

ORPHAZYME RAISES EUR 14 MILLION TO FURTHER EXPAND ITS CLINICAL DEVELOPMENT PROGRAMME

Orphazyme ApS today announced the successful completion of a EUR 14M financing round. Two new investors, LSP and the ALS Investment Fund joins the investor syndicate comprising Kurma Partners, Idivest Partners, Novo A/S, Sunstone Capital and Aescap Venture.

CEO Anders Hinsby said: "We are very pleased to welcome LSP and the ALS Investment Fund to our group of investors. The additional funding gives us the opportunity to develop our lead programme, arimoclomol, in additional indications, such as Gaucher disease. We see a great potential for arimoclomol, highlighted by the recently reported successful Phase 1/2 trials in two protein misfolding diseases: SOD1-ALS and sporadic Inclusion Body Myositis. Our focus is now to conduct robust trials to move arimoclomol expediently towards registration in these debilitating diseases"

Orphazyme is developing therapeutics for lysosomal storage- and protein misfolding diseases. Arimoclomol is in Phase 3 development for the treatment of Niemann-Pick disease type C. The current financing will support clinical development in two additional protein misfolding diseases: Gaucher disease, a lysosomal storage disease, and sporadic Inclusion Body Myositis, a currently untreatable progressive muscle wasting disease in people over the age of 50. During the first half of 2017 the company will also decide on progressing the programme in familial Amyotrophic Lateral Sclerosis.

LSP Managing Partner Martijn Kleijwegt was, following the transaction, elected to the Board of Directors.

Martijn Kleijwegt said: "LSP shares Orphazyme's passion for tackling orphan diseases with a high unmet need. I look forward to joining the Board of Directors at a time when the company is pushing the boundaries and bringing its technology to a wider range of diseases that have devastating consequences for the sufferers and their loved ones."

About Orphazyme ApS

Orphazyme ApS is a Danish biotech company that develops paradigm-changing medicines for the treatment of protein misfolding diseases with well-characterised mechanisms of disease. The lead programme is in development as a treatment for the lysosomal storage disease Niemann-Pick disease type C. This is one of a family ~50 debilitating genetic disorders that often affect children, most of whom are currently untreatable. For more information, please visit www.orphazyme.com.

About LSP

LSP is an independent European investment firm, providing financing for private and public life sciences companies. LSP's mission is to connect investors to inventors, focusing on unmet medical needs. Since the late 1980s, LSP's management has invested in about 100 innovative enterprises, many of which have grown to become leaders of the global life sciences industry. With over €1 billion of investment capital raised to date and offices in Amsterdam, Munich and Boston, LSP is one of Europe's leading life sciences investors. The LSP Health Economics Fund invests in innovative products that can increase the quality of health care, while reducing the cost of care. For more information, please visit: www.lspvc.com.

About ALS Investment Fund

The ALS Investment Fund supports and finances biotech companies that develop drugs and diagnostics for Amyotrophic Lateral Sclerosis (ALS), also known as Lou Gehrig's disease). The Fund was initiated by 3 Dutch ALS patients and is supported by a top-tier European VC firm, Life Sciences Partners. The ALS Investment Fund invests globally, but focuses on Europe and the US. The fund brings depth of disease-specific knowledge and a deep network in the space. More information is available at www.alsinvestmentfund.com.