

September 28, 2023

Translation

MEDIPAL HOLDINGS CORPORATION
JCR Pharmaceuticals Co., Ltd.**MEDIPAL HOLDINGS and JCR Pharmaceuticals Conclude Two Agreements
on Mucopolysaccharidosis Type IIIB Treatment**

September 28, 2023 -- MEDIPAL HOLDINGS CORPORATION (HQ: Chuo-ku, Tokyo; Representative Director, President and CEO: Shuichi Watanabe; hereinafter, "MEDIPAL") and JCR Pharmaceuticals Co., Ltd. (HQ: Ashiya, Hyogo Prefecture; Chairman and President: Shin Ashida; hereinafter, "JCR") announced today that the Board of Directors of both companies conclude two agreements on JR-446, a blood-brain barrier (BBB) penetrating form of α -N-acetylglucosaminidase that was developed using JCR's proprietary J-Brain Cargo[®] technology, for the treatment of Mucopolysaccharidosis Type IIIB (MPS IIIB or Sanfilippo syndrome type B).

In October 2022, both companies entered into a partnership for the development of certain drugs for the treatment of ultra-rare diseases. JR-446 is an additional program to this partnership.

Under the ex-Japan licensing agreement, MEDIPAL will acquire an exclusive commercialization right of JR-446 with sublicensing rights, including clinical development, manufacturing, and marketing. As regards to the Japan collaboration agreement, MEDIPAL will support JCR in the clinical development of JR-446 in Japan, including distribution of investigational drugs, disease awareness and conducting clinical trials. MEDIPAL will receive a certain amount of profit under a profit-sharing scheme based on the sales in Japan.

In accordance with both agreements, JCR is scheduled to receive upfront payments. In addition, JCR will receive royalties dependent on the sales outside Japan and milestone payments when relevant milestone events are achieved in the development progress in Japan. JCR is responsible for sales in Japan and will book sales.

JR-446 is a novel drug developed based on the J-Brain Cargo[®] technology, which has been clinically validated through the approval of IZCARGO[®] for the treatment of MPS II in Japan in 2021. Animal studies have previously confirmed delivery of JR-446 into the CNS (Central Nervous System) and somatic organs and significant reduction of substrate caused by the deficiency in the enzyme that causative for the onset of MPS IIIB. As there is no established standard of care for the treatment of MPS IIIB, JR-446 is believed to have the potential to become a transformative treatment of MPS IIIB.

MEDIPAL and JCR recognize the urgency to develop treatment for MPS IIIB for which there is no

established treatment. It is also noted that there are potentially undiagnosed and misdiagnosed MPS IIIB patients in Japan and overseas due to the nature of the disease being confused with different diseases.

Additional explanation

About the J-Brain Cargo® platform technology

JCR Pharmaceuticals has developed a proprietary BBB-penetrating technology J-Brain Cargo®, to bring biotherapeutics into the CNS. The technology allows a target substance to penetrate the BBB using transferrin receptors which appear on the surface of endothelial cells in brain capillaries. The first drug developed based on this technology and approved in Japan for the treatment of MPS II (mucopolysaccharidosis type II) is IZGARGO® (INN: pabinafusp alfa).

About Sanfilippo syndrome (MPS III)

Sanfilippo syndrome is a collection of autosomal recessive disease caused by a deficiency of enzymes that metabolize certain mucopolysaccharides within the body. The disease is classified into four subtypes (A, B, C, and D) by the deficiency in the respective enzyme, causing accumulation of heparan sulfate in tissues throughout the body, including the brain. While MPS III is a heterogeneous disease, the rapidly progressive form of the disease frequently affects neurocognitive development, which peaks around 2 or 3 years of age, before subsequently deteriorating, resulting, among other signs and symptoms, in a complete loss of speech by the age of 7 or 8. Progression further gives rise to symptoms such as sleep disorders, hepatosplenomegaly, seizures, impaired mobility, vision impairment, and neurobehavioral abnormalities.

About MEDIPAL HOLDINGS CORPORATION (As of March 31, 2023)

Representative: Shuichi Watanabe, Representative Director, President and CEO
Headquarters: 3-1-1, Kyobashi, Chuo-ku, Tokyo, Japan
Established: May 6, 1923
Employees (consolidated): 12,801
Paid-in capital: 22,398 million yen
Scope of business: As a holding company, MEDIPAL HOLDINGS CORPORATION controls, administers, and supports the operating activities of companies in which it holds shares in the Prescription Pharmaceutical Wholesale Business; the Cosmetics, Daily Necessities and OTC Pharmaceutical Wholesale Business; and the Animal Health Products and Food Processing Raw Materials Wholesale Business, and conducts business development for the MEDIPAL Group.

About JCR Pharmaceuticals Co., Ltd. (As of March 31, 2023)

Representative: Shin Ashida, Chairman, President and CEO
Headquarters: 3-19, Kasuga-cho, Ashiya, Hyogo, Japan
Established: September 13, 1975
Employees (consolidated): 854
Paid-in capital: 9,061 million yen
Scope of business: Manufacture, selling, buying, import/export of pharmaceutical products, regenerative medicines and drug substances. Selling, buying, import/export of medical devices and laboratory instruments.

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