

New incentive program for the Board of Directors of Orphazyme

Copenhagen, Denmark, September 21, 2020 – Orphazyme A/S (ORPHA.CO) (the “Company”), a late-stage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases, today announced that the Company introduces a new Board Incentive Program for the Company’s members of the Board of Directors, as approved at the Company’s Extraordinary General Meeting held on September 21, 2020.

The purpose of the Board Incentive Program is to attract and retain qualified members of the Board of Directors as share-based incentives are commonly used among competing international biotech and pharmaceutical companies. Further, the Board Incentive Program reflects the objective of a motivated and lasting value creation for the shareholders.

The Board Incentive Program is based on restricted share units (“RSUs”) which entitle the participant, subject to vesting of the RSUs, to be allocated a number of shares equivalent to the number of RSUs vested at maximum price per RSU equal to the par value of the Company’s shares.

The RSUs will have a vesting period from the date of grant and until approval of the annual report at the annual general meeting in the Company in the following year. Vesting are not conditional on any financial or performance criteria, but is, however, conditional upon (i) the continued membership by the participant on the Company’s Board of Directors until vesting and (ii) the initiation or completion of an offering and listing of American Depositary Shares in the United States. In addition, exercise of the RSUs are contingent on the participant not exercising his or her RSUs granted in March 2020 and approved at the Annual General Meeting on March 26, 2020, which will subsequently lapse and no longer be exercisable. Upon vesting, the RSUs may be exercised within a period of twelve months.

Based on the current number of participants in the Board Incentive Program, the program will comprise 22,993 shares in total, including an on-boarding grant to a new board member in accordance with the Company’s Remuneration Policy. The theoretical fair value of each RSU has been estimated at DKK 81.70 calculated on the basis of a Black-Scholes model.

For additional information, please contact

Orphazyme A/S

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About Orphazyme A/S

Orphazyme is a late-stage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases. The company is harnessing amplification of Heat-Shock Proteins (or HSPs) in order to develop and commercialize novel therapeutics for diseases caused by protein misfolding, protein aggregation, and lysosomal dysfunction, including lysosomal storage diseases and neuromuscular degenerative diseases. Arimoclomol, the company’s lead candidate, is in clinical development for four orphan diseases: Niemann-Pick disease Type C (NPC), Amyotrophic Lateral Sclerosis (ALS), sporadic Inclusion Body Myositis (sIBM) and Gaucher disease. Orphazyme is headquartered in Denmark and has operations in the U.S. and Switzerland. Orphazyme’s shares are listed on Nasdaq Copenhagen (ORPHA.CO).

Forward-looking statements

This company announcement may contain certain forward-looking statements, including with respect to the terms, timing and completion of the proposed offering. 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. 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,” “could”, 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. 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.