

Orphazyme A/S
Attn.: CFO Anders Vadsholt
Ole Maaløes Vej 3
DK-2200 Copenhagen N
Denmark

11 February 2020

MAJOR SHAREHOLDER ANNOUNCEMENT REGARDING ORPHAZYME A/S

This major shareholder announcement regarding Orphazyme A/S, Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark, registration (CVR) number 32266355, (the “**Company**”) is made pursuant to Section 38 of the Danish Capital Markets Act.

LSP V Coöperatieve U.A., a cooperative with excluded liability and official seat in Amsterdam, the Netherlands, with registered office address at Johannes Vermeer, Plein 9, 1071 DV Amsterdam, the Netherlands, and registered with the Dutch trade register under number 61888575 (“**LSP**”), hereby notifies the Company that:

On 11 February 2020, LSP has increased its indirect shareholding in the Company as a consequence of having been re-delivered 1,050,000 shares of a nominal value of DKK 1 lent to the Company through Danske Bank A/S for purposes of the Company's delivery of shares to investors in the Company's private placement offering. LSP has not participated in the private placement offering and has only facilitated the loan of the shares for purposes of the Company's offering of shares in the private placement offering.

Accordingly and following the dilution resulting from the Company's issuance of new shares in the private placement offering, LSP holds directly and indirectly 2,710,829 shares of a nominal value of DKK 1 each in the Company, corresponding to 10.03% of the total share capital and voting rights. LSP holds directly 279,157 shares of a nominal value of DKK 1 each in the Company, corresponding to 1.03% of the total share capital and voting rights, and LSP holds indirectly through Orpha Pooling B.V. 2,431,672 shares of a nominal value of DKK 1 each in the Company, corresponding to 9.00% of the total share capital and voting rights.

Orpha Pooling B.V. is a private limited liability company and has its official seat in Amsterdam, the Netherlands, with registered office address at Johannes Vermeer, Plein 9, 1071 DV Amsterdam, the Netherlands, and is registered with the Dutch trade register under number 67827055. The shareholders of Orpha Pooling B.V. are (i) LSP (88.24% of the total share capital and voting rights), and (ii) ALS Invest 2 B.V. (11.76% of the total share capital and voting rights), a private limited liability company, having its official seat in Amsterdam, the Netherlands, and its registered office address at Eerste Weteringdwaarsstraat 54 E, 1017TP Amsterdam, the Netherlands, registered with the Dutch trade register under number 67804187.

LSP is a closed end investment fund managed by LSP V Management B.V. LSP V Management B.V. is a private limited liability company and has its official seat in Amsterdam, the Netherlands, with registered office address at Johannes Vermeer, Plein 9, 1071 DV Amsterdam, the Netherlands, and is registered with the Dutch trade register under number 60800542. LSP V Management B.V. is a subsidiary of LSP Management Group B.V. LSP Management Group B.V. is a limited liability company has its official seat in Amsterdam, the Netherlands, with registered office address at Johannes Vermeer, Plein 9, 1071 DV Amsterdam, the Netherlands, and is registered with the Dutch trade register under number 34288020.

This major shareholder notification is made by LSP V Coöperatieve U.A. as the controlling shareholder of Orpha Pooling B.V.

Best regards,

LSP V Coöperatieve U.A.