NEWS RELEASE

NIPPON SHINYAKU CO., LTD.

September 4th, 2019

Launch of Defitelio® injection 200mg for the treatment of sinusoidal obstruction syndrome (hepatic veno-occlusive disease)

Nippon Shinyaku Co., Ltd (HQ: Minami-ku, Kyoto, President: Shigenobu Maekawa) announced as of today that it has launched Defitelio® injection 200mg (defibrotide sodium/NS-73) for the treatment of sinusoidal obstruction syndrome [(SOS), hepatic veno-occlusive disease (VOD)].

SOS (VOD) is a life-threatening complication caused by the intensive therapy a patient receives before hematopoietic stem cell transplantation (HSCT), and after chemotherapy or radiation therapy. Features of SOS (VOD) include elevation of bilirubin level, weight gain, tender hepatomegaly, ascites, and jaundice. In severe cases, SOS (VOD) can cause multi-organ dysfunction or failure, leading to a high mortality rate. Since no clinically effective drug for SOS (VOD) is currently available in Japan, Defitelio has been developed as a new treatment option.

Defitelio is thought to work by stabilizing and protecting vascular endothelium, and normalizing the coagulation/fibrinolysis balance.

Nippon Shinyaku licensed in Defitelio from Jazz Pharmaceuticals plc in 2017 and obtained the marketing approval based on the results from the Japanese investigator sponsored clinical studies and the global studies in June, 2019. Defitelio is commercially available in 35 countries including U.S. and European countries as the first approved therapy for the treatment of severe hepatic SOS (VOD) which develops in patients following HSCT. The Ministry of Health, Labour and Welfare (MHLW) granted an orphan drug designation of Defitelio in September, 2018.

Nippon Shinyaku will contribute to the treatment of patients with SOS (VOD) in Japan by appropriately deliverin g medical information on Defitelio to medical facilities.

Summary of Defitelio

Brand Name	Defitelio Injection 200mg
Generic Name	defibrotide sodium
Date of approval	June 18th, 2019
Date of NHI reimbursement price listing	September 4th, 2019
Date of launch	September 4th, 2019
Number of approval	30100AMX00006000
Dosage Forms and Strengths	Aqueous Injection fluid: defibrotide sodium 200 mg/2.5 mL in a vial
Indications and Usage	sinusoidal obstruction syndrome (SOS)/ hepatic veno-occlusive disease (VOD)
Dosage and administration	Administer defibrotide sodium 6.25 mg/kg as a 2-hour intravenous infusion 4 times a day
NHI reimbursement price	JPY53,108/vial (200mg/2.5mL)
Packaging unit	2.5mL x 10 vials

ABOUT JAZZ PHARMACEUTICALS PLC

Jazz Pharmaceuticals plc (Nasdaq: JAZZ), a global biopharmaceutical company, is dedicated to developing life-changing medicines for people with limited or no options. As a leader in sleep medicine and with a growing hematology/oncology portfolio, Jazz has a diverse portfolio of products and product candidates in development, and is focused on transforming biopharmaceutical discoveries into novel medicines. For more information, please visit www.jazzpharmaceuticals.com.

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