FOR IMMEDIATE RELEASE

Tokyo, January 22, 2021

OrphanPacific, Inc.
Torii Pharmaceutical Co., Ltd.

OrphanPacific Inc. Receives Manufacturing and Marketing Approval of ORLADEYO Capsules 150mg for Suppression of HAE Attacks in Japan

OrphanPacific, Inc.(OrphanPacific) and Torii Pharmaceutical Co., Ltd. (Torii) announced that OrphanPacific has today received manufacturing and marketing approval for ORLADEYO Capsules 150mg (generic name: berotralstat hydrochloride) (hereinafter “ORLADEYO”), a plasma kallikrein inhibitor indicated for the suppression of the onset of attacks in acute hereditary angioedema (HAE) in Japan.

ORLADEYO is a drug developed in Japan by BioCryst Pharmaceuticals, Inc. ("BioCryst") and earned the Sakigake designation in October 2015, and the orphan drug designation in December 2018. The New Drug Application was filed by OrphanPacific under the contract with BioCryst.

ORLADEYO is a new oral agent that is designed to supress the onset of acute HAE attacks by specifically inhibiting the bradykinin-producing enzyme. The Phase 3 clinical studies conducted in Japan confirmed the efficacy and safety of ORLADEYO in HAE patients for the suppression of the onset of acute HAE attacks.

OrphanPacific and Torii expect ORLADEYO to be a new option for the treatment of HAE in Japan. Under the terms of the November 2019 agreement between BioCryst and Torii, ORLADEYO will be sold exclusively by Torii in Japan. The drugs’ launch date will be announced as soon as a decision is reached for the National Health Insurance Drug Price.

Approval Summary

Product Name	: ORLADEYO capsule 150mg
Generic Name	: berotralstat hydrochloride
Indications	: Suppression of the onset of acute attacks in patients with hereditary angioedema
Dosage and Administration	: The recommended dose of berotralstat for adults and children aged 12 years or older is 150mg (1 capsule)

taken orally once daily.

Manufacturing and Distributor : OrphanPacific, Inc.

Distributor : Torii Pharmaceutical Co., Ltd.

ABOUT hereditary angioedema (HAE)

HAE is a genetic disorder that develops by overproduction of bradykinin, a vasodilator substance, due to C1 inhibitor deficiency or functional decline and causes sudden edema to various regions of the body such as extremities, face, throat or digestive organ, resulting in swelling and pain that may affect the patients' quality of daily life.

Contact for OrphanPacific, Inc.
Corporate Planning Department
OrphanPacific, Inc. Tokyo: +81-3-6779-8151
E-mail: info@orphanpacific.com

Contact for Torii Pharmaceutical Co., Ltd.:
Corporate Planning Department
Torii Pharmaceutical Co., Ltd. Tokyo: +81-3-3231-6814
E-mail: webmaster@torii.co.jp