

Company announcement

No. 10/2017

Orphazyme A/SOle Maaløes Vej 3
DK-2200 Copenhagen Nwww.orphazyme.com
Company Registration No. 32266355

16 November 2017

Major shareholder announcement

With reference to company announcement no. 5/2017 dated 16 November 2017 regarding the result of the Initial Public Offering (the "Offering") of Orphazyme A/S ("Orphazyme"), Orphazyme hereby announces that it has received the following notifications pursuant to section 29 of the Danish Securities Trading Act regarding the existing major shareholders' direct and indirect respective holdings of shares in Orphazyme.

Prior to the completion of the Offering, the major shareholders of Orphazyme were the Novo Nordisk Foundation through Novo Holdings A/S (33.1%); Aescap Venture Management B.V. through Coöperative Aescap Venture I U.A. (15.2%); Sunstone Life Science Ventures A/S through Sunstone LSV General Partner II ApS and Sunstone Life Science Ventures Fund II K/S (15.6%); LSP Management Group B.V. through LSP V Management B.V., LSP V Coöperatieve U.A. and Orpha Pooling B.V. (21.0%); Idinvest Partners through FCPI Idinvest Patrimoine n°3, FCPI Idinvest Patrimoine n°4, FCPI Objectif Innovation Patrimoine n°6 and FCPI Objectif Innovation Patrimoine n°7 (7.2%); and Kurma Partners through Kurma Biofund II (6.6%).

Novo Holdings A/S has reduced its shareholding in Orphazyme as a consequence of (i) the dilution resulting from the issue of 7,500,000 new shares in Orphazyme of a nominal value of DKK 1 each in connection with the Offering; and (ii) the lending of 1,125,000 shares in Orphazyme of a nominal value of DKK 1 each by Novo Holdings A/S to Joint Global Coordinators for the purposes of delivery of shares to investors as a result of the overallotment option in connection with the Offering (the "Overallotment Option"). Regardless of whether the Overallotment Option is exercised, Novo Holdings A/S will have an equivalent number of shares redelivered, either by way of existing shares or new shares issued by Orphazyme.

Aescap Venture Management B.V., Sunstone LSV General Partner II ApS, LSP Management Group B.V., Idinvest Partners and Kurma Partners have reduced their respective indirect shareholdings in Orphazyme as a consequence of the dilution resulting from the issue of 7,500,000 new shares in Orphazyme of a nominal value of DKK 1 each in connection with the Offering.

Moreover, Kurma Biofund II and LSP V Coöperatieve U.A. have increased their respective direct shareholdings in Orphazyme by having subscribed for an additional number of shares in Orphazyme in connection with the Offering.

Subsequent to the Offering, Orphazyme has received the following major shareholder announcements:

Novo Holdings A/S

Following completion and settlement of the above transactions in connection with the Offering, Novo Holdings A/S' holding of shares in Orphazyme will be a total of 2,705,832 shares (i.e. excluding lending shares pursuant to the Overallotment Option) of a nominal value of DKK 1 each, corresponding to 13.6% of the total share capital and voting rights.

Novo Holdings A/S is a limited liability company organized under the laws of Denmark under CVR no. 24257630 with its registered office at Tuborg Havnevej 19, 2900 Hellerup, Denmark. Novo Holdings A/S is wholly-owned by the Novo Nordisk Foundation.

The Novo Nordisk Foundation is a foundation organised under the laws of Denmark under CVR no. 10582989 with its registered office at Tuborg Havnevej 19, DK-2900 Hellerup, Denmark.

Aescap Venture

Following completion and settlement of the Offering, Aescap Venture Management B.V.'s indirect holding of shares in Orphazyme through Coöperative Aescap Venture I U.A. will be a total of 1,765,605 shares of a nominal value of DKK 1 each, corresponding to 8.9% of the total share capital and voting rights.

Coöperative Aescap Venture I U.A. is a limited liability cooperative association organised under the laws of the Netherlands, registered under registration number 34257886 with its registered address at Science Park 406, 1098 XH Amsterdam, the Netherlands. Aescap Venture I U.A. is managed by Aescap Venture Management B.V.

Aescap Venture Management B.V. is a limited liability company organised under the laws of the Netherlands, registered under registration number 32108991 with its registered address at Science Park 406, 1098 XH Amsterdam, the Netherlands.

Sunstone Life Science Ventures

Following completion and settlement of the Offering, Sunstone Life Science Ventures A/S' indirect holding of shares in Orphazyme through Sunstone LSV General Partner II ApS and Sunstone Life Science Ventures Fund II K/S will be a total of 1,804,405 shares of a nominal value of DKK 1 each, corresponding to 9.1% of the total share capital and voting rights.

Sunstone Life Science Ventures Fund II K/S is organised under the laws of Denmark under CVR no. 30582268 with its registered address at Lautrupsgade 7, 5., 2100 Copenhagen Ø, Denmark. Sunstone Life Science Venture Fund II K/S is managed by Sunstone LSV General Partner II ApS.

Sunstone LSV General Partner II ApS is organised under the laws of Denmark under CVR no. 30575245 with its registered address at Lautrupsgade 7, 5., 2100 Copenhagen Ø, Denmark. Sun-stone LSV General Partner II ApS is a subsidiary of Sunstone Life Science Ventures A/S.

Sunstone Life Science Ventures A/S is organised under the laws of Denmark under CVR no. 33859198 with its registered address at Lautrupsgade 7, 5., 2100 Copenhagen Ø, Denmark.

LSP

Following completion and settlement of the Offering, (i) LSP V Coöperatieve U.A.'s direct holding of 279,157 shares of a nominal value of DKK 1 each in Orphazyme will correspond to 1.4% of the total share capital and voting rights; and (ii) Orpha Pooling B.V.'s direct holding of 2,431,672 shares of a nominal value of DKK 1 each in Orphazyme will correspond to 12.2% 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 V Coöperatieve U.A. (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 V Coöperatieve U.A. is a cooperative with excluded liability 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 61888575. LSP V Coöperatieve U.A. 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.

Idinvest Partners

Following completion and settlement of the Offering, Idinvest Partners' indirect holding of shares in Orphazyme through the FCPI Idinvest Patrimoine n°3, FCPI Idinvest Patrimoine n°4, FCPI Objectif Innovation Patrimoine n°6 and FCPI Objectif Innovation Patrimoine n°7 will be below 5% of the total share capital and voting rights. Consequently, Idinvest Partners will no longer be a major shareholder of Orphazyme pursuant to section 29 of the Danish Securities Trading Act.

Idinvest Partners manages its investment in Orphazyme through FCPI Idinvest Patrimoine n°3, FCPI Idinvest Patrimoine n°4, FCPI Objectif Innovation Patrimoine n°6 and FCPI Objectif Innovation Patrimoine n°7, which are French *Fonds Communs de Placement dans l'Innovation*, governed by the laws of France, having their registered office at 117 Avenue des Champs-Élysées, 75 008 Paris, France, registered with the Registry of Trade and Companies of Paris under number 414 735 175.

Idinvest Partners is a French *société anonyme* governed by the laws of France, having its registered office at 117 Avenue des Champs-Élysées, 75 008 Paris, France, registered with the Registry of Trade and Companies of Paris under number 414 735 175.

Kurma Partners

Following completion and settlement of the Offering, Kurma Partners' indirect holding of shares in Orphazyme through Kurma Biofund II will be below 5% of the total share capital and voting rights. Consequently, Kurma Partners and Kurma Biofund II will no longer be a major shareholder of Orphazyme pursuant to section 29 of the Danish Securities Trading Act.

Kurma Biofund II, a professional private equity fund represented by its management company Kurma Partners, a French *société anonyme* (corporation), with its registered office at 24 rue Royale – 75008 Paris, registered with the Paris Trade and Companies Registry under number 510 043 136.

For additional information, please contact

Orphazyme

Anders Vadsholt, CFO

+45 28 98 90 55

About Orphazyme

Orphazyme is a Danish biotech company with a late stage orphan drug pipeline, developing new treatment options for orphan protein misfolding diseases. The Company was founded in 2009 based on early scientific discovery in heat shock proteins ("HSPs"). Since inception, the Company has translated scientific discovery into a late stage clinical development programme. The Company is headquartered in Copenhagen and currently has 30 employees.

The Company focuses on severe and mostly fatal diseases with a high unmet need, and with a particularly strong commitment to neuromuscular diseases and a group of severe genetic diseases called lysosomal storage diseases. The Company plans to pursue development of its lead candidate through to registration in the EU and the United States after which launch and commercialisation is expected to be undertaken by the Company.

The lead candidate arimoclomol is in development as a potential treatment for four orphan diseases; two neuromuscular diseases, sporadic Inclusion Body Myositis ("sIBM") and Amyotrophic Lateral Sclerosis ("ALS"), and two lysosomal storage diseases, Niemann Pick type C ("NPC") and Gaucher disease.