolerated dose at intervals of 7 days or more. The maximum is 1.6 mg per dose, and oral administration is performed twice daily after meals at each dose.
For pediatric patients aged two years or older, the starting dose in the table below are usually administered orally twice daily after meals. While confirming the tolerability, the maintenance dose is determined by increasing the dose to the maximum tolerated dose according to the table below at intervals of 7 days or more. It is not allowed to exceed the maximum dose in the table below. In any dose, and oral administration is performed twice daily after meals at each dose.
<table>
<tr><th>Body weight (kg)</th><th>Starting dose (per dose)</th><th>Increasing dose (per dose)</th><th>Maximum dose (per dose)</th></tr>
<tr><td>≥9kg to <25kg</td><td>0.1mg</td><td>0.1mg</td><td>0.8mg</td></tr>
<tr><td>≥25 to <50 kg</td><td>0.15mg</td><td>0.15mg</td><td>1.2mg</td></tr>
<tr><td>≥50 kg</td><td>0.2mg</td><td>0.2mg</td><td>1.6mg</td></tr>
</table>
< Chronic thromboembolic pulmonary hypertension for which surgical treatment is not indicated or which remained/recurred after surgical treatment >
The usual dose for adults is started at 0.2 mg per dose, administered orally twice daily after meals. While confirming the tolerability, the maintenance dose is determined by increasing the 0.2 mg per dose to the maximum tolerated dose at intervals of 7 days or more. The maximum is 1.6 mg per dose, and oral administration is performed twice daily after meals at each dose.</td></tr>
<tr><td>NHI reimbursement price</td><td>JPY 6,272.20 /tablet</td></tr>
<tr><td>Packaging unit</td><td>30 tablets [10 tablets (PTP) X 3]</td></tr>
</table>

Contact

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