

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

6 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 6 February 2020, Novo has decreased its shareholding in the Company due to the lending of 2,021,673 shares of a nominal value of DKK 1 each to the Company through Danske Bank A/S for purposes of the Company’s delivery of shares to investors in a private placement offering. Novo does not participate in the private placement offering and is only facilitating the loan of the shares for purposes of the Company’s offering of shares in the private placement offering.

Accordingly and following the stock loan, Novo holds no shares in the Company, corresponding to 0% 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 no shares in the Company, corresponding to 0% of the total share capital and voting rights.

Best regards,

Novo Holdings A/S