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PRICING STATEMENT RELATED TO THE OFFERING OF SHARES IN ORPHAZYME A/S

This document constitutes a pricing statement relating to the Offering of Offer Shares as described in the offering circular (the “Offering Circular”) published by Orphazyme A/S (“Orphazyme”) on 6 November 2017 and should be read in conjunction with the Offering Circular. Terms used and not defined herein have the meaning ascribed to them in the Offering Circular.

The Offer Period closed on 15 November 2017 at 00:01 a.m. (CET) for retail investors in Denmark for individual orders up to and including DKK 3 million each and at 11 a.m. (CET) for institutional investors, as announced through Nasdaq Copenhagen A/S (“Nasdaq Copenhagen”) in Orphazyme’s company announcement no. 3/2017 dated 14 November 2017.

The final Offer Price and other Offering statistics are set forth below:

1. OFFERING STATISTICS

Offer Price per Offer Share	DKK 80
Total number of Offer Shares offered (assuming the Overallotment Option is not exercised)	7,500,000
Total number of Overallotment Shares ⁽¹⁾	1,125,000
Total number of Offer Shares offered (assuming the Overallotment Option is exercised in full)	8,625,000
Market capitalisation of Orphazyme at the Offer Price (excluding the Overallotment Option) ⁽²⁾	DKK 1,594 million
Approximate aggregate gross proceeds receivable by the Company (assuming the Overallotment Option is not exercised)	DKK 600 million
Approximate aggregate gross proceeds receivable by the Company (assuming the Overallotment Option is exercised in full)	DKK 690 million
Percentage of outstanding share capital owned by Orphazyme’s existing shareholders following the Offering (assuming the Overallotment Option is not exercised)	64.5%
Percentage of outstanding share capital owned by Orphazyme’s existing shareholders following the Offering (assuming the Overallotment Option is exercised in full)	61.0%

(1) Additional new Shares available for subscription by the Joint Global Coordinators, on behalf of the Managers, at the Offer Price pursuant to the Overallotment Option granted by Orphazyme.

(2) Including a capital increase of nominally DKK 838,092 due to the exercise of Pre-IPO Warrants (as further described below). Assuming the Overallotment Option is exercised in full, the market capitalization of Orphazyme at the Offer Price would DKK 1,684 million.

2. SHARE CAPITAL INFORMATION

Orphazyme's share capital immediately following completion of the Offering (excluding the Overallotment Option) will be as follows:

	<u>No. of Shares</u>	<u>Nominal Value ⁽¹⁾</u>
Issued share capital ⁽²⁾	19,928,184	DKK 19,928,184

(1) Each Share of Orphazyme has a nominal value of DKK 1.

(2) Including a capital increase of nominally DKK 838,092 due to the exercise of Pre-IPO Warrants (as further described below).

The first day of trading and official listing on Nasdaq Copenhagen is expected to be today 16 November 2017 subject to the Offering not being withdrawn prior to settlement and completion of the Offering. The Offer Shares are delivered in the form of Temporary Purchase Certificates under the temporary ISIN (DK0060911055). On 22 November 2017, the Temporary Purchase Certificates will automatically be exchanged in VP SECURITIES A/S for a corresponding number of Shares in Orphazyme delivered under the permanent ISIN (DK0060910917) end of day. In connection with the Temporary Purchase Certificates being automatically exchanged for Shares, the Temporary Purchase Certificates will cease to exist. All existing shareholders have undertaken an obligation not to trade in the existing Shares until the Temporary Purchase Certificates have been exchanged for Shares and the Shares have been admitted to trading on Nasdaq Copenhagen.

The first day of trading of the Temporary Purchase Certificates on Nasdaq Copenhagen under the symbol "ORPHA TEMP" is expected to be today 16 November 2017 under the temporary ISIN (DK0060911055), and the last day of trading of the Temporary Purchase Certificates on Nasdaq Copenhagen is expected to be 20 November 2017. The Shares are expected to be admitted to trading and official listing on Nasdaq Copenhagen under the permanent ISIN (DK0060910917) and under the symbol "ORPHA" on 21 November 2017. Consequently, after the trading has closed on Nasdaq Copenhagen on 20 November 2017, all future trading in Orphazyme's shares on Nasdaq Copenhagen will continue and settle under the permanent ISIN (DK0060910917) and under the symbol "ORPHA".

Payment for and settlement of the Offer Shares are expected to take place on 20 November 2017 by way of delivery of Temporary Purchase Certificates under the temporary ISIN (DK0060911055) against payment in immediately available funds in Danish kroner in book-entry form to investors' accounts with VP SECURITIES A/S and through the facilities of Euroclear Bank S.A./N.A. and Clearstream Banking S.A.

Upon completion of the Offering and after payment for the Temporary Purchase Certificates representing the Offer Shares, the capital increase relating to the new Shares to be issued by the Company pursuant to the Offering will be registered with the Danish Business Authority, which is expected to take place on 20 November 2017.

If the Offering is terminated or withdrawn, the Offering and any associated arrangements will lapse, all submitted orders will be automatically cancelled, any monies received in respect of the Offering will be returned to the investors without interest (less any transaction costs) and admission to trading and/or official listing of the Temporary Purchase Certificates or the Shares on Nasdaq Copenhagen will be cancelled. Consequently, any trades in the Temporary Purchase Certificates and/or Shares effected on or off the market before settlement of the Offering may subject investors to liability for not being able to deliver the Temporary Purchase Certificates and/or Shares sold, and

investors who have sold or acquired Temporary Purchase Certificates and/or Shares on or off the market may incur a loss. All dealings in the Temporary Purchase Certificates and/or Offer Shares prior to settlement of the Offering are for the account of, and at the sole risk of, the parties concerned.

3. 2017 CAPITAL STRUCTURE ADJUSTMENT

As described in the Offering Circular, the capital structure of Orphazyme was in preparation of the Offering adjusted by way of a merger of the three previous share classes into one combined with an issue of bonus Shares in order to account for the now abolished preference shares (the “2017 Capital Structure Adjustment”). In connection with the 2017 Capital Structure Adjustment, the class B and C preference shares of Orphazyme were converted into Shares on a 1:1 ratio. In order to account for the preferential rights attached to the preference shares, a directed issue of bonus Shares using free reserves of Orphazyme was carried out at par value in favour of the preference shareholders, assuming an Offer Price in the top end of the Offer Price Range.

4. EXERCISE OF PRE-IPO WARRANTS

As described in the Offering Circular, a number of Pre-IPO Warrants could be exercised (or, if not exercised, lapse without compensation) in connection with the Offering. A total of 838,092 Pre-IPO Warrants have been exercised and, accordingly, Orphazyme’s nominal share capital will be increased by DKK 838,092. Such capital increase is expected to be registered with the Danish Business Authority on or around 20 November 2017.

5. OWNERSHIP STRUCTURE

The table below sets forth the ownership percentages in Orphazyme as at the date hereof; and immediately following the completion of the Offering, assuming (a) the Overallotment Option is exercised in full and (b) the Overallotment Option is not exercised at all.

The information in the table below includes indirect holdings through legal entities (e.g. private holding companies) to the extent relevant. In case the shareholding percentages do not sum to 100%, this is due to rounding.

Shareholders	Shares held as at the date hereof		Shares held after completion of the Offering assuming the Overallotment Option is exercised in full		Shares held after completion of the Offering assuming the Overallotment Option is not exercised	
	Number of Shares	%	Number of Shares	%	Number of Shares	%
Novo Holdings ⁽¹⁾	3,830,832	33.1%	3,830,832	18.2%	3,830,832	19.2%
Aescap Venture	1,765,605	15.2%	1,765,605	8.4%	1,765,605	8.9%
Sunstone Capital	1,804,405	15.6%	1,804,405	8.6%	1,804,405	9.1%
Orpha Pooling B.V. ⁽²⁾ (LSP and ALS Invest)	2,431,672	21.0%	2,710,829	12.9%	2,710,829	13.6%
Idinvest	833,362	7.2%	833,362	4.0%	833,362	4.2%
Kurma Biofund II	765,947	6.6%	905,525	4.3%	905,525	4.5%
Kansas Life Sciences Development Inc.	16,691	0.1%	16,691	0.1%	16,691	0.1%
UCL Business PLC	16,578	0.1%	16,578	0.1%	16,578	0.1%
Founders ⁽³⁾	125,000	1.1%	125,000	0.6%	125,000	0.6%
Warrant holders ⁽⁴⁾	-	-	838,092	4.0%	838,092	4.2%
New shareholders	-	-	8,206,265	39.0%	7,081,265	35.5%
Total	11,590,092	100%	21,053,184	100%	19,928,184	100%

Existing Board of Directors

Georges Gemayel	-	-	87,758	0.4%	87,758	0.4%
Bo Jesper Hansen	-	-	79,445	0.4%	79,445	0.4%
Martijn Kleijwegt	-	-	-	-	-	-
Martin Bonde	-	-	46,009	0.2%	46,009	0.2%
Martin Rahbek Kornum	18,750	0.2%	41,476	0.2%	41,476	0.2%
Nanna Lüneborg	-	-	-	-	-	-
Patrick J.H. Krol	-	-	-	-	-	-
Rémi Droller	-	-	-	-	-	-
Sten Verland	-	-	-	-	-	-

New Board Members

Anders Hedegaard	-	-	6,250	0.0%	6,250	0.0%
Catherine Moukheibir	-	-	7,980	0.0%	7,980	0.0%

Executive Management

Anders Hinsby	18,750	0.2%	204,596	1.0%	204,596	1.0%
Anders Vadsholt	-	-	127,806	0.6%	127,806	0.6%

Key Employees

Thomas Blaettler	-	-	110,377	0.5%	110,377	0.6%
Thomas Kirkegaard Jensen	68,750	0.6%	229,285	1.1%	229,285	1.2%

(1) As described in the Offering Circular, Novo Holdings has pursuant to a stock lending agreement made 1,125,000 Shares available to the Joint Global Coordinators for purposes of delivery of the Offer Shares to investors in connection with the Overallotment Option. Regardless of whether the Overallotment Option is exercised, Novo Holdings will have an equivalent number of Shares redelivered, either by way of existing Shares or new Shares issued by the Company. Accordingly, in the table above, Novo Holdings' holding of Shares does not reflect the lending of those 1,125,000 Shares pursuant to the Overallotment Option.

(2) Including 279,157 Shares of a nominal value of DKK 1 each subscribed for by LSP V Coöperatieve U.A. in connection with the Offering.

(3) Excluding any Pre-IPO Warrants exercised and/or Investment Shares subscribed for by the founders in connection with the Offering as part of Orphazyme's long-term incentive programme (as otherwise set out below with respect to Anders Hinsby, Martin Rahbek Kornum and Thomas Kirkegaard Jensen).

(4) A capital increase of nominally DKK 838,092 is expected to be carried out on 20 November 2017 due to the exercise of Pre-IPO Warrants (as further described above).