

Company announcement No. 06/2023 Orphazyme A/S Ole Maaløes Vej 3 DK-2200 Copenhagen N

www.orphazyme.com Company Registration No. 32266355

Major shareholder announcement

Copenhagen, Denmark, May 12, 2023 – Orphazyme A/S (ORPHA.CO) (the "Company"), hereby announces the receipt of notification pursuant to Section 38 of the Danish Capital Markets Act from Scandinavian Investment Group A/S that Scandinavian Investment Group A/S holds shares, corresponding to more than 5% of the total share capital and voting rights in the Company.

The Company has today become aware that each of Scandinavian Investment Group A/S, MH Investment ApS and Nordic Compound Invest A/S hold slightly below 10% of the total share capital and voting rights in the Company, see also company announcements no. 4 and 5 of 9 May 2023.

For additional information, please contact

Orphazyme A/S

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