

Company announcement

No. 11/2021

Orphazyme A/S Ole Maaløes Vej 3 DK-2200 Copenhagen N

www.orphazyme.com Company Registration No. 32266355

Reporting of transactions in Orphazyme's shares made by persons discharging managerial responsibilities

Copenhagen, Denmark, March 25, 2021 – Pursuant to the Market Abuse Regulation article 19, Orphazyme A/S, CVR no. 32266355 ("Orphazyme"), hereby notifies receipt of information of the following transactions in Orphazyme's shares by persons discharging managerial responsibilities in Orphazyme:

1	Details of the Reporting Person / Closely A	Associated Person	
a)	Name	Catherine Moukheibir	
2	Reason for the notification		
a)	Position/status	Member of the Board of Directors	
b)	Initial notification/Amendment	Initial notification	
3	Details of the Company, emission allowand	ce market participant, auction platform, auctioneer	
	or auction monitor		
a)	Name	Orphazyme A/S	
b)	LEI code	549300250ZD2GGSQ7L42	
4	Details of the transaction(s): section to be	repeated for (i) each type of instrument; (ii) each	
	type of transaction; (iii) each date; and (iv) each place where transactions have been		
	conducted		
a)	Description of the financial instrument, type of	Shares	
	instrument and identification code	ISIN DK0060910917	
b)	Nature of the transaction	Exercise of Restricted Share Units	
c)	Price(s) and volume(s)	Price(s) Volume(s)	
٥,		DKK 61.1895 1,927	
d)	Aggregated information	Aggregated volume: 1,927 shares	
		Aggregated price: DKK 117,912.17	
		Price per share (volume weighted average): DKK 61.1895	
e)	Date of the transaction	March 24, 2021	
f)	Place of the transaction	Outside a trading venue	



1	Details of the Reporting Person / Closely A	Associated Person
a)	Name	Martijn Kleijwegt
2	Reason for the notification	
a)	Position/status	Member of the Board of Directors
b)	Initial notification/Amendment	Initial notification
3	Details of the Company, emission allowand or auction monitor	ce market participant, auction platform, auctioneer
a)	Name	Orphazyme A/S
b)	LEI code	549300250ZD2GGSQ7L42
4		repeated for (i) each type of instrument; (ii) each r) each place where transactions have been
a)	Description of the financial instrument, type of instrument and identification code	Shares ISIN DK0060910917
b)	Nature of the transaction	Exercise of Restricted Share Units
c)	Price(s) and volume(s)	Price(s) Volume(s) DKK 61.1895 1,927
d)	Aggregated information	Aggregated volume: 1,927 shares Aggregated price: DKK 117,912.17 Price per share (volume weighted average): DKK 61.1895
e)	Date of the transaction	March 24, 2021
f)	Place of the transaction	Outside a trading venue
4		repeated for (i) each type of instrument; (ii) each) each place where transactions have been
a)	Description of the financial instrument, type of	Shares
	instrument and identification code	
	instrument and identification code	ISIN DK0060910917
b)	instrument and identification code Nature of the transaction	ISIN DK0060910917 Sale of shares to LSP V Cöoperatieve U.A.
b)		
c)	Nature of the transaction	Sale of shares to LSP V Cöoperatieve U.A. Price(s) Volume(s)
c)	Nature of the transaction Price(s) and volume(s)	Sale of shares to LSP V Cöoperatieve U.A. Price(s) Volume(s) EUR - 1,927 Aggregated volume: 1,927 shares Aggregated price: EUR 1



1	Details of the Reporting Person / Closely A	Associated Person
a)	Name	LSP V Cöoperatieve U.A.
2	Reason for the notification	
a)	Position/status	Closely Associated Person to Martijn Kleijwegt, member of the Board of Directors
b)	Initial notification/Amendment	Initial notification
3	Details of the Company, emission allowand or auction monitor	ce market participant, auction platform, auctioneer
a)	Name	Orphazyme A/S
	LEI code	54930025OZD2GGSQ7L42
D)		
4		repeated for (i) each type of instrument; (ii) each
	type of transaction; (iii) each date; and (iv	v) each place where transactions have been
4	type of transaction; (iii) each date; and (iv	v) each place where transactions have been
4	type of transaction; (iii) each date; and (iveconducted Description of the financial instrument, type of instrument and identification code	Shares
4 a)	type of transaction; (iii) each date; and (iveconducted Description of the financial instrument, type of instrument and identification code Nature of the transaction	Shares ISIN DK0060910917
4 a) b) c)	type of transaction; (iii) each date; and (iveconducted Description of the financial instrument, type of instrument and identification code Nature of the transaction	Shares ISIN DK0060910917 Purchase of shares from Martijn Kleijwegt Price(s) Volume(s)
4 a) b) c)	type of transaction; (iii) each date; and (iveconducted Description of the financial instrument, type of instrument and identification code Nature of the transaction Price(s) and volume(s)	Shares ISIN DK0060910917 Purchase of shares from Martijn Kleijwegt Price(s) Volume(s) EUR - 1,927 Aggregated volume: 1,927 shares Aggregated price: EUR 1

For additional information, please contact

Orphazyme A/S

Anders Vadsholt, Interim CEO & CFO

+45 28 98 90 55

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. Arimoclomol, the company's lead candidate, is in clinical development for four orphan diseases: Niemann-Pick disease type C (NPC), Amyotrophic Lateral Sclerosis (ALS), Inclusion Body Myositis (IBM) and Gaucher disease. Orphazyme is headquartered in Denmark and has operations in the U.S. and Switzerland. Orphazyme's shares are listed on Nasdaq U.S. (ORPH) and Nasdaq Copenhagen (ORPHA).

About arimoclomol

Arimoclomol is an investigational drug candidate that amplifies the production of Heat Shock Proteins (HSPs). HSPs can rescue defective misfolded proteins, clear protein aggregates, and improve the function of lysosomes. Arimoclomol is administered orally and has now been studied in seven phase 1, four phase 2 and one pivotal phase 2/3 trial. Arimoclomol is in clinical development for NPC, Gaucher Disease, IBM, and ALS. Arimoclomol has received orphan drug designation (ODD) for NPC, IBM, and ALS in the US and EU. Arimoclomol has received fast-track designation (FTD) from the U.S. Food and Drug Administration (FDA) for NPC, IBM and ALS. In addition, arimoclomol has received breakthrough therapy designation (BTD) and rare-pediatric disease designation (RPDD) from the FDA for NPC.



Forward-looking statement

This company announcement may contain certain forward-looking statements, including in respect of the anticipated commercialization of arimoclomol. 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, including the risk that applicable regulatory authorities fail to approve arimoclomol on the anticipated timeline or at all. 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.