

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

Amsterdam, 1 October 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 1 October 2020, LSP has reduced its direct and indirect shareholding in the Company as a consequence of the dilution resulting from the issue of 7,616,146 new ordinary shares in Orphazyme of a nominal value of DKK 1 each in connection with the initial public offering of American Depositary Shares (“ADSs”) in the U.S. and a concurrent private placement of ordinary shares in Europe. LSP has not participated in the initial public offering in the U.S. or the private placement in Europe.

Accordingly, LSP holds directly and indirectly 2,710,829 shares of a nominal value of DKK 1 each in the Company, corresponding to 7.81% 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 0.80% 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 7.01% 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 Weteringdwarsstraat 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.