

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.

Novo Holdings A/S, registration (CVR) number 24257630, Tuborg Havnevej 19, DK-2900 ("**Novo**"), hereby notifies the Company that:

On 11 February 2020, Novo has increased its shareholding in the Company as a consequence of having been redelivered 2,021,673 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. Novo 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, Novo holds 2,021,673 shares of a nominal value of DKK 1 each in the Company, corresponding to 7.5% of the total share capital and voting rights.

This major shareholder notification is made on behalf of Novo Nordisk Fonden, registration (CVR) number 10582989, Tuborg Havnevej 19, DK-2900. Novo Nordisk Fonden holds 100% of the shares in Novo and holds accordingly indirectly 2,021,673 shares of a nominal value of DKK 1 each in the Company, corresponding to 7.5% of the total share capital and voting rights.

Best regards,

Novo Holdings A/S