

Company announcement No. 36/2022

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<u>www.orphazyme.com</u> Company Registration No. 32266355

Orphazyme reports financial results in Interim Report First Half 2022

Copenhagen, Denmark, September 26, 2022 – Orphazyme A/S (ORPHA.CO) (the "Company") today announces its Interim Report First Half 2021 for the period January 1 – June 30, 2022.

Anders Vadsholt, Chief Executive Officer and Chief Financial Officer said, "During the first half of 2022, we executed the sale of substantially all of Orphazyme's assets, including the continuing development of arimoclomol, to KemPharm Denmark A/S, a wholly owned subsidiary of KemPharm, Inc. Our employees demonstrated unwavering commitment throughout this difficult time, and I am delighted that many of them are pursuing their mission to bring new treatment options to patients with NPC at KemPharm. We are now focused on exploring ways to realize value from the Company's remaining assets and may engage in activities and investments related to biopharmaceutical research and development, participate in partnerships or cooperate with other businesses."

Corporate Updates First Half 2022

- In January 2022, Christophe Bourdon, Chief Executive Officer (CEO), resigned from his position as CEO of Orphazyme to take on the role of CEO at another company. Anders Vadsholt was appointed CEO of Orphazyme, effective March 1, 2022, in addition to his position as Chief Financial Officer (CFO).
- In February 2022, Orphazyme was notified of a negative trend vote by the Committee for Medicinal Products for Human Use (CHMP) of the European Medicine's Agency (EMA) relating to its marketing authorization application (MAA) for arimoclomol in Niemann-Pick disease type C, or NPC, in Europe. The Company withdrew the MAA in March 2022, ahead of the CHMP's final vote.
- On March 11, 2022, the Company's board of directors (the "Board of Directors") filed to initiate incourt restructuring proceedings under the Danish Insolvency Act with the aim of exploring whether a
 basis could be established for all or part of Orphazyme's operations to continue, including a basis for
 injecting further capital, and/or a basis for a sale of all or parts of its assets.
- In March 2022, the Company voluntarily delisted American Depositary Shares (ADSs) representing Orphazyme's ordinary shares from the Nasdaq Global Select Market in the United States, effective March 31, 2022.
- In May 2022, the Company signed an agreement to sell substantially all Orphazyme's assets and business activities, including those relating to the development and approval of arimoclomol and the full claw back liability related to the French early access program, to KemPharm Denmark A/S ("KemPharm"), a wholly owned subsidiary of KemPharm, Inc., for a total amount of USD 12.8 million in cash and assumed liabilities estimated to equal approximately USD 5.2 million (the "Sale of Assets").
- Stephanie Okey, Carrolee Barlow and Martin Bonde stepped down from their positions as members of the Board of Directors as of May 23, 2022.
- The restructuring proposal was approved on May 30, 2022, by Orphazyme's known creditors and affirmed by the Danish Maritime and Commercial High Court; the in-court restructuring proceedings were discontinued on May 30, 2022, and the Sale of Assets was completed on May 31, 2022.
- In June 2022, Bo Jesper Hansen was elected Chairman of the Board of Directors and John Sommer Schmidt and Anders Vadsholt were elected to the Board of Directors. Georges Gemayel, former Chairman of the Board of Directors and Andrew Mercieca, member of the Board of Directors did not stand for re-election at the Company's Annual General Meeting.

Financial Highlights First Half 2022

Unless otherwise stated, comments in this announcement refer to H1 performance from continuing operations. From January 1, 2022, the business operations and activities that were part of the Sale of Assets agreement with KemPharm have been reclassified as discontinued operations. 2021 figures have been restated according to IFRS.



- Operating expenses for continuing operations for the six months ended June 30, 2022 totaled DKK 27.3 million compared to DKK 57.3 million for the same period in 2021.
- Loss from continuing operations totaled DKK 24.4 million compared DKK 55.0 million for the same period in 2021
- The net result from operations held for sale (discontinued operations) was a profit of DKK 87.4 million compared to a net loss of DKK 408.8 million for the same period in 2021, primarily due to the Sale of Assets to KemPharm.
- The total comprehensive profit/loss (both continuing and discontinued operations) for the first half 2022
 was a net profit of DKK 62.5 million, or DKK 1.79 per share (basic and diluted), compared to a total
 comprehensive loss of DKK 463.7 million, or DKK (13.27) per share (basic and diluted) for the same period
 in 2021.
- As of June 30, 2022, Orphazyme held cash totaling DKK 49.5 million compared to DKK 102.3 million as of December 31, 2021.

Subsequent Events

- As announced in April 2022, the Deposit Agreement among the Company, The Bank of New York Mellon, as depositary, and owners and American Depositary Receipt (ADRs) holders was terminated, effective July 6, 2022.
- On August 11, 2022, the U.S. District Court for the Northern District of Illinois granted in part and denied in part defendants' motion to dismiss the amended complaint in the putative class action lawsuit, captioned *Busic v. Orphazyme A/S*, et al., No. 21- cv-03640 (N.D. III.). The Court dismissed all claims under the Securities Act of 1933 relating to the Company's registration statement for its IPO of ADSs and dismissed as inactionable a number of challenged statements for the lead plaintiff's claims under the Securities Exchange Act of 1934. On August 26, 2022, the Court amended its August 11, 2022 order to also dismiss certain claims against certain individual defendants. The Court's dismissal of claims in its August 11 and 26, 2022 orders was without prejudice to the filing of a further amended complaint. On September 8, 2022, the lead plaintiff filed a second amended complaint. Defendants have until October 6, 2022 to respond to the second amended complaint. Management does not believe these claims have any merit and believe that the outcome will not materially affect the Company's financial position.

Outlook

As announced September 25, 2022 (Company announcement No. 35/2022), Orphazyme has adjusted its outlook for 2022, as published on June 7, 2022. The change is driven by the separation of activities into continuing and discontinued operations. The discontinued activities are related to the sale of assets to KemPharm A/S. The continuing business activities are the limited ongoing operational business activities with only two employees. For the full-year 2022, we anticipate a loss from continuing operations in the range DKK 40 – 45 million and a positive result from discontinued operations relating to the Sale of Assets to KemPharm of DKK 87.4 million (previously operating profit in the range DKK 10 – 30 million). As previously communicated, we expect to end 2022 with more than DKK 30 million in cash and equivalents.

For additional information, please contact

Orphazyme A/S

Anders Vadsholt, Chief Executive Officer and Chief Financial Officer

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Forward-looking statement

This company announcement may contain certain forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995, including in respect of the company's anticipated operating plans and performance, financial position and ability to operate as a going concern. 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 actions by regulatory agencies, effects of the global COVID-19 pandemic, and the impact of the Company's Sale of Assets. These statements may include, without limitation, any statements preceded by, followed by, or including words such as "target," "believe," "expect," "aim," "intend," "may," "anticipate," "estimate," "plan," "project," "will," "can have," "likely," "should," "would," and other words and terms of similar meaning or the negative thereof. Forward-looking statements are subject to inherent risks and uncertainties beyond the Company's control that could cause the Company's actual results, performance, or achievements to be materially different from the expected results, performance, or achievements expressed or implied by such forward-looking statements. In light of these risks and uncertainties, and other risks and uncertainties that are described in the Risk Factors section and other sections of the Company's Annual Report on Form 20-F, filed with the Securities and Exchange



Commission (SEC) on March 2, 2021, and other filings that the Company makes with the SEC from time to time (which are available at http://www.sec.gov), the events and circumstances discussed in such forward-looking statements may not occur, and the Company's actual results could differ materially and adversely from those anticipated or implied thereby. Although the Company's forward-looking statements reflect the good faith judgment of its management, these statements are based only on facts and factors currently known by the Company. As a result, you are cautioned not to rely on these forward-looking statements. 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.

Attachments

36-2022 Interim Report First Half 2022