To Whom it may concern

January 30, 2026

Ohara Pharmaceutical Co., Ltd.

Notice of Filing for Manufacturing and Marketing Authorization
Dordaviprone Hydrochloride (Domestic Development Code: OP-10)

Ohara Pharmaceutical Co., Ltd. (Head Office: Koka City, Shiga Prefecture; Representative Director and CEO: Seiji Ohara; hereinafter referred to as "Ohara") today announced that it has filed an application in Japan for the manufacturing and marketing authorization of dordaviprone hydrochloride (domestic development code: OP-10; hereinafter referred to as "OP-10"). The planned indication is for the treatment of patients with diffuse midline glioma harboring an H3 K27M mutation.

OP-10 is considered to induce apoptosis in tumor cells through activation of the mitochondrial protease ClpP (Caseinolytic protease P) and antagonistic activity against dopamine D2 receptors, leading to metabolic disruption within tumor cells, mitochondrial damage, and activation of the integrated stress response pathway. [1)]

In the United States, Jazz Pharmaceuticals plc (NASDAQ: JAZZ; hereinafter "Jazz") obtained accelerated approval in August 2025 for dordaviprone for the treatment of adult and pediatric patients aged 1 year and older with diffuse midline glioma harboring an H3 K27M mutation whose disease has progressed following prior therapy (U.S. brand name: MODEYSO).

In 2019, Ohara entered into a licensing agreement with Oncoceutics, Inc. (Subsequently acquired by Chimerix/Jazz) for the development and commercialization of OP-10 in Japan and has continued its domestic development efforts, including the implementation of clinical studies in Japanese patients. Based on clinical study results in both Japanese and non-Japanese patients with diffuse midline glioma, Ohara has submitted its application for manufacturing and marketing authorization of OP-10.

Ohara remains committed to making OP-10 available to patients as quickly as possible and will continue to exert every effort to address significant unmet medical needs.

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About Diffuse Midline Glioma with H3 K27M Mutation

Diffuse midline glioma is a collective term for infiltrative gliomas that arise in or near the midline structures of the central nervous system, including the diencephalon (thalamus and hypothalamus), brainstem (midbrain, pons, and medulla), and spinal cord. The majority of these tumors harbor the H3 K27M mutation, in which lysine at position 27 of the N-terminal tail of histone H3.3 or H3.1 is replaced by methionine. In the 2016 World Health Organization (WHO) Classification of Tumours of the Central Nervous System, "diffuse midline glioma, H3 K27M–mutant" was defined as a Grade IV tumor, representing the highest level of malignancy. [2)]

The standard treatment approach for diffuse midline glioma with H3 K27M mutation consists of maximal safe tumor resection followed by radiotherapy whenever possible. However, due to the tumor location, surgical resection is often not performed, as surgery may worsen neurological symptoms. In addition, the therapeutic effect of radiotherapy is temporary, and tumor regrowth commonly occurs.[3) 4)]

At present, no drugs have been approved in Japan for the treatment of diffuse midline glioma with H3 K27M mutation.

References

1) Prabhu VV, et al. ONC201 and imipridones: Anti-cancer compounds with clinical efficacy. Neoplasia. 2020, 22(12), 725-744.

2) Louis DN, et al. The 2016 World Health Organization classification of tumors of the central nervous system: a summary. Acta Neuropathol. 2016, 131, 803-820.

3) The Japan Neurosurgical Society; The Japanese Society of Pathology. Clinical and Pathological Guidelines for the Management of Brain Tumors, 5th Edition. Kanehara & Co., Ltd., 2023.

4) The Japan Society for Neuro-Oncology; The Japan Neurosurgical Society. Guidelines for the Diagnosis and Treatment of Brain Tumors: Pediatric Brain Tumor Edition (2022). Kanehara & Co., Ltd., 2022.

About Ohara Pharmaceutical Co., Ltd.

Ohara Pharmaceutical Industrial Co., Ltd. is a pharmaceutical company built on three core pillars: the development of innovative drugs centered on orphan drugs for rare diseases, a generic pharmaceuticals business, and global operations with a particular focus on Africa.

Guided by its mission, "Everything from the patient's perspective—delivering trusted pharmaceuticals for the future of healthcare," Ohara pursues innovation not only in treatment, but also across prevention, diagnosis, and aftercare, while responding to changes in the healthcare environment and striving to improve treatment outcomes.

Ohara aims to enhance the quality of healthcare by providing Total Healthcare Solutions and is committed to delivering reliable pharmaceuticals and services that earn the trust of patients worldwide.