

**Company announcement**

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Inside information

**Orphazyme A/S in restructuring**

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Company Registration No. 32266355

## **Orphazyme A/S under In-Court-Restructuring to sell substantially all of its assets and business activities to KemPharm, Inc.**

**Copenhagen, Denmark, 15 May 2022**

- KemPharm to acquire Orphazyme assets, including those relating to the development and approval of arimoclomol, for a total of USD 12.8 million in cash and assumed liabilities estimated to equal approximately USD 5.2 million
- The majority of Orphazyme's approximately 20 current employees will become employees at KemPharm
- KemPharm intends to continue to pursue approval of arimoclomol as a treatment option for NPC

Orphazyme A/S in restructuring (ORPHA.CO; ORPH) ("Orphazyme" or the "Company"), a late-stage biopharmaceutical company developing arimoclomol for Niemann-Pick disease type C (NPC), announces today that it has signed an agreement to sell substantially all of the Company's assets and business activities to KemPharm Denmark A/S, a wholly owned subsidiary of KemPharm Inc. (KMPH: NASDAQ, NY). KemPharm is a specialty pharmaceutical company focused on the discovery and development of novel treatments for rare central nervous system diseases.

Under the agreement, KemPharm intends to retain substantially all of Orphazyme's current employees, to continue the early access programs with arimoclomol, and to continue to pursue the potential approval of arimoclomol as a treatment option for NPC. KemPharm will pay Orphazyme a total of USD 12.8 million in cash and assume liabilities estimated to equal approximately USD 5.2 million. Completion of the transaction is expected to result in full or very high coverage to creditors with undisputed claims based on the claims filed during the restructuring. The deal is subject to approval by Orphazyme's creditors and the Danish bankruptcy court, and it is expected to be completed on or before 1 June 2022.

"Since Orphazyme is under in-court restructuring, the primary objective was to secure a deal that satisfies our obligations towards the creditors including our employees. We are pleased that this has been achieved. In addition, we have secured the continued pursuit of developing arimoclomol in the hope of making it available for NPC patients, which has been our driving motivation since the foundation of the company," stated Georges Gemayel, Chairman of the Board of Directors of Orphazyme.

Travis C. Mickle, President and CEO of KemPharm, stated: "NPC is a devastating disease and there is a profound need for an effective treatment to help patients. We believe the efficacy signal for arimoclomol is convincing, and that there is a viable regulatory pathway to obtain regulatory marketing approvals. KemPharm has had significant experience with challenging regulatory situations, and we welcome the opportunity to work together with the team to resubmit the regulatory applications and make arimoclomol available to all who could benefit from the treatment."

Following completion of the deal, Orphazyme will no longer have any ongoing operational business activities.

**For additional information, please contact****Orphazyme A/S in restructuring**

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**About KemPharm**

KemPharm is a specialty pharmaceutical company focused on the discovery and development of novel treatments for rare central nervous system (CNS) diseases through its proprietary LAT<sup>®</sup> (Ligand Activated Therapy) platform technology. KemPharm utilizes its proprietary LAT<sup>®</sup> platform technology to generate improved prodrug versions of FDA-approved drugs as well as to generate prodrug versions of existing compounds that may have applications for new disease indications. KemPharm's prodrug product candidate pipeline is focused on the high need areas of idiopathic hypersomnia (IH) and other CNS/rare diseases. In addition, the U.S. Food and Drug Administration (FDA) has approved AZSTARYS<sup>®</sup>, a once-daily treatment for ADHD in patients ages six years and older containing KemPharm's prodrug, serdexmethylphenidate (SDX), which is being commercialized by Corium, Inc. in the U.S., and APADAZ<sup>®</sup>, an immediate-release combination product containing benzhydrocodone, KemPharm's prodrug of hydrocodone, and acetaminophen, which is being commercialized by KVK-Tech, Inc. in the U.S. For more information on KemPharm and its pipeline of prodrug product candidates visit [www.kempharm.com](http://www.kempharm.com) or connect with us on [Twitter](#), [LinkedIn](#), [Facebook](#) and [YouTube](#).

**About Orphazyme**

Orphazyme is a late-stage biopharmaceutical company developing arimoclochol for Niemann-Pick disease type C (NPC). Orphazyme is headquartered in Denmark. Orphazyme's shares are listed on Nasdaq Copenhagen (ORPHA).

**About arimoclochol**

Arimoclochol is an investigational drug candidate that amplifies the production of heat shock proteins (HSPs). HSPs can rescue defective misfolded proteins and improve the function of lysosomes. Arimoclochol is administered orally, and has now been studied in 10 Phase 1, four Phase 2, and three pivotal Phase 2/3 trials. Arimoclochol has received Orphan Drug Designation (ODD) for NPC in the US and EU. Arimoclochol has received Fast-Track Designation (FTD), Breakthrough Therapy Designation (BTD), and Rare Pediatric Disease Designation (RPDD) from the U.S. Food and Drug Administration (FDA) for NPC. On June 17, 2021, Orphazyme received a Complete Response Letter from the FDA regarding its New Drug Application for arimoclochol for the treatment of NPC. The company has requested a type B-meeting to be held early Q3 2022.

**Forward-looking statement**

This company announcement may contain certain forward-looking statements under the U.S. Private Securities Litigation Reform Act of 1995 and otherwise, including forward-looking statements about the Company's sale of substantially all of its assets and business activities to KemPharm Inc. Although the Company believes its expectations are based on reasonable assumptions, all statements other than statements of historical fact included in this company announcement about future events are subject to (i) change without notice and (ii) factors beyond the Company's control, including pursuant to regulatory or judicial intervention. Except as required by law, the Company assumes no obligation to update these forward-looking statements publicly, or to update the reasons actual results could differ materially from those anticipated in the forward-looking statements, even if new information becomes available in the future.