Offering of up to 10,781,250 shares

(a public limited liability company incorporated in Denmark under registration (CVR) no. 32266355)

This document relates to the initial public offering of up to 10,781,250 new shares ("**Offer Shares**") of DKK 1 nominal value each (and together with the Overallotment Shares (as defined below) the "**Offering**") of Orphazyme A/S (the "**Company**" or "**Orphazyme**"). The Company is offering such number of Offer Shares as will raise gross proceeds of up to approximately DKK 690 million (hereof DKK 90 million pursuant to the Overallotment Option (as defined herein)). Assuming completion of the Offering, the Company's registered share capital will increase by a nominal value of up to DKK 9,375,000 as a result of the issue of Offer Shares (excluding the Overallotment Option). The exact number of Offer Shares will be determined based on a book-building process. As used herein, "Shares" shall refer to all outstanding shares of the Company at any given time.

Skandinaviska Enskilda Banken, Danmark, branch of Skandinaviska Enskilda Banken AB (Publ.), Sweden; Vækstfonden; BI Asset Management Fondsmæglerselskab A/S on behalf of certain clients; Handelsbanken, branch of Svenska Handelsbanken AB (Publ.), Sweden; and Spar Nord Bank A/S have in connection with the Offering subject to certain conditions undertaken to subscribe for Offer Shares as "**Cornerstone Investors**" for a total subscription amount of DKK 230 million, corresponding to approximately 38.3% of the Offering (excluding the Overallotment Option). The final allocation within the respective ranges of commitment undertaken by the Cornerstone Investors shall be at the sole discretion of Orphazyme.

The Offering consists of (i) an initial public offering to retail and institutional investors in Denmark (the "**Danish Offering**"); and (ii) private placements to institutional investors in certain other jurisdictions (excluding the United States) (the "**International Offering**"). The Offering outside the United States will be made in compliance with Regulation S under the U.S. Securities Act of 1933, as amended (the "**U.S. Securities Act**") ("**Regulation S**").

The Company has granted the Joint Global Coordinators, on behalf of the Managers (as defined herein) an option (the "**Overallotment Option**") to subscribe for up to 1,406,250 additional new Shares in the aggregate at the Offer Price (the "**Overallotment Shares**"), exercisable, in whole or in part, from the date of Admission (as defined herein) until 30 calendar days thereafter, solely to cover overallotments or short positions, if any, incurred in connection with the Offering. If the Overallotment Option is exercised, the term "Offer Shares" shall also include the Overallotment Shares.

You are advised to examine all the risks and legal requirements described in this Offering Circular that might be relevant in connection with an investment in the Offer Shares. Investing in the Offer Shares involves a high degree of risk. See "*Risk Factors*" beginning on page 34 for a discussion of certain risks that prospective investors should consider before investing in the Offer Shares.

OFFER PRICE RANGE: DKK 64 - DKK 80 PER OFFER SHARE

The offer price at which the Offer Shares will be sold (the "**Offer Price**") is expected to be between DKK 64 and DKK 80 per share (the "**Offer Price Range**") and will be determined through a book-building process. The Offer Price will be determined by the Company in consultation with the Joint Global Coordinators, and is expected to be announced through Nasdaq Copenhagen A/S ("**Nasdaq Copenhagen**") no later than 8:00 a.m. (CET) on 17 November 2017.

The offer period (the "**Offer Period**") will commence on 6 November 2017 and will close no later than 16 November 2017 at 12:00 p.m. (noon) (CET). The Offer Period may be closed prior to 16 November 2017; however, the Offer Period will not be closed in whole or in part before 15 November 2017 at 00.01 a.m. (CET). The Offer Period in respect of applications for purchases of amounts up to, and including, DKK 3 million may be closed before the remainder of the Offering is closed. If the Offering is closed before 16 November 2017, the first day of trading may be moved forward accordingly. Any such early closing, in whole or in part, will be announced through Nasdaq Copenhagen.

Payment for and settlement of the Offer Shares are expected to take place on or around 21 November 2017 (the "**Settlement Date**") by way of delivery of temporary purchase certificates under the temporary ISIN DK0060911055 (the "**Temporary Purchase Certificates**") against payment in immediately available funds in Danish kroner in book-entry form to investors' accounts with VP SECURITIES A/S ("**VP Securities**") and through the facilities of Euroclear Bank S.A./N.A., as operator of the Euroclear System ("**Euroclear**") and Clearstream Banking, S.A. ("**Clearstream**"). Subject to completion of the Offering and registration of the new Offer Shares with the Danish Business Authority, the Temporary Purchase Certificates will automatically be exchanged in VP Securities for a corresponding number of Shares, which are expected to be delivered two business days after the Settlement Date under the permanent ISIN DK0060910917 in book-entry form to the holder of the Temporary Purchase Certificates' account with VP Securities and through the facilities of Euroclear and Clearstream. If the Offering is closed before 16 November 2017 (i.e. the Settlement Date), the delivery of Temporary Purchase Certificates, the automatic exchange of Temporary Purchase Certificates for Shares and the first day of trading and official listing of the Shares on Nasdaq Copenhagen may be moved forward accordingly. The Offering may be withdrawn after Admission and until settlement of the Offering. All dealings in the Temporary Purchase Certificates and/or the Offer Shares prior to settlement of the Offering will be for the account of, and at the sole risk of, the parties involved. Registration of the new Offer Shares issued by the Company with the Danish Business Authority will take place following completion of the Offering on the Settlement Date, which is expected to take place on 21 November 2017.

Prior to the Offering, there has been no public market for the Temporary Purchase Certificates or the Shares. Application has been made for the Temporary Purchase Certificates to be admitted to trading on Nasdaq Copenhagen (the "**Admission**") under the symbol "ORPHA TEMP" and for the Shares to be admitted to trading and official listing on Nasdaq Copenhagen under the symbol "ORPHA". The Admission is subject to, among other things, Nasdaq Copenhagen's approval of the distribution of the Offer Shares, the Offering not being withdrawn prior to the settlement of the Offering and the Company making an announcement to that effect. The first day of trading and official listing on Nasdaq Copenhagen is expected to be 17 November 2017 subject to the Offering not being withdrawn prior to settlement and completion of the Offering. The first day of trading of the Temporary Purchase Certificates on Nasdaq Copenhagen is expected to be 17 November 2017 and the last day of trading of the Temporary Purchase Certificates on Nasdaq Copenhagen is expected to be 21 November 2017. The first day of trading of the Shares on Nasdaq Copenhagen under the permanent ISIN is expected to be 22 November 2017. In connection with the Temporary Purchase Certificates being automatically exchanged for Shares, the Temporary Purchase Certificates will cease to exist.

This document has been prepared under Danish law in compliance with the requirements set out in the Danish Consolidated Act no. 251 of 21 March 2017 on securities trading (the "Danish Securities Trading Act"), the Executive Order no. 1257 of 6 November 2015 on prospectuses for securities admitted to trading in a regulated market and for offering to the public of securities of at least €5,000,000, as amended (the "Danish Executive Order on Prospectuses") and as supplemented by Regulation (EU) 2017/1129 of 14 June 2017, as well as Commission Regulation (EC) no. 809/2004 of 29 April 2004, as amended (the "Prospectus Regulation"). This document does not constitute an offer to sell or the solicitation of an offer to subscribe for or buy any of the Offer Shares in any jurisdiction to any person to whom it would be unlawful to make such an offer in such a jurisdiction.

The Offer Shares have not been and will not be registered under the U.S. Securities Act and are being offered and sold outside the United States in compliance with Regulation S. For certain restrictions on transfer of the Offer Shares, see "*Transfer Restrictions*". The distribution of this document and the offer of the Offer Shares in certain jurisdictions are restricted by law. Persons into whose possession this document comes are required by the Company, the Main Shareholders and the Managers to inform themselves about and to observe such restrictions. For a description of certain restrictions on offers of Offer Shares and on distribution of this document, see "*Selling Restrictions*".

Joint Global Coordinators and Joint Bookrunners

Carnegie **Danske Bank**

Co-Lead Manager

Oddo

The date of this Offering Circular is 6 November 2017

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Introduction

Notice to Investors in the United States

The Offer Shares have not been recommended by any U.S. federal or state securities commission or regulatory authority. Furthermore, the foregoing authorities have not confirmed the accuracy or determined the adequacy of this Offering Circular. Any representation to the contrary is a criminal offence in the United States.

The Offer Shares have not been and will not be registered under the U.S. Securities Act and will be offered outside the United States in compliance with Regulation S. For certain restrictions on transfer of the Offer Shares, see "*Transfer Restrictions*".

European Economic Area ("EEA") Restrictions

In any Member State of the EEA other than Denmark that has implemented the Prospectus Directive, this Offering Circular is only addressed to, and is only directed at, investors in that EEA Member State who fulfil the criteria for exemption from the obligation to publish a prospectus, including qualified investors, within the meaning of the Prospectus Directive as implemented in each such EEA Member State.

This Offering Circular has been prepared on the basis that all offers of Offer Shares, other than the offer contemplated in Denmark, will be made pursuant to an exemption under the Prospectus Directive, as implemented in Member States of the EEA, from the requirement to produce a prospectus for offers of Offer Shares. Accordingly, any person making or intending to make any offer within the EEA of Offer Shares which is the subject of the placement contemplated in this Offering Circular should only do so in circumstances in which no obligation arises for the Company, the Main Shareholders or any of the Managers to produce a prospectus for such offer. Neither the Company, the Main Shareholders nor the Managers have authorised, nor do the Company, the Main Shareholders or the Managers authorise, the making of any offer of Offer Shares through any financial intermediary, other than offers made by Managers which constitute the final placement of Offer Shares contemplated in this Offering Circular.

The Offer Shares have not been, and will not be, offered to the public in any Member State of the European Economic Area that has implemented the Prospectus Directive, excluding Denmark (a "**Relevant Member State**"). Notwithstanding the foregoing, an offering of the Offer Shares may be made under the following exemptions under the Prospectus Directive as implemented in that Relevant Member State:

- to any qualified investor as defined in the Prospectus Directive;
- to fewer than 150 natural or legal persons (other than qualified investors as defined in the Prospectus Directive), subject to obtaining the prior consent of the Joint Global Coordinators for any such offer; or
- in any other circumstances falling within Article 3(2) of the Prospectus Directive;

provided that no such offer of Offer Shares shall result in a requirement for the publication by the Company, the Main Shareholders or any Manager of a prospectus pursuant to Article 3 of the Prospectus Directive or a supplemental prospectus pursuant to Article 16 of the Prospectus Directive as supplemented by Commission Delegated Regulation (EC) no. 382/2014 of 7 March 2014.

For the purposes of this provision, the expression an "offer to the public" in relation to any Offer Shares in any Relevant Member State means the communication in any form and by any means of sufficient information on the terms of the Offering and the Offer Shares so as to enable an investor to decide to purchase Offer Shares, as that definition may be varied in that Relevant Member State by any measure implementing the Prospectus Directive in that Relevant Member State, and the expression "**Prospectus Directive**" means Directive 2003/71/EC (and amendments thereto to the extent implemented in the Relevant Member State), and includes any relevant implementing measure in the Relevant Member State.

United Kingdom Restrictions

Offers of the Offer Shares pursuant to the Offering are only being made to persons in the United Kingdom who are "qualified investors" or otherwise in circumstances which do not require publication by the Company of a prospectus pursuant to section 85(1) of the UK Financial Services and Markets Act 2000.

Any investment or investment activity to which the Offering Circular relates is available only to, and will be engaged in only with persons who, (i) are investment professionals falling within Article 19(5); or (ii) fall within Article 49(2)(a) to (d) ("high net worth companies, unincorporated associations, etc."), of the UK Financial Services and Markets Act 2000 (Financial Promotion) Order 2005 or other persons to whom such investment or investment activity may lawfully be made available (together, "**relevant persons**"). Persons who are not relevant persons should not take any action on the basis of the Offering Circular and should not act or rely on it.

Responsibility Statement

The Company's Responsibility
The Company is responsible for this Offering Circular in accordance with Danish law.

The Company's Statement
We hereby declare that we, as the persons responsible for this Offering Circular on behalf of the Company, have taken all reasonable care to ensure that, to the best of our knowledge and belief, the information contained in this Offering Circular is in accordance with the facts and does not omit anything likely to affect the import of its contents.

Copenhagen, 6 November 2017

Orphazyme A/S

Board of Directors

Georges Gemayel
Chairman

Bo Jesper Hansen
Deputy Chairman

Martijn Kleijwegt
Board Member

Martin Bonde
Board Member

Martin Rahbek Kornum
Board Member

Nanna Lüneborg
Board Member

Patrick J.H. Krol
Board Member

Rémi Droller
Board Member

Sten Verland
Board Member

Georges Gemayel: Professional board member
Bo Jesper Hansen: Professional board member
Martijn Kleijwegt: Founder and Managing Partner at LSP
Martin Bonde: Chief Executive Officer at Vaccibody AS
Martin Rahbek Kornum: Co-founder of Orphazyme and senior patent counsel at Coloplast A/S
Nanna Lüneborg: Principal at Novo Holdings A/S
Patrick J.H. Krol: Managing Partner at Aescap Venture
Rémi Droller: Managing Partner at Kurma Partners
Sten Verland: Partner and co-founder at Sunstone Life Science Ventures

Executive Management

Anders Mørkeberg Hinsby
CEO

Anders Vadsholt
CFO

Summary

Danish Summary
The Danish summary below is a translation of the English summary beginning on page 20. In the event of any discrepancies between the Danish and the English version, the English version shall prevail.

Dansk Resumé
Det danske resumé nedenfor er en oversættelse af det engelske resumé, som begynder på side 20. I tilfælde af uoverensstemmelse mellem det danske og det engelske resumé, skal det engelske resumé have forrang.

Resumé

Resuméer består af oplysningskrav, der benævnes "Elementer". Disse Elementer er nummereret i afsnit A—E (A.1—E.7). Dette resumé indeholder alle de Elementer, der skal være indeholdt i et resumé for denne type værdipapir og udsteder i henhold til Prospektforordningen nr. 486/2012 med senere ændringer. Da nogle Elementer ikke kræves medtaget, kan der forekomme huller i nummereringen af Elementerne. Selvom et Element skal indsættes i resuméet på grund af typen af værdipapir og udsteder, er det muligt, at der ikke kan gives nogen relevante oplysninger om Elementet. I så fald indeholder resuméet en kort beskrivelse af Elementet med angivelsen "ikke relevant".

Afsnit A – Indledning og advarsler

A.1 Advarsel til investorer

Dette resumé bør læses som en indledning til Prospektet.

Enhver beslutning om investering i de Udbudte Aktier bør træffes af investoren på baggrund af Prospektet som helhed.

Hvis en sag vedrørende oplysningerne i Prospektet indbringes for en domstol i henhold til national lovgivning i medlemsstaterne i det Europæiske Økonomiske Samarbejdsområde, kan den sagsøgende investor være forpligtet til at betale omkostningerne i forbindelse med oversættelse af Prospektet, inden sagen indledes.

Kun de personer, som har indgivet resuméet, herunder eventuelle oversættelser heraf, kan ifalde et civilretligt erstatningsansvar, men kun såfremt resuméet er misvisende, ukorrekt eller uoverensstemmende, når det læses sammen med de øvrige dele af Prospektet, eller hvis det ikke, når det læses sammen med Prospektets øvrige dele, indeholder nøgleoplysninger som hjælp til investorernes overvejelser om, hvorvidt de vil investere i de Udbudte Aktier.

A.2 Tilsagn til formidlere

Ikke relevant. Der er ikke indgået nogen aftale vedrørende anvendelse af Prospektet i forbindelse med et efterfølgende salg eller en endelig placering af de Udbudte Aktier. Selskabet er således ikke indforstået med, at Prospektet bruges i forbindelse med finansielle formidleres efterfølgende salg eller endelige placering af værdipapirer.

Afsnit B – Udsteder

B.1 Juridisk og kommercielt navn

Selskabet er registreret med det juridiske navn Orphazyme A/S og har ingen binavne.

B.2 Domicil, retlig form, indregistreringsland

Selskabet har hjemsted i Københavns Kommune på adressen Ole Maaløes Vej 3, 2200 København N og blev stiftet i Danmark som et anpartsselskab i henhold til dansk ret den 19. juni 2009 og senere omdannet til et aktieselskab den 20. oktober 2017.

B.3 Nuværende virksomhed og hovedaktiviteter

Orphazyme er et dansk biotekselskab med en sen klinisk pipeline vedrørende behandling af sjældne sygdomme, som udvikler nye behandlingsmuligheder for sjældne protein-misfoldningssygdomme. Selskabets førende lægemiddelkandidat, arimoclomol, stimulerer produktionen af cellernes protein-redningssystem, heat shock-proteiner ("**HSP'er**"), som hjælper de misfoldede proteiner med enten at genskabe deres funktionelle form eller, hvis dette ikke kan opnås, at fjerne proteinerne fra cellerne ved hjælp af cellernes genbrugssystem, lysosomerne, så de ikke længere danner toksiske aggregater.

Arimoclomol udvikles som en potentiel behandling af fire sjældne sygdomme: To neuromuskulære sygdomme, sporadisk inklusionslegeme myositis ("**sIBM**") og amyotrofisk lateral sklerose ("**ALS**"), og to lysosomale ophobnings-sygdomme, Niemann-Picks sygdom type c ("**NPC**") og Gauchers sygdom. Arimoclomol har vist sig at være veltolereret i syv fase I-forsøg og tre fase II-forsøg, hvor tolerabiliteten var sammenlignelig med den for placebo. Arimoclomol fås i kapselform, er i stand til at passere blodhjernebarrieren og har en biotilgængelighed på 80-90%, og der er opnået klinisk proof of concept (validering), baseret på retningsbestemt forbedring på tværs af uafhængige endepunkter, i de neuromuskulære sygdomme samt prækliniske data, der understøtter udviklingen af arimoclomol i de lysosomale ophobningssygdomme.

Selskabet forventer i øjeblikket at have afsluttet tre potentielle registreringsstudier ved udgangen af 2020, og den første potentielle markedsføringsgodkendelse forventes i 2020.

B.4a Beskrivelse af de væsentligste nyere tendenser, der påvirker Selskabet og de sektorer, inden for hvilke Selskabet opererer

Orphazyme har til hensigt at lancere nye behandlinger af alvorlige og invaliderende, sjældne sygdomme – indledningsvist inden for området protein-misfoldningssygdomme. De neuromuskulære og lysosomale ophobnings-sygdomme fratager progressivt patienterne deres grundlæggende funktioner og medfører i mange tilfælde for tidlig død.

Sjældne sygdomme ("orphan diseases") rammer en lille andel af befolkningen. For mange af de sjældne sygdomme findes der ingen eller kun få behandlingsmuligheder, og der er derfor et stort behov for at udvikle nye behandlinger. Orphazyme udvikler medicin, som potentielt kan forbedre livet for de patienter, der lider af disse sygdomme, hvor der er begrænsede eller slet ingen alternative behandlingsmuligheder.

For at fremme og give incitament til at forske i og udvikle lægemiddelprodukter til sjældne sygdomme, trods det lave antal patienter og de store udviklingsomkostninger, vedtog man i USA i 1983 Orphan Drug Act for at undgå, at disse sygdomme bliver overset. Orphan Drug Act blev en succes og førte til implementeringen af ordninger for lægemidler til sjældne sygdomme ("orphan drugs") på andre store markeder, herunder f.eks. i Japan i 1993 og i EU i 2000. Under orphan drug-ordningerne kan myndighederne tildele et lægemiddelprodukt såkaldt orphan drug-designation. Selskaber, som udvikler lægemidler med orphan drug-designation, er berettiget til en lang række fordele, herunder muligheden for gratis rådgivning fra FDA og EMA samt visse økonomiske fordele som f.eks. skattefradrag for forsknings- og udviklingsomkostninger samt fritagelse for eller reducerede gebyrer for regulatoriske ansøgninger. Hvis en lægemiddelkandidat med orphan drug-designation godkendes af de regulatoriske myndigheder eller myndighedsorganer ved afslutningen af kliniske forsøg, kan de respektive myndigheder beslutte, hvorvidt det farmaceutiske produkt skal tildeles orphan drug-status. Med orphan drug-status har det pågældende lægemiddel markedseksklusivitet i syv år i USA og ti år i EU[1].

[1] EMA European Medicines Agency, Marketing authorisation and market exclusivity, http://www.ema.europa.eu/ema/index.jsp?curl=pages/regulation/general/general_content_000392.jsp& and U.S. Food & Drug Administration, Code of Federal Regulations Title 21, https://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfcfr/cfrsearch.cfm?cfrpart=316&showfr=1&source=govdelivery&subpartnode=21%3A5.0.1.1.6.4&utm_medium=email&utm_source=govdelivery

Da orphan drugs er rettet mod sjældne, svært invaliderende og/eller livstruende sygdomme med mindre patientgrupper, kan disse lægemidler sælges til en højere pris end almindelige lægemidler. I 2016 var den gennemsnitlige årlige omkostning pr. patient i USA ca. fem gange højere for orphan drugs end for lægemidler mod større sygdomme[2]. Der er mange faktorer, som påvirker prisen på orphan drugs og almindelige lægemidler, herunder sygdomsbyrde, samfundets omkostningsbesparelser, behandlingsbehov, klinisk behandlingseffekt, bivirkninger, tolerabilitet, antal patienter berørt af sygdommen, omkostninger forbundet med udvikling og fremstilling, samt forhandlinger med betalere/forsikringsselskaber.

I 2016 steg det globale salg af orphan drugs med 12,2% på årsbasis og nåede USD 114 mia., mens salget af lægemidler mod større sygdomme (eksklusive generiske lægemidler) steg med 2,4% i samme periode og udgjorde USD 578 mia. Det samlede salg af receptpligtig medicin (eksklusive generiske lægemidler) steg med 3,9% i 2016 og udgjorde i alt USD 692 mia. Markedet for orphan drugs forventes at vokse til USD 209 mia. inden 2022, svarende til en gennemsnitlig årlig vækstrate på 11,1% i perioden 2017-2022, hvilket er næsten dobbelt så høj vækst som for det samlede marked for receptpligtig medicin i samme periode[3] .

B.5 Beskrivelse af Koncernen og Selskabets plads i Koncernen

Ikke relevant. Selskabet er ikke en del af en koncernstruktur.

B.6 Personer, som direkte eller indirekte har en andel i udsteders kapital eller stemmerettigheder eller kontrollerer Selskabet

Ved gennemførelsen af Kapitalomstruktureringen 2017 (under forudsætning af en Udbudskurs svarende til den højeste kurs i Udbudskursintervallet) og forud for Optagelsen vil Selskabets Aktier være ejet af Novo Holdings (33,1%), Aescap Venture (15,2%), Sunstone Capital (15,6%), Orpha Pooling B.V. (et joint venture mellem LSP og ALS Invest) (21,0%), Idinvest (7,2%) og Kurma Biofund II (6,6%) (samlet benævnt "**Hovedaktionærerne**") samt UCL Business PLC (0,1%) og Kansas Life Sciences Development Inc. (0,1%). De resterende Aktier (1,1%) ejes indirekte af Selskabets grundlæggere (CSO Thomas Kirkegaard Jensen; CEO Anders Hinsby; professor Marja Helena Jaattela samt medlem af den Nuværende Bestyrelse (som defineret i dette Prospekt) og medstifter, Martin Rahbek Kornum) gennem deres respektive 100%-ejede holdingselskaber.

Hvis Udbudskursen fastsættes under en kurs svarende til den højeste kurs i Udbudskursintervallet, vil der blive udstedt et yderligere antal bonusaktier kort før Optagelsen til de tidligere præferenceaktionærer med henblik på at tage højde for Udbudskursen som led i Kapitalomstruktureringen 2017. Under forudsætning af en Udbudskurs svarende til den laveste kurs i Udbudskursintervallet vil der blive udstedt op til 1.621.968 stk. bonusaktier a nom. DKK 1. Ovenstående ejerandele vil derfor blive tilpasset i overensstemmelse hermed.

I alt 1.293.293 Pre-IPO Warrants (som defineret i dette Prospekt) kan udnyttes (eller bortfalder hvis de ikke udnyttes) i forbindelse med Udbuddet. Den maksimale nominelle kapitalforhøjelse som følge af udnyttelsen af Pre-IPO Warrants er begrænset til DKK 866.965. Kapitalforhøjelsen forbundet med en eventuel udnyttelse af Pre-IPO Warrants forventes at finde sted i forbindelse med afvikling af Udbuddet, dvs. omkring Afviklingsdatoen den 21. november 2017.

Alle Aktier har samme rettigheder og er ligestillet bl.a. med hensyn til stemmeret.

Bortset fra som anført ovenfor er Selskabet ikke bekendt med, at nogen person direkte eller indirekte ejer en andel af Selskabets aktiekapital eller stemmerettigheder, der skal indberettes efter dansk ret.

B.7 Udvalgte regnskabs- og virksomhedsoplysninger

Nedenstående opstillinger viser et resumé af regnskabsoplysninger for Orphazymes virksomhed. Selskabet har uddraget resuméet af resultatopgørelsen for regnskabsårene 2016 og 2015 af det offentliggjorte Reviderede Regnskab (som defineret i dette Prospekt) for 2016 og det reviewede, ikke-reviderede Sammenligningsregnskab for 2015 (som defineret i dette Prospekt), som er medtaget på side F-17 i Prospektet. Selskabet har uddraget resuméet af resultatopgørelsen for 1. halvår 2017 og 2016 og resuméet af balancen pr. 30. juni 2017 af det ikke-reviderede sammendrag af delårsregnskabet, som er medtaget på side F-5 - F-6 i Prospektet.

[2] Orphan Drug Report 2015, Evaluate Pharma; Orphan Drug Report 2017, Evaluate Pharma
[3] Orphan Drug Report 2017, Evaluate Pharma

Orphazyme fører sine regnskaber og optegnelser i DKK, og det reviderede regnskab er udarbejdet i overensstemmelse med internationale regnskabsstandarder ("**IFRS**") udstedt af International Accounting Standards Board ("**IASB**") og godkendt af den Europæiske Union samt yderligere krav i årsregnskabsloven. Disse data bør læses sammen med Selskabets regnskab og tilhørende noter, som er medtaget i Prospektet.

Resultatopgørelse

	1. halvår		Kalenderår	
(DKK '000)	**1. halvår 2017**	**1. halvår 2016**	**2016**	**2015**
Forsknings- og udviklingsomkostninger	(46.870)	(27.875)	(55.817)	(45.865)
Administrationsomkostninger	(7.972)	(2.696)	(7.703)	(7.220)
Driftsunderskud	**(54.842)**	**(30.571)**	**(63.520)**	**(53.085)**
Finansielle poster	(122)	35	85	(317)
Resultat før skat	**(54.964)**	**(30.536)**	**(63.435)**	**(53.402)**
Skattegodtgørelse	2.841	2.750	5.500	5.688
Periodens resultat	**(52.123)**	**(27.786)**	**(57.935)**	**(47.714)**

Balance

(DKK '000)	Pr. 30. juni 2017	Pr. 31. december 2016	Pr. 31. december 2015
Aktiver			
Langfristede aktiver			
Materielle anlægsaktiver	1.007	987	1.487
Tilgodehavende skat	5.591	2.750	2.750
Deposita	357	310	211
Langfristede aktiver i alt	**6.955**	**4.047**	**4.448**
Kortfristede aktiver			
Tilgodehavende skat	5.500	5.500	5.875
Tilgodehavende fra kapitalforhøjelse	91.319	–	–
Andre tilgodehavender	5.104	3.421	644
Periodeafgrænsningsposter	3.245	4.624	5.970
Likvide beholdninger	33.589	14.349	68.015
Kortfristede aktiver i alt	138.757	27.894	80.504
Aktiver i alt	**145.712**	**31.941**	**84.952**
Passiver			
Egenkapital			
Egenkapital i alt	**121.297**	**17.509**	**74.143**
Kortfristede aktiver			
Leverandørgæld	7.614	4.718	2.447
Gæld til aktionærer	223	–	–
Anden gæld	16.578	9.714	8.362
Kortfristede forpligtelser i alt	24.415	14.432	10.809
Passiver i alt	**145.712**	**31.941**	**84.952**

Pengestrømsopgørelse

	1. halvår		Kalenderår	
(DKK '000)	**1. halvår 2017**	**1. halvår 2016**	**2016**	**2015**
Driftsaktiviteter				
Nettoresultat før skat	(54.842)	(30.571)	(63.435)	(53.402)
Justering vedr. afstemning af resultat før skat til pengestrømme fra driftsaktiviteter				
Aktiebaseret omkostning	–	–	–	–
Af- og nedskrivninger	291	287	706	554
Gevinst/tab på afhændelse af aktiver	–	–	33	–
Ændring i andre tilgodehavender	(1.730)	(1.032)	(2.876)	(277)
Ændring i forudbetalinger	1.379	1.296	1.347	(4.969)
Ændringer i leverandørgæld	2.896	1.134	2.271	858
Ændringer i anden gæld	6.821	2.284	1.352	4.330
Pengestrømme fra skat	–	–	5.875	6.250
Renteudgifter, netto	(122)	35	–	241
Pengestrømme til driftsaktivitet (netto)	**(45.307)**	**(26.567)**	**(54.727)**	**(46.414)**
Investeringsaktiviteter				
Investeringer i materielle anlægsaktiver	(311)	–	(238)	(495)
Pengestrømme til investeringsaktiviteter (netto)	**(311)**	**–**	**(238)**	**(495)**
Finansieringsaktiviteter				
Kapitalbidrag fra aktionærerne	65.431	–	1.330	85.970
Likviditet fra konvertible lån	–	–	–	–
Banklån	–	463	–	–
Omkostninger forbundet med kapitalbidrag	(573)	–	(30)	(215)
Pengestrømme fra finansieringsaktiviteter (netto)	**64.858**	**463**	**1.300**	**85.755**
Netto ændring i likvide beholdninger	**19.240**	**(26.104)**	**(53.665)**	**38.846**
Likvide beholdninger primo perioden	14.349	68.014	68.014	29.169
Likvide beholdninger ultimo perioden	**33.589**	**41.910**	**14.349**	**68.015**

Sammenligning mellem 1. halvår 2017 og 1. halvår 2016

Drifts resultat

Forsknings- og udviklingsomkostningerne udgjorde i alt DKK 46,9 mio. i 1. halvår 2017 i forhold til DKK 27,9 mio. i samme periode i 2016. Stigningen skyldes primært igangsættelsen af fase II/III-forsøget vedrørende NPC medio 2016 og omkostninger til forberedelser til et fase II-forsøg i Indien vedrørende Gauchers sygdom. Perioden var desuden påvirket af omkostninger til fremstilling af arimoclomol til fase III-forsøget i sIBM.

Administrationsomkostningerne udgjorde DKK 8,0 mio. i 1. halvår 2017 i forhold til DKK 2,7 mio. i samme periode i 2016. Stigningen skyldes primært omkostninger til advokater, revisorer og konsulenter til forberedelse af børsnoteringen ("**Børsnoteringen**") samt rekruttering af nye ledelsesmedlemmer og yderligere personale. En del af omkostningerne forbundet med Børsnoteringen og den forventede udstedelse af nye Aktier er indregnet i forudbetalinger, indtil Udbuddet er gennemført. Pr. 30. juni 2017 har Selskabet i alt indregnet forudbetalinger vedrørende Børsnoteringen på DKK 1,4 mio.

Pr. 30. juni 2017 havde Orphazyme likvide beholdninger på DKK 33,6 mio. mod likvide beholdninger på DKK 14,3 mio. pr. 31. december 2016. Stigningen afspejler kapitalforhøjelsen på DKK 65,4 mio. i 1. halvår 2017 og opvejes til dels af omkostninger forbundet med Orphazymes forretningsaktiviteter, herunder omkostninger til igangværende og planlagte kliniske forsøg. I 1. halvår 2017 udgjorde pengestrømme fra driftsaktiviteter DKK -45,3 mio. mod DKK -26,6 mio. i 1. halvår 2016. Selskabet har øget sine pengestrømme fra finansieringsaktiviteter via kapitalforhøjelser fra både nuværende og nye investorer.

Sammenligning mellem regnskabsårene 2016 og 2015

Forsknings- og udviklingsomkostningerne udgjorde i alt DKK 55,8 mio. i 2016 mod DKK 45,9 mio. i 2015. Stigningen skyldes primært igangsættelsen af fase II/III-forsøget vedrørende NPC medio 2016 og omkostninger forbundet med nye medarbejdere til at understøtte det stigende aktivitetsniveau.

Administrationsomkostningerne udgjorde DKK 7,7 mio. i 2016 sammenlignet med DKK 7,2 mio. i 2015. Stigningen fra 2015 til 2016 skyldtes rekruttering af nye ledelsesmedlemmer ved udgangen af 2016.

Pr. 31. december 2016 havde Orphazyme likvide beholdninger på DKK 14,3 mio. mod likvide beholdninger på DKK 68,0 mio. pr. 31. december 2015. Faldet kunne henføres til omkostninger forbundet med Orphazymes forretningsaktiviteter, herunder omkostninger til igangværende og planlagte kliniske forsøg.

B.8 Udvalgte vigtige proforma-regnskabsoplysninger

Ikke relevant. Prospektet indeholder ingen proforma-regnskabsoplysninger, da der ikke har været nogen transaktioner, som medfører en væsentlig (defineret som mere end 25%) bruttoændring i relevante nøgletal som f.eks. balancesum, nettoomsætning eller nettoresultat.

B.9 Resultatforventninger eller -prognoser

Selskabet forventer et nettounderskud for 2017 på DKK 125 mio. - DKK 135 mio. Orphazyme har ingen omsætning, og omkostningerne vedrører primært forskning og udvikling. Skattegodtgørelse og indtægter fra offentlige tilskud har en positiv indvirkning på årets nettoresultat. Orphazymes resultat for 2017 vil kunne afvige markant fra denne prognose.

B.10 Forbehold i revisionspåtegningen vedrørende historiske finansielle oplysninger

Ikke relevant. Revisionspåtegningerne på de historiske regnskabsoplysninger i Prospektet er afgivet uden forbehold.

B.11 Forklaring, hvis Selskabets arbejdskapital ikke er tilstrækkelig til at dække Selskabets nuværende behov

Det er Orphazymes opfattelse, af den arbejdskapital, som er til rådighed pr. prospektdatoen, ikke er tilstrækkelig til at dække det aktuelle arbejdskapitalbehov i en periode på 12 måneder efter prospektdatoen. Som yderligere beskrevet i "*Baggrund for Udbuddet og Anvendelse af provenu*" har Orphazyme til hensigt at finansiere sine aktiviteter i 12 måneder efter prospektdatoen og datoen for Optagelsen med en del af provenuet fra Udbuddet. Hvis Udbuddet ikke gennemføres, vil Orphazyme søge alternative finansieringsmuligheder i samarbejde med eksisterende aktionærer.

Afsnit C – Værdipapirer

C.1 En beskrivelse af typen og klassen af Udbudte Aktier, herunder fondskode

Aktierne er ikke opdelt i aktieklasser:
Permanent ISIN-kode for Aktierne: DK0060910917
Midlertidig ISIN-kode for de Midlertidige Købsbeviser: DK0060911055

C.2 Valuta for de Udbudte Aktier

De Udbudte Aktier vil være denomineret i danske kroner ("**DKK**").

C.3 Antallet af udstedte og fuldt indbetalte Aktier og af udstedte, men ikke fuldt indbetalte Aktier

Før Udbuddets gennemførelse vil Selskabets aktiekapital have en nominel værdi på DKK 12.310.967, fordelt på 12.310.967 stk. Aktier a DKK 1 eller multipla heraf, under forudsætning af en Udbudskurs svarende til midtpunktet i Udbudskursintervallet (og derved en udstedelse af 720.875 stk. bonusaktier forud for Optagelsen som led i Kapitalomstruktureringen 2017), som alle vil blive udstedt og fuldt indbetalt.

C.4 En beskrivelse af Aktiernes rettigheder

Alle Aktier, herunder de Udbudte Aktier, har samme rettigheder, og de Udbudte Aktier er ligestillet med alle andre Aktier med hensyn til stemmeret, fortegningsret, indløsning, konvertering og restriktioner eller begrænsninger i henhold til Selskabets vedtægter ("**Vedtægterne**") eller med hensyn til ret til udbytte eller provenu i tilfælde af opløsning eller likvidation. I henhold til Selskabets Vedtægter er ingen Aktier omfattet af særlige rettigheder, restriktioner eller begrænsninger.

Hvert aktiebeløb med en nominel værdi på DKK 1 giver én stemme på Selskabets generalforsamling samt ret til at modtage udloddet udbytte.

Enhver aktionær har ret til at få behandlet et bestemt emne på generalforsamlingen, såfremt aktionæren skriftligt fremsætter krav derom over for Bestyrelsen senest seks uger før generalforsamlingen.

C.5 En beskrivelse af eventuelle indskrænkninger i Aktiernes omsættelighed

Ikke relevant. Aktierne er frit omsættelige omsætningspapirer, og der gælder ingen indskrænkninger i Aktiernes omsættelighed i henhold til Selskabets Vedtægter eller dansk ret.

C.6 Optagelse til handel på et reguleret marked

De Midlertidige Købsbeviser er søgt optaget til handel på Nasdaq Copenhagen under symbolet "ORPHA TEMP", og Aktierne er søgt optaget til handel og officiel notering på Nasdaq Copenhagen under symbolet "ORPHA". Optagelsen er bl.a. betinget af gennemførelse af Nasdaq Copenhagens godkendelse af spredningen af de Udbudte Aktier, valg af de Nye Bestyrelsesmedlemmer (som defineret i dette Prospekt), at Udbuddet ikke trækkes tilbage før afvikling af Udbuddet, og at Selskabet offentliggør en meddelelse herom.

Hvis Udbuddet lukkes før den 16. november 2017, vil Optagelsen, Afviklingsdatoen, levering af de Midlertidige Købsbeviser, automatisk ombytning af Midlertidige Købsbeviser til Aktier og Aktiernes første handels- og officielle noteringsdag på Nasdaq Copenhagen blive fremrykket tilsvarende.

Betaling for og afvikling af de Udbudte Aktier forventes at finde sted omkring den 21. november 2017 i form af levering af Midlertidige Købsbeviser. Betinget af gennemførelse af Udbuddet og registrering af de Udbudte Aktier i Erhvervsstyrelsen vil de Midlertidige Købsbeviser automatisk blive ombyttet i VP Securities til et tilsvarende antal Aktier, der forventes leveret den 23. november 2017. Første handels- og officielle noteringsdag på Nasdaq Copenhagen forventes at være den 17. november 2017, betinget af at Udbuddet ikke trækkes tilbage før afvikling og gennemførelse af Udbuddet. Første handelsdag for de Midlertidige Købsbeviser på Nasdaq Copenhagen forventes at være den 17. november 2017, og sidste handelsdag for de Midlertidige Købsbeviser på Nasdaq Copenhagen forventes at være den 21. november 2017. Første handelsdag for Aktierne på Nasdaq

Copenhagen i den permanente ISIN-kode forventes at være den 22. november 2017. I forbindelse med de Midlertidige Købsbevisers automatiske ombytning til Aktier vil de Midlertidige Købsbeviser ophøre med at eksistere.

C.7 En beskrivelse af udbyttepolitik

Selskabet har ikke erklæret udbytte eller foretaget udbytteudlodninger i de seneste to regnskabsår. Selskabet har aktuelt til hensigt at anvende alle tilgængelige ressourcer samt eventuel omsætning til Selskabets nuværende og fremtidige aktiviteter. Pr. prospektdatoen forventer Selskabet ikke at foretage udbytteudlodninger i den nærmeste fremtid.

Afsnit D – Risici

D.1 Nøgleoplysninger om de vigtigste risici, der er specifikke for Selskabet eller dettes branche

De nedenfor omtalte risikofaktorer og usikkerheder omfatter de risici, som Orphazymes ledelse på nuværende tidspunkt vurderer som værende væsentlige, men det er ikke de eneste risikofaktorer og usikkerheder, Orphazyme er eksponeret mod. Der er yderligere risikofaktorer og usikkerheder, herunder risici som Orphazyme på nuværende tidspunkt ikke er bekendt med, eller som ledelsen på nuværende tidspunkt anser for uvæsentlige, som kan opstå eller blive væsentlige i fremtiden, og som kan føre til et fald i de Udbudte Aktiers værdi, og til at hele eller en del af det investerede beløb mistes. Risikofaktorerne er ikke nævnt i prioriteret rækkefølge efter vigtighed eller sandsynlighed.

- Orphazyme kan blive negativt berørt af konkurrence fra andre life science-virksomheder, der udvikler andre behandlinger til de samme sygdomme, som Orphazyme udvikler produkter imod.
- Ændringer i det regulatoriske miljø og compliance-regler kan få væsentlig negativ indvirkning på Selskabet.
- Prisen på og efterspørgslen efter farmaceutiske produkter kan blive påvirket af globale økonomiske faktorer. Orphazymes evne til at fastsætte priser og dermed skabe omsætning fra produkter, det måtte udvikle, afhænger af den gældende og fremtidige tilskuds- og lægemiddelprispolitik og regler derfor.
- Selskabet har ingen produkter, som er godkendt til kommercielt salg, har aldrig genereret indtægter og kan lide væsentlige underskud i fremtiden, hvilket gør det svært at vurdere dets fremtidige levedygtighed.
- Orphazyme er på nuværende tidspunkt meget afhængig af arimoclomol.
- De kliniske forsøg, der gennemføres til afprøvning af Selskabets produkter, vil måske ikke vise de ønskede resultater, og kan blive forsinket eller blive dyrere end forventet.
- Orphazymes produkter kan medføre uønskede bivirkninger eller besidde andre egenskaber, som kan forsinke eller forhindre, at de bliver godkendt af myndighederne, begrænse den kommercielle profil eller medføre væsentligt negative konsekvenser efter en eventuel regulatorisk godkendelse.
- Udfordringer i forbindelse med rekruttering af patienter til kliniske forsøg og indgåelse af aftaler med investigatorer og hospitaler kan få væsentlig negativ indvirkning på Selskabet.
- Orphazyme er yderst afhængig af at opnå og opretholde fornødne regulatoriske godkendelser.
- Hvis Selskabet ikke er i stand til at opnå orphan drug-designation eller markedseksklusivitet for sine produktkandidater eller ikke kan få gavn af den dermed forbundne markedseksklusivitet, vil det få væsentlig negativ indvirkning på Orphazyme.
- Risici forbundet med fremstilling samt forskning og udvikling, herunder brug af farlige stoffer og kemi- og produktionskontrol ("**CMC**"), kan få væsentlig negativ indvirkning på Selskabet.
- Selskabet er afhængig af, at eksterne leverandører leverer bestemte licenser, produkter og tjenesteydelser og kliniske forsøg, og eventuelle problemer med Selskabets væsentlige eksterne leverandører kan gribe forstyrrende ind i Selskabets virksomhed og drift.
- Orphazyme er afhængig af en vellykket kommercialisering af sine produkter.
- Selskabet kan fremover søge at indgå samarbejdsaftaler med tredjemand om udvikling og kommercialisering af sine produkter. Hvis sådanne samarbejdsaftaler ikke er succesfulde, vil Selskabet muligvis ikke kunne udnytte produkternes markedspotentiale.
- Uønskede hændelser, produktansvar og andre krav kan få væsentlig negativ indvirkning på Orphazyme.
- Ændringer i gældende lovgivning og regulering i de jurisdiktioner, hvor Selskabet har aktiviteter, eller involvering i retstvister kan få væsentlig negativ indvirkning på Selskabets virksomhed, finansielle stilling, resultat og fremtidsudsigter.

- Selskabet er udsat for risici vedrørende databeskyttelse, brud på cybersikkerhed og manglende overholdelse af regler for behandling af personoplysninger og sikkerhedskrav vedrørende data.
- Selskabet vil måske ikke være i stand til at tiltrække, integrere, styre og fastholde kompetent personale eller nøglemedarbejdere.
- Risici forbundet med at få tildelt en priority review voucher.
- Ændringer i valutakurser og renter kan få negativ indvirkning på Selskabets driftsresultat.
- Selskabet kan i fremtiden få behov for yderligere kapital, som måske ikke vil være tilgængelig på forretningsmæssigt gunstige vilkår, om overhovedet.
- De fremadrettede finansielle oplysninger indeholdt i dette Prospekt kan afvige væsentligt fra Selskabets faktiske resultater, og investorer bør ikke tillægge de fremadrettede regnskabsoplysninger for megen vægt.
- Hvis Orphazyme ikke er i stand til at opnå og opretholde beskyttelse af relevante immaterielle rettigheder, vil det få væsentlig negativ indvirkning på værdien af Orphazymes produkter.
- Orphazyme er muligvis ikke i stand til at håndhæve eller beskytte sine immaterielle rettigheder tilstrækkeligt.
- Orphazyme kan blive mødt af påstande om krænkelse og anden anfægtelse fra tredjemands side.
- De tidligere arbejdsgivere for Orphazymes medarbejdere og konsulenter vil muligvis forsøge at gøre rettigheder til Orphazymes immaterielle rettigheder gældende.
- Selskabet vil ikke søge at beskytte sine immaterielle rettigheder i alle jurisdiktioner verden over og vil muligvis ikke være i stand til i tilstrækkelig grad at håndhæve sine immaterielle rettigheder selv i de jurisdiktioner, hvor der søges beskyttelse.
- Hvis Orphazyme ikke er i stand til at sikre fortroligheden af visse oplysninger, kan det få væsentlig negativ indflydelse på værdien af Selskabets produkter og teknologi.

D.3 Nøgleoplysninger om de vigtigste risici vedrørende de Udbudte Aktier

- Efter Udbuddet vil Hovedaktionærerne fortsat være større aktionærer og kan kontrollere eller på anden måde påvirke Selskabets vigtige dispositioner.
- Der er en begrænset mængde Aktier i fri handel.
- Selskabets Aktier har ikke tidligere været handlet offentligt, og kursen kan være volatil og variere.
- Forskelle i valutakurser kan få væsentlig negativ indvirkning på værdien af aktiebeholdninger eller værdien af udbetalt udbytte.
- Selskabet har til hensigt at tilbageholde alle likvide midler og eventuelle fremtidige indtægter og geninvestere dem i Selskabet, og derfor vil aktionærernes evne til at opnå et afkast på deres investering måske afhænge af en stigning i kursen på Aktierne.
- Hvis værdipapir- eller brancheanalytikere ikke offentliggør analyser eller offentliggør upræcise eller ugunstige analyser om Selskabets virksomhed, kan det medføre et fald i Aktiernes kurs og handelsvolumen.
- Fremtidige salg af Aktier efter Udbuddet kan medføre et fald i Aktiernes markedskurs.
- Udstedelsen af yderligere Aktier i Selskabet til finansiering af den fremtidige drift, udvikling og kommercialisering af produkter eller til finansiering af virksomhedskøb, et eventuelt aktieincitaments- eller aktieoptionsprogram m.v. kan udvande de eksisterende aktiebesiddelser.
- Amerikanske og andre udenlandske aktionærer vil muligvis ikke kunne deltage i fremtidige aktieudbud.
- Investors rettigheder som aktionær er underlagt dansk ret, som i visse henseender afviger fra aktionærrettighederne i henhold til retsreglerne i andre lande.
- Omdannelsen til et børsnoteret selskab vil øge Selskabets omkostninger og kan få en forstyrrende indvirkning på Selskabets ordinære virksomhedsdrift.
- Udbuddet kan blive tilbagekaldt efter, at de Midlertidige Købsbeviser er optaget til handel, og indtil der er sket afvikling af Udbuddet.

Afsnit E – Udbud

E.1 Udbuddets samlede nettoprovenu og anslåede udgifter

Nettoprovenuet til Selskabet fra salget af nye Aktier, der udstedes af Selskabet i forbindelse med Udbuddet, forventes at være ca. DKK 635 mio. (heraf DKK 85 mio. i henhold til Overallokeringsretten) efter fradrag af provision og estimerede omkostninger til Udbuddet, som Selskabet skal betale, under de forudsætninger, som er anført i Prospektet.

De samlede omkostninger i forbindelse med Udbuddet, herunder provisioner og honorarer (faste og diskretionære), som Selskabet skal betale til Emissionsbankerne, andre rådgiverhonorarer og omkostninger, skønnes at udgøre ca. DKK 55 mio. (under forudsætning af fuld udnyttelse af Overallokeringsretten).

Selskabet har endvidere indgået aftale om at betale en salgsprovision til kontoførende institutter (dog undtaget Emissionsbankerne) svarende til 0,25% af Udbudskursen på de Udbudte Aktier, der tildeles til ordrer med en kursværdi til og med DKK 3 mio. afgivet gennem de kontoførende institutter (undtagen Emissionsbankerne), der betales af Selskabet beregnet forholdsmæssigt i forhold til det antal Udbudte Aktier, der sælges).

E.2a Baggrund for Udbuddet og anvendelse af provenu, forventet nettoprovenu

Udbuddet forventes at understøtte Orphazymes driftsmæssige strategi, styrke Orphazymes offentlige og kommercielle profil og give Orphazyme forbedret adgang til de offentlige kapitalmarkeder og en bred kreds af nye danske og internationale aktionærer.

Orphazyme anslår, at nettoprovenuet fra Udbuddet vil udgøre ca. DKK 550 mio., ekskl. Overallokeringsretten. Hvis Joint Global Coordinators udnytter Overallokeringsretten fuldt ud, anslår Selskabet, at nettoprovenuet til Selskabet fra Udbuddet vil udgøre ca. DKK 635 mio. Disse estimater er med forbehold for de forudsætninger, der er beskrevet i "*Udbuddet – Omkostninger i forbindelse med Udbuddet*".

Orphazymes baggrund for Udbuddet er at rejse kapital til at understøtte virksomheden. Selskabet har til hensigt at allokere nettoprovenuet fra Udbuddet sammen med den eksisterende likviditet pr. 30. september 2017, i alt DKK 649 mio., ekskl. Overallokeringsretten, som følger:

- Ca. DKK 135-155 mio. til finansiering af gennemførelsen af et fase II/III-forsøg med arimoclomol til behandling af sIBM.
- Ca. DKK 165-185 mio. til finansiering af gennemførelsen af et fase II/III-forsøg med arimoclomol til behandling af ALS.
- Ca. DKK 75-85 mio. til finansiering af gennemførelsen af et fase II/III-forsøg med arimoclomol til behandling af NPC.
- Ca. DKK 65-75 mio. til finansiering af gennemførelsen af et fase II-forsøg med arimoclomol til behandling af Gauchers sygdom.
- Ca. DKK 85-95 mio. til finansiering af generelle forsknings- og udviklingsaktiviteter, hvoraf ca. 50% allokeres til finansiering af forskningsstøtte til igangværende kliniske forsøg og tilsyn med centrale laboratorier, ca. 40% til finansiering af aktiviteter, der kan føre til indlevering af NDA-ansøgninger (new drug application), og ca. 10% til finansiering af fremskridt inden for forsknings- og leadprojekter til præklinisk udvikling.
- Det resterende beløb, DKK 65-75 mio. til finansiering af forberedelse til registrerings- og kommercielle aktiviteter, til finansiering af arbejdskapitalen samt til generelle selskabs- og administrative formål, som kan omfatte ansættelse af yderligere personale, anlægsinvesteringer og omkostninger forbundet med at drive en børsnoteret virksomhed.

Hvis Overallokeringsretten (maks. DKK 85 mio. i nettoprovenu) udnyttes, vil provenuet blive anvendt til yderligere at understøtte fremskridt inden for forsknings- og leadprojekter til præklinisk udvikling samt registrerings- og kommercielle aktiviteter.

Hvis der opnås positive resultater fra fase II/III-forsøget med arimoclomol til behandling af NPC, kan Orphazyme vælge af reallokere midler til at understøtte ansøgnings- og registreringsaktiviteter vedrørende arimoclomol til behandling af NPC, hvilket vil føre til en hurtigere anvendelse af provenuet fra Udbuddet. I dette scenarie kan Selskabet få behov for at rejse yderligere kapital, opnå gældsfinansiering eller søge at indgå partnerskaber eller andre finansieringsordninger for at have finansiering til at gennemføre forsøgene i de øvrige indikationer samtidig med gennemførelsen af ansøgnings- og registreringsaktiviteterne samt forberede sig til en senere kommercialisering. Hvis der opnås negative resultater fra fase II/III-forsøget med arimoclomol til behandling af NPC, vil Orphazyme anvende de resterende midler til at gennemføre de planlagte forsøg i de øvrige tre indikationer.

Ovenstående forventede anvendelse af nettoprovenuet fra Udbuddet udgør Selskabets aktuelle hensigter baseret på de nuværende planer og forretningsmæssige forhold. Pr. datoen for Prospektet kan Orphazyme ikke med sikkerhed forudsige alle detaljerne omkring anvendelsen af nettoprovenuet fra Udbuddet eller de beløb, som Selskabet faktisk vil bruge på ovennævnte formål. De faktiske omkostninger kan afvige væsentligt fra disse skøn, og Selskabet kan vurdere, at det er nødvendigt eller tilrådeligt at omfordele nettoprovenuet inden for de ovenfor beskrevne kategorier eller at anvende dele deraf til andre formål. Omfanget og den tidsmæssige placering af den faktiske anvendelse af nettoprovenuet vil variere som følge af en lang række forhold, herunder den relative succes og omkostninger forbundet med dets forsknings-, prækliniske og kliniske udviklingsprogrammer, og hvorvidt Orphazyme indgår samarbejdsaftaler og/eller strategiske partnerskaber med eksterne parter.

E.3 Udbudsbetingelser

Selskabet udbyder op til 10.781.250 stk. nye Aktier (inkl. Overallokeringsaktier) med henblik på at rejse et bruttoprovenu på op til DKK 690 mio. (heraf DKK 90 mio. i henhold til Overallokeringsretten). Under forudsætning af gennemførelse af Udbuddet vil Selskabets registrerede aktiekapital stige med op til nominelt DKK 9.375.000 som følge af udstedelsen af Udbudte Aktier (ekskl. Overallokeringsretten). Det præcise antal Udbudte Aktier fastsættes på grundlag af en bookbuilding-proces.

Selskabet har givet Joint Global Coordinators en Overallokeringsret til, på vegne af Emissionsbankerne, at tegne op til 1.406.250 stk. Overallokeringsaktier (nye Aktier), som kan udnyttes helt eller delvist fra datoen for Optagelsen og indtil 30 kalenderdage derefter, alene til dækning af eventuel overallokering eller korte positioner i forbindelse med Udbuddet. Med henblik på levering af de Udbudte Aktier til investorer i forbindelse med Overallokeringsretten har Novo Holdings indvilget i at stille 1.406.250 stk. eksisterende Aktier til rådighed. Orphazyme har indvilget i at udstede op til et tilsvarende antal nye Aktier, som Joint Global Coordinators vil tegne og tilbagelevere til Novo Holdings.

Udbuddet består af: 1) børsnotering og et offentligt udbud til private og institutionelle investorer i Danmark og 2) en privatplacering til institutionelle investorer i visse andre jurisdiktioner (ekskl. USA). Udbuddet uden for USA foretages i overensstemmelse med Regulation S i U.S. Securities Act.

Skandinaviska Enskilda Banken, Danmark, filial af Skandinaviska Enskilda Banken AB (Publ.), Sverige; Vækstfonden; BI Asset Management Fondsmæglerselskab A/S på vegne af visse kunder; Handelsbanken, filial af Svenska Handelsbanken AB (Publ.), Sverige; og Spar Nord Bank A/S har i forbindelse med Udbuddet på visse betingelser forpligtet sig til som Cornerstone-Investorer at tegne Udbudte Aktier for et samlet tegningsbeløb på DKK 230 mio., svarende til ca. 38,3% af Udbuddet (ekskl. Overallokeringsretten).

Udbudskursintervallet forventes at udgøre mellem DKK 64 og DKK 80 pr. Udbudt Aktie og vil blive fastsat ved en bookbuilding-proces. Udbudskursen og det præcise antal Udbudte Aktier, der skal sælges, fastsættes af Bestyrelsen i samråd med Joint Global Coordinators og forventes offentliggjort via Nasdaq Copenhagen senest den 17. november 2017 kl. 8.00 (dansk tid).

Udbudskursintervallet kan blive justeret i løbet af bookbuilding-processen. Hvis Udbudskursintervallet justeres, vil Selskabet meddele dette via Nasdaq Copenhagen og offentliggøre et tillæg til dette Prospekt. Efter offentliggørelsen af et sådant tillæg har investorer, der har indleveret købsordrer på Udbudte Aktier i Udbuddet, to handelsdage til at tilbagekalde deres købsordre. I dette tilfælde vil meddelelsen om Udbudskursen først blive offentliggjort, når fristen for udnyttelse af retten til tilbagekaldelse er udløbet. Udbudskursen kan således ligge uden for Udbudskursintervallet.

Udbudsperioden løber fra den 6. november 2017 til senest den 16. november 2017 kl. 12.00 (dansk tid). Udbudsperioden kan lukkes før den 16. november, men hel eller delvis lukning af Udbudsperioden vil dog tidligst finde sted den 15. november 2017 kl. 00.01 (dansk tid). Hvis Udbuddet lukkes før den 16. november 2017, kan meddelelsen om Udbudskursen, tildeling og Optagelse blive fremrykket tilsvarende. Udbudsperioden for købsordrer for beløb til og med DKK 3 mio. kan lukkes før resten af Udbuddet. En sådan tidligere hel eller delvis lukning offentliggøres i givet fald via Nasdaq Copenhagen.

Der skal som minimum tegnes 1 stk. Udbudt Aktie. Der gælder intet maksimalt tegningsbeløb i Udbuddet. Antallet af aktier begrænses dog til antallet af Udbudte Aktier i Udbuddet.

Ordrer fra danske investorer om tegning for beløb til og med DKK 3 mio. skal afgives på den ordreblanket, der er indeholdt i Prospektet. Ordreblanketten skal indsendes til investors eget kontoførende institut i løbet af Udbudsperioden eller en eventuelt kortere periode, der måtte blive offentliggjort via Nasdaq Copenhagen. Ordrer er bindende og kan ikke ændres eller annulleres. Ordrer kan afgives med en maksimumkurs i DKK pr. Udbudt Aktie. Hvis Udbudskursen overstiger maksimumkursen pr. Udbudt

Aktie, der er anført på ordreblanketten, vil der ikke blive tildelt Udbudte Aktier til investor. Hvis der ikke er angivet en maksimumkurs, anses ordren for afgivet til Udbudskursen. Alle ordrer, der er afgivet til en kurs lig med Udbudskursen eller en højere kurs, afregnes til Udbudskursen efter eventuel tildeling. Ordrer skal afgives for et antal Udbudte Aktier eller for et samlet beløb afrundet til nærmeste kronebeløb. Der kan kun indleveres én ordreblanket for hver VP-konto. For at en ordre er bindende, skal den udfyldte og underskrevne ordreblanket indsendes til investors eget kontoførende institut i så god tid, at det kontoførende institut kan behandle og fremsende ordren, således at den modtages af Danske Bank A/S senest den 16. november 2017 kl. 12.00 (dansk tid) eller på det eventuelle tidligere tidspunkt, hvor Udbuddet lukkes.

Investorer, som ønsker at afgive ordrer på tegning for beløb over DKK 3 mio., kan afgive interessetilkendegivelse til en eller flere af Emissionsbankerne i løbet af Udbudsperioden. Disse investorer kan i Udbudsperioden løbende ændre eller tilbagekalde deres interessetilkendegivelser, men disse interessetilkendegivelser bliver bindende ordrer ved udløbet af Udbudsperioden. Umiddelbart efter fastsættelsen af Udbudskursen vil investorerne få tildelt et antal Midlertidige Købsbeviser repræsenterende Udbudte Aktier til Udbudskursen inden for rammerne af investors sidst afgivne eller justerede interessetilkendegivelse. Alle ordrer, der er afgivet til en kurs lig med Udbudskursen eller en højere kurs, afregnes til Udbudskursen efter eventuel tildeling.

Hvis de samlede antal Aktier, der er afgivet ordrer på i Udbuddet, overstiger antallet af Udbudte Aktier, vil der blive foretaget reduktion på følgende måde:

- Ved ordrer med en kursværdi til og med DKK 3 mio. foretages matematisk reduktion.
- Ved ordrer med en kursværdi på mere end DKK 3 mio. sker der individuel tildeling. Joint Global Coordinators vil tildele de Udbudte Aktier efter aftale herom med Bestyrelsen.
- Op til 3.593.750 stk. Udbudte Aktier vil blive reserveret til Cornerstone-Investorerne til tegning til Udbudskursen i forbindelse med Udbuddet.
- Op til 93.750 stk. Udbudte Aktier vil blive reserveret til visse medlemmer af Bestyrelsen til tegning i forbindelse med Udbuddet til Udbudskursen.
- Op til 21.875 stk. Udbudte Aktier vil blive reserveret til Direktionen og Nøglemedarbejderne til tegning til Udbudskursen som en investering i forbindelse med Orphazymes langsigtede incitamentsprogram.
- Op til 93.750 stk. Udbudte Aktier vil blive reserveret til Orphazymes medarbejdere til tegning i forbindelse med Udbuddet til Udbudskursen.

Efter Udbudsperiodens udløb modtager investorerne normalt en opgørelse over det eventuelle antal Midlertidige Købsbeviser repræsenterende Udbudte Aktier, der er tildelt dem, og værdien heraf til Udbudskursen, medmindre andet aftales mellem investor og det pågældende kontoførende institut.

Betaling for og afvikling af de Udbudte Aktier forventes at finde sted på Afviklingsdatoen i form af elektronisk levering af Midlertidige Købsbeviser mod kontant betaling i danske kroner til investorernes konti hos VP Securities og gennem Euroclear og Clearstream.

Betinget af gennemførelse af Udbuddet og registrering af de nye Udbudte Aktier i Erhvervsstyrelsen vil de Midlertidige Købsbeviser automatisk blive ombyttet i VP Securities til et tilsvarende antal Aktier, der forventes leveret elektronisk to hverdage efter Afviklingsdatoen til indehaveren af de Midlertidige Købsbevisers konto hos VP Securities og gennem Euroclear og Clearstream. Hvis Udbuddet lukkes før den 16. november 2017, kan Afviklingsdatoen, levering af de Midlertidige Købsbeviser, automatisk ombytning af Midlertidige Købsbeviser til Aktier og Aktiernes første handels- og officielle noteringsdag på Nasdaq Copenhagen blive fremrykket tilsvarende. Al handel med Midlertidige Købsbeviser og/eller de Udbudte Aktier forud for afvikling af Udbuddet sker for de involverede parters egen regning og risiko.

E.4 Væsentlige interesser i Udbuddet, herunder interessekonflikter

Visse medlemmer af Bestyrelsen samt Direktionen og Nøglemedarbejderne er, eller vil efter udnyttelsen af Pre-IPO Warrants blive, indirekte aktionærer i Selskabet eller har økonomiske interesser deri, hvorved de har interesser i Udbuddet. Der er dog ingen medlemmer af Bestyrelsen eller Direktionen eller Nøglemedarbejderne, der direkte eller indirekte ejer mere end 5% af Selskabets aktiekapital.

Direktionen, Nøglemedarbejderne og visse medlemmer af Bestyrelsen ejer Pre-IPO Warrants, som Selskabet har tildelt som led i Pre-IPO warrantprogrammet. Disse Pre-IPO Warrants kan udnyttes i forbindelse med Udbuddet, og dermed har Direktionen, Nøglemedarbejderne og visse medlemmer af Bestyrelsen interesser i Udbuddet.

Skandinaviska Enskilda Banken, Danmark, filial af Skandinaviska Enskilda Banken AB (Publ.), Sverige; Vækstfonden; BI Asset Management Fondsmæglerselskab A/S på vegne af visse kunder; Handelsbanken, filial af Svenska Handelsbanken AB (Publ.), Sverige; og Spar Nord Bank A/S har i forbindelse med Udbuddet med forbehold for visse betingelser forpligtet sig til at tegne Udbudte Aktier som Cornerstone-Investorer og vil således have en direkte økonomisk interesse i Udbuddet.

Emissionsbankerne og deres respektive tilknyttede virksomheder har været involveret i transaktioner med og leveret forskellige ydelser relateret til forretningsbankvirksomhed, investeringsbankvirksomhed, finansiel rådgivning og andre ydelser Orphazyme, og Emissionsbankerne og deres respektive tilknyttede virksomheder leverer i øjeblikket og kan i fremtiden levere sådanne ydelser til Orphazyme. For visse af disse transaktioner og ydelser gælder det, at deling af information er underlagt restriktioner af hensyn til fortrolighed, interne procedurer eller gældende regler og forskrifter. Emissionsbankerne har modtaget og vil modtage sædvanligt honorar og provision for disse transaktioner og ydelser og vil muligvis få interesser, der ikke er forenelige med eller potentielt kunne være i modstrid med potentielle investorers og Selskabets interesser. Danske Bank A/S forventes også at fungere som långiver i henhold til en lånefacilitet til Direktionen og Nøglemedarbejderne vedrørende deres tegning af Investeringsaktier (som defineret i dette Prospekt) i forbindelse med Orphazymes langsigtede incitamentsprogram.

E.5 Sælgende Aktionærer og lockup-aftaler

Sælgende Aktionærer
Udbuddet består af nye Aktier udbudt af Selskabet. Ingen af Selskabets eksisterende aktionærer udbyder Aktier i forbindelse med Udbuddet.

Lockup-Aftaler
Hovedaktionærerne har indgået aftale med Emissionsbankerne om, at de, bortset fra som beskrevet nedenfor, i en periode, der begynder pr. datoen for dette Prospekt og slutter på det tidligste tidspunkt af 1) datoen for Selskabets offentliggørelse af resultaterne fra det igangværende fase II/III-forsøg i NPC, som i øjeblikket ventes i 3. kvartal 2018 (men tidligst 180 dage efter Optagelsen), eller 2) 360 dage efter Optagelsen, ikke uden Emissionsbankernes forudgående skriftlige samtykke vil: 1) udbyde, pantsætte, sælge, indgå aftale om at sælge, sælge nogen option eller indgå aftale om at købe, købe nogen option eller indgå aftale om at sælge, tildele nogen option, ret eller warrant til at købe, udlåne, få Selskabet til at udstede Aktier, eller på anden måde, direkte eller indirekte, overdrage eller afhænde (eller offentliggøre en sådan disposition) nogen af deres Aktier på tidspunktet for Optagelsen (ekskl. Udbudte Aktier tegnet i forbindelse med Udbuddet) (**"Lockup-aktier"**), eller nogen værdipapirer, der kan konverteres til, udnyttes til eller ombyttes til sådanne Lockup-aktier, 2) indgå nogen swap eller anden disposition, der helt eller delvist overdrager nogen af de økonomiske konsekvenser i forbindelse med ejerskab af Lockup-aktier, uanset om sådanne transaktioner beskrevet under pkt. 1) eller 2) afregnes ved levering af sådanne Lockup-aktier eller sådanne andre værdipapirer, kontant eller på anden måde, eller 3) fremsætte begæring om generalforsamling i Selskabet, eller indkalde eller tage skridt til at indkalde til generalforsamling med henblik på at stille forslag om at bemyndige Selskabet til at udstede Aktier eller warrants til tegning af Aktier eller offentliggøre hensigt om at indgå i nogen af foranstående transaktioner.

Selskabet har indgået aftale med Emissionsbankerne om stort set samme begrænsninger som anført ovenfor i en periode på 180 dage fra Optagelsen med visse undtagelser.

Medlemmerne af Bestyrelsen på tidspunktet for Optagelsen (med undtagelse af de Nye Bestyrelsesmedlemmer), Direktionen og Nøglemedarbejderne har indgået aftale med Emissionsbankerne om, at de i en periode på 360 dage fra Optagelsen for så vidt angår Aktier ejet på tidspunktet for Optagelsen (og eventuelle Aktier tegnet som resultat af udnyttelsen af Pre-IPO Warrants, hvis relevant) vil være underlagt stort set tilsvarende begrænsninger som Selskabet og Hovedaktionærerne som anført ovenfor.

Derudover har alle Selskabets eksisterende aktionærer pr. datoen for dette Prospekt påtaget sig en forpligtelse til ikke at handle med eksisterende Aktier, indtil de Midlertidige Købsbeviser er ombyttet til Aktier, og Aktierne er optaget til handel og officiel notering på Nasdaq Copenhagen.

E.6 Beløb og procentdel for umiddelbar udvanding som følge af Udbuddet

De eksisterende Aktier, som udstedes pr. prospektdatoen, vil blive udvandet i forbindelse med Udbuddet af udstedelsen af op til 10.781.250 stk. nye Aktier, svarende til en nominel værdi på DKK 10.781.250, under forudsætning af fuld udnyttelse af Overallokeringsretten. Efter Udbuddets gennemførelse udgør de eksisterende Aktier, der er udstedt og udestående, 57,9% af Selskabets aktiekapital, under forudsætning af fuld tegning af alle nye Aktier i forbindelse med Udbuddet (herunder fuld

udnyttelse af Overallokeringsretten) samt udnyttelse af alle Pre-IPO Warrants, der optjenes i forbindelse med Udbuddet, samt justeret for Kapitalomstruktureringen 2017 (forudsat at Udbudskursen svarer til midtpunktet i Udbudskursintervallet og dermed en udstedelse af 720.875 stk. bonusaktier forud for Optagelsen som led i Kapitalomstruktureringen 2017). Selskabets nettokapital pr. 30. september 2017 udgjorde DKK 94.500.000, svarende til en indre værdi pr. Aktie på DKK 18,5, svarende til DKK 8,2 pr. Aktie baseret på Selskabets kapitalstruktur pr. prospektdatoen. Indre værdi pr. Aktie beregnes ved at dividere den samlede nettokapital med det samlede antal udstedte Aktier. Baseret på Selskabets nettokapital pr. 30. september 2017 og de samme forudsætninger som beskrevet ovenfor og justeret for de anslåede omkostninger i forbindelse med Udbuddet ville Selskabets indre værdi pr. Aktie ved Udbuddets gennemførelse udgøre DKK 32,1.

E.7 Anslåede udgifter, som investor pålægges af Selskabet eller de Sælgende Aktionærer

Ikke relevant. Hverken Selskabet eller Emissionsbankerne vil pålægge investorerne omkostninger. Investorerne skal betale sædvanlige transaktions- og ekspeditionsgebyrer til deres kontoførende institutter.

English Summary

Summaries are made up of disclosure requirements known as "Elements". These Elements are numbered in Sections A—E (A.1—E.7). This summary contains all the Elements required to be included in a summary for this type of security and issuer under the Prospectus Regulation no. 486/2012, as amended. Because some Elements are not required to be addressed, there may be gaps in the numbering sequence of the Elements. Even though an Element may be required to be inserted in the summary because of the type of security and issuer, it is possible that no relevant information can be given regarding the Element. In this case, a short description of the Element is included in the summary with the mention of "not applicable".

Section A – Introduction and Warnings

A.1 Warning to investors

This summary should be read as an introduction to this Offering Circular.

Any decision to invest in the Offer Shares should be based on consideration of the Offering Circular as a whole by the investor.

Where a claim relating to the information contained in the Offering Circular is brought before a court, under the national legislation of the European Economic Area member states, the plaintiff investor might have to bear the costs of translating this Offering Circular before the legal proceedings are initiated.

Civil liability attaches only to those persons who have tabled the summary, including any translation thereof, but only if this summary is misleading, inaccurate or inconsistent when read together with the other parts of the Offering Circular or it does not provide, when read together with the other parts of the Offering Circular, key information in order to aid investors when considering whether to invest in the Offer Shares.

A.2 Consent for intermediaries

Not applicable. No agreement has been made in regard to the use of the Offering Circular in connection with a subsequent resale or final placement of the Offer Shares. Accordingly, the Company does not consent to the use of the Offering Circular for subsequent resale or final placement of securities by financial intermediaries.

Section B – Issuer

B.1 Legal and commercial name

The Company is registered with the legal name Orphazyme A/S and does not have any secondary names.

B.2 Domicile, legal form, country of incorporation

The Company has its registered office at Ole Maaløes Vej 3, DK-2200 Copenhagen N in the municipality of Copenhagen, Denmark and was incorporated in Denmark as a private limited liability company under the laws of Denmark on 19 June 2009 and later converted into a Danish public limited liability company on 20 October 2017.

B.3 Current operations and principal activities

Orphazyme is a Danish biotech company with a late stage orphan drug pipeline, developing new treatment options for orphan protein misfolding diseases. The Company's lead candidate, arimoclomol, stimulates the production of the cells' protein rescue system, the heat shock proteins ("**HSPs**"), which helps the misfolded proteins to either get back to their functional shape or, if this cannot be achieved, removing them from the cells by means of the cells' recycling system, the lysosomes, so they no longer form toxic aggregates.

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, and arimoclomol has been well-tolerated in seven phase I trials and three phase II trials with tolerability comparable to that of a placebo. Arimoclomol is orally available, readily crosses the blood brain barrier, has a bioavailability of 80-90%, and a clinical proof-of-concept, based on directional benefit across independent efficacy endpoints, has been achieved in the neuromuscular diseases and pre-clinical data supporting the advancement of arimoclomol in the lysosomal storage diseases.

Currently, the Company expects to have completed three potential registration studies by the end of 2020 with the first potential marketing authorisation expected in 2020.

B.4a Description of the most significant recent trends affecting the Company and the industries in which it operates

Orphazyme aims to introduce novel therapies for the treatment of serious and debilitating orphan diseases initially within the field of protein misfolding diseases. The neuromuscular and lysosomal storage diseases progressively deprive patients of basic abilities, and in many cases, result in premature death.

Orphan diseases are rare conditions that affect a small part of the population. Many orphan diseases have no, or only few, treatment options, and there is therefore high need for new therapies to be developed. Orphazyme is developing medicine with the potential to improve the lives of those suffering from such diseases with limited or no availability of alternative treatment options.

To encourage and incentivise the research and development of pharmaceutical products for rare diseases, despite the small number of patients and the significant development costs, the Orphan Drug Act was passed in the United States in 1983 to avoid neglect of such diseases. The success of the Orphan Drug Act led to the implementation of orphan drug regimes in other key markets as well, for example Japan in 1993 and the EU in 2000. Under the orphan drug regimes, authorities can grant a pharmaceutical product a so-called orphan drug designation. Developers of drugs with an orphan drug designation are entitled to several advantages, including the possibility of free of charge advice from the FDA and EMA and certain financial benefits, such as R&D tax credits and exemptions or reductions in regulatory submission fees. If a drug candidate with orphan drug designation is approved by the regulatory authorities or authorised bodies upon completion of clinical trials, the respective authority can decide whether the pharmaceutical product should receive orphan drug status. Orphan drug status provides the orphan drug with market exclusivity for seven and 10 years in the United States and the EU, respectively[4].

As orphan drugs target rare, severely disabling and/or life-threatening diseases with small patient populations, such drugs are usually priced at a premium to non-orphan drugs. In 2016, the average annual cost per patient in the United States was approximately five times higher for orphan drugs compared to non-orphan drugs[5]. Many factors affect the pricing of orphan drugs and non-orphan drugs, such as the burden of disease, cost avoidance by society, extent of unmet need, clinical efficacy, side effects, tolerability, number of patients affected by the disease, the cost of development and manufacturing as well as negotiation with payers/insurers.

In 2016, worldwide orphan drug sales increased by 12.2% year-on-year and reached USD 114 billion, whereas non-orphan drugs (excluding generics) grew by 2.4% in the same period, amounting to USD 578 billion. The total prescription drug sales (excluding generics) grew by 3.9% in 2016 and amounted to a total of USD 692 billion. The orphan drug market is estimated to grow to USD 209 billion by 2022, corresponding to a cumulative average growth rate ("**CAGR**") of 11.1% in the period 2017-2022, almost double the growth of the overall prescription pharmaceutical product market during the same period[6].

B.5 Description of the Group and the Company's position within the Group

Not applicable. The Company is not part of a group structure.

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[4] EMA European Medicines Agency, Marketing authorisation and market exclusivity, http://www.ema.europa.eu/ema/index.jsp?curl=pages/regulation/general/general_content_000392.jsp& and U.S. Food & Drug Administration, Code of Federal Regulations Title 21, https://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfcfr/cfrsearch.cfm?cfrpart=316&showfr=1&source=govdelivery&subpartnode=21%3A5.0.1.1.6.4&utm_medium=email&utm_source=govdelivery

[5] Orphan Drug Report 2015, Evaluate Pharma; Orphan Drug Report 2017, Evaluate Pharma

[6] Orphan Drug Report 2017, Evaluate Pharma

B.6 Persons who, directly or indirectly, have an interest in the issuer's capital or voting rights or have control over the Company

Upon completion of the 2017 Capital Structure Adjustment (assuming an Offer Price at the top end of the Offer Price Range) and prior to Admission, the Company's Shares will be held by Novo Holdings (33.1%); Aescap Venture (15.2%); Sunstone Capital (15.6%); Orpha Pooling B.V. (a joint venture between LSP and ALS Invest) (21.0%); Idinvest (7.2%); and Kurma Biofund II (6.6%) (together referred to as the "**Main Shareholders**") as well as UCL Business PLC (0.1%) and Kansas Life Sciences Development Inc. (0.1%). The remaining Shares (1.1%) are indirectly held by the founders of the Company (CSO Thomas Kirkegaard Jensen; CEO Anders Hinsby; Professor Marja Helena Jaattela; and member of the Existing Board of Directors (as defined herein), and co-founder Martin Rahbek Kornum) through their respective wholly-owned holding companies.

In case the Offer Price is set below the top end of the Offer Price Range, an additional number of bonus Shares will be issued shortly before Admission in favour of the former preference shareholders in order to account for the Offer Price as part of the 2017 Capital Structure Adjustment. Assuming an Offer Price at the bottom end of the Offer Price Range, up to 1,621,968 bonus Shares with a nominal value of DKK 1 each will be issued. Consequently, the ownership stakes set out above may be adjusted accordingly.

A total of 1,293,293 Pre-IPO Warrants (as defined herein) may be exercised (or lapse if not exercised) in connection with the Offering. The maximum nominal capital increase resulting from the exercise of the Pre-IPO Warrants is limited to DKK 866,965. The capital increase related to exercise of Pre-IPO Warrants, if any, is expected to take place in connection with settlement of the Offering, i.e. on or around the Settlement Date being 21 November 2017.

All Shares have the same rights and rank pari passu in respect of, inter alia, voting rights.

Other than set out above, the Company is not aware that any person directly or indirectly owns an interest in the Company's share capital or voting rights that would be notifiable under Danish law.

B.7 Selected financial and business information

The following tables present summary financial data for Orphazyme's business. The Company has derived the summary statement of profit or loss data for the years ended 31 December 2016 and 2015 from the statutory Audited Financial Statements (as defined herein) for the period 1 January – 31 December 2016 and reviewed unaudited 2015 Comparative Financial Statements (as defined herein) for the period 1 January – 31 December 2015, included on pages F-17 elsewhere in this Offering Circular. The Company has derived the summary statement of profit or loss data for the six months ended 30 June 2017 and 2016 and the summary statement of financial position data as of 30 June 2017 from the unaudited condensed Interim Financial Statements, included on pages F-5 - F-6 elsewhere in this Offering Circular.

Orphazyme maintains its books and records in DKK and prepares its audited financial statements in accordance with International Financial Reporting Standards ("**IFRS**"), issued by the International Accounting Standards Board ("**IASB**") and adopted by the European Union and additional requirements in the Danish Financial Statements Act. You should read this data together with our financial statements and related notes appearing elsewhere in this Offering Circular.

Statement of profit or loss

	Six months period ended 30 June		Twelve months ended 31 December	
(TDKK)	**H1 2017**	**H1 2016**	**2016**	**2015**
Research and development expenses	(46,870)	(27,875)	(55,817)	(45,865)
Administrative expenses	(7,972)	(2,696)	(7,703)	(7,220)
Operating loss	**(54,842)**	**(30,571)**	**(63,520)**	**(53,085)**
Net financials	(122)	35	85	(317)
Loss before tax	**(54,964)**	**(30,536)**	**(63,435)**	**(53,402)**
Income tax benefit	2,841	2,750	5,500	5,688
Net loss for the period	**(52,123)**	**(27,786)**	**(57,935)**	**(47,714)**

Statement of financial position data

(TDKK)	As of 30 June 2017	As of 31 December 2016	As of 31 December 2015
Assets			
Non-current assets			
Property, plant and equipment	1,007	987	1,487
Corporation tax receivable	5,591	2,750	2,750
Deposits	357	310	211
Total non-current assets	**6,955**	**4,047**	**4,448**
Current assets			
Corporation tax receivable	5,500	5,500	5,875
Receivable capital increase	91,319	–	–
Other receivables	5,104	3,421	644
Prepayments	3,245	4,624	5,970
Cash and cash equivalents	33,589	14,349	68,015
Total current assets	138,757	27,894	80,504
Total assets	**145,712**	**31,941**	**84,952**
Equity and liabilities			
Equity			
Total equity	**121,297**	**17,509**	**74,143**
Current liabilities			
Trade payables	7,614	4,718	2,447
Payables to shareholders	223	–	–
Other payables	16,578	9,714	8,362
Total current liabilities	24,415	14,432	10,809
Total equity and liabilities	**145,712**	**31,941**	**84,952**

Statement of cash flow data

	Six months period ended 30 June		Twelve months ended 31 December	
(TDKK)	**H1 2017**	**H1 2016**	**2016**	**2015**
Operating activities				
Net loss before tax	(54,842)	(30,571)	(63,435)	(53,402)
Adjustments to reconcile loss before tax to cash flows from operating activities				
Share-based expense	–	–	–	–
Depreciation and write-down	291	287	706	554
Gain/loss on sale and disposal of assets	–	–	33	–
Change in other receivables	(1,730)	(1,032)	(2,876)	(277)
Change in prepayments	1,379	1,296	1,347	(4,969)
Change in trade payables	2,896	1,134	2,271	858
Change in other payables	6,821	2,284	1,352	4,330
Cash flows from taxes	–	–	5,875	6,250
Interest paid, net	(122)	35	–	241
Net cash used in operating activities	**(45,307)**	**(26,567)**	**(54,727)**	**(46,414)**
Investing activities				
Investment in property, plant and equipment	(311)	–	(238)	(495)
Net cash used in investing activities	**(311)**	**–**	**(238)**	**(495)**
Financing activities				
Capital contributions from shareholders	65,431	–	1,330	85,970
Cash from convertible loan	–	–	–	–
Bank loans	–	463	–	–
Expenses related to capital contributions	(573)	–	(30)	(215)
Net cash provided by financing activities	**64,858**	**463**	**1,300**	**85,755**
Net change in cash and cash equivalents	**19,240**	**(26,104)**	**(53,665)**	**38,846**
Cash and cash equivalents at the beginning of the period	14,349	68,014	68,014	29,169
Cash and cash equivalents at the end of the period	**33,589**	**41,910**	**14,349**	**68,015**

Comparison of six months period ended 30 June 2017 and 30 June 2016

Results of operations

Research and development costs totaled DKK 46.9 million in the six months period ended 30 June 2017 compared to DKK 27.9 million in the six months period ended 30 June 2016. The increase was mainly due to initiation of the NPC phase II/III trial in mid 2016 and the costs for the preparation of a phase II trials in India for Gaucher disease. Furthermore, the period is impacted by cost of manufacturing arimoclomol for the sIBM phase III trial.

Administrative expenses were DKK 8.0 million in the six months period ended 30 June 2017 compared to DKK 2.7 million in the six months period ended 30 June 2016. The increase is mainly due to costs for lawyers, auditors and consultants for preparing the initial public offering ("**IPO**"), as well as the hiring of new members of management and additional staff. Part of the costs directly associated with the IPO and the expected issuance of new Shares is recognised within prepayments until the Offering takes place. As at 30 June 2017, the Company has in total recognised prepayments related to the IPO of DKK 1.4 million.

As of 30 June 2017, Orphazyme had cash and cash equivalents of DKK 33.6 million compared with cash and cash equivalents of DKK 14.3 million as of 31 December 2016. The increase reflects the capital increase of DKK 65.4 million in the six months period ended 30 June 2017 and is partly offset by the costs associated with Orphazyme's business activities, including costs of current and planned clinical trials. In the six months period ended 30 June 2017, operating activities has net cash outflow of DKK 45.3 million, compared to DKK 26.6 million in the six months period ended 30 June 2016. The Company has increased cash flow from financing activities through capital increases from both current and new investors.

Comparison of results for the years ended 31 December 2016 and 2015

Research and development costs totaled DKK 55.8 million in 2016 compared to DKK 45.9 million in 2015. The increase was mainly due to the initiation of the NPC phase II/III trial in mid 2016 and cost of new employees to support the increase in the level of activity.

Administrative expenses were DKK 7.7 million in 2016 compared to DKK 7.2 million in 2015. The increase from 2015 to 2016 was due to the hiring of new members of management at the end of 2016.

As of 31 December 2016, Orphazyme had cash and cash equivalents of DKK 14.3 million compared with cash and cash equivalents of DKK 68.0 million as of 31 December 2015. The decrease reflects the costs associated with Orphazyme's business activities, including costs of current and planned clinical trials.

B.8 Selected key pro forma financial information

Not relevant. No pro forma financial information is presented in the Offering Circular as there have not been any transactions that result in a significant (defined as more than 25%) gross change in relevant indicators such as total assets, net revenue or net profit.

B.9 Profit forecast or estimate

The Company expects a net loss for 2017 of DKK 125 million - DKK 135 million. Orphazyme has no revenue and costs are primarily related to research and development. Income tax benefit and income from public grants have a positive effect on the net result for the year. Orphazyme's result of operations for 2017 could deviate materially from this forecast.

B.10 Qualifications in the audit report on the historical financial information

Not applicable. The audit reports on the historical financial information included in the Offering Circular have been issued without qualifications.

B.11 Explanation if the issuer's working capital is not sufficient for the Company's present requirements

In the opinion of Orphazyme, the working capital available as of the date of this Offering Circular is not sufficient for its present working capital needs for the twelve months following the date of this Offering Circular. As further described in "*Background to the Offering and Use of Proceeds*", Orphazyme intends to finance its operations for the twelve months following the date of this Offering Circular and the date of Admission using part of the proceeds from the Offering. In case the Offering is not completed, Orphazyme will seek alternative methods of finance in cooperation with its existing shareholders.

Section C – Securities

C.1 A description of the type and the class of the Offer Shares, including any security identification number

The Shares are not divided into share classes.
Permanent ISIN for the Shares: DK0060910917
Temporary ISIN for the Temporary Purchase Certificates: DK0060911055

C.2 Currency of the Offer Shares

The Offer Shares will be denominated in Danish kroner ("**DKK**").

C.3 The number of Shares issued and fully paid and issued but not fully paid

Before completion of the Offering, the Company's share capital will have a nominal value of DKK 12,310,967, divided into 12,310,967 Shares of DKK 1 each or multiples thereof, assuming an Offer Price at the midpoint of the Offer Price Range (and thereby an issue of 720,875 bonus Shares prior to Admission as part of the 2017 Capital Structure Adjustment), which will all be issued and fully paid up.

C.4 A description of the rights attached to the Shares

All Shares, including the Offer Shares, have the same rights and the Offer Shares will rank pari passu with all other Shares in the Company in respect of voting rights, preemption rights, redemption, conversion and restrictions or limitations according to the articles of association of the Company (the "**Articles of Association**") or eligibility to receive dividends or proceeds in the event of dissolution and liquidation. No Shares carry special rights, restrictions or limitations pursuant to the Company's Articles of Association.

Each Share with a nominal value of DKK 1 gives the holder the right to one vote at the Company's general meetings and to receive distributed dividends.

Every shareholder is entitled to have specific business transacted at the general meeting, provided that the shareholder submits a written request to that effect to the Board of Directors not later than six weeks before the date of the general meeting.

C.5 A description of any restrictions on the free transferability of the Shares

Not applicable. The Shares are negotiable instruments and no restrictions under the Company's Articles of Association or Danish law apply to the transferability of the Shares.

C.6 Admission to trading on a regulated market

Application has been made for the Temporary Purchase Certificates to be admitted to trading on Nasdaq Copenhagen under the symbol "ORPHA TEMP" and for the Shares to be admitted to trading and official listing on Nasdaq Copenhagen under the symbol "ORPHA". The Admission is subject to, among other things, Nasdaq Copenhagen's approval of the distribution of the Offer Shares, the election of the New Board Members (as defined herein), the Offering not being withdrawn prior to the settlement of the Offering and the Company making an announcement to that effect.

If the Offering is closed before 16 November 2017, the Admission, the Settlement Date, the delivery of Temporary Purchase Certificates, the automatic exchange of Temporary Purchase Certificates for Shares and the first day of trading and official listing of the Shares on Nasdaq Copenhagen may be moved forward accordingly.

Payment for and settlement of the Offer Shares are expected to take place on or around 21 November 2017 by way of delivery of Temporary Purchase Certificates. Subject to completion of the Offering and registration of the Offer Shares with the Danish Business Authority, the Temporary Purchase Certificates will automatically be exchanged in VP Securities for a corresponding number of Shares, which are expected to be delivered on 23 November 2017. The first day of trading and official listing on Nasdaq Copenhagen is expected to be 21 November 2017 subject to the Offering not being withdrawn prior to settlement and

completion of the Offering. The first day of trading of the Temporary Purchase Certificates on Nasdaq Copenhagen is expected to be 17 November 2017 and the last day of trading of the Temporary Purchase Certificates on Nasdaq Copenhagen is expected to be 21 November 2017. The first day of trading of the Shares on Nasdaq Copenhagen under the permanent ISIN is expected to be 22 November 2017. In connection with the Temporary Purchase Certificates being automatically exchanged for Shares, the Temporary Purchase Certificates will cease to exist.

C.7 A description of dividend policy

The Company has not declared or made any dividend payments for the last two financial years. Currently, the Company intends to use all available financial resources as well as revenue, if any, for purposes of the Company's current and future business. As of the date hereof, the Company does not expect to make dividend payments within the foreseeable future.

Section D – Risks

D.1 Key information on the key risks that are specific to the Company or its industry

The risks and uncertainties discussed below are those that Orphazyme's management currently views as material, but these risks and uncertainties are not the only ones that it faces. Additional risks and uncertainties, including risks that are not known to Orphazyme at present or that its management currently deems immaterial, may also arise or become material in the future, which could lead to a decline in the value of the Offer Shares and a loss of part or all of your investment. The following risk factors are not listed in any particular order of priority as to significance or probability.

- Orphazyme may be adversely affected by competition from other life sciences companies developing other treatments for similar diseases to those targeted by Orphazyme's products in development.
- Changes in the regulatory and compliance environment may have a significant adverse impact on the Company.
- The pricing and demand for pharmaceutical products may be affected by global economic factors. Orphazyme's ability to determine prices and thus generate revenue from any products that it may develop will depend on enacted and future reimbursement and drug pricing policies and regulations.
- The Company has no products approved for commercial sale, has never generated any revenue and may incur significant losses in the future, which makes it difficult to assess its future viability.
- Orphazyme is currently highly dependent on arimoclomol.
- Clinical trials being conducted to test the Company's products may not show the desired results or may be delayed or more be costly than anticipated.
- Orphazyme's products may cause undesirable side effects or have other properties that could delay or prevent their regulatory approval, limit the commercial profile, or result in significant negative consequences following regulatory approval, if any.
- Challenges in recruiting patients for clinical trials and concluding agreements with investigators and hospitals may have a material negative impact on the Company.
- Orphazyme is highly dependent on obtaining and maintaining required regulatory approvals.
- If the Company is unable to obtain orphan product designation or marketing exclusivity for its product candidates or is unable to benefit from the associated marketing exclusivity, such will have a material adverse effect on Orphazyme.
- Risks relating to manufacturing and research & development, including use of hazardous materials as well as chemistry and manufacturing controls ("**CMC**"), may have a material adverse effect on the Company.
- The Company is dependent on third-party vendors to provide certain licenses, products and services, clinical trials, and the Company's business and operations could be disrupted by any problems with its significant third-party vendors.
- Orphazyme is dependent on successful commercialization of its products.
- The Company may in the future seek to enter into collaborations with third parties for the development and commercialization of its products. If such collaborations are not successful, the Company may not be able to capitalize on the market potential of the products.
- Adverse events, product liability and other claims may have material adverse effects on Orphazyme.
- Changes in applicable laws and regulation in the jurisdictions in which the Company operates, or involvement in legal disputes, may have a material adverse effect on the Company's business, financial condition, results and prospects.

- The Company faces risks related to data privacy concerns, cyber security breaches and failure to comply with privacy regulations and security requirements relating to data.
- The Company may not be able to attract, integrate, manage and retain qualified personnel or key employees.
- Risks relating to priority review voucher.
- The Company's results of operations may be adversely affected by changes in foreign currency exchange rates and interest rates.
- The Company may require additional capital in the future, which may not be available to it on commercially favourable terms, or at all.
- The projected financial information included in this Offering Circular may differ materially from the Company's actual results, and investors should not place undue reliance on it.
- If Orphazyme is unable to obtain and maintain protection for relevant intellectual property rights, the value of Orphazyme's products will be significantly and adversely affected.
- Orphazyme may not be able to successfully enforce and defend its intellectual property rights.
- Orphazyme may face infringement claims and other challenges by third parties.
- The previous employers of Orphazyme's employees and consultants may attempt to assert rights over Orphazyme's intellectual property.
- The Company will not seek to protect its intellectual property rights in all jurisdictions throughout the world and may not be able to adequately enforce its intellectual property rights even in the jurisdictions where protection is sought.
- If Orphazyme is unable to protect the confidentiality of certain information, the value of its products and technology could be materially adversely affected.

D.3 Key information on the key risks relating to the Offer Shares

- Following the Offering, the Main Shareholders will continue to be large shareholders and may control or otherwise influence important actions the Company takes.
- There is limited free float in the Shares.
- The Company's Shares have not previously been publicly traded, and their price may be volatile and fluctuate.
- Differences in exchange rates may materially adversely affect the value of shareholdings or dividends paid.
- The Company intends to retain all available funds and any future earnings and reinvest such in the Company and, consequently, the shareholders' ability to achieve a return on their investment may depend on appreciation in the price of the Shares.
- If securities or industry analysts do not publish research or publish inaccurate or unfavorable research about the Company's business, the price of the Shares and their trading volume could decline.
- Future sales of Shares after the Offering may cause a decline in the market price of the Shares.
- The issuance of additional Shares in the Company to fund future operations, development and commercialisation of its products, or to fund acquisitions, any share incentive or share option plan or otherwise may dilute existing shareholdings.
- Shareholders in the United States and other non-Danish jurisdictions may not be able to participate in future equity offerings.
- Investors' rights as shareholders will be governed by Danish law and differ in some respects from the rights of shareholders under the laws of other countries.
- Transformation into a listed public company will increase the Company's costs and may disrupt the regular operations of its business.
- The Offering may be withdrawn after Admission to trading of the Temporary Purchase Certificates and until settlement of the Offering.

Section E – Offer

E.1 Total net proceeds of the Offer and estimated expenses

The net proceeds to the Company from the sale of new Shares to be issued by the Company pursuant to the Offering are expected to be approximately DKK 635 million (hereof DKK 85 million pursuant to the Overallotment Option), after deduction of commissions and estimated Offering expenses payable by the Company, with the assumptions set forth in this Offering Circular.

The total expenses in relation to the Offering, including commissions and fees (fixed and discretionary) payable by the Company to the Managers, other advisor fees and expenses, are estimated to be approximately DKK 55 million (assuming full exercise of the Overallotment Option).

Further, the Company has agreed to pay a selling commission to account-holding banks (unless such account-holding bank is a Manager) equivalent to 0.25% of the Offer Price of the Offer Shares that are allocated in respect of orders of up to and including DKK 3 million submitted through the account-holding banks (except for the Managers) to be paid proportionally by the Company based on the number of Offer Shares, respectively, that are sold).

E.2a Reasons for the Offer and use of proceeds, estimated net amount of the proceeds

The Offering is expected to support Orphazyme's operational strategy, advance Orphazyme's public and commercial profile, and provide Orphazyme with improved access to public capital markets and a diversified base of new Danish and international shareholders.

Orphazyme estimates that the net proceeds from the Offering will be approximately DKK 550 million, excluding the Overallotment Option. If the Joint Global Coordinators exercise the Overallotment Option in full, the Company estimates that the net proceeds to the Company from the Offering will be approximately DKK 635 million. These estimates are subject to the assumptions set forth in "*The Offering—Costs of the Offering*".

Orphazyme's reason for the Offering is to raise funds to support its business. The Company intends to allocate the net proceeds from the Offering, together with its existing cash resources as of 30 September 2017, in total of DKK 649 million, excluding the Overallotment Option, as follows:

- approximately DKK 135-155 million to fund the completion of a phase II/III trial with arimoclomol for the treatment of sIBM;
- approximately DKK 165-185 million to fund the completion of a phase II/III trial with arimoclomol for the treatment of ALS;
- approximately DKK 75-85 million to fund the completion of a phase II/III trial with arimoclomol for the treatment of NPC;
- approximately DKK 65-75 million to fund the completion of a phase II trial with arimoclomol for the treatment of Gaucher disease;
- approximately DKK 85-95 million to fund general research and development activities whereof approximately 50% is allocated to funding of research support for ongoing clinical trials and central lab oversight, approximately 40% to fund NDA enabling activities and approximately 10% to fund the advancement of discovery and lead projects into pre-clinical development; and
- the remainder amount, DKK 65-75 million, to fund preparing for registration and commercial activities, working capital, and for general corporate and administrative purposes, which may include the hiring of additional staff, capital expenditures, and the costs of operating as a public company.

In case the Overallotment Option (maximum DKK 85 million in net proceeds) is exercised, the proceeds will be used to further support the advancement of discovery and lead projects into preclinical development and registration and commercial activities.

In the event of positive results from the phase II/III trial with arimoclomol for the treatment of NPC, Orphazyme may choose to reallocate funds to facilitate filing and registration activities in arimoclomol for NPC, thus accelerating the use of proceeds from the Offering. In this scenario, the Company may need to raise additional funds, obtain debt financing or seek partnerships or other financing arrangements in order to have funds to complete the studies in the remaining indications simultaneously with pursuing the filing and registration activities as well as prepare for the later commercialisation. In case of negative results from the phase II/III trial with arimoclomol for the treatment of NPC, Orphazyme intends to use its remaining funds to pursue completion of the planned studies within the remaining three indications.

The foregoing expected use of the net proceeds from the Offering represents the Company's current intentions based upon present plans and business conditions. As of the date hereof, Orphazyme cannot predict with certainty all of the particulars of the use of the net proceeds of the Offering or the amounts that the Company will actually spend on the purposes set forth above. Actual expenditures may vary substantially from these estimates and the Company may find it necessary or advisable to reallocate the net proceeds within the above-described categories or to use portions thereof for other purposes. The amounts and timing of our actual use of net proceeds will vary based on numerous factors, including the relative success and cost of its research, pre-clinical and clinical development programs, and whether Orphazyme enters into collaborations and/or strategic partnerships with third parties.

E.3 Terms and conditions of the Offer

The Company is offering up to 10,781,250 new Shares (including the Overallotment Shares) in order to raise gross proceeds of up to DKK 690 million (hereof DKK 90 million pursuant to the Overallotment Option). Assuming completion of the Offering, the Company's registered share capital will increase by a nominal value of up to DKK 9,375,000 as a result of the issue of Offer Shares (excluding the Overallotment Option). The exact number of Offer Shares will be determined based on a book-building process.

The Company has granted the Joint Global Coordinators an Overallotment Option, on behalf of the Managers, to subscribe for up to 1,406,250 Overallotment Shares (new Shares), exercisable, in whole or in part, from the date of Admission until 30 calendar days thereafter, solely to cover overallotments or short positions, if any, incurred in connection with the Offering. For purposes of delivery of the Offer Shares to investors in connection with the Overallotment Option, Novo Holdings has agreed to make 1,406,250 existing Shares available. Orphazyme has agreed to issue up to a corresponding number of new Shares that the Joint Global Coordinators will subscribe for and redeliver to Novo Holdings.

The Offering consists of: (i) an initial public offering to retail and institutional investors in Denmark and (ii) private placements to institutional investors in certain other jurisdictions (excluding the United States). The Offering outside the United States will be made in compliance with Regulation S under the U.S. Securities Act.

Skandinaviska Enskilda Banken, Danmark, branch of Skandinaviska Enskilda Banken AB (Publ.), Sweden; Vækstfonden; BI Asset Management Fondsmæglerselskab A/S on behalf of certain clients; Handelsbanken, branch of Svenska Handelsbanken AB (Publ.), Sweden; and Spar Nord Bank A/S have in connection with the Offering subject to certain conditions undertaken to subscribe for Offer Shares as Cornerstone Investors for a total subscription amount of DKK 230 million, corresponding to approximately 38.3% of the Offering (excluding the Overallotment Option).

The Offer Price Range is expected to be between DKK 64 and DKK 80 per Offer Share and will be determined through a book-building process. The Offer Price and the exact number of Offer Shares to be sold will be determined by the Board of Directors in consultation with the Joint Global Coordinators, and is expected to be announced through Nasdaq Copenhagen no later than 8:00 a.m. (CET) on 17 November 2017.

The Offer Price Range may be adjusted during the book-building process. If the Offer Price Range is adjusted, the Company will make an announcement through Nasdaq Copenhagen and publish a supplement to this Offering Circular. Following publication of such supplement, investors who have submitted orders to purchase Offer Shares in the Offering will have two trading days to withdraw their purchase offer. In such an event, the announcement of the Offer Price will not be published until the period for exercising such withdrawal rights has ended. The Offer Price may thus be outside of the Offer Price Range.

The Offer Period will commence on 6 November 2017 and will close no later than 16 November 2017 at 12:00 p.m. (noon) (CET). The Offer Period may be closed prior to 16 November 2017; however, the Offer Period will not be closed in whole or in part before 15 November 2017 at 00:01 (CET). If the Offering is closed before 16 November 2017, the announcement of the Offer Price, allocation and the Admission may be moved forward accordingly. The Offer Period in respect of applications for purchases of amounts up to, and including, DKK 3 million may be closed before the remainder of the Offering is closed. Any such earlier closing, in whole or in part, will be announced through Nasdaq Copenhagen.

The minimum subscription amount is one Offer Share. No maximum subscription amount applies to the Offering. However, the number of shares is limited to the number of Offer Shares in the Offering.

Applications by Danish investors to subscribe for amounts of up to and including DKK 3 million should be made by submitting the application form enclosed in the Offering Circular to the investor's own account-holding bank during the

Offer Period or such shorter period as may be announced through Nasdaq Copenhagen. Applications are binding and cannot be altered or cancelled. Bids may be made at a maximum price per Offer Share in Danish kroner. If the Offer Price exceeds the maximum price per Offer Share stated in the application form, then no Offer Shares will be allocated to the investor. Where no maximum price per share has been indicated, applications will be deemed to be made at the Offer Price. All applications made at a price equivalent to the Offer Price, or a higher price, will be settled at the Offer Price following allotment, if any. Applications should be made for a number of Offer Shares or for an aggregate amount rounded to the nearest Danish kroner amount. Only one application will be accepted from each account in VP Securities. For binding orders, the application form must be submitted to the investor's own account-holding bank in complete and executed form in due time to allow the investor's own account-holding bank to process and forward the application to ensure that it is in the possession of Danske Bank A/S, no later than 12:00 p.m. (noon) (CET) on 16 November 2017, or such earlier time at which the Offering is closed.

Investors who wish to apply to subscribe for amounts of more than DKK 3 million can indicate their interest to one or more of the Managers during the Offer Period. During the Offer Period, such investors can continuously change or withdraw their declarations of interest, but these declarations of interest become binding applications at the end of the Offer Period. Immediately following the determination of the Offer Price, investors will be allocated a number of Temporary Purchase Certificates representing Offer Shares at the Offer Price within the limits of the investor's most recently submitted or adjusted declaration of interest. All applications made at a price equivalent to the Offer Price, or a higher price, will be settled at the Offer Price following allotment, if any.

In the event that the total amount of shares applied for in the Offering exceeds the number of Offer Shares, reductions will be made as follows:

- With respect to applications for amounts of up to and including DKK 3 million, reductions will be made mathematically.
- With respect to applications for amounts of more than DKK 3 million, individual allocations will be made. The Joint Global Coordinators will allocate the Offer Shares after agreement upon such allocations with the Board of Directors.
- Up to 3,593,750 Offer Shares will be reserved for the Cornerstone Investors to subscribe for at the Offer Price in connection with the Offering.
- Up to 93,750 Offer Shares will be reserved for certain members of the Board of Directors to subscribe for in connection with the Offering at the Offer Price.
- Up to 21,875 Offer Shares will be reserved for the Executive Management and Key Employees to subscribe for at the Offer Price as an investment in connection with Orphazyme's long-term incentive programme.
- Up to 93,750 Offer Shares will be reserved for Orphazyme's employees to subscribe for in connection with the Offering at the Offer Price.

Following the expiration of the Offer Period, investors will normally receive a statement indicating the number of Temporary Purchase Certificates representing Offer Shares allocated, if any, and the equivalent value at the Offer Price unless otherwise agreed between the investor and the relevant account-holding bank.

Payment for and settlement of the Offer Shares are expected to take place on the Settlement Date by way of delivery of Temporary Purchase Certificates against payment in immediately available funds in Danish kroner in book-entry form to investors' accounts with VP Securities and through the facilities of Euroclear and Clearstream.

Subject to completion of the Offering and registration of the new Offer Shares with the Danish Business Authority, the Temporary Purchase Certificates will automatically be exchanged in VP Securities for a corresponding number of Shares, which are expected to be delivered two business days after the Settlement Date in book-entry form to the holder of the Temporary Purchase Certificates' account with VP Securities and through the facilities of Euroclear and Clearstream. If the Offering is closed before 16 November 2017, the Settlement Date, the delivery of Temporary Purchase Certificates, the automatic exchange of Temporary Purchase Certificates for Shares and the first day of trading and official listing of the Shares on Nasdaq Copenhagen may be moved forward accordingly. All dealings in the Temporary Purchase Certificates and/or the Offer Shares prior to settlement of the Offering will be for the account of, and at the sole risk of, the parties involved.

E.4 Material interests in the Offer, including conflicts of interest

Certain members of the Board of Directors as well as the Executive Management and Key Employees are, or will upon exercise of Pre-IPO Warrants become, indirect shareholders in the Company or hold economic interests therein and therefore have an interest in the Offering. However, no member of the Board of Directors or Executive Management or any of the Key Employees, directly or indirectly, hold more than 5% of the Company's share capital.

The Executive Management, Key Employees and certain members of the Board of Directors hold Pre-IPO Warrants granted by the Company as part of its pre-IPO warrant programme. These Pre-IPO Warrants may be exercised in connection with the Offering and, accordingly, the Executive Management, Key Employees and certain members of the Board of Directors have an interest in the Offering.

Skandinaviska Enskilda Banken, Danmark, branch of Skandinaviska Enskilda Banken AB (Publ.), Sweden; Vækstfonden; BI Asset Management Fondsmæglerselskab A/S on behalf of certain clients; Handelsbanken, branch of Svenska Handelsbanken AB (Publ.), Sweden; and Spar Nord Bank A/S have in connection with the Offering subject to certain conditions undertaken to subscribe for Offer Shares as Cornerstone Investors and, accordingly, will have a direct economic interest in the Offering.

The Managers and their respective affiliates have engaged in transactions with and performed various commercial banking, investment banking, financial advisory and other services for Orphazyme, and the Managers and their respective affiliates are currently providing and may in the future provide such services for Orphazyme. With respect to certain of these transactions and services, the sharing of information is restricted for reasons of confidentiality, internal procedures or applicable rules and regulations. The Managers have received and will receive customary fees and commissions for these transactions and services and may come to have interests that may not be aligned or could potentially conflict with potential investors' and the Company's interests. Danske Bank A/S is expected to also be a lender under a loan facility in favour of the Executive Management and Key Employees with respect to their subscription for Investment Shares (as defined herein) in connection with Orphazyme's long-term incentive programme.

E.5 Selling Shareholders and Lock-Up Arrangements

The Selling Shareholders
The Offering consists of new Shares offered by the Company. None of the Company's existing shareholders are offering Shares in connection with the Offering.

Lock-Up Arrangements
The Main Shareholders have agreed with the Managers that they will not, except as set forth below, for a period starting on the date hereof and ending on the earlier of (i) the Company's publication of the results of the ongoing NPC phase II/III trial, currently expected for Q3 2018 (however, not earlier than 180 days after Admission), or (ii) 360 days after Admission, without the prior written consent of the Managers: (i) offer, pledge, sell, contract to sell, sell any option or contract to purchase, purchase any option or contract to sell, grant any option, right or warrant to purchase, lend, cause the Company to issue Shares, or otherwise transfer or dispose of (or publicly announce such action), directly or indirectly, any of their Shares held as of Admission (excluding any Offer Shares subscribed for in connection with the Offering) ("**Lock-Up Shares**"), or any securities convertible into or exercisable or exchangeable for such Lock-Up Shares; (ii) enter into any swap or other arrangement that transfers to another, in whole or in part, any of the economic consequences of ownership of any Lock-Up Shares, whether any such transactions described in clause (i) or (ii) above are to be settled by delivery of such Lock-Up Shares or such other securities, in cash or otherwise; or (iii) propose any general meeting of the Company, or convene or take action to convene any general meeting for the purpose of proposing, any resolution of the Company authorizing the issue of any Shares or warrants to subscribe for Shares or publicly announce any intention to enter into any of the foregoing transactions.

The Company has agreed with the Managers to substantially the same restrictions set forth above for a period of 180 days from Admission subject to certain exemptions.

The members of the Board of Directors at the time of Admission (other than the New Board Members), Executive Management and the Key Employees have agreed with the Managers that, for a period of 360 days from Admission, in respect of the Shares held as of Admission (and any Shares subscribed for as a result of exercise of Pre-IPO Warrants, if applicable), they will be subject to substantially the same restrictions as those of the Company and Main Shareholders as set forth above.

Moreover, all existing shareholders of the Company as of the date hereof have undertaken an obligation not to trade in the existing Shares until the Temporary Purchase Certificates have been exchanged for Shares and the Shares have been admitted to trading and official listing on Nasdaq Copenhagen.

E.6 The amount and percentage of immediate dilution resulting from the Offering

The existing Shares issued as of the date hereof will be diluted in connection with the Offering by the issuance of up to 10,781,250 new Shares, corresponding to a nominal value of DKK 10,781,250, assuming full exercise of the Overallotment Option. Following completion of the Offering, the existing Shares issued and outstanding will make up 57.9% of the Company's share capital, assuming full subscription for all new Shares issued in connection with the Offering (including full exercise of the Overallotment Option) as well as exercise of all Pre-IPO Warrants vesting in connection with the Offering, and adjusting for the 2017 Capital Structure Adjustment (assuming an Offer Price at the midpoint of the Offer Price Range and, thereby, an issue of 720,875 bonus Shares prior to Admission as part of the 2017 Capital Structure Adjustment). The Company's net capital as of 30 September 2017 was DKK 94,500,000, corresponding to a net book value per Share of DKK 18.5, corresponding to DKK 8.2 per Share based on the Company's capital structure as of the date hereof. The net book value per Share is calculated by dividing the total net capital by the total number of Shares issued. Based on the Company's net capital as of 30 September 2017 and the same assumptions as described above as well as adjusting for the estimated costs related to the Offering, the Company's net book value per Share as of completion of the Offering would be DKK 32.1.

E.7 Estimated expenses charged to the investor by the Company or the Selling Shareholders

Not applicable. None of the Company or the Managers will charge expenses to investors. Investors will have to bear customary transaction and handling fees charged by their account-holding banks.

Risk Factors

An investment in the Offer Shares involves a high degree of financial risk. You should carefully consider all information in this Offering Circular, including the risks described below, before you decide to buy the Offer Shares. This section addresses both risks associated with the industry in which the Company operates and the specific risks associated with its business. If any such risks were to materialise, the Company's business, results of operations, financial condition and/or prospects could be materially and adversely affected, resulting in a decline in the value of the Offer Shares and a loss of part or all of your investment. Further, this section describes certain risks relating to the Offering and the Offer Shares which could also adversely impact the value of the Offer Shares. With respect to forward-looking statements that involve risks and uncertainties, please see "Special Notice regarding Forward-Looking Statements".

The risks and uncertainties discussed below are those that Orphazyme's management currently views as material, but these risks and uncertainties are not the only ones that it faces. Additional risks and uncertainties, including risks that are not known to Orphazyme at present or that its management currently deems immaterial, may also arise or become material in the future, which could lead to a decline in the value of the Offer Shares and a loss of part or all of your investment. The following risk factors are not listed in any particular order of priority as to significance or probability.

Risks Relating to the Company's Industry and the Market

1. Orphazyme may be adversely affected by competition from other life sciences companies developing other treatments for similar diseases to those targeted by Orphazyme's products in development.
The industry in which Orphazyme operates is highly competitive. The pharmaceutical industry is subject to global competition and swift technogical advances. For some of the diseases currently targeted by the Company's products in development there are at present limited other treatment options available. However, the Company is exposed to potential competition from biotech, pharmaceutical and other companies which are developing or may initiate development of competing products. Some of the Company's competitors may have a substantially stronger financial position and other resources available and, accordingly, such competitors may develop more effective or affordable products in a more rapid or efficient manner or may achieve commercialization of their products earlier or in a better manner than Orphazyme. These competing products may gain wider acceptance within the market and could render the Company's products obsolete or limit the ability of the Company to generate revenues, which could materially adversely affect the Company's business, financial condition, results and prospects.

The Company may face heightened competition from gene therapy, alternative treatment forms, and, after expiry of patent protection for the Company's products, also generics. The Company is aware of several pharmaceutical and biopharmaceutical companies that have successfully commercialized products or have commenced clinical trials of products addressing areas which are being targeted by the Company, including Edaravone to treat ALS marketed by Mitsubishi Tanabe Pharma in the United States under the brand name Radicava® and in Japan under the brand name Radicut®, Riluzole (generic, off patent) to treat ALS marketed by (amongst others) Covis Pharma in the United States and Aventis Pharma in the EU under the brand name Rilutek®, Masitinib developed by AB Science SA to treat ALS and in respect of which successful phase III trials have been reported as completed, Tirasemtiv to treat ALS which is being studied in phase III trials by Cytokinetics Inc., Miglustat marketed by Actelion Pharmaceuticals (Johnson & Johnson) under the brand name Zavesca in the US and EU to treat Gaucher's disease and which is also approved and marketed in the EU to treat NPC and in respect of which there are reports of off-label use to treat NPC, VTS-270 (a cyclodextrin) currently being studied by Sucampo Pharmaceuticals (Vtesse, Inc.) in clinical phase I/IIa trials to treat NPC and Trappsol® Cyclo™ another cyclodextrin currently being developed by CTD Holding, Inc. (Sphingo Biotechnology, Inc.) in clinical phase I/IIa trials to treat NPC. A new oral substrate reduction therapy currently being developed by Genzyme under the name GZ/SAR402671 and i.a. reported to be studied in phase II clinical trials for the treatment of Gaucher's disease.

If the Company is unable to respond effectively to competition, future demand for its products may materially decrease, which could have a material adverse effect on its business, financial condition, results of operations or prospects.

2. Changes in the regulatory and compliance environment may have a significant adverse impact on the Company.
The pharmaceutical and biotech industry is subject to a wide range of laws, as well as regulations laid down by the FDA, the EMA and other regulatory authorities, on matters such as orphan drugs, clinical trials, use of data, animal testing, approval processes, requirements to production, marketing, sales, pricing, pharmacovigilance and intellectual property rights. Regulatory changes in these and other areas in jurisdictions in which the Company develops, tests, produces, and intends to market and sell its products may have material adverse effects on the Company's business, financial condition, results and prospects. Such changes, which are outside of the Company's control, may cause the Company to incur significant costs, revise, delay or stop all or part of its development program, operations or products or

adopt new processes and procedures in order to comply with new laws or regulation, and may negatively impact how the Company is able to develop, attest, produce, market and sell its products, for instance by making it more costly and demanding in terms of resources to develop or obtain approval for the Company's products.

On 29 March 2017, the United Kingdom initiated Article 50 of the Lisbon Treaty for the United Kingdom to leave the European Union and the United Kingdom is scheduled to leave the European Union on 29 March 2019 ("Brexit"). As part of Brexit, EMA currently situated in London, is expected to relocate to the EU. There is a risk that the relocation process will interrupt current administrative routines and occupy resources which may lead to delays in EMAs handling of the Company's applications and generally adversely affect the Company's dealings with EMA. Further, there is considerable doubt resulting from lack of precedent and the complexity of the UK and EU's intertwined legal regimes as to how Brexit will impact the life sciences and healthcare sector in Europe including the Company, including with respect to ongoing clinical trials. The impact will to a large extent depend on the model and means by which the United Kingdom's relationship with the European Union is maintained post Brexit. By way of example following Brexit, United Kingdom will no longer be covered by the centralized procedure for obtaining EU-wide marketing authorization from the EMA and unless a specific agreement is entered into, this will necessitate instead a separate national authorization for the United Kingdom of medicinal products the process for which is currently unclear. Brexit may adversely affect and delay the Company's ability to market and sell any medicinal products in the United Kingdom.

3. The pricing and demand for pharmaceutical products may be affected by global economic factors. Orphazyme's ability to determine prices and thus generate revenue from any products that it may develop will depend on enacted and future reimbursement and drug pricing policies and regulations.
Altered macro-economic factors may adversely affect pharmaceutical companies, including Orphazyme. For instance, a decline in the economy could put pressure on payers, including authorities, insurance companies and hospitals, resulting in a lower willingness to pay for pharmaceutical products and may also lead to changes in areas such as national subsidies, prescription regulations and distribution terms which may have a negative impact on Orphazyme. The Company bases its development activities and commercial strategy on estimates of the number of patients who may benefit from and hence be medically eligible for a particular treatment. These estimates are subject to great uncertainty and may prove to be too optimistic. Further, even if the number of medically eligible patients is correctly estimated, the number of patients who will ultimately receive a particular treatment may be greatly reduced as a result of financial and political considerations as described above.

The successful commercialization of Orphazyme's product candidates will depend in part on the extent to which governmental authorities and health insurers establish adequate coverage and reimbursement levels and pricing policies. Changes in applicable regulations limiting reimbursements for any of Orphazyme's product candidates would have a material adverse effect on Orphazyme's business, financial condition, results and prospects.

In the United States and the other principal markets in which Orphazyme may in the future sell its products, if approved, there is continued economic, regulatory and political pressure to promoting changes in healthcare systems with the stated ambitions of containing healthcare costs and/or expanding access to healthcare. Already enacted legislation in the United States has introduced cost-reduction measures and other provisions that could decrease the coverage and price that Orphazyme may receive for any approved products. Further, new initiatives are expected to continue to be introduced and may likely introduce additional reductions in health care funding, which could have a material adverse effect on Orphazyme's customers and accordingly, its financial operations. In the EU, provision of healthcare, including the establishment and operation of health services and the pricing and reimbursement of medicinal products, is almost exclusively a matter for national, and not EU, law and policy. National governments and health service providers have different priorities and approaches to the delivery of health care and the pricing and reimbursement of products in that context. In general, however, the healthcare budgetary constraints in most EU member states have resulted in restrictions on the pricing and reimbursement of medicines and such measures are expected to continue, which could affect Orphazyme's ability to commercialize any products for which it obtains marketing approval.

In addition, the pricing of the Company's products may be affected by a number of factors, including the burden of disease, extent of unmet need, clinical efficacy, side effects, tolerability, number of patients affected by the disease, as well as the cost of development and manufacturing, all of which may adversely affect the price, Orphazyme may be able to obtain for its products.

Risks Relating to the Company's Business

4. The Company has no products approved for commercial sale, has never generated any revenue and may incur significant losses in the future, which makes it difficult to assess its future viability.
Orphazyme is a clinical stage biotechnology company not yet having had any products approved for commercial sale. Biopharmaceutical product development is a highly speculative undertaking and involves a substantial degree of risk, which extends to risks related to the

regulatory approval process for drug candidates. To date, the Company has focused on research and development activities and, in particular, on developing its lead product candidate in development, as described in "*Business*". Going forward, the Company expects to continue to incur significant losses from its operations. As of 30 June 2017, the Company had an accumulated deficit of DKK 264 million. Substantially all of the losses have resulted from expenses incurred in connection with research and development programs and from general and administrative costs ("**G&A activities**").

None of the Company's products in development have been approved for commercial sale, and no revenues have been generated so far. It is expected that annual operating expenses will increase over the next several years, as the research and development efforts are expanded, and due to the additional costs of being a listed public company.

5. Orphazyme is currently highly dependent on arimoclomol.
The Company is currently conducting pre-clinical studies and clinical trials based on the arimoclomol molecule. Even though the Company is also in the discovery phase with respect to new molecular entities, the Company is highly dependent on arimoclomol.

The clinical and commercial success of the Company's products in development will depend on a number of factors, including the timely completion of the ongoing and future clinical trials; the ability to establish and obtain regulatory approval for commercial-scale manufacturing processes for the products in development; whether the products' safety, tolerability and efficacy profiles will be satisfactory to the regulatory authorities to warrant marketing approval; the timely receipt of necessary marketing approvals from the regulatory authorities; whether the regulatory authorities require additional clinical trials prior to approval to market the products; the incidence and severity of adverse side effects of the products; the ability to successfully commercialize the products; achieving and maintaining compliance with all applicable regulatory requirements; acceptance of the products as safe and effective by patients and the medical community; the availability, perceived advantages, relative cost, relative safety and relative efficacy of alternative and competing treatments; obtaining and sustaining an adequate level of coverage and reimbursement for the products by third-party payors; the effectiveness of marketing, sales and distribution strategies and operations; the ability of third-party manufacturers to manufacture and supply the products and to develop, validate and maintain commercially viable manufacturing processes; enforcing intellectual property rights in and to the products, avoiding interference, opposition, derivation or similar proceedings with respect to the Company's patent rights, and avoiding other challenges to the patent rights and patent infringement claims, and continued acceptable safety profiles of the products following approval, if approved.

Many of these factors are beyond the Company's control, for instance results of clinical trials conducted by the Company or others, and the regulatory submission process and potential threats to the Company's intellectual property rights. Should one or more of these risks materialize, it is likely to have an adverse effect on the Company's business, financial condition, results and prospects.

Further, although the safety profile, i.e. risks to patients, of arimoclomol has been explored in pre-clinical studies and phase I clinical trials as well as in phase II trials in ALS and sIBM, there is always a risk that hitherto unknown safety concerns may arise in connection with the use of pharmaceutical compounds such as arimoclomol.

Should such risks relating to safety materialize, it is likely to have a material adverse effect on the Company's business, financial condition, results and prospects.

The Company acquired arimoclomol (and certain other molecules), including pre-clinical and clinical data, intellectual property rights and other assets, including contractual rights and obligations relating to arimoclomol from the United States based biopharmaceutical company CytRx Corporation ("**CytRx**") in 2011. Through the purchase, the Company became party to a number of related research, development and licensing contracts, and undertook to be responsible for all related obligations under these contracts going forward. There is a risk that the Company did not at the time of the acquisition or in the period since manage to properly identify and assess all risks relating to the acquired assets such as obligations, liabilities, defects or other shortcomings and that these will materialize at a later stage. If this should occur, it may materially harm the operations of the Company and the Company will have only limited recourse against the seller.

6. Clinical trials being conducted to test the Company's products may not show the desired results or may be delayed or more costly than anticipated.
Prior to launching a pharmaceutical product on the market, its safety and efficacy for treatment of patients must be ascertained through execution of certain pre-clinical studies and clinical trials conducted in accordance with the International Council for Harmonisation of Technical Requirements for Pharmaceuticals for Human Use (ICH), the FDA's, the EMA's and other applicable regulatory authorities' legal requirements, regulations and guidelines, including good laboratory practices ("**GLP**"), an international standard meant to harmonize the conduct and quality of non-clinical studies and the reporting of findings, as well as good clinical practices ("**GCP**"), an international ethical and scientific quality standard for designing, conducting, recording and reporting trials that involve the participation of human subjects. Conducting such trials is complex, costly and time-consuming, and neither the results nor the timing can be predicted with any certainty. Certain of the

clinical trials currently sponsored by the Company relate to pediatric diseases for which there are additional regulatory requirements, for instance in relation to documentation. The performance of clinical trials is associated with risks relating to, for instance, designing the trials in the most appropriate manner, complying with regulatory requirements, entering into agreements with investigators and hospitals and recruitment of patients. A number of companies in the pharmaceutical, biopharmaceutical and biotechnology industries have suffered significant setbacks in advanced clinical trials due to lack of efficacy or due to adverse safety profiles. A randomised, double-blinded, placebo-controlled phase II clinical trial in SOD1 ALS was completed in 2016; however, based on the mechanism of action of arimoclomol that is expected to be relevant across the broad ALS patient population, a trend effect in an open label extension phase II trial in sporadic ALS and encouraging feedback from FDA, the trial design for the phase II/III clinical trial is being designed with a view to support a marketing authorisation in the broader ALS indication. This amendment of the trial strategy may increase the risks related to the trial.

Designing and conducting clinical trials for orphan drugs involves additional risks relating to for instance the relevant indications not being well characterized and experience with treatment of orphan diseases being limited. Such risks may for instance cause delays, increase in costs compared to budgets. Quality deficits in the execution of trials could result in trials being suspended or discontinued. The result could be that Orphazyme is not granted necessary regulatory approvals for further clinical trials and/or relevant authorisations required for the marketing and sale of its products. If these risks materialize, it could materially adversely affect the Company's business, financial condition, results and prospects.

There is a risk that the clinical trials currently sponsored by Orphazyme will not confirm previous results or will not demonstrate sufficient evidence of safety and efficacy to ensure the granting of the requisite regulatory approvals, in particular as the Company thus far has only conducted relatively small phase II clinical trials that were not powered for efficacy. This could mean that the clinical trials would not lead to pharmaceutical products that could be commercialized. Adverse or inconclusive results may, despite initially promising results, result in Orphazyme's products not being approved for marketing and sale, and there is a risk that additional clinical trials will be required resulting in increased costs, signficiant delays to the filing with regulatory authorities, a filing for a narrower indication or that the Company may ultimately have to abandon the commercialization of one or more of its products. All Orphazyme's current clinical trials are studying the same chemical compound – arimoclomol – but within different indications. There is a risk, therefore, that any unexpected findings, including but not limited to serious adverse events, in one clinical trial may have a "spill-over" effect on other trials, in particular if the finding is related to the compound as such, for instance in relation to safety and tolerability. Any finding in one trial, therefore, may have the implication that it halts or significantly delays the Company's entire clinical development portfolio. As clinical product development can be affected by unforeseen delays, increased costs, unexpected adverse events, unforeseen suspensions and unfavourable results, these circumstances could have a material adverse effect on the Company's business, financial condition, results and prospects.

All of the Company's clinical trials have been relatively small, each with less than 100 persons, and have advanced through phase I and phase II, which may entail additional risks, including hitherto unidentified low incidence safety risks, safety risks associated with high dose long term treatment or lack of efficacy given that none of the trials was powered for efficacy. In particular, to date, the Company has had no experience testing arimoclomol in patients with NPC and Gaucher disease. If these risks were to materialize, it could materially adversely affect the Company's business, financial condition, results and prospects.

7. Orphazyme's products may cause undesirable side effects or have other properties that could delay or prevent their regulatory approval, limit the commercial profile, or result in significant negative consequences following regulatory approval, if any.
The Company and/or regulatory bodies may suspend clinical trials at any time, if it is believed that patients, who participate in such clinical trials are being exposed to unacceptable health risks resulting in an unfavourable risk benefit assessment and/or serious unexpected adverse events. Undesirable side effects caused by the Company's products could cause the Company or regulatory authorities to interrupt, delay or halt clinical trials and could result in a more restrictive use permitted by the authorities or the delay or denial of approval by regulatory authorities. In the event that trials conducted reveal an unacceptable severity and prevalence of adverse side effects, such trials could be suspended or terminated and regulatory authorities could order the Company to cease further development of or deny approval of the products in development for any or all targeted indications. The treatment-related side effects could affect patient recruitment or the ability of enrolled patients to complete the trial or result in potential product liability claims. Even after receiving approval, the products may later exhibit adverse effects that could prevent their widespread use and/or necessitate their withdrawal from the market. Any of these events could have a material adverse effect on the Company's business, reputation, financial condition, results and prospects.

To date, a total of 248 human subjects have been exposed to arimoclomol, including 112 healthy volunteers who have received between 50 mg and 1800 mg arimoclomol daily and 136 patients suffering from sIBM, ALS and NPC. 82 ALS patients received arimoclomol across the 12-week phase II dose ranging trial ("**AALS-001**") and the six-month open-label extension trial ("**AALS-001-OL**"). In an investigator-initiated trial in SOD1-ALS, 19 patients were treated with between 300 mg and 600 mg arimoclomol daily for 12 months. In another investigator-initiated trial in sIBM 16 patients were treated with 300 mg arimoclomol daily for four months. As of the date hereof, no safety risks

from taking arimoclomol have been identified in seven phase I trials and three phase II trials, and, overall, tolerability (i.e. degree to which an overt adverse effect can be tolerated by the subject/patient) of arimoclomol was comparable to a placebo. Potential mild tolerability side effects identified were diarrhea, nausea and dry mouth. In the trials these side effects did not result in treatment discontinuation. Arimoclomol is an inhibitor of the organic cation transporter 2 ("**OCT2**"), and, thus, inhibits OCT2 dependent transport of creatinine. Thereby, arimoclomol can lead to a transient decrease in creatinine clearance and increase in serum creatinine that is not considered a safety risk. In the trials, serum creatinine increases were still within normal limits and the values went back to baseline levels while subjects continued treatment with arimoclomol.

In the AALS-001 trial, nine serious adverse events ("**SAEs**"), including three deaths, were reported in the placebo-controlled main part of the study; two incidences of pulmonary embolus; two incidences of myasthenia/ALS progression; one incidence of apnea; one incidence of bone fracture; one incidence of dysphagia; and one incidence of respiratory disorder; one incidence of thrombophlebitis. All SAEs were determined to be either unlikely or not related to trial medication. There were no treatment related deaths. In the AALS-001-OL, the open label extension of the trial, 13 patients had SAEs. There was one SAE, pulmonary embolism, which was deemed possibly related to trial medication. All other SAEs were deemed not related or unlikely related to the trial medication. In the phase II SOD1-ALS trial adverse events, occurred with similar frequency in the two treatment groups, and were largely considered unrelated to trial drug by the site investigators. 22 SAEs were reported (15 in the placebo group and seven in the arimoclomol-treated group), none of which were considered related to trial drug. A single participant stopped arimoclomol treatment because of a skin rash that was deemed probably related to trial drug. In the sIBM trial involving, 16 patients on active treatment and eight patients on placebo there was no difference in the rate, type and severity of adverse events between the arimoclomol and the placebo group. One SAE of high blood pressure was noted in the arimoclomol group. In the ongoing phase II/III trial in NPC, two patients developed urticarial and angioedema, respectively, the treatment allocation remains blinded until the end of the pivotal part of the trial.

While none of the above events has led to the identification of a safety risk that would be considered associated with the treatment with arimoclomol, given that still a limited number of patients have been exposed to date for a limited time and daily dose, rare safety risks or risks associated with higher doses cannot be excluded. Often, uncommon risks are only identified when thousands of patients have been treated long term, not uncommonly after years on the market.

8. Challenges in recruiting patients for clinical trials and concluding agreements with investigators and hospitals may have a material negative impact on the Company.
The diseases for which Orphazyme's products are currently being developed are rare and, consequently, patient groups relevant for testing Orphazyme's products are limited in size and located across many jurisdictions, see the relevant descriptions under "*Industry*" and "*Business*". Therefore, even though the Company through its contract research organisations ("**CROs**") cooperates closely with relevant physicians treating these patient groups, finding and recruiting the appropriate number of patients for the clinical trials, as well as patients with a profile appropriate for the clinical trials, may be challenging. Should clinical trials in indications similar to the Company's products be initiated, it could negatively affect the possibility for the Company of recruiting patients. If Orphazyme cannot find the necessary number of appropriate patients to complete its clinical trials, this could have a material negative impact on the Company's business, financial condition, results and prospects.

In connection with testing of its products, Orphazyme is dependent on being able to enter into agreements with CROs conducting the clinical trials with respect to the products. Orphazyme through its CROs is in close ongoing dialogue with those physicians, who are relevant as investigators; however, if Orphazyme is not able through its CROs to enter into the necessary agreements on clinical trials, it may have a significant negative impact on the Company. Further, if the counterparties to the Company-sponsored clinical trial agreements do not carry out their obligations or do so within the agreed deadlines, the clinical trials may be delayed, terminated or unsuccessful which may have a material negative impact on the Company's business, financial condition, results and prospects.

9. Orphazyme is highly dependent on obtaining and maintaining required regulatory approvals.
Before Orphazyme can start commercializing its products, a number of regulatory registrations and approvals must be obtained. For instance, approvals from the authorities and ethical committees as well as consents from patients participating in the clinical trials are required before initiating pre-clinical studies and clinical trials, and marketing authorisations must be obtained from the relevant authorities. Obtaining regulatory registrations and approvals is highly complex and time-consuming. If required regulatory registrations or approvals are delayed, denied or withdrawn, it is likely to have a material adverse effect on the Company's business, financial condition, results and prospects.

Even after a pharmaceutical product has been approved for marketing, a number of regulatory requirements must still be met in order for the approval to be upheld, for instance pharmacovigilance, reporting on adverse effects and requirements relating to labelling and marketing of the product. Should the Company be unable to comply with applicable regulatory requirements, the Company may be subject

to fines, withdrawal of regulatory approvals, recall of products, suspension of manufacturing, other operational restrictions, criminal sanctions and damage claims all of which can have a material adverse effect on the Company's business, financial condition, results and prospects.

10. If the Company is unable to obtain orphan product designation or marketing exclusivity for its product candidates or is unable to benefit from the associated marketing exclusivity, such will have a material adverse effect on Orphazyme.
Certain diseases are so rare that drug developers are reluctant to develop them under usual marketing conditions since this does not allow the recovery of the capital invested for the research and development. In order to promote research and development of treatment of rare diseases, regulatory authorities in some jurisdictions, including the United States and EU may designate drugs for relatively small patient populations as so-called "orphan drugs". Such orphan drug designation provides the drug developing company with a number of financial and other incentives.

In the EU, the European Commission may designate a product candidate as an orphan medicinal product, if it is a medicine for the diagnosis, prevention or treatment of life-threatening or very serious conditions that affects not more than five in 10,000 persons in the European Union, or it is unlikely that marketing of the medicine would generate sufficient returns to justify the investment needed for its development. In the EU, it is a further requirement that there are no other satisfactory treatments or, if such exist, that the product candidate must be of or must bring significant benefit to those affected by the disease. Under the US Orphan Drug Act, the FDA may designate a product candidate as an orphan drug, if it is a drug intended to treat a rare disease or condition, which is generally defined as a patient population of fewer than 200,000 individuals annually in the United States.

Generally, if a product candidate with an orphan drug designation receives the first marketing approval for the indication for which it has such designation, the product is entitled to a period of marketing exclusivity, which, subject to certain exceptions precludes the EMA and other national drug regulators in the EU, from accepting the marketing application for a similar medicinal product for the same indication or for the United States precludes the FDA from approving the marketing application of the same drug for the same indication for that time period.

The applicable period is ten years in the EU and seven years in the United States. The period can be reduced to six years in the EU, if a product no longer meets the criteria for orphan drug designation or if the product is sufficiently profitable so that market exclusivity is no longer justified. In the EU, orphan exclusivity may also be extended for an additional two years (i.e., a maximum of 12 years' orphan exclusivity), if the product is approved on the basis of a dossier that includes pediatric clinical trial data generated in accordance with an approved paediatric investigation plan. Orphan drug exclusivity may be lost in the United States, if the FDA determines that the request for designation was materially defective or if the manufacturer is unable to assure sufficient quantity of the product to meet the needs of patients with the rare disease or condition. For further information, please see "*Industry*".

It is an important component of Orphazyme's business strategy to seek orphan drug status where available for its product candidates. Orphazyme received orphan drug designation for arimoclomol for the treatment of Niemann-Pick disease type C by the FDA and EMA; ALS by the FDA and EMA; and sIBM by EMA. In September 2017, Orphazyme made an application to the FDA for orphan drug designation for arimoclomol for the treatment of sIBM. Although the Company is reasonably comfortable that its application to the FDA satisfies the criteria for obtaining orphan drug designation by the FDA, there is a risk that the FDA will not grant the requested orphan drug designation or that such designation will be delayed. Moreover, orphan drug status may not ensure that Orphazyme has market exclusivity in a particular market and there is no assurance it will be able to receive orphan drug designation for any additional product candidates. Further, the granting of a request for orphan drug designation does not alter the standard regulatory requirements and process for obtaining marketing approval.

Even if Orphazyme obtains orphan drug exclusivity for a product candidate, the exclusivity thus created may not effectively protect the product from competition, because exclusivity can be suspended under certain circumstances. In the United States, even after an orphan drug is approved, the FDA can subsequently approve another drug for the same condition, if the FDA concludes that the later drug is clinically superior in that it is shown to be safer, more effective or makes a major contribution to patient care. In the EU, orphan exclusivity will not prevent a marketing authorization being granted for a similar medicinal product in the same indication, if the new product is safer, more effective or otherwise clinically superior to the first product or if the marketing authorization holder of the first product is unable to supply sufficient quantities of the product. If Orphazyme's competitors are able to obtain orphan product exclusivity for their products in the same indications for which Orphazyme is developing its product candidates, the Company may not be able to have its products approved by the applicable regulatory authority within a significant period of time, or at all.

Should an orphan drug designation be revoked or should the market exclusivity period be suspended, shortened or even revoked, it could have a material adverse impact on Orphazyme's business, financial condition, results and prospects.

Specifically, the Company is dependent on retaining and obtaining orphan drug designations for IBM and ALS. Further, the Company is dependent on the conversion of orphan drug designations into orphan drug status for arimoclomol for the treatment of IBM and ALS after marketing approval, see the relevant description in "*Business*". This dependency is a result of the Company's general patent protection for the composition of matter coverage expiring in 2020, as the market exclusivity resulting from a grant of orphan drug status will effectively prolong the period during which the products are protected. The patent protection for treatment of lysosomal diseases expires in 2029. Should the Company not be able to obtain or maintain orphan drug status for IBM and ALS or for other products, this may have a material adverse effect on Orphazyme's business, financial condition, results and prospects.

11. Risks relating to manufacturing and research & development, including use of hazardous materials as well as chemistry and manufacturing controls ("CMC"), may have a material adverse effect on the Company.
Due to the chemical ingredients of pharmaceutical products and the nature of the manufacturing process, Orphazyme, its employees and third party contractors are subject to safety reporting requirements, environmental regulations and, going forward, additional requirements following potential receipt of marketing approval. Should Orphazyme fail to comply with applicable rules and regulations, the Company could be subject to criminal sanctions and substantial liability or could be required to suspend or modify its operations. Further, if any of the Company's employees or third party contractors perform acts or omissions that are considered unethical, criminal or otherwise contrary to applicable laws and regulations and/or internal guidelines, Orphazyme's reputation may be harmed which could have an adverse effect on the Company's business, financial condition, results and prospects, and Orphazyme may not be able to manufacture sufficient quantities of its product candidates in a cost-effective or timely manner. Manufacturing includes the production, formulation and stability testing of an active pharmaceutical ingredient and its formulation into pharmaceutical products, such as capsules or tablets. Any delays in production would delay Orphazyme's pre-clinical studies and human clinical trials, which could adversely affect its business, financial condition, results and prospects.

Orphazyme does not have its own manufacturing facility and currently does not intend to develop any such manufacturing capacity. Orphazyme is therefore dependent on third parties for manufacturing its products. Orphazyme has already contracted with third parties for the development and manufacture of its product candidates and may be required also in the future to enter into contracting arrangements with third parties to manufacture its product candidates for large-scale, pre-clinical studies and/or clinical trials and for marketing and sale (after approval). Orphazyme may not be able to make the transition from development-scale to commercial production of arimoclomol or from laboratory-scale to development-scale of new molecules. Orphazyme may need to develop additional manufacturing resources, enter into collaborative arrangements with other parties, who have established manufacturing capabilities, or have other third parties manufacture its products on a contract basis. Orphazyme may not have access on acceptable terms to the necessary and substantial financing that would be required to scale-up production and develop effective commercial manufacturing processes and technologies. Orphazyme may not be able to enter into collaborative or contracting arrangements on acceptable terms with parties that will meet its requirements for quality, quantity and timeliness.

Any manufacturing of pharmaceutical products is subject to a number of regulatory requirements, for instance testing, quality control and documentation. Orphazyme is dependent on its contract manufacturing partners appropriately handling CMC and the costs of compliance may be high. Manufacturing facilities must be approved by the authorities and will be subject to regular audits by the authorities. Such audits may lead to suspension of manufacturing and interfere with product supply and distribution. If Orphazyme's existing or future contract manufacturing partners do not manufacture the products properly and otherwise fulfil their contractual and regulatory obligations to deliver agreed quantities of products in a timely manner and in the correct quality, it could have a material adverse effect on Orphazyme's business, financial condition, results and prospects.

In its research and development the Company uses hazardous materials, such as Rotenone. The Company cannot completely eliminate the risk of accidential contamination or injury from the use, storage, handling or disposal of these materials. The Company may be held liable for any such accident or injury which may exeed any insurance coverage and which could have a negative impact on the Company's business, financial condition, results and prospects. Although Orphazyme believes that it holds all permits required for its use of hazardous materials, any failure to comply with applicable laws and regulations could result in fines, suspension of permits or authorisations or claims for damages.

12. The Company is dependent on third-party vendors to provide certain licenses, products and services, clinical trials, and the Company's business and operations could be disrupted by any problems with its significant third-party vendors.
Orphazyme utilises a number of third-party suppliers and service providers to supply services critical for the Company, such as contract research services, contract manufacturing services and IT services. Disruptions to the business, financial stability or operations of these suppliers and service providers, including due to strikes, labour disputes or other disruptions to the workforce, or to their willingness and ability to produce or deliver the products and provide the services Orphazyme requires in accordance with Orphazyme's and the authorities' requirements, could affect the Company's ability to develop and market its products on a timely basis, which could materially harm the

Company. If these suppliers and service providers were unable or unwilling to continue providing their products or services in the manner expected, or at all, Orphazyme could encounter difficulty finding alternative suppliers. Even if the Company is able to secure alternative appropriate suppliers in a timely manner, the Company's costs could increase significantly. Any of these events could adversely affect the Company's business, financial condition, results and prospects.

Specifically, Orphazyme is dependent on agreements with external parties, who carry out the clinical trials sponsored by Orphazyme. If these external parties do not carry out their obligations towards the Company or do not meet expected deadlines, if the parties need to be replaced, or if the quality or accuracy of the clinical data they obtain is compromised, ongoing and planned clinical trials may be extended, delayed or terminated which could have a material negative impact on the Company's business, financial condition, results and prospects.

13. Orphazyme is dependent on successful commercialization of its products.
In order to be successful, Orphazyme must commercialize its products, once the relevant authorisations for marketing and sale have been obtained. Whether commercialization is successful will depend on factors such as the Company's ability to succesfully execute its business strategy and attract and build-up the internal resources necessary to effectively market the Company's products. Even if the Company's products are commercialized, market acceptance for the products may be less than estimated, and/or competitors may successfully obstruct commercialization efforts. Competition in the pharmaceutical industry is intense, and the degree of market acceptance for any of the Company's products (if and when commercialized) will depend on a number of factors, including cost-effectiveness, alternative treatment methods, changes in physicians' treatment preferences, reimbursement policies of governments and third-party payers, and marketing and distribution support for the Company's products. If commercialization is not successful, it will have material adverse effect on the Company's business, financial condition, results and prospects.

Orphazyme does not currently have in-house capabilities for sales, marketing and distribution but intends to develop such capabilities in order to market its products (if approved) directly through its own sales and marketing force in selected geographic areas, including the United States and Europe. In order to implement this strategy of commercializing in-house, the Company must develop a sales and marketing organisation and establish distribution capability. This entails recruiting additional managerial, operational, financial and other employees, which would be expensive and time-consuming and could delay product launches. Further, if Orphazyme is not able to attract and retain personnel with the required competencies within sales, marketing and distribution or enter into collaborations with third parties having such competencies, it could have a material adverse effect on the Company's business, financial condition, results and prospects.

14. The Company may in the future seek to enter into collaborations with third parties for the development and commercialization of its product. If such collaborations are not successful, the Company may not be able to capitalize on the market potential of the products.
The Company may in the future enter into collaboration agreements with third-party collaborators, i.e. by introducing a license right or a distribution agreement, for development and commercialization of existing or other products to address market opportunities that require large development investments and/or special expertise in selected geographic areas, as well as to share the financial risks involved in drug development and commercialisation of certain of its product candidates.

The Company has no signifianct experience in entering into major collaboration and/or licence agreements. The Company may be unable to attract partners for collaboration agreements and/or the terms of those collaboration agreements, which the Company chooses to enter into may not be favourable to the Company. This may be a result of factors such as general market demand for particular products or products within specific therapeutic areas, results of clinical trials relating to the products or market competition.

If the Company is not successful in efforts to enter into future partnership agreements, the Company's business, financial condition, results and prospects may be negatively affected. Even if the Company is successful in entering into collaboration agreements, such agreements may not lead to development or commercialization of the products in the most efficient manner or at all.

With any future collaboration agreements, the Company expects to have limited control over the amount and timing of resources that such collaborators dedicate to the development or commercialization of the products. The ability to generate revenue from these arrangements will depend on such collaborators' abilities to successfully perform the functions assigned to them in these arrangements. The Company's potential partners may have significant discretion in determining how to pursue planned activities and the Company may have limited control over the quality and nature of the efforts and resources that such a partner applies to the collaboration as well as the branding and marketing of Orphazyme and its products. The Company cannot be certain that any collaborations will be scientifically or commercially successful or that the Company will receive revenues from any collaboration agreements.

Any of these events could adversely affect the Company's business, financial condition, results and prospects.

15. Adverse events, product liability and other claims may have material adverse effects on Orphazyme.
Companies in the pharmaceutical industry, such as Orphazyme, are generally subject to risks related to potential adverse events and product liability litigation. When conducting clinical trials, there is always the risk of participants being injured or experiencing adverse effects. Any such adverse effects may have a material negative impact on the clinical trials and thereby on the Company's business, financial condition, results and prospects.

Testing for adverse events is part of clinical testing of pharmaceutical products. Adverse events may occur in connection with clinical testing of products, but can also occur once the product is on the market. Adverse events must be handled in accordance with regulatory law and practice. If adverse effects occur during clinical testing, the Company may have to conduct additional testing, which will cause delays in the Company's development program and result in increased costs for the Company, or ultimately lead to the Company abandoning the development of the product in question. If adverse events occur once the product is on the market, there is a risk that Orphazyme may have to recall and destroy products, and ultimately there is a risk of fines, suspension or withdrawal of regulatory approvals and the Company becoming involved in litigation. The occurrence of any such event could have a material adverse effect on the Company's business, financial condition, results and prospects.

Product liability risks are inherent in development, marketing and sale of pharmaceutical products. Although Orphazyme is not currently subject to any product liability claims, such claims could arise at a later date. Litigation could be time-consuming for the Company and lead to significant costs and losses for the Company.

Orphazyme has taken out product liability insurance in respect of all clinical trials it has performed and is performing with respect to its products. If Orphazyme obtains marketing authorization for one or more of its products, it intends to take out insurance coverage appropriate for the commercialization of its products. However, there can be no assurance that such insurance cover will be available on reasonable commercial terms or that it will prove adequate. If sufficient insurance cover is not obtained covering, for instance, product liability, Orphazyme could be subject to significant liabilities which could have material negative impact on the Company's business, financial condition, results and prospects.

Orphazyme may from time to time become involved in various litigation matters and governmental or regulatory investigations, prosecutions or similar matters arising out of its current or future business. Orphazyme cannot accurately anticipate the liabilities which may adversely affect its business, and its insurance or indemnities may not cover all claims that may be asserted against it, and any claims asserted against it, regardless of merit or eventual outcome, may harm Orphazyme's reputation.

There is no guarantee that Orphazyme will be successful in defending itself in future litigation or similar matters under various laws. Should the ultimate judgments or settlements in any such future litigation or investigation significantly exceed Orphazyme's insurance coverage, they could have a material adverse effect on the Company's business, financial condition, results and prospects.

16. Changes in applicable laws and regulation in the jurisdictions in which the Company operates, or involvement in legal disputes, may have a material adverse effect on the Company's business, financial condition, results and prospects.
Future legal or regulatory changes in jurisdictions where the Company currently operates, or in such jurisdictions in which the Company may choose to operate in the future, could materially and adversely affect the Company's business, financial condition, results and prospects, including by imposing regulatory and operational restrictions and compliance obligations on the Company's business, reducing the Company's revenue or increasing the Company's expenses. For instance, changes in applicable laws in the following areas may have an impact on Orphazyme's operations: Orphan drugs; clinical trials; use of data; animal testing; regulatory approval processes; requirements to production; marketing, sales and pricing or pharmaceutical products; pharmacovigilance and other regulatory requirements; and intellectual property rights.

Orphazyme is not currently involved in any legal disputes with third parties or regulatory authorities and to Orphazyme's knowledge no such disputes are threatening. However, the Company may in the future become involved in legal disputes, proceedings and investigations which may be costly and time-consuming and any claims asserted against Orphazyme, regardless of merit or eventual outcome, may harm its reputation. There is no guarantee that Orphazyme will be successful in defending itself in its any future unknown litigation or similar matters under various laws. Should the ultimate judgments or settlements in any such future litigation or investigation exceed Orphazyme's insurance coverage, they could have a material adverse effect on the Company's business, financial condition, results and prospects.

17. The Company faces risks related to data privacy concerns, cyber security breaches and failure to comply with privacy regulations and security requirements relating to data.
The Company processes sensitive personal data, including information from clinical trials, and health data obtained in connection with reporting of adverse events. The Company is subject to data protection laws, privacy requirements and other regulatory restrictions in the various jurisdictions in which the Company operates.

The Company's failure to keep apprised of, and comply with, privacy, data use and security laws, standards and regulations, including, for instance, unauthorized disclosure of or access to data, could result in the suspension or revocation of the Company's approvals or registrations, the limitation, suspension or termination of services or the imposition of administrative, civil or criminal penalties, including fines which may, after the EU General Data Protection Regulation enters into force in May 2018, be as high as or up to EUR 20 million for serious infringements or 4% of the annual worldwide turnover of an undertaking. In addition, such failure or non-compliance may cause existing or potential partners, including hospitals, physicians and patients to cease interacting with the Company, and could damage the Company's reputation and brand. In addition, to the extent more restrictive laws, rules or security requirements relating to business and personal data are adopted in the future in the various jurisdictions in which the Company operates, such changes could have an adverse impact on the Company by increasing its costs or imposing restrictions on its business processes. Accordingly, the Company's failure to keep apprised of, and comply with, privacy, data use and security laws, standards and regulations could have a material adverse effect on the Company's reputation, business, financial condition, results and prospects. The Company's financial exposure to any actual or alleged breach of such regulations or standards may either not be insured against or not fully covered through any insurance maintained by the Company.

Cyber security attacks on the Company's servers, information systems and databases, or third party servers, information systems and databases on which the Company's information is stored, could compromise the security of its data or could cause interruptions in the operations of its businesses. Notwithstanding safeguards, cyber security breaches, internal security breaches, physical security breaches or other unauthorised or accidental access to the Company's servers, other information systems or databases could result in tampering with, or the theft or publication of, sensitive information or the deletion or modification of data, or could otherwise cause interruptions in the Company's operations.

The tampering with, disruption to, or the theft or publication of, sensitive information or the deletion or modification of records held either in the Company's systems or the systems of others to which the Company has access, could subject the Company to increased costs and exposure to litigation. The loss of confidential information could result in the payment of damages and reputational harm and have a material adverse effect on the Company's business, financial condition, results of operations and prospects.

The Company's financial exposure from the items referenced above may either not be insured against or not fully covered through any insurance maintained by the Company and could have a material adverse effect on the Company's business, financial condition, results and prospects.

18. The Company may not be able to attract, integrate, manage and retain qualified personnel or key employees.

The success of Orphazyme's business depends on its ability to successfully develop and commercialize its products. Since Orphazyme's organization currently consists of a limited number of employees with additional personnel hires planned for the years to come, Orphazyme's ability to successfully develop and commercialize its products will depend on recruiting a range of specialist personnel, particularly in the areas of development of new products, planning and managing clinical programs and commercialization of pharmaceutical products, and also requires Orphazyme to retain and develop the necessary qualified personnel who can provide the needed expertise to support Orphazyme's business and operations. The market for qualified personnel is competitive and Orphazyme may not succeed in recruiting personnel to, for instance, commercialize its products as currently envisaged, or it may fail to effectively replace current personnel who depart with qualified or effective successors. Orphazyme's effort to retain and develop personnel may also result in significant additional expenses, which could adversely affect its profitability. Orphazyme cannot assure that key personnel, including its senior management such as the CEO, the CFO, the CMO or the CSO, will continue to be employed or that it will be able to attract and retain qualified personnel in the future. Failure to retain or attract key personnel could have a material adverse effect on the Company's business, financial condition, results and prospects.

19. Risks relating to priority review voucher.

As further described, see description under "*Business*", the Company intends to seek, where possible, to obtain rare pediatric disease priority review vouchers from the FDA, although this is not part of the Company's core strategy. The priority review voucher program was created by the US Congress in 2007 to encourage development of drugs for neglected tropical diseases, but only a limited number of priority review vouchers have been awarded by the FDA. Any voucher award by the FDA requires i.a. that rare pediatric disease drug itself receives approval from the FDA and there are additional criteria that must be fulfilled. If such priority review vouchers are granted, the Company may use the voucher for its own FDA approval processes or decide to sell the voucher to other biotech or pharmaceutical companies. There is no established market for priority review vouchers and disclosed sales prices may not be indicative of the current value of vouchers. The term of the rare pediatric disease priority review vouchers program expires in 2020 and may not be renewed or may be amended or canceled prior to such expiry. Hence it may be unavailable to the Company even if all requirements are met. Further, the award of a voucher will trigger an obligation to market the relevant rare pediatric disease product within one year from FDA approval or else the FDA may revoke the voucher. Finally, a voucher award subjects the Company to post marketing reporting obligations to the FDA.

20. The Company's results of operations may be adversely affected by changes in foreign currency exchange rates and interest rates.
Substantially all of Orphazyme's income is expected to be in USD and EUR, while part of its operating costs is currently denominated in DKK, although in the future such DKK denominated operating costs are likely to constitute a smaller percentage of the total operating costs. Orphazyme does not currently have in place hedging contracts to cover its currency risks and, accordingly, fluctuations in DKK against, in particular USD, could have an adverse effect on the Company's business, financial position, results and prospects.

The Company's interest rate risk mainly derives from the fact that following the Offering, the Company will hold a large cash position. Significant negative changes in interest rates could therefore affect the value of the Company's funds and any placement thereof and may thereby adversely affect the Company's business, financial condition, results and prospects.

21. The Company may require additional capital in the future, which may not be available to it on commercially favourable terms, or at all.
The Company is currently loss-making and the Company may need to raise additional capital. The Company has so far been financed by funds invested by the Company's shareholders with a shareholder base which has expanded since 2010. Based on the current operating plan and the existing capital resources together with the minimum proceeds from the Offering, the Company expects to be able to fund its operating plan for at least 12 months. However, the operating plan may change as a result of many factors currently unknown, and it may be necessary to seek additional funds sooner than planned. The future funding requirements will depend on many factors, including, but not limited to, the progress, timing, scope, results and costs of pre-clinical studies and clinical trials for the Company's products in development, including the ability to enroll patients in a timely manner for clinical trials as well as the time and cost necessary to obtain regulatory approvals for the Company's products. In addition, funding requirements will also depend on the progress in commercialization and promotion of the Company's products and the efforts to develop and commercialize the Company's other existing products in development as well as the manufacturing, selling and marketing costs associated with the products, including the cost and timing of building sales and marketing capabilities. This extends to the sales price and the availability of adequate third-party coverage and reimbursement for the Company's products; the number and scope of pre-clinical and discovery programs that the Company may decide to pursue or initiate, the time and cost necessary to respond to technological and market developments and the costs of filing, prosecuting, maintaining, defending and enforcing any patent claims and other intellectual property rights, including litigation costs and the outcome of such litigation, including costs of defending any claims of infringement brought by others in connection with the development, manufacture or commercialization of the Company's products.

The Company may seek to raise new capital in the future through loans, public or private debt or equity financings by issuing additional Shares or other preferred financing shares, debt or equity securities convertible into Shares, or rights to acquire these securities, and exclude the pre-emption rights pertaining to the then-outstanding Shares.

Any additional financing that the Company could seek may not be available on favourable terms or at all, which could adversely affect the Company's future plans and its ability to execute its strategy and could have a material adverse effect on the Company's business, financial condition, results and prospects.

22. The projected financial information included in this Offering Circular may differ materially from the Company's actual results, and investors should not place undue reliance on it.
The financial projections set forth in this Offering Circular, including under "*Prospective Financial Information for the Financial Year Ending 31 December 2017*" and elsewhere, are Orphazyme's projections for the financial year 2017. The "*Prospective Financial Information for the Financial Year Ending 31 December 2017*" includes financial projections that qualify as profit forecasts. For profit forecasts, the Prospectus Regulation requires Orphazyme, among other things, to disclose the principal assumptions on which Orphazyme bases the forecast and to include a report prepared by Orphazyme's independent auditors, EY (as defined herein), on such forecasts and assumptions. EY did not make any assessment as to whether the assumptions underlying these financial projections are well-founded or whether such financial projections are realisable. Orphazyme has prepared its financial projections in accordance with the Prospectus Regulation. These financial projections are based upon a number of assumptions, which are inherently subject to significant business, operational, economic and other risks, many of which are outside of Orphazyme's control. Accordingly, such assumptions may change or may not materialise at all. In addition, unanticipated events may adversely affect the actual results and cash flows that the Company achieves in future periods whether or not its assumptions relating to the financial year 2017 otherwise prove to be correct. As a result, the Company's actual results and cash flows may vary materially from these projections, and investors should not place undue reliance on them. See also "*Special Notice Regarding Forward-Looking Statements*".

Risks Relating to Intellectual Property Rights

23. If Orphazyme is unable to obtain and maintain protection for relevant intellectual property rights, the value of Orphazyme's products will be significantly and adversely affected.
Orphazyme's success is to a large extent dependent on its ability to obtain and maintain patents and other intellectual property rights for Orphazyme's products. Orphazyme's ability to obtain and maintain such rights may be influenced by a number of factors. For instance, patents issued or licensed to Orphazyme may be challenged and/or be invalid or unenforceable or circumvented. Other risks include patents not being issued to Orphazyme based on applications that are currently pending or the scope of the claims being narrowed during the examination process; future products not being patentable; the scope of any patent protection not being sufficiently broad to exclude other competitors; and that others may claim rights to patents and other proprietary rights which Orphazyme holds or licenses.

The patent position of biotech and pharmaceutical companies, including Orphazyme, is generally uncertain and comprises complex legal and factual issues. If Orphazyme fails to obtain and maintain patent protection for Orphazyme's products, Orphazyme could lose its competitive advantage, and the competition Orphazyme faces would increase, which would have a material adverse effect on the Company's business, financial condition, results and prospects.

24. Orphazyme may not be able to successfully enforce and defend its intellectual property rights.
The enforcement and defense of Orphazyme's intellectual property rights, including patent rights, through legal or administrative proceedings may be costly and time-consuming, may divert Orphazyme's personnel from their usual responsibilities and may provide Orphazyme's competitors and others with insights into Orphazyme's proprietary rights. Moreover, there can be no assurance that Orphazyme will have sufficient financial or other resources to conduct such enforcement or defense actions. An adverse determination in any litigation or other proceeding could put one or more of Orphazyme's patents at risk of being invalidated or interpreted narrowly and could put its pending patent applications at risk of not being issued. The occurrence of any of the above could have a material adverse effect on the Company's business, financial condition, results and prospects.

25. Orphazyme may face infringement claims and other challenges by third parties.
Orphazyme may be subject to time-consuming infringement actions and could incur significant costs if third parties believe that Orphazyme's products, or the methods used to manufacture or use them, infringe patents or other proprietary rights held by such third parties. Orphazyme and its patent advisers, when performing freedom to operate searches as part of the development of a patent strategy or preparing and processing patent applications, may fail to identify relevant prior art. Should Orphazyme be met with infringement claims or other challenges of its intellectual property rights by third parties, an adverse outcome could be costly, time-consuming and may subject Orphazyme to significant liabilities, and force the Company to curtail or cease the development, marketing and sale of some or all of its products or lead to significant costs for developing non-infringing products or licensing technology or products from the party claiming infringement (which license may not be available on commercially reasonable terms or at all) which could have a material adverse effect on the Company's business, financial condition, results and prospects.

26. The previous employers of Orphazyme's employees and consultants may attempt to assert rights over Orphazyme's intellectual property.
The vast majority of Orphazyme's employees and consultants were previously employed at universities or biopharmaceutical or pharmaceutical companies, including competitors and potential competitors to Orphazyme. The Company may be subject to claims that it or these employees have, inadvertently or otherwise, used or disclosed intellectual property, trade secrets or other proprietary information of their former employers. Such claims may lead to material costs for the Company, or an inability of the Company to protect or use its intellectual property rights, which could have a material adverse effect on the Company's business, financial condition, results and prospects.

27. The Company will not seek to protect its intellectual property rights in all jurisdictions throughout the world and may not be able to adequately enforce its intellectual property rights even in the jurisdictions where protection is sought.
Filing, prosecuting and defending patents on the products in all countries and jurisdictions throughout the world would be prohibitively expensive, and the intellectual property rights in some countries could be less extensive than those in the EU or the United States, assuming that rights are obtained in the EU and the United States. Competitors may use the Company's technologies in such jurisdictions to develop their own products and, further, may export otherwise infringing products to territories where the Company has patent protection, but enforcement is not as strong as that in the EU or the United States.

In addition, the laws of some countries do not protect intellectual property rights to the same extent as in the EU and the United States. Many companies have encountered significant problems in protecting and defending intellectual property rights in certain jurisdictions. The legal systems of some countries, particularly developing countries, do not favor the enforcement of patents and other intellectual property protection, especially those relating to biopharmaceuticals or biotechnologies. This could make it difficult to stop the infringement of the Company's

patents, if obtained, or the misappropriation of other intellectual property rights. For example, many countries have compulsory licensing laws under which a patent owner under certain conditions must grant licenses to third parties. In addition, many countries limit the enforceability of patents against third parties, including government agencies or government contractors. In these countries, patents may provide limited or no benefit. Patent protection must ultimately be sought on a country-by-country basis, which is an expensive and time-consuming process with uncertain outcomes.

Such lack of patent protection may lead to material costs for the Company, or an inability of the Company to protect or use its intellectual property rights, which could have a material adverse effect on the Company's business, financial condition, results and prospects.

28. If Orphazyme is unable to protect the confidentiality of certain information, the value of its products and technology could be materially adversely affected.

Orphazyme's know-how, trade secrets and other intellectual property, including the ability to protect its intellectual property, are significant to the Company. In addition to patented products and technology, Orphazyme relies upon unpatented proprietary technology processes, know-how and data that Orphazyme regards as trade secrets. Orphazyme seeks to protect its trade secrets in part through confidentiality agreements with employees, consultants and third parties. These agreements may be breached, and Orphazyme may not have adequate remedies for any such breach. In addition, Orphazyme's trade secrets may otherwise become known or be independently developed by competitors in a manner providing Orphazyme with no practical recourse against the competing parties. If any such events were to occur, there could be a material adverse effect on the Company's business, financial condition, results and prospects.

Risks Relating to the Offering

29. Following the Offering, the Main Shareholders will continue to be shareholders and may be able to influence important actions the Company takes.

Following the Offering, the Main Shareholders will continue to be shareholders and may be able to influence the outcome of decisions at general meetings and some will remain represented on the Board of Directors, which may influence important actions the Company takes. This concentration of share ownership could have the effect of delaying, postponing or preventing a change of control in the Company, and impact mergers, consolidations, acquisitions or other forms of combinations, which may or may not be desired by other shareholders. No assurances can be given that the interests of the Main Shareholders, or investors directly or indirectly controlling the Main Shareholders, will not differ from the interests of other shareholders. The interests of the Main Shareholders may not be aligned with the interests of minority shareholders with respect to such voting decisions.

30. There is limited free float in the Shares.

The Main Shareholders' and Cornerstone Investors' respective shareholdings following the completion of the Offering may affect the demand for the Shares. If the Main Shareholders and Cornerstone Investors continue to hold on to their respective holdings of Shares, this may affect the liquidity of the Shares, may impair the ability of investors to sell their Shares at the times or volumes they may wish to do so and may increase the volatility of the price of the Shares. In addition, the Main Shareholders' and Cornerstone Investors' share ownership may adversely affect the trading price of the Shares because investors often perceive disadvantages in owning shares in companies with large shareholders.

31. The Company's Shares have not previously been publicly traded, and their price may be volatile and fluctuate.

There is currently no public market for the Company's Temporary Purchase Certificates and/or Shares, and an active and liquid trading market may not develop or be sustained after the Offering. If an active and liquid trading market does not develop or is not sustained, the liquidity and trading price of the Temporary Purchase Certificates and/or Shares could be materially and adversely affected, and investors may have difficulty selling their Temporary Purchase Certificates and/or Shares. The market price of the Temporary Purchase Certificates and/or Shares may subsequently vary from the Offer Price and may be higher or lower than the price paid by investors. The trading price of the Temporary Purchase Certificates and/or Shares may fluctuate in response to many factors, including extraneous factors beyond the Company's control, which may include, but are not limited to, variations in the Company's and its competitors' actual or anticipated operating results; variations in operating results that vary from the expectations of securities analysts that follow the Company's shares and investors; guidance, if any, that the Company provides to the public, any changes in this guidance, or the Company's failure to meet this guidance; substantial trading in the Company's shares; changes in general economic or market conditions or trends in the Company's industry or the economy as a whole; the public's response to press releases or other public announcements by the Company or third parties; changes in laws and regulation, announcements relating to litigation or governmental investigations (including those of competition authorities); announcements by the Company, its competitors or third parties; future changes to accounting principles; general market and economic conditions and events such as system failures and disruptions; technology changes or changes in patient behavior; natural disasters, war or acts of terrorism. In addition, Nasdaq Copenhagen or the global securities markets may experience significant price and volume fluctuations, as they have done in recent years, which may have a material adverse effect on the market price of the Temporary Purchase Certificates and/or Shares and create a risk that investors may not be able to sell their Temporary Purchase Certificates and/or Shares at the Offer Price or a higher price.

32. Differences in exchange rates may materially adversely affect the value of shareholdings or dividends paid.
The Shares will be denominated in Danish kroner only, and any dividends will be paid in Danish kroner. As a result, shareholders outside Denmark may experience material adverse effects on the value of their shareholding and their dividends when converted into other currencies if the Danish kroner depreciates against the relevant currency.

33. The Company intends to retain all available funds and any future earnings and reinvest such in the Company and, consequently, the shareholders' ability to achieve a return on their investment may depend on appreciation in the price of the Shares.
The Company has never declared or paid any cash dividends, and it is the intention to retain all available funds and any future earnings to fund the development and expansion of the Company's business. Therefore, the shareholders are not likely to receive any dividends on their Shares for the foreseeable future and the success of an investment in Shares will depend upon any future appreciation in its value. Consequently, investors may need to sell all or part of their holdings of Shares after price appreciation, which may never occur, as the only way to realize any future gains on their investment. There is no guarantee that the Shares will appreciate in value or even maintain the price at which investors have purchased them. Investors seeking cash dividends should not purchase the Shares.

34. If securities or industry analysts do not publish research or publish inaccurate or unfavorable research about our business, the price of the Shares and their trading volume could decline.
The trading market for the Shares depends in part on the research and reports that securities or industry analysts publish about the Company. As a newly listed public company, it is expected that a limited number of securities analysts publish research reports about the Company. In the future, if no or only limited securities or industry analysts cover the Company, the trading price for the Shares would be negatively impacted. If one or more of the analysts who covers Orphazyme downgrades the Shares or publishes inaccurate or unfavorable research about the Company, the price of Shares would likely decline. If one or more of these analysts ceases coverage of the Company or fails to publish reports on the Company regularly, or downgrades our Shares, demand for Shares could decrease, which could cause the price of the Shares or their trading volume to decline.

35. Future sales of Shares after the Offering may cause a decline in the market price of the Shares.
The market price of the Shares could decline as a result of sales of Shares by the Company, the Main Shareholders, Cornerstone Investors, Executive Management or the Company's other shareholders after the Offering or the perception that these sales could occur. These sales also may make it difficult for the Company to issue equity securities in the future at a time and a price that the Company deems appropriate. Following the Offering, the Company, the Main Shareholders and certain members of the Company's management will be subject to certain contractual lock-up provisions, in each case for a limited period only and subject to important exceptions. After the expiry of the applicable lock-up periods, see "*Plan of Distribution—Lock-up Arrangements*", the Main Shareholders and certain members of the Company's management could sell their respective holdings of Shares in whole or in part. In addition, the Company could offer to sell new Shares in public or private transactions. Any such future sales by the Company could dilute the ownership interests of the Company's then-existing shareholders, and sales by the Company, the Main Shareholders, Executive Management or the Company's other shareholders, or the perception that such sales could occur, could adversely affect the trading price of the Shares.

36. The issuance of additional Shares in the Company to fund future operations, development and commercialisation of its products, or to fund acquisitions, any share incentive or share option plan or otherwise may dilute existing shareholdings.
The Company may seek to raise financing to fund future operations, development and commercialisation of its products, or to fund acquisitions and other growth opportunities, invest in its business, or for general corporate purposes. In particular, this funding need may increase and occur sooner than the Company currently expects, for instance if the Company experiences delays or other issues with its drug development. The Company may, for these and other purposes, such as in connection with share incentive and warrant and/or share option plans, issue additional equity or convertible equity securities. For a description of the Company's pre-IPO warrant programme and contemplated share-based long-term incentive programme, see "*Board of Directors, Exective Management and Key Employees—Incentive programmes*". As a result, the Company's existing shareholders may suffer dilution in their percentage ownership or the price of the Shares may be adversely affected.

37. Shareholders in the United States and other non-Danish jurisdictions may not be able to participate in future equity offerings.
The Articles of Association provide for pre-emptive rights to be granted to shareholders, unless such rights are disapplied by a shareholder resolution at a general meeting or the Shares are issued on the basis of an authorisation to the Board of Directors under which the Board of Directors may disapply the pre-emption rights. See "*Description of the Shares and Share Capital—Authorisation to Increase the Share Capital*" for a description of the authorisation to increase the share capital which has been granted to the Board of Directors.

However, securities laws of certain jurisdictions may restrict the Company's ability to allow participation by shareholders in future offerings. In particular, shareholders in the United States or in certain other jurisdictions may not be entitled to exercise these rights unless either the rights and the Shares are registered under the U.S. Securities Act, or the rights and the Shares are offered pursuant to an exemption from, or transaction not subject to, the registration requirements of the U.S. Securities Act, or equivalent local securities laws.

In such cases, shareholders resident in such non-Danish jurisdictions may experience a dilution of their shareholding, possibly without such dilution being offset by any compensation received in exchange for subscription rights. The Company cannot assure prospective investors that any exemption from such overseas securities law requirements would be available to enable shareholders in the United States or certain other jurisdictions to exercise their pre-emption rights or, if available, that the Company will utilise any such exemption.

38. Investors' rights as shareholders will be governed by Danish law and differ in some respects from the rights of shareholders under the laws of other countries.

As a listed public company, the Company will be organised under the laws of Denmark. The rights of holders of the Shares will be governed by the Articles of Association and by Danish law. These rights may differ in some respects from the rights of shareholders in corporations organised outside Denmark. In addition, it may be difficult for investors to prevail in a claim against the Company under, or to enforce liabilities predicated upon, the securities laws of jurisdictions outside Denmark.

39. Transformation into a listed public company will increase the Company's costs and may disrupt the regular operations of its business.

The Company expects to incur additional legal, regulatory, finance, accounting, investor relations and other administrative expenses as a result of having publicly traded Shares. The additional demands associated with being a public company may disrupt regular operations of the Company's business by diverting the attention of some of the Company's senior management team away from operational activities to management and administrative oversight, adversely affecting the Company's ability to attract and complete business opportunities and increasing the difficulty in both retaining professionals and managing and growing the Company's businesses. In addition, failure to comply with any laws or regulations applicable to the Company as a public company may result in legal proceedings and/or regulatory investigations, and may cause reputational damage. Any of these effects could harm the Company's business, financial condition, results and prospects.

40. The Offering may be withdrawn after Admission to trading of the Temporary Purchase Certificates and until settlement of the Offering.

As described in "*The Offering—Withdrawal of the Offering*", the Underwriting Agreement (as defined herein) contains a provision entitling the Joint Global Coordinators to terminate the Offering (and the arrangements associated with it) after admission of the Temporary Purchase Certificates to trading on Nasdaq Copenhagen (expected on or around 17 November 2017) and prior to settlement of the Offering by delivery and payment of the Temporary Purchase Certificates representing the Offer Shares (expected on or around 21 November 2017). Such termination rights may only be exercised under certain circumstances, including force majeure and material changes in the financial condition of the Company's business. Such termination rights will lapse upon settlement of the Offering, currently expected to take place on 21 November 2017, except in respect of the Overallotment Shares. The termination rights of the parties to the Underwriting Agreement will lapse, in respect of the Overallotment Shares, upon settlement of the subscription for the Overallotment Shares, if the Overallotment Option is exercised.

Nasdaq Copenhagen's approval of the Admission is subject to such termination rights not having been exercised after pricing and prior to settlement of the Offering (excluding any termination rights in respect of the Overallotment Option).

The Underwriting Agreement contains closing conditions which the Company believes are customary for offerings such as the Offering. In addition, the Company has given customary representations and warranties to the Joint Global Coordinators. The completion of the Offering is dependent on compliance with all of the closing conditions set forth in the Underwriting Agreement. If one or more closing conditions are not met, the Joint Global Coordinators may, at their discretion, withdraw the Offering.

If the Offering is terminated or withdrawn, the Offering and any associated arrangements will lapse, all submitted orders will be automatically cancelled, any monies received in respect of the Offering will be returned to the investors without interest (less any transaction costs) and admission to trading and/or official listing of the Temporary Purchase Certificates or the Shares on Nasdaq Copenhagen will be cancelled. Consequently, any trades in the Temporary Purchase Certificates and/or Shares effected on or off the market before settlement of the Offering may subject investors to liability for not being able to deliver the Temporary Purchase Certificates and/or Shares sold, and investors who have sold or acquired Temporary Purchase Certificates and/or Shares on or off the market may incur a loss. All dealings in the Temporary Purchase Certificates and/or Offer Shares prior to settlement of the Offering are for the account of, and at the sole risk of, the parties concerned.

Important Notice relating to the Offering Circular

In this Offering Circular, the "**Company**" or "**Orphazyme**" refers to Orphazyme A/S registered under (CVR) no. 32266355.

No representation or warranty, express or implied, is made by Carnegie Investment Bank, filial af Carnegie Investment Bank AB (publ), Sverige, or Danske Bank A/S (together, the "**Joint Global Coordinators**", the "**Joint Bookrunners**"), or Oddo BHF SCA (the "**Co-Lead Manager**" and together with the Joint Global Coordinators, the "**Managers**") as to the accuracy or completeness of any information contained in this Offering Circular.

The information in this Offering Circular is as of the date printed on the front of the cover, unless expressly stated otherwise. The delivery of this Offering Circular at any time does not imply that there has been no change in Orphazyme's business or affairs since the date hereof or that the information contained herein is correct as of any time subsequent to the date hereof. In the event of any changes to the information in this Offering Circular that may affect the valuation of the Offer Shares during the period from the date of announcement to the first day of trading, such changes will be announced pursuant to the rules of the Danish Executive Order on Prospectuses, *inter alia*, which governs the publication of prospectus supplements.

For purposes of the Offering, the Company has prepared a prospectus in English (including a Danish translation of the summary) in compliance with the standards and requirements of Danish law. The registration document, which forms a part of this Offering Circular, has been prepared in accordance with Annex XXV of the Prospectus Regulation, which is applicable to small- and medium-sized enterprises.

In making an investment decision, investors must rely on their own assessment of the Company and the terms of this Offering, as described in this Offering Circular, including the merits and risks involved. Any purchase of the Offer Shares should be based on the assessments of the information in the Offering Circular that the investor in question may deem necessary, including the legal basis and consequences of the Offering, and including possible tax consequences that may apply, before deciding whether or not to invest in the Offer Shares. Investors should rely only on the information contained in this Offering Circular, including the risk factors described herein.

The Offering will be completed under Danish law, and none of the Main Shareholders, the Managers or the Company have taken any action or will take any action in any jurisdiction with the exception of Denmark that may result in a public offering of the Offer Shares.

No person has been authorised to give any information or make any representation not contained in this Offering Circular and, if given or made, such information or representation must not be relied upon as having been authorised by the Managers or the Company. Neither the Company nor the Managers accept any liability for any such information or representation.

The distribution of this Offering Circular and the offer or sale of the Offer Shares in certain jurisdictions are restricted by law. By purchasing Offer Shares, investors will be deemed to have made certain acknowledgements, representations and agreements as described in this Offering Circular. Prospective investors should be aware that they may be required to bear the financial risks of any such investment for an indefinite period of time. No action has been or will be taken by the Managers or the Company to permit a public offering in any jurisdiction other than Denmark. Persons into whose possession this Offering Circular may come are required by the Managers and the Company to inform themselves about and to observe such restrictions. This Offering Circular may not be used for, or in connection with, any offer to, or solicitation by, anyone in any jurisdiction or under any circumstances in which such offer or solicitation is not authorised or is unlawful. For further information with regard to restrictions on offers and sales of the Offer Shares and the distribution of this Offering Circular, see "*Selling Restrictions*". This Offering Circular does not constitute an offer to sell or a solicitation of an offer to buy any of the Offer Shares in any jurisdiction to any person to whom it would be unlawful to make such an offer. This Offering Circular may not be forwarded, reproduced or in any other way redistributed by anyone but the Managers and the Company. Investors may not reproduce or distribute this Offering Circular, in whole or in part, and investors may not disclose the content of this Offering Circular or use any information herein for any purpose other than considering the purchase of Offer Shares. Investors agree to the foregoing by accepting delivery of this Offering Circular.

The Managers are acting for the Company and no one else in relation to the Offering and admission to trading and/or official listing of the Temporary Purchase Certificates and/or the Shares on Nasdaq Copenhagen. The Managers will not be responsible to anyone other than the Company for providing the protections afforded to clients of the Managers, or for providing advice in relation to the Offering and admission to trading and/or official listing of the Temporary Purchase Certificates and/or the Shares on Nasdaq Copenhagen.

Stabilisation

IN CONNECTION WITH THE OFFERING, DANSKE BANK AS THE STABILISING MANAGER, OR ITS AGENTS, ON BEHALF OF THE MANAGERS, MAY ENGAGE IN TRANSACTIONS THAT STABILISE, MAINTAIN OR OTHERWISE AFFECT THE PRICE OF THE SHARES FOR UP TO 30 DAYS FROM THE COMMENCEMENT OF TRADING OF THE TEMPORARY PURCHASE CERTIFICATES ON NASDAQ COPENHAGEN. SPECIFICALLY, THE MANAGERS MAY OVERALLOT OFFER SHARES OR EFFECT TRANSACTIONS WITH A VIEW TO SUPPORTING THE MARKET PRICE OF THE SHARES AT A LEVEL HIGHER THAN THAT WHICH MIGHT OTHERWISE PREVAIL. THE STABILISING MANAGER AND ITS AGENTS ARE NOT REQUIRED TO ENGAGE IN ANY OF THESE ACTIVITIES AND, AS SUCH, THERE IS NO ASSURANCE THAT THESE ACTIVITIES WILL BE UNDERTAKEN; IF UNDERTAKEN, THE STABILISING MANAGER OR ITS AGENTS MAY END ANY OF THESE ACTIVITIES AT ANY TIME AND THEY MUST BE BROUGHT TO AN END AT THE END OF THE 30-DAY PERIOD MENTIONED ABOVE. SAVE AS REQUIRED BY LAW OR REGULATION, THE STABILISING MANAGER DOES NOT INTEND TO DISCLOSE THE EXTENT OF ANY STABILISATION TRANSACTIONS UNDER THE OFFERING. SEE "*PLAN OF DISTRIBUTION*".

Special Notice regarding Forward-Looking Statements

Certain statements in this Offering Circular constitute forward-looking statements. Forward-looking statements are statements (other than statements of historical fact) relating to future events and Orphazyme's anticipated or planned financial and operational performance. The words "targets", "believes", "expects", "aims", "intends", "plans", "seeks", "will", "may", "might", "anticipates", "would", "could", "should", "continues", "estimates" or similar expressions or the negatives thereof, identify certain of these forward-looking statements. Other forward-looking statements can be identified in the context in which the statements are made. Forward-looking statements appear in a number of places in this Offering Circular, including, without limitation, under the headings "*Summary*", "*Risk Factors*", "*Dividends and Dividend Policy*", "*Business*" and "*Operating and Financial Review*", and include, among other things, statements addressing matters such as:

- Orphazyme's future results of operations, in particular, the statements relating to Orphazyme's expectations and estimates for the financial year 2017;
- Orphazyme's product pipeline and commercialization thereof;
- Orphazyme's business strategy, plans and objectives for future products and services, future operations and events;
- the competitive environment in which Orphazyme operates;
- Orphazyme's financial condition;
- Orphazyme's working capital, cash flow and capital expenditure; and
- general economic trends and trends in Orphazyme's industry.

Although Orphazyme believes that the expectations reflected in these forward-looking statements are reasonable, such forward-looking statements involve known and unknown risks, uncertainties and other important factors that could cause Orphazyme's actual results, performance, achievements or industry results to differ materially from any future results, performance or achievements expressed or implied by such forward-looking statements. Such risks, uncertainties and other important factors include, among others:

- competition from other life sciences companies developing other treatments for similar diseases to those targeted by Orphazyme's products in development;
- changes in the regulatory and compliance environment;
- pricing and demand for pharmaceutical products;
- Orphazyme's ability to determine prices and thus generate revenue from any products that it may develop will depend on enacted and future reimbursement and drug pricing policies and regulations;
- the clinical and commercial success of Orphazyme's products in development;
- the results and timing of clinical trials conducted regarding Orphazyme's products in development, including the risk of delays with product development and/or clinical trials;
- undesirable side effects caused by Orphazyme's products;
- challenges in recruiting patients for clinical trials and concluding agreements with investigators and hospitals;
- obtaining and maintaining required regulatory approvals;
- obtaining and maintaining orphan product designation or marketing exclusivity for its product candidates;
- risks relating to manufacturing, including use of hazardous materials as well as CMC;
- Orphazyme's relationship with certain third-party vendors;
- Orphazyme's ability to successfully commercialize its products;
- the success of potential collaborations with third parties for the development and commercialization of Orphazyme's products;
- risks related to data privacy concerns, cyber security breaches and failure to comply with privacy regulations and security requirements relating to data;
- Orphazyme's ability to attract, integrate, manage and retain qualified personnel or key employees;
- material changes in certain foreign currency exchange rates and interest rates;
- risk related to intellectual property rights; and
- other factors referenced in this Offering Circular.

Should one or more of these risks or uncertainties materialise, or should any underlying assumptions prove to be incorrect, Orphazyme's actual financial condition, cash flows or results of operations could differ materially from what is described herein as anticipated, believed, estimated or expected. Orphazyme urges investors to read the sections of this Offering Circular entitled "*Risk Factors*", "*Business*", "*Operating and Financial Review*" and "*Prospective Financial Information for the Financial Year Ending 31 December 2017*" for a more complete discussion of the factors that could affect Orphazyme's future performance and the industry in which Orphazyme operates.

Orphazyme does not intend, and does not assume any obligation, to update any forward-looking statements contained herein, except as may be required by law or the rules of Nasdaq Copenhagen. All subsequent written and oral forward-looking statements attributable to Orphazyme or to persons acting on Orphazyme's behalf are expressly qualified in their entirety by the cautionary statements referred to above and contained elsewhere in this Offering Circular.

Enforcement of Civil Liabilities and Service of Process

Orphazyme is organised under the laws of Denmark. As a result, it may not be possible for investors to effect service of process upon the Company or any of its respective directors and officers or to enforce against any of the aforementioned parties a judgement obtained in a court outside Denmark.

Presentation of Financial and Certain other Information

This Offering Circular presents historical financial information comprising selected income statements, statements of financial position, statement of changes in equity and cash flow statements derived from Orphazyme's:

- audited financial statements at 31 December 2016 and for the financial year 1 January 2016 – 31 December 2016 with comparative figures for the period 1 July 2015 – 31 December 2015 (six month conversion period) and the period 1 July 2014 – 30 June 2015 (12 months period), prepared in accordance with the International Financial Reporting Standards as adopted by the European Union ("**IFRS**") and additional requirements of the Danish Financial Statements Act as included in the statutory annual report for 2016 and on pages F-14 - F-36 of this Offering Circular (the "**Audited Financial Statements**");
- reviewed unaudited financial statements for the period 1 January 2015 to 31 December 2015 comprising "Statement of Profit or Loss and Other Comprehensive Income", "Statement of Financial Position", "Statement of Changes in Shareholders Equity" and "Statement of Cash Flows" prepared based on the Company's accounting policies for recognition and measurement as set out on pages F-37 - F-46 of this Offering Circular (the "**2015 Comparative Financial Statements**"); and
- reviewed unaudited condensed interim financial statements for the period 1 January 2017 to 30 June 2017 with unaudited and non-reviewed comparative figures for the periode 1 January 2016 to 30 June 2016 prepared in accordance with the International Accounting Standard 34 on "Interim Financial Reporting" ("**IAS 34**") as set out in pages F-3 - F-13 of this Offering Circular (the "**Interim Financial Statements**").

The above financial statements have been audited or reviewed, as applicable, by Ernst & Young Godkendt Revisionspartnerselskab ("**EY**") to the extent stated in their independent auditor's report appearing therein. The functional currency of Orphazyme is translated into Danish kroner.

Orphazyme has previously undertaken statutory financial reporting based on a financial year covering the period from 1 July to 30 June; however, with effect from the calendar year 2016, Orphazyme's financial year has been converted to the period 1 January to 31 December. Accordingly, the 2015 Comparative Financial Statements have been prepared in order to present Orphazyme's historical financial information on a comparative basis with the statutory financial statements for the calendar year 2016. Consequently, the presentation of Orphazyme's financial performance and position in sections "*Business*", "*Selected Historical Financial and Operating Information*" and "*Operating and Financial Review*" hereof is based on (i) the audited financial statements for the financial year 2016 compared to the reviewed unaudited financial statements for the financial year 2015, and (ii) the reviewed interim financial statements for the period 1 January 2017 to 30 June 2017 compared to the non-reviewed unaudited comparative figures for the period 1 January 2016 to 30 June 2016 prepared in accordance with IAS 34. However, specific overview presentations of certain key financial figures in sections "*Selected Historical Financial and Operating Information*" and B.7 of the "*Summary*" hereof will also include historical financial information for the financial periods 1 July 2014 to 30 June 2015 and 1 July to 31 December 2015 accompanied by relevant references to the F-pages included in this Offering Circular.

Rounding adjustments

Rounding adjustments have been made in calculating some of the financial information included in this Offering Circular. As a result, figures shown as totals in some tables may not be exact arithmetic aggregations of the figures that precede them.

Foreign Currency Presentation

Foreign Currency Presentation

Orphazyme publishes its financial information in Danish kroner. Unless Orphazyme notes otherwise, all amounts in this Offering Circular are expressed in Danish kroner.

As used herein, references to (i) "Danish kroner" or "DKK" are to the Danish kroner, the lawful currency of Denmark; (ii) "euro", "EUR" or "€" are to the euro, the lawful currency of the participating member states in the Third Stage of the European and Monetary Union of the Treaty Establishing the European Community; (iii) "GBP" or "£" are to the British sterling pound, the lawful currency of the United Kingdom; and (iv) "U.S. dollar", "USD" or "$" are to the United States dollar, the lawful currency of the United States of America.

Exchange Rates

DKK/USD

The following table sets forth, for the periods and dates indicated, the average, high, low and period end U.S. dollar buying rates expressed in Danish kroner per one U.S. dollar, such data having been provided by the Danish Central Bank. The Danish Central Bank fixes exchange rates on the basis of information obtained from a number of central banks on a daily conference call hosted by the European Central Bank at 2:15 p.m. (CET). The average rates for each period represent the daily average of the U.S. dollar buying rates for such period.

	Reference Rates of Danish kroner per USD 1.00			
Calendar Year	**Average**	**High**	**Low**	**Period End**
2015	6.73	7.08	6.18	6.83
2016	6.73	7.17	6.43	7.05
2017 (through 1 November 2017)	6.65	7.16	6.17	6.41
Month				
January through 30 June 2017	6.88	7.16	6.52	6.52
July 2017	6.46	6.56	6.34	6.34
August 2017	6.30	6.36	6.17	6.29
September 2017	6.24	6.34	6.17	6.30
October 2017	6.33	6.41	6.28	6.39
November 2017 (through 1 November 2017)	6.41	6.41	6.41	6.41

DKK/EUR

The following table sets forth, for the periods and dates indicated, the average, high, low and period-end euro buying rates expressed in Danish kroner per one euro, such data having been provided by the Danish Central Bank. The Danish Central Bank fixes exchange rates on the basis of information obtained from a number of central banks on a daily conference call hosted by the European Central Bank at 2:15 p.m. (CET). The average rates for each period represent the daily average of the euro buying rates for such period. The exchange rate of Danish kroner per euro is regulated by the exchange rate mechanism, a system originally established in 1979 for controlling exchange rates within the monetary system of the EU. Under this system, Denmark sets its central exchange rate to 7.46 kroner per euro and allows fluctuations of the exchange rate within a 2.25% band. This means that the exchange rate can fluctuate from a high of DKK 7.63 per EUR 1.00 to a low of DKK 7.29 per EUR 1.00. If the market-determined floating exchange rate rises above or falls below the band, the Danish Central Bank must intervene.

	Reference Rates of Danish kroner per EUR 1.00			
Calendar Year	**Average**	**High**	**Low**	**Period End**
2015	7.46	7.47	7.43	7.46
2016	7.45	7.46	7.43	7.43
2017 (through 1 November 2017)	7.44	7.44	7.43	7.44
Month				
January through 30 June 2017	7.44	7.44	7.43	7.44
July 2017	7.44	7.44	7.44	7.44
August 2017	7.44	7.44	7.44	7.44
September 2017	7.44	7.44	7.44	7.44
October 2017	7.44	7.44	7.44	7.44
November 2017 (through 1 November 2017)	7.44	7.44	7.44	7.44

DKK/GBP

The following table sets forth, for the periods and dates indicated, the average, high, low and period-end British pound buying rates expressed in Danish kroner per GBP, such data having been provided by the Danish Central Bank. The average rates for each period represent the daily average of the GBP buying rates for such period.

	Reference Rates of Danish kroner per GBP 1.00			
Calendar Year	**Average**	**High**	**Low**	**Period End**
2015	10.28	10.72	9.49	10.11
2016	9.11	10.19	8.22	8.68
2017 (through 1 November 2017)	8.50	8.92	8.00	8.52
Month				
January through 30 June 2017	8.64	8.92	8.40	8.46
July 2017	8.39	8.48	8.30	8.32
August 2017	8.16	8.32	8.00	8.09
September 2017	8.32	8.50	8.08	8.44
October 2017	8.36	8.47	8.25	8.47
November 2017 (through 1 November 2017)	8.52	8.52	8.52	8.52

Exchange Controls and Other Limitations Affecting Shareholders of a Danish Company

There is no legislation in Denmark that restricts the export or import of capital (except for certain investments in specific areas as well as transactions involving entities or individuals subject to sanctions, all in accordance with applicable resolutions adopted by the United Nations and the European Union), including, but not limited to, foreign exchange controls, or which affects the remittance of dividends, interest or other payments to non-resident holders of the Offer Shares. As a measure to prevent money laundering and financing of terrorism, persons travelling into or out of Denmark carrying amounts of money (including, but not limited to, cash, traveller's cheques and securities) worth the equivalent of EUR 10,000 or more must declare such amounts to the Danish tax authorities when travelling into or out of Denmark.

Available Information

Copies of the following documents may be inspected and obtained during usual business hours on any day (excluding Saturdays, Sundays and Danish public holidays) at the Company's registered office at Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark during the period in which this Offering Circular is in effect:

(i) the Company's memorandum of association and the Articles of Association;
(ii) the Audited Financial Statements;
(iii) the 2015 Comparative Financial Statements and the Interim Financial Statements as included in this Offering Circular; and
(iv) this Offering Circular.

These documents are also, subject to certain restrictions with respect to the Offering Circular, available on Orphazyme's website.

The Danish Consolidated Act no. 1089 of 14 September 2015 on limited liability companies, as amended (the "**Danish Companies Act**") requires the Company to make its statutory annual reports, including the audited financial statements, available to its shareholders on the Company's website three weeks before the Company's annual general meeting. At the same time, the Company is required to send the notice convening the general meeting to registered shareholders who have so requested.

Market and Industry Information

This Offering Circular contains statistics, data and other information relating to markets, market sizes, market shares, market positions and other industry data pertaining to Orphazyme's business and markets. Unless otherwise indicated, such information is based on Orphazyme's analysis of multiple sources, including market studies that Orphazyme commissioned from Defined Health, a Cello Health business and Medical Marketing Economics, as well as the U.S. Food and Drug Administration, the European Medicines Agency and Evaluate Pharma.

While Orphazyme can confirm that information from external sources has been accurately reproduced, Orphazyme has not independently verified and cannot give any assurances as to the accuracy of market data as presented in this Offering Circular that was extracted or derived from these external sources. As far as Orphazyme is aware and able to ascertain from this information, no facts have been omitted which would render the information provided inaccurate or misleading.

Industry publications or reports generally state that the information they contain has been obtained from sources believed to be reliable, but the accuracy and completeness of such information is not guaranteed. Market data and statistics are inherently predictive and subject to uncertainty and not necessarily reflective of actual market conditions. Such statistics are based on market research, which itself is based on sampling and subjective judgements by both the researchers and the respondents.

As a result, prospective investors should be aware that statistics, data, statements and other information relating to markets, market sizes, market shares, market positions and other industry data in this Offering Circular (and projections, assumptions and estimates based on such information) may not be reliable indicators of the Company's future performance and the future performance of the industry in which it operates. Such indicators are necessarily subject to a high degree of uncertainty and risk due to the limitations described above and to a variety of other factors, including those described under "*Risk Factors*" and elsewhere in this Offering Circular.

Expected Timetable of Offering and Financial Calendar

Expected Timetable of Principal Events (CET)

Offer Period starts	Monday 6 November 2017
Offer Period will not be closed in whole or in part before	Wednesday 15 November 2017 at 00:01 a.m.
Offer Period expires	Thursday 16 November 2017 at 12:00 p.m. (noon)
Publication of the pricing statement containing the Offer Price and number of Offer Share	Friday 17 November 2017 before 8:00 a.m.
First day of trading and official listing on Nasdaq Copenhagen (subject to the Offering not being withdrawn prior to settlement and completion of the Offering)	Friday 17 November 2017, 9:00 a.m.
First day of trading of the Temporary Purchase Certificates on Nasdaq Copenhagen under the temporary ISIN (subject to the Offering not being withdrawn prior to settlement and completion of the Offering)	Friday 17 November 2017 at 9:00 a.m.
Completion of the Offering, including settlement of the Offer Shares (excluding the Overallotment Option, unless exercised by that date) by way of delivery of Temporary Purchase Certificates	Tuesday 21 November 2017
Registration of the share capital increase regarding the new Shares to be issued by the Company pursuant to the Offering	Tuesday 21 November 2017
Last day of trading of the Temporary Purchase Certificates on Nasdaq Copenhagen under the temporary ISIN	Tuesday 21 November 2017
First day of trading of the Shares on Nasdaq Copenhagen under the permanent ISIN	Wednesday 22 November 2017 at 9:00 a.m.
Automatic exchange of the Temporary Purchase Certificates for Shares in VP Securities	Thursday 23 November 2017

Financial Calendar

The Company's financial year runs from 1 January through 31 December. Financial reporting will be published on a half-yearly basis. The Company currently expects to publish financial reports according to the following schedule (which is subject to changes, as may be announced via Nasdaq Copenhagen, if relevant):

Deadline for submitting shareholder proposals to the Annual General Meeting	Wednesday 28 February 2018
Financial report for the full year ending 31 December 2017	Thursday 15 March 2018
Annual General Meeting 2018	Thursday 12 April 2018
Interim report for the period ending 30 June 2018	Tuesday 28 August 2018

Background and the Offering and Use of Proceeds

The Offering is expected to support Orphazyme's operational strategy, advance Orphazyme's public and commercial profile, and provide Orphazyme with improved access to public capital markets and a diversified base of new Danish and international shareholders.

Orphazyme estimates that the net proceeds from the Offering will be approximately DKK 550 million, excluding the Overallotment Option. If the Joint Global Coordinators exercise the Overallotment Option in full, the Company estimates that the net proceeds to the Company from the Offering will be approximately DKK 635 million. These estimates are subject to the assumptions set forth in "*The Offering—Costs of the Offering*".

Orphazyme's reason for the Offering is to raise funds to support its business. The Company intends to allocate the net proceeds from the Offering, together with its existing cash resources as of 30 September 2017, in total of DKK 649 million, excluding the Overallotment Option, as follows:

- approximately DKK 135-155 million to fund the completion of a phase II/III trial with arimoclomol for the treatment of sIBM;
- approximately DKK 165-185 million to fund the completion of a phase II/III trial with arimoclomol for the treatment of ALS;
- approximately DKK 75-85 million to fund the completion of a phase II/III trial with arimoclomol for the treatment of NPC;
- approximately DKK 65-75 million to fund the completion of a phase II trial with arimoclomol for the treatment of Gaucher disease;
- approximately DKK 85-95 million to fund general research and development activities whereof approximately 50% is allocated to funding of research support for ongoing clinical trials and central lab oversight, approximately 40% to fund NDA enabling activities and approximately 10% to fund the advancement of discovery and lead projects into pre-clinical development; and
- the remainder amount, DKK 65-75 million, to fund preparing for registration and commercial activities, working capital, and for general corporate and administrative purposes, which may include the hiring of additional staff, capital expenditures, and the costs of operating as a public company.

In case the Overallotment Option (maximum DKK 85 million in net proceeds) is exercised, the proceeds will be used to further support the advancement of discovery and lead projects into preclinical development and registration and commercial activities.

In the event of positive results from the phase II/III trial with arimoclomol for the treatment of NPC, Orphazyme may choose to reallocate funds to facilitate filing and registration activities in arimoclomol for NPC, thus accelerating the use of proceeds from the Offering. In this scenario, the Company may need to raise additional funds, obtain debt financing or seek partnerships or other financing arrangements in order to have funds to complete the studies in the remaining indications simultaneously with pursuing the filing and registration activities as well as prepare for the later commercialisation. In case of negative results from the phase II/III trial with arimoclomol for the treatment of NPC, Orphazyme intends to use its remaining funds to pursue completion of the planned studies within the remaining three indications.

The foregoing expected use of the net proceeds from the Offering represents the Company's current intentions based upon present plans and business conditions. As of the date hereof, Orphazyme cannot predict with certainty all of the particulars of the use of the net proceeds of the Offering or the amounts that the Company will actually spend on the purposes set forth above. Actual expenditures may vary substantially from these estimates and the Company may find it necessary or advisable to reallocate the net proceeds within the above-described categories or to use portions thereof for other purposes. The amounts and timing of our actual use of net proceeds will vary based on numerous factors, including the relative success and cost of its research, pre-clinical and clinical development programs, and whether Orphazyme enters into collaborations and/or strategic partnerships with third parties.

Dividends and Dividend Policy

General

All Shares, including the Offer Shares, have the same rights and the Offer Shares will rank *pari passu* with all other Shares, including in respect of eligibility to receive dividends and participate in share buybacks. Upon the issuance and registration of the new Shares to be issued by the Company pursuant to the Offering with the Danish Business Authority (which is expected to take place on completion of the Offering), the new Shares will be entitled to receive dividends to the extent any dividends are declared and payable with respect to the new Shares.

Dividend Policy and Share Buybacks

The Company has not declared or made any dividend payments for the last two financial years. Currently, the Company intends to use all available financial resources as well as revenue, if any, for purposes of the Company's current and future business. As of the date hereof, the Company does not expect to make dividend payments within the foreseeable future.

Any future determination related to our dividend policy and the declaration of any dividends will be made at the discretion of the Board of Directors and will depend on a number of factors, including the Company's results of operations, financial condition, future prospects, contractual restrictions, restrictions imposed by applicable law and other factors the Board of Directors deems relevant. There can be no assurances that Orphazyme's performance will facilitate dividend payments, and, in particular, the Company's ability to pay dividends may be impaired if any of the risks described in this Offering Circular were to occur. See "*Risk Factors*".

As an alternative, or in addition to, making dividend payments, the Company's Board of Directors may initiate share buybacks. The decision by the Board of Directors to engage in share buybacks, if any, will be made in accordance with the factors applicable to dividend payments set forth above.

The information on the Company's policies relating to dividend and share buybacks constitutes forward-looking statements. Forward-looking statements are not guarantees of future financial performance, and the Company's actual dividends or share buybacks could differ materially from those expressed or implied by such forward-looking statements as a result of many factors, including those described under "*Special Notice regarding Forward-Looking Statements*" and "*Risk Factors*".

Legal and Regulatory Requirements

Dividends

In accordance with the Danish Companies Act, dividends, if any, are declared with respect to a financial year at the annual general meeting of shareholders in the following year at the same time as the statutory annual report which includes the audited financial statements for that financial year is approved.

Further, the Company's general meeting may resolve to distribute interim dividends or authorise the Company's Board of Directors to decide on the distribution of interim dividends. A resolution to distribute interim dividends within six months after the date of the balance sheet as set out in the Company's latest adopted annual report shall be accompanied by a balance sheet from either the Company's latest annual report or an interim balance sheet which must be reviewed by the Company's auditors. If the decision to distribute an interim dividend is resolved more than six months after the date of the balance sheet as set out in the Company's latest adopted annual report, an interim balance sheet must be prepared and reviewed by the Company's auditors. The balance sheet or the interim balance sheet, as applicable, must in each case show that sufficient funds are available for distribution.

Dividends may not exceed the amount proposed or recommended by the Company's Board of Directors. Moreover, dividends and interim dividends may only be made out of distributable reserves and may not exceed what is considered sound and adequate with regard to the Company's financial condition and such other factors as the Company's Board of Directors may deem relevant.

Dividends paid to the Company's shareholders may be subject to withholding tax. See "*Taxation*" for a description of Danish withholding taxes in respect of dividends declared on the Company's Shares and certain other Danish income tax considerations relevant to the purchase or holding of Shares.

Share buybacks

In accordance with the Danish Companies Act, share buybacks, if any, may only be carried out by the Board of Directors using funds that could have been distributed as dividends at the latest annual general meeting. Any share buyback shall as a main rule be carried out in accordance with an authorisation granted by the general meeting. The authorisation shall be granted for a specific period of time which may not exceed five years. The authorisation shall specify the maximum permitted value of treasury shares as well as the minimum and maximum amount that the Company may pay as consideration for such shares.

As of the date of this Offering Circular, the Board of Directors is authorised in the period until 2 November 2022 to approve the acquisition of treasury shares, on one or more occasions, with a total nominal value of up to 10% of the share capital of the Company, for so long as the Company's holding of treasury shares after such acquisition does not exceed 10% of the Company's share capital. The consideration may not deviate more than 10% from the official price quoted on Nasdaq Copenhagen at the time of the acquisition.

Share buybacks will be deemed a sale of shares for Danish tax purposes and as a general rule are not subject to Danish withholding tax, provided that the Company is admitted to trading on a regulated market. See "*Taxation*" for a description of Danish income tax considerations relevant to the purchase or holding of Shares.

Other requirements

Dividends, if any, will be paid in accordance with the rules of VP Securities, as in force from time to time, and will be paid to the shareholders' accounts with their account-holding banks in Danish kroner to those recorded as beneficiaries. Settlement of the Offering, including registration of the share capital increase relating to the new Offer Shares with the Danish Business Authority, is expected to take place within two business days after the announcement of the Offer Price and allocation, and is expected to be on 21 November 2017. Registration through the holder's account-holding bank will take place as soon as practically possible thereafter.

Dividends not claimed by shareholders are forfeited in favour of the Company, normally after three years, under the general rules of Danish law or statute of limitations.

Under the Company's Articles of Association and applicable Danish law, there are no dividend restrictions or special procedures for non-Danish resident holders of Shares.

Capitalisation and Indebtedness

The following table sets forth the capitalisation, indebtedness and cash, cash equivalents, and securities of Orphazyme as of 30 September 2017:

- on an actual basis reflecting the carrying amounts on the balance sheet of the Company; and
- on an adjusted basis reflecting the use of net proceeds of the Offering (assuming full exercise of the Overallotment Option) as described in "*Background to the Offering and Use of Proceeds*"; the 2017 Capital Structure Adjustment (assuming an Offer Price at the midpoint of the Offer Price Range and, thereby, an issue of 720,875 bonus Shares prior to Admission as part of the 2017 Capital Structure Adjustment); and exercise of all Pre-IPO Warrants in connection with the Offering.

See "*Description of the Shares and Share Capital*" for information relating to the Company's issued share capital and number of outstanding Shares. You should read this table in conjunction with the Company's Audited Financial Statements and Interim Financial Statements included elsewhere in this Offering Circular and "*Operating and Financial Review*".

	As of 30 September 2017		
DKK million	**Actual**	**Adjustments**	**As Adjusted**
Cash and cash equivalents	98.9	635	733.9
Total cash and cash equivalent assets	**98.9**	**635**	**733.9**
Bank debt	0	0	0
Trade payables	1.8	0	1.8
Other payables	22.9	0	22.9
Current financial debt	**24.7**	**0**	**24.7**
Of which is secured	0	0	0
Of which is guaranteed	0	0	0
Of which is unsecured/unguaranteed	24.7	0	24.7
Non-current financial debt	**0**	**0**	**0**
Of which is secured	0	0	0
Of which is guaranteed	0	0	0
Of which is unsecured/unguaranteed	0	0	0
Total financial indebtedness	**24.7**	**0**	**24.7**
Net financial indebtedness	**(74.2)**	**(635)**	**(709.2)**
Share capital	5.1	17.7	22.8
Share premium	218.9	617.3	836.2
Accumulated deficit	(129.5)	0	(129.5)
Total equity	**94.5**	**635**	**729.5**

Other than as specifically set out above, all of Orphazyme's interest-bearing liabilities are unsecured and unguaranteed.

Orphazyme may in the future need additional capital and may seek to obtain further financing through raising new equity capital or debt financing.

Orphazyme has no reason to believe that there has been any material change to its actual capitalisation since 30 September 2017, other than the 2017 Capital Structure Adjustment (as reflected above) and/or changes resulting from the ordinary course of business.

Working capital statement

In the opinion of Orphazyme, the working capital available as of the date of this Offering Circular is not sufficient for its present working capital needs for the 12 months following the date of this Offering Circular. As further described in "*Background to the Offering and Use of Proceeds*", Orphazyme intends to finance its operations for the 12 months following the date of this Offering Circular and the date of Admission using part of the proceeds from the Offering. In case the Offering is not completed, Orphazyme will seek alternative methods of finance in cooperation with its existing shareholders.

Industry

This Offering Circular contains statistics, data and other information relating to markets, market sizes, market shares, market positions and other industry data pertaining to the Company's business and markets. Unless otherwise indicated, such information is based on the Company's analysis of sources as listed in "Market and Industry Information". Such information has been accurately reproduced, and, as far as the Company is aware from such information, no facts have been omitted which would render the information provided inaccurate or misleading.

Overview

Orphazyme is a Danish biotech company with a late stage orphan drug pipeline based in Copenhagen, Denmark, aiming to introduce novel therapies for the treatment of serious and debilitating rare diseases within the field of protein misfolding diseases, exemplified by neuromuscular diseases and lysosomal storage diseases.

The Company's technology is based on amplifying human cells' defense against protein aggregation and misfolding. This defense consists of a system of proteins known as *heat shock proteins*, which work by rescuing proteins from misfolding and aggregation.

The Company's current lead program, arimoclomol, works by increasing the body's own production of heat shock proteins in cells experiencing stress or toxicity. Orphazyme has ongoing or planned clinical stage trials for the 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 ("**Gaucher**"). These four diseases are rare, often severe diseases with limited or no current treatment options and arimoclomol has orphan drug designation with the US Food and Drug Administration (the "**FDA**") for ALS and NPC and the European Medicines Agency (the "**EMA**") for NPC, ALS and sIBM. While arimoclomol is currently being developed for treatment of the above diseases, available data from pre-clinical studies indicate that the compound may potentially also be developed for treating other related diseases (e.g. Parkinson's disease as well as a range of other lysosomal storage diseases).

The orphan drug industry

Orphan drugs

Orphan drugs denote pharmaceutical products that target rare and often severe diseases. The diseases are in general serious, debilitating and often fatal if not treated. Rarity is defined in legal and regulatory frameworks and varies across geographical regions. In the United States, EU and Japan, orphan disease populations are defined based on prevalence:[7]

- USA: < 200,000 patients (< 6.37 in 10,000 individuals based on the United States population of 314 million)
- EU: < 250,000 patients (<5 in 10,000 individuals based on EU population of 514 million)
- Japan: < 50,000 patients (< 4 in 10,000 individuals based on Japan population of 128 million)

The National Organization for Rare Disorders ("**NORD**") currently estimates that 30 million Americans suffer from 7,000 rare diseases and Eurordis estimates that more than 6,000 rare diseases may affect more than 30 million EU citizens.

To encourage and incentivize the research and development ("**R&D**") of pharmaceutical products for rare diseases despite the small number of patients and the significant development costs, the regulatory authorities or authorized bodies in the United States, the EU, Japan and other countries can grant a pharmaceutical product a so-called *orphan drug designation*. Developers of medicine with an orphan drug designation are entitled to several advantages, including the possibility of free of charge advice from the FDA and EMA and under specific circumstances certain financial benefits, such as R&D tax credits (while not part of the Company's current planning) and exemptions or reductions in regulatory submission fees. If a drug candidate with orphan drug designation is approved by the regulatory authorities or authorized bodies upon completion of the clinical trials, the respective authority can provide the pharmaceutical product with orphan drug status. Orphan drug status provides the orphan drug with market exclusivity for seven and 10 years in the United States and the EU, respectively.[8] For further information on advantages in development of orphan drugs, please refer to the "*—Differentiating factors for orphan drugs*" section below.

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[7] FDA, EMA and Pharmaceuticals and Medical Devices Agency

[8] EMA European Medicines Agency, Marketing authorisation and market exclusivity, http://www.ema.europa.eu/ema/index.jsp?curl=pages/regulation/general/general_content_000392.jsp& and U.S. Food & Drug Administration, Code of Federal Regulations Title 21, https://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfcfr/cfrsearch.cfm?cfrpart=316&showfr=1&source=govdelivery&subpartnode=21%3A5.0.1.1.6.4&utm_medium=email&utm_source=govdelivery

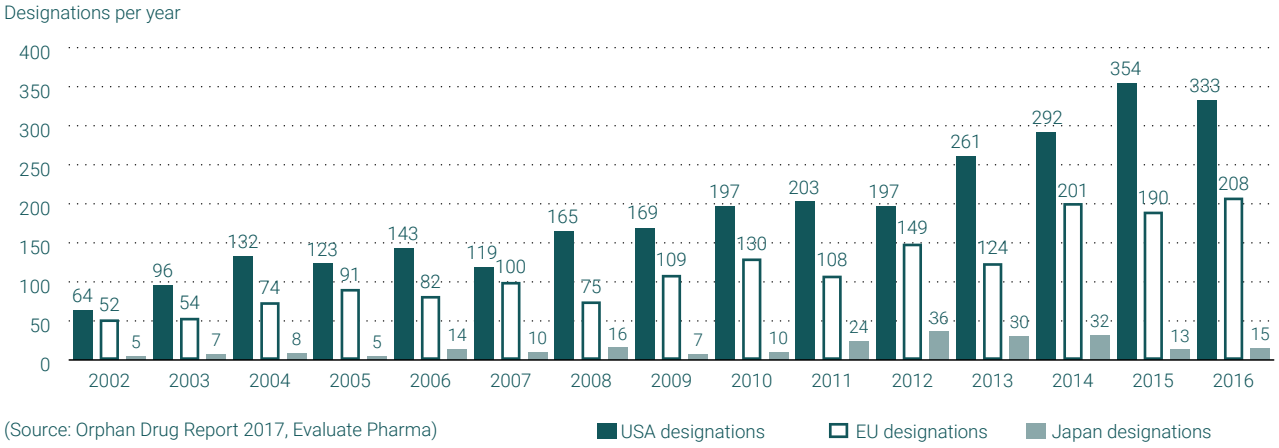
Background to orphan drugs

The Orphan Drug Act was passed in the United States in 1983 ("**Orphan Drug Act**") to avoid neglect of less common diseases. The Orphan Drug Act was set up to incentivize the development of pharmaceutical products for severe diseases affecting so few individuals that it would not be considered profitable otherwise. Orphan diseases are rare conditions that affect a small percentage of the population. Many orphan diseases have no treatment options, and there is high unmet need for new therapies to be developed.

The Orphan Drug Act is considered to have been a success. In the decade before the Orphan Drug Act was passed, only ten treatments for rare diseases were developed in the United States. Today, more than 600 orphan drugs have been approved and almost 4,000 pharmaceutical product candidates have received orphan drug designations by the FDA. In 2016, the FDA approved 22 novel drugs, of which nine were classified as orphan drugs.[9]

The success of the Orphan Drug Act led to the implementation of orphan drug regimes in other key markets as well, for example Japan in 1993 and the EU in 2000. In 2016, the cumulative number of pharmaceutical product candidates with orphan drug designations given in the United States, the EU and Japan were 3,976; 1,824 and 387, respectively.[10]

Worldwide orphan drug designations



(Source: Orphan Drug Report 2017, Evaluate Pharma)

Differentiating factors for orphan drugs

There are several factors contributing to the growing number of approvals of orphan drugs. The orphan drug designation and orphan drug status present a series of advantages for the developer of the drug (the "**sponsor**") during the development phase. In addition, after approval these factors may contribute to a favorable commercialization position.

Advantages in the development phase

Upon receipt of an orphan designation, sponsors benefit from a range of incentives related to the regulatory process with the FDA, EMA or other relevant regulatory bodies. These include, among others, protocol assistance from the FDA and EMA to ensure alignment between the company (sponsor) and the regulatory authority. The development of orphan drugs is subject to the same guidelines as other pharmaceutical products and substantial evidence of safety and efficacy must be documented in all instances. However, an important advantage is the recognition by the FDA and EMA of the inherent challenges related to the investigation of orphan drug candidates and the fact that traditional approaches used in trials of non-orphan diseases are not always feasible for orphan drugs. The FDA and EMA have issued guidelines that deal specifically with the issues likely to be encountered by developers of orphan drugs and suggest meetings throughout the development process with the sponsors to agree on a feasible design and extent of the trials. A continuous dialogue with the FDA and EMA is particularly useful in ensuring that development of an orphan drug meets the expectations of the regulatory authorities or authorised bodies. This is reflected in the median time from filing to approval with the FDA which is, on average, three months shorter for orphan drugs compared to non-orphan drugs.[11]

[9] FDA https://www.fda.gov/drugs/developmentapprovalprocess/druginnovation/ucm483775.htm
[10] Orphan Drug Report 2017, Evaluate Pharma
[11] Orphan Drug Report 2015, Evaluate Pharma

Orphan drugs appear to have a higher likelihood of approval compared to non-orphan drug candidates. According to findings produced in a scientific study from 2014, the likelihood of ultimate approval of an orphan drug candidate in the phase I stage is 32.9% compared to 10.4% across all drug candidates.[12] For drugs at the phase II stage, approval rates increase to 37.9% and 16.2%, respectively, and for the phase III stage approval rates increase further to 54.2% and 50.0%, respectively. Furthermore, the results are even more profound for non-oncology related orphan drugs (the focus of Orphazyme); where the numbers increase to 44.5% for phase I, 50.4% for phase II and 62.1% for phase III.[13]

The regulatory advantages offered to developers of orphan drugs also include a reduction of the R&D costs compared to costs associated with the development of non-orphan drugs. Developers of drug candidates with an orphan drug designation may be entitled to a 50% US tax credit on R&D costs for the designated drug, R&D grants in the United States for phase I to phase III clinical trials and a reduction/relief on certain fees. Due to the rarity of disease, in many cases one adequate and well-controlled phase III trial is considered sufficient for marketing authorisation, compared to the standard requirement of two phase III trials or one very large phase III trial for non-orphan drugs. Clinical trials in orphan diseases usually involve fewer patients than trials in non-orphan diseases. As a result the average cost of a phase III clinical trial for an orphan drug is in general approximately half that of a non-orphan drug.[14]

Advantages in the marketing phase

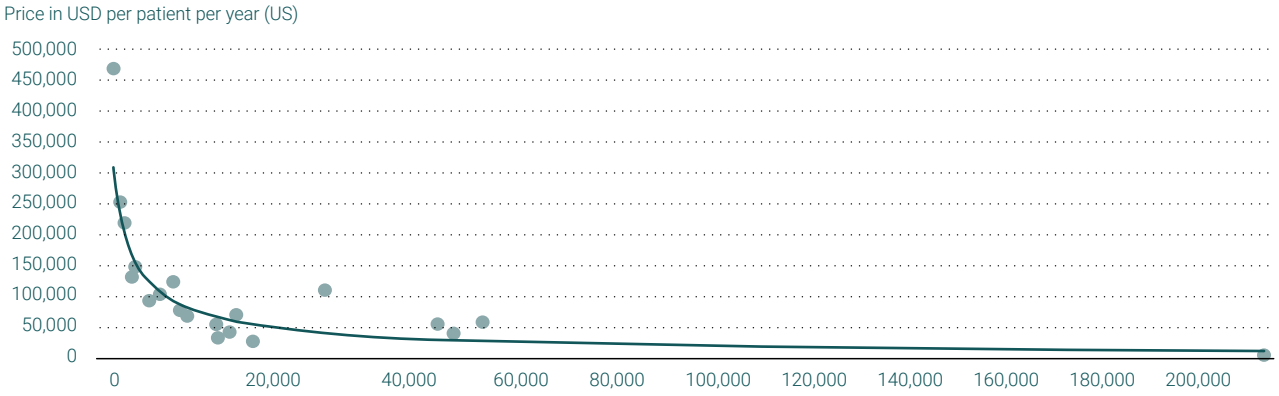
Upon completion of clinical trials, drug candidates may receive orphan drug status by the respective authority approving the drug. Upon the receipt of such status, the orphan drug is entitled to a seven-year market exclusivity in the United States and a ten-year market exclusivity in the EU. In addition, an approved paediatric investigation plan ("PIP") in the EU makes the sponsors eligible for a two-year extension of the 10-year period and six months in the United States. During the period of market exclusivity, the orphan drug is protected from similar drugs being approved for the same therapeutic indication, unless a safer, more effective or otherwise clinically superior product addressing the same therapeutic indication is introduced. Also, due to the fact that a relatively low number of patients are affected by these rare diseases, the patients are often cared for by a well-defined small number of specialist physicians practicing at a relatively small number of hospitals or clinics per country, making it possible to target these physicians with a small sales and marketing organization.

Generally, orphan drugs are priced at a premium to non-orphan drugs. As orphan drugs target rare and often life-threatening diseases, high prices are essential to ensure an expected return on investment for the risk taken by pharmaceutical companies. Many factors affect the pricing of orphan drugs and non-orphan drugs such as the burden of disease, extent of unmet need, clinical efficacy, side effects, tolerability, number of patients affected by the disease, the cost of development and manufacturing as well as negotiation with payers/insurers. In the United States, pharmaceutical companies negotiate pricing directly with payers/insurance companies and most of these provide access to orphan drugs, including the public insurance programmes Medicare and Medicaid. In the EU, access to the market is gained through a centralized procedure with EMA's Committee for Medicinal products for Human Use ("**CHMP**"), while prices are negotiated separately with the authorities in the individual countries in which the product is marketed.

In 2016, the average annual cost per patient in the Unites States was approximately five times higher for orphan drugs compared to non-orphan drugs. Furthermore, a market study indicates an inverse correlation between the number of patients and annual price per patient when considering the 20 most sold orphan drugs in the United States in 2016. The inverse correlation is further enhanced when considering the top 10 selling drugs in the United States in 2016 that treated fewer than 10,000 patients.[15]

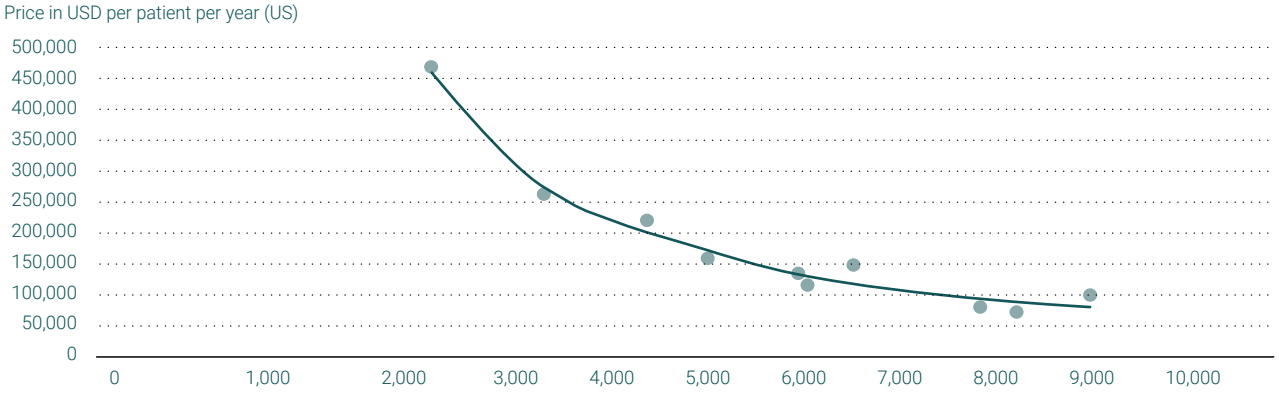
[12] Hay et al, Clinical development success rates for investigational drugs, Nature Biotechnology, 2014
[13] Hay et al, Clinical development success rates for investigational drugs, Nature Biotechnology, 2014
[14] Orphan Drug Report 2015, Evaluate Pharma; Orphan Drug Report 2017, Evaluate Pharma
[15] Orphan Drug Report 2015, Evaluate Pharma; Orphan Drug Report 2017, Evaluate Pharma

Price per patient and number of patients for the 20 most sold orphan drugs (by sales) in the United States, 2016



(Source: Orphan Drug Report 2017, Evaluate Pharma)

Price per patient and number of patients for the 10 most sold orphan drugs (by sales) in the United States, 2016 (fewer than 10,000 patients treated)



(Source: Orphan Drug Report 2017, Evaluate Pharma)

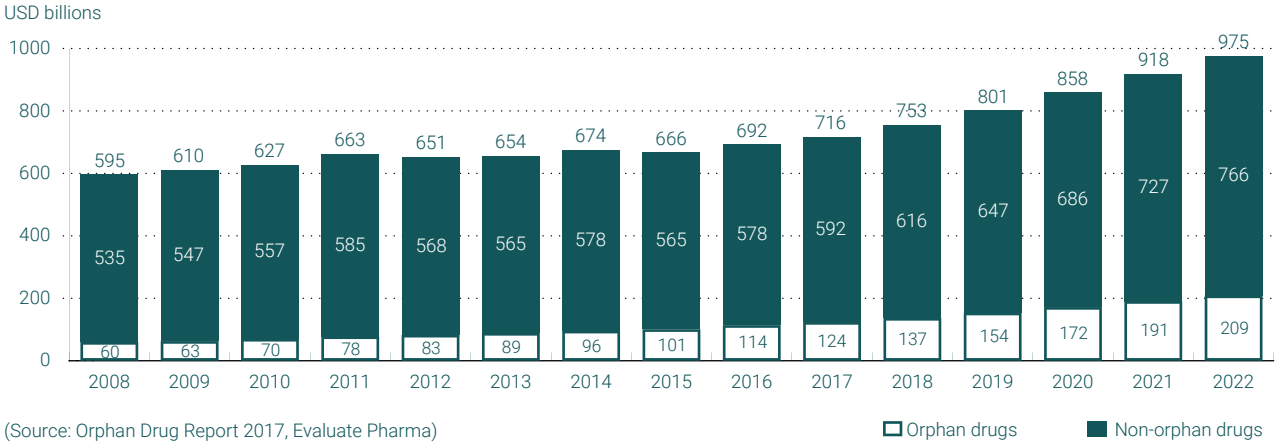
Overview of the orphan drug market

The orphan drug market has shown substantial growth over the last couple of years. In 2016, worldwide orphan drug sales increased by 12.2% year-on-year and reached USD 114 billion, whereas non-orphan drugs (excluding generics) grew by 2.4% in the same period, amounting to USD 578 billion. The total prescription drug sales (excluding generics) grew by 3.9% in 2016 and amounted to a total of USD 692 billion. The orphan drug market is estimated to grow to USD 209 billion by 2022, corresponding to a CAGR of 11.1% in the period 2017-2022, almost double the growth of the overall prescription pharmaceutical product market during the same period.[16]

The development of the worldwide orphan drug market indicates that governments' initiatives have been successful. In addition, it reflects that advances in R&D have improved pharmaceutical companies' ability to develop treatments for rare diseases.

[16] Orphan Drug Report 2017, Evaluate Pharma

Worldwide orphan drugs and prescription drugs sales (actual and estimated), excluding generics



(Source: Orphan Drug Report 2017, Evaluate Pharma)

Neuromuscular disorders[17]

Neuromuscular disorders encompass a range of conditions affecting a part of the neuromuscular system and impairing the functioning of the muscles, thereby causing muscle weakness and fatigue. In particular, the disorders affect the nerve cells (so-called neurons) that send messages to control the voluntary muscles (i.e. muscles one can control, such as arms and legs) and the muscles themselves. As the disorders manifest, the neurons become unhealthy or die, resulting in a loss of communication between the nervous system and the muscles. Weakening of the muscles may lead to aches, pains, movement problems and may affect the heart function and ability to breathe. The causes of neuromuscular disorders vary by type, but the diseases targeted by Orphazyme are characterized by proteins misfolding and clumping together (protein aggregation) prohibiting proper recycling. Protein aggregation can cause cell stress and eventually cell death. Among the group of neuromuscular disorders, Orphazyme currently conducts clinical trials for the diseases sIBM and ALS.

Sporadic Inclusion Body Myositis ("sIBM")

Introduction to sIBM

sIBM is an acquired, rare and slowly progressing muscle disorder which becomes apparent during adulthood. Among individuals older than 50 years it is the most common muscle wasting disorder. The disease is generally characterised by progressive weakness and degeneration (atrophy) of the muscles, especially those of the arms and legs, particularly the quadriceps. sIBM typically affects the ability to grab or manipulate objects, causes trouble walking or rising and can progress to cause severe disability. In addition, difficulty swallowing (dysphagia) due to weakness of throat muscles may occur. sIBM typically presents itself later in life with 87% of patients experiencing onset of symptoms at 50 years of age or more, and earlier symptoms usually only being recognized retrospectively. In most cases, the disease progresses relentlessly over 10 to 15 years until the affected patient has lost mobility entirely.

The cause of sIBM is unknown. It is possible that multiple immunological, genetic and environmental factors and age-related factors (degenerative factors) are all involved in the development of the disorder. Two distinct processes have been identified in individuals with sIBM – one autoimmune (immunologic) and one degenerative – and it is possible that these processes are related. Autoimmune disorders occur when the body's immune system mistakenly attacks healthy tissue. Numerous factors support that sIBM likely may be an autoimmune disorder. Especially, the presence of certain inflammatory white blood cells in the muscle tissue of affected individuals and has led sIBM to be classified as an autoimmune inflammatory muscle disease. However, sIBM does not respond to conventional therapies used to treat autoimmune disorders (e.g. corticosteroids or immunosuppressive drugs), suggesting that other factors play a critical role in development of sIBM. In addition to inflammatory signs, muscle tissue of individuals with sIBM shows degenerative changes. Specifically, the muscle tissue of affected individuals contains compartments containing abnormal clumps of a wide range of different proteins. These clumps (or protein aggregates), called 'inclusion bodies', give the disorder its name. The significant degenerative component has led some researchers to argue that sIBM is primarily a degenerative muscle disorder and therefore protein misfolding and aggregation a viable target for therapy.

[17] The information in this section is principally derived from management information based on research and market reports

The exact prevalence of sIBM is unknown. However, it is estimated at 5-10 individuals per 1,000,000 in the general population in the United States, but studies have reported values ranging from 4.9-70 individuals per 1,000,000. Further, a recent meta-analysis of nine sIBM prevalence studies concludes that prevalence is approximately 24.8 per million in the United States and the major European countries or higher, corresponding to at least 17,000 patients.[18] Based on a few epidemiological studies, the prevalence of sIBM appears to vary considerably between different populations and racial groups, which suggest that genetic factors may play a role in determining susceptibility to the disease. For example, sIBM appears most common among Caucasians where data from studies indicates that prevalence in individuals at the age of 50 of above could be 20-70 individuals per 1,000,000, corresponding to approximately 6,000 cases in the United States. sIBM also appears to have a relatively high prevalence in people of Japanese descent, where the number of patients have increased in recent years. Though awareness of the disorder is growing, many expert clinicians believe it remains underdiagnosed. One of the confounding factors to the unknown prevalence of sIBM is that it is often misdiagnosed. The most common misdiagnosis is polymyositis because the immunologic hallmarks are similar, particularly early in the disease course. The insidious onset and slow progression of the disease process accounts, in part, for the misdiagnosis. In a long-term observation study of sIBM conducted in two research centers, it was discovered that 30% of patients initially received an incorrect diagnosis.

Diagnosis of sIBM is not straight forward, leading to a diagnostic delay in many cases of more than 10 years from onset of the disease, in particular when patients do not consult neuromuscular experts. The slow progression of the disease, patient assumption that initial weakness is due to aging misdiagnosis and the fact that no treatment is available to the patients today, as described above, are some of the factors contributing to the delay in diagnosis. Today, the diagnosis of sIBM is made based on thorough clinical evaluation, careful patient history and a variety of specialized tests, such as muscle biopsy (a procedure in which a tiny amount of muscle tissue is surgically removed and studied under a microscope to detect characteristic changes that indicate sIBM).

Treatment options for sIBM and unmet need

As of the date hereof, there are no effective treatments for sIBM. No therapeutic agent has shown efficacy in preventing, halting or reversing the progression of sIBM and therefore, no drugs have been approved for sIBM. In particular, the disorder has not shown to respond to conventional therapies for autoimmune disorders, such as corticosteroids or immunosuppressive drugs (drugs suppressing the immune system, such as azathioprine, methotrexate, cyclosporine, and cyclophosphamide). Therefore, the standard treatment option for sIBM consists only of supportive therapy (namely physical, speech and occupational therapy). Physical therapy aims to build strength in unaffected or mildly affected muscles, while speech therapists monitor the ability to swallow, as chocking or aspiration due to dysphagia is common.

As such, there is a significant unmet need for treatment options for sIBM. Although life expectancy for sIBM patients is similar to that of the general population, quality of life is severely affected. Ten to 15 years post-symptom onset, most patients will require assistance with basic daily activities, and some become wheelchair bound or bedridden. Irrespective of the course of treatment, most patients still progress (more than 80% in most studies) and there is no evidence that any form of treatment slows progression. The disease can be an indirect cause of death due to respiratory failure (aspiration, dysphagia or cachexia) or infection, particularly those of the respiratory tract. In the terminal phase of illness, some patients have requested euthanasia due to unbearable suffering or terminal deep sedation has been applied due to dysphagia and cachexia.

Competition for treatment of sIBM

Only a few investigational therapies are being studied for patients with sIBM. Orphazyme has identified one development program that, in addition to Orphazyme's own program, is ongoing for the sIBM disease. Milo Biotechnology is conducting a phase I/IIa trial with treatment using follistatin gene transfer. Follistatin is a protein that increases muscle mass and strength. Results from the trial have not yet been published but thus far very preliminary clinical data suggests a potential benefit on the 6 minute walk test.[19]

Earlier in 2017, Novartis halted their Bimagrumab study concerning sIBM after the phase IIb/III trial failed to meet its primary endpoint. Bimagrumab is a fully human monoclonal antibody developed to treat pathological muscle loss and weakness.[20] Bimagrumab works through a completely different mechanism of action than arimoclomol.

[18] Aoife C et al, Journal of Neuromuscular Diseases 2017
[19] Company analysis and clinicaltrials.gov
[20] FierceBiotech, Novartis 'breakthrough' muscle drug bimagrumab flunks a late-stage trial, 21 April 2016

Market for treatment of sIBM

The size of the patient population in Europe and the United States is not fully elucidated but has been conservatively estimated to be between 7,000 and 15,000 individuals.[21] However, the recent meta-analysis of sIBM prevalence estimates a patient population of approximately 24.8 per million or 17,000 individuals in the United States and the major European countries.[22] Many clinicians believe that sIBM is underdiagnosed and thus the number of patients is likely higher than current estimates. If an effective treatment for sIBM becomes available, it is likely that awareness of the disease increases and patients may may be diagnosed earlier in their disease. In addition, this may also lead to an increase in the prevalence of sIBM through identification of patients currently undiagnosed or misdiagnosed. However, it is unknown by how much the estimated prevalence would increase.[23]

Amyotrophic Lateral Sclerosis ("ALS")

Introduction to ALS

ALS, also called Lou Gehrig's disease, is a rapidly progressive and invariably fatal neurological disease. It attacks neurons responsible for controlling voluntary muscles. ALS results in muscle weakness, progressive disability and eventually death, typically from respiratory failure. Time from onset to mortality is typically short in the range of two to five years. However, ALS is a distinctly heterogeneous disease and the clinical course is variable, with some patients dying within one year from symptom onset and other living for more than a decade. Muscle weakness results from progressive degeneration of motor neurons in different parts of the central nervous system, where upper and lower neurons selectively die. The cause of damage to the neurons is unknown, but several theories have been proposed, including glutamate toxicity, protein misfolding and oxidative stress.

Despite being classified as a rare disease, ALS is considered one of the more common neuromuscular diseases worldwide. The incidence of ALS is estimated at between 1-3 per 100,000 individuals per year globally. The patient population in Europe and the United States is estimated to be approximately 50,000 patients. While the mean age of onset is between 55 and 65 years, symptoms can begin at any adult age. The disease occurs more frequently in men than women, whereas prevalence is roughly the same throughout the world (4-5.4 per 100,000 individuals) with no ethnic, racial or socioeconomic differences.

Around 10 percent of patients with ALS have ALS associated with pathogenic mutations (commonly referred to as 'familial ALS') and the remaining 90 percent have a sporadic onset of ALS (i.e. without any identified genetic component). In about 20 percent of the cases of familial ALS (i.e. two percent of the total ALS population), mutations in the gene coding for the copper-zinc superoxide dismutase ("**SOD1**") enzyme have been found. SOD1 is an enzyme with antioxidant properties important in the protection against free radicals, a highly reactive oxygen species that can damage cellular components via oxidative stress. Defects in SOD1 may cause ALS via two mechanisms: inhibition of SOD1 enzymatic activity and the formation of toxic SOD1 protein aggregates. Arimoclomol has thus far been tested in two phase II trials, one dose-ranging trial in sporadic ALS and one trial in ALS associated with pathogenic SOD1 mutations. For further information, please refer to the *"Business—Arimoclomol—ALS"* section.

In Western countries, the rate of diagnosis is high, but initial symptoms of ALS can be subtle at first, and it may take 12 to 14 months to be accurately diagnosed. Early symptoms of ALS may resemble those of other diseases and therefore diagnosis may largely be a matter of excluding other conditions. For that, certain diagnostic procedures, such as magnetic resonance imaging or a test to detect electrical activity in muscles (electromyography) may be utilized. ALS is characterized by degeneration of both the upper and lower motor neurons. Some patients with ALS may initially present only with findings due to degeneration of the upper motor neurons. Lower motor neuron degeneration usually appears within three to five years in these patients.[24]

Treatment options for ALS and unmet need

As of the date hereof, there are a limited number of available treatments for ALS and these only impart a modest effect. The focus of medical care is to give symptomatic management of patients with mild to moderate disease and easing (palliative) intervention in patients with severe or terminal disease. The care of ALS patients is often provided in multidisciplinary clinics, with a team comprised by respiratory therapist, physiotherapist, occupational therapist, dietician, speech consultant and social worker.

Until recently, the only pharmaceutical product used for modifying ALS was Rilutek (riluzole) developed by Sanofi, which was the first drug

[21] Defined Health for Orphazyme, 2017
[22] Aoife C et al, Journal of Neuromuscular Diseases 2017
[23] Defined Health for Orphazyme, 2017
[24] Defined Health for Orphazyme, 2017

to be approved by the FDA for the treatment of the disease more than 20 years ago. Riluzole was tested in two confirmatory studies, where it showed to prolong median survival/time to respiratory failure by approximately 60 to 90 days. In both studies, the results of the pre-defined per protocol analysis were negative, but statistical significance was reached in post-hoc analysis. Side effects like nausea and severe fatigue are common with Riluzole treatment and where frequently cited as the reasons for discontinuing treatment, whereas other and more severe side effects are rarer.

In May 2017, the FDA approved Radicava (edaravone), which is the first approved ALS treatment option for more than 20 years in the United States. Radicava, developed by Mitsubishi Tanabe Pharma Corporation, is an intravenous infusion treatment for ALS. The comprehensive clinical development program for Radicava in ALS has spanned 13 years. The pivotal phase III trial (MCI186-19) evaluated 137 people in Japan with ALS and formed a basis for the FDA approval of Radicava. Data from the study demonstrated that patients receiving Radicava for six months experienced significantly less decline in physical function compared to placebo – by 33 percent measured by the ALS Functional Rating Scale-Reviced (ALSFRS-R), a validated rating instrument for monitoring the progression of disability in patients with ALS. The Company believes that the approval of Radicava, based on a study with relatively few people, provides evidence of the support from the FDA for development of drugs for the treatment of ALS, which could also be relevant for Orphazyme. As of the date hereof, an application for a marketing authorization with respect to Radicava has not been filed in Europe.[25]

With the current treatment options there is still a major need for new effective treatments for patients with ALS in order to improve the clinical course of the disease and to further extend survival.

Competition for treatment of ALS

In addition to the current treatment options for ALS, a number of pharmaceutical products are being developed to treat ALS. In addition to arimoclomol, masitinib developed by AB Science and tirasemtiv developed by Cytokinetics are the most advanced products in development. AB Science is developing masitinib in multiple diseases, including ALS, where a phase III trial has been completed. The trial compared efficacy and safety in combination with Riluzole versus placebo and showed effect on ALSFRS-R, Amyotrophic Lateral Sclerosis Assessment Questionnaire (ALSAQ), which measure the subjective well-being of patients and Forced Vital Capacity (FVC), which is an indicator of survival and disease progression. Cytokinetics is developing tirasemtiv for the treatment of ALS and certain other debilitating diseases. In phase II tirasemtiv showed decline in FVC, but not in ALSFRS. Tirasemtiv is currently the subject of a large phase III trial called VITALITY-ALS, which is designed to assess effects versus placebo on respiratory function and muscle strength. Results from the trial are expected in the fourth quarter of 2017. In addition to arimoclomol, masitnib and tirasemtiv multiple early and mid-stage trials for treatment of ALS exist.[26]

Market for treatment of ALS

The CDC (Centers for Disease Control and Prevention) estimates that there are approximately 16,000 prevalent cases of ALS in the United States and that approximately 5,000 new cases are diagnosed each year. The number of total cases for the largest five markets in the EU (Germany, France, Italy, Spain and the United Kingdom) is estimated to be slightly higher and for the broader European geography the estimate is approximately 40,000 prevalent cases. Hence, the conservatively estimated market for the United States and Europe is approximately 50,000 cases. In addition, Japan is a large market for ALS and is estimated to be between 8,000 and 14,000 cases.[27]

The newly approved drug for treatment of ALS, Radicava, has an indicated cost of USD 1,000 for each infusion. The ALS Association has estimated an annual treatment cost of USD 146,000.

Lysosomal storage diseases[28]

Lysosomal storage diseases are inherited metabolic disorders where enzyme deficiencies result in an accumulation of toxic materials in the cells of the body. These deficiencies are often caused by mutations leading to misfolding and degradation of the enzymes. Lysosomes are membrane-bound compartments of enzymes, located in the body's cells, used to break down fats, proteins and other large molecules into the respective building blocks. Loss of lysosomal enzyme activity due to their misfolding and dysfunction prohibits the lysosomes from performing their normal function and results in accumulation of metabolites in the cells, why these diseases are referred to as "lysosomal storage diseases".

[25] ENCALS invites MT Pharma to conduct a trial in Europe, European Network to Cure ALS, https://www.encals.eu/encals-statement-edaravone/
[26] Company analysis and clinicaltrials.gov
[27] Mehta et al, MMWR Surveill Summ 2016, Chio et al., Neuroepidemiology, 2013 and Defined Health for Orphazyme, 2017
[28] The information in this section is principally derived from management information based on research and market reports

Lysosomal storage diseases are comprised of more than 50 different diseases, that may affect different parts of the body, e.g. the brain, central nervous system, skeleton, skin and heart, and new disorders continue to be discovered. Although clinical trials are in progress, only a few approved treatments are currently available for a few of the lysosomal storage diseases such as Gaucher Type I, Pompe disease, Fabry disease, MPS I, MPS II, MPS IV, MPS VI, LAL-D, etc. Among the group of lysosomal storage diseases, Orphazyme is currently conducting clinical research with arimoclomol for NPC.

Niemann-Pick disease ("NPC")

Introduction to NPC

NPC disease is a rare, genetic and progressive disease that impairs the ability of the body to move cholesterol and other fatty substances (lipids) inside the cells. The result is an accumulation of lipids within the body's tissue, including the brain tissue, causing damage to the affected areas. The symptoms upon onset of NPC vary from fatality during the first months after birth to a progressive disorder not diagnosed until adulthood. The disease affects neurologic and psychiatric functions as well as various internal organs. Symptoms of NPC usually occur during middle to late childhood as e.g. difficulties in drawing and writing, loss of previously acquired speech skills and difficulties in swallowing. During this period, affected individuals may also develop impairment of intellectually ability, psychiatric disturbances and progressive loss of memory. Symptoms include enlargement of the liver and/or spleen and lung diseases. Systemic symptoms of NPC are more common in infancy or childhood and the rate of progression is usually much slower in individuals with onset of symptoms during adulthood. NPC is usually fatal and the majority of individuals with the disease die before the age of 20. Adults diagnosed with NPC are more likely to present with dementia or psychiatric symptoms.

The NPC disease is caused by mutations on one of two genes, NPC1 or NPC2. Approximately 95% of people with the disease have mutations in NPC1. Genetic diseases are determined by the combination of the pair of genes for a particular trait received from the father and the mother. NPC is an autosomal recessive disorder (i.e. two copies of an abnormal gene must be present in order for the disease or trait to develop). Although uncertainty exists about the function of the NPC1 and NPC2 protein products, they are known to be involved in the trafficking of large molecules inside the cells of the body. Hence, a mutated gene may lead to insufficient protein production and, as a consequence, an abnormal accumulation e.g. cholesterol and/or other fatty materials and sugars in the organs most commonly affected, such as the liver, spleen and brain.

As many cases of NPC go misdiagnosed or undiagnosed, it is difficult to determine the disease's true frequency. Incidence of NPC between 0.7-0.8 per 100,000 individuals is frequently referenced. In addition, 1 to 9 per 100,000 is reported on Orphanet and 0.1 per 10,000 is reported in the EMA orphan drug designation documentation on Zavesca, an approved treatment for NPC.[29] According to a Niemann-Pick foundation (NNPDF), NPC has an estimated 500 cases diagnosed annually worldwide. NNPDF notes that it is believed that the number of people affected is higher. Other Niemann-Pick foundations (INPDA and NPUK) say incidence of 1 in 120,000 is widely reported and that recent evidence[30] suggests that this may be an underestimate.

NPC is often initially diagnosed as a learning disability, mild retardation, and delayed development of fine motor skills and it is common to spend several years seeking a diagnosis before NPC is identified. The diagnosis of NPC is made upon the characteristic symptoms, as described above, obtained from clinical evaluations and tests to evaluate a protein's function or the presence of accumulated byproducts (biochemical tests) and to evaluate if the NPC1 or NPC2 gene is mutated (gene sequencing). However, physicians' limited experience with NPC often results in delayed diagnosis.

Treatment options for NPC and unmet need

The majority of current treatment options are palliative and only directed towards the specific symptoms apparent in each individual (e.g. referral to a speech therapist to optimise the swallowing function, prescription of anti-seizure medications to prevent seizures and prescription of melatonin to treat insomnia and other sleep problems caused by the disease) and may require the coordinated efforts of a team of specialists.

Only one drug, Zavesca (miglustat) owned by Actelion Pharmaceuticals, is currently being marketed for NPC and only in certain jurisdictions. Studies have found beneficial effects of Zavesca on lipid trafficking defects as it reduces the accumulation of substrates in the brain and delays the progression of neurological symptoms, although only demonstrated for eye movements. A range of safety and tolerability problems are known to be associated with Zavesca, including weight loss, decreased appetite, diarrhoea, nausea and thrombocytopenia. Zavesca has not been approved by the FDA for treatment of NPC, but it is approved for the treatment of Gaucher type I disease in the US.

[29] Medical Marketing Economics for Orphazyme, 2016
[30] A. Wassif, Christopher et al. High incidence of unrecognized visceral/neurological late-onset Niemann-Pick disease, type C1, predicted by analysis of massively parallel sequencing data sets. Genetics in Medicine 18, 41–48 (2016)

Zavesca is approved for treatment of NPC in Europe, Canada, Australia, New Zealand, and several countries in Asia and South America as Zavesca and in Japan as Brazaves.[31]

Due to the limited availability and efficacy and side effects of the existing treatment option, the Company believes a significant unmet need for treatment of NPC continues to exist.

Competition for treatment of NPC

In addition to Zavesca, the only approved treatment of NPC, studies are currently being performed to test the safety and efficacy of other treatment options. In addition to Orphazyme's phase II/III trial with arimoclomol, two treatment options have been identified by the Company as being in development; VTS-270 in phase IIb/III (by Sucampo Pharmaceuticals) and Trappsol in phase I/IIa (by CTD Holding).

Market for treatment of NPC

Based on the prevalence described above the number of potential NPC patients in the United States and in the EU is conservatively estimated to between 1,000 and 2,000 individuals in total. Diagnostic challenges may affect the number of potential patients. However, a treatment option could also increase awareness of the disease and assist in identifying more cases.

The only established sales data available for treatment of NPC is based on Zavesca. In 2016, Actelion Pharmaceuticals' global sales from Zavesca were CHF 104 million, which however also related to Gaucher as described below.[32]

Gaucher

Introduction to Gaucher

Gaucher is a rare, inherited metabolic disorder causing certain sugar (glucose) containing fat (lipids and especially glycolipids) to abnormally accumulate in the lysosomes of cells, especially within the bone marrow, spleen and liver, due to the lack of a certain enzyme (glucocerebrosidase). The symptoms vary greatly from patient to patient with some patients having few or no symptoms while others may experience significant complications. The usual symptoms of Gaucher include an abnormally enlarged liver and/or spleen (hepatosplenomegaly), low levels of circulating red blood cells (anemia) and blood cells promoting clotting (thrombocytopenia) and skeletal abnormalities. Like NPC, Gaucher is an autosomal recessive disorder.

Three distinct forms of Gaucher have been identified to date based on the absence or presence of neurological complications and the extent of such. Gaucher type 1 is at the outset characterised by a lack of neurological complications and usually result in a low level of blood clotting cells (thrombocytopenia) and a low level of red blood cells in circulation (anemia) causing easy bruising and chronic fatigue, respectively. In addition, affected individuals may experience an abnormally enlarged liver and/or spleen (hepatosplenomegaly) and skeletal anomalies. Gaucher type 2 (so-called acute neuronopathic Gaucher disease) is characterised by onset in the early months of life and entails neurological complications arising from the accumulation of a certain lipid (glucocerebroside) in the brain. The symptoms include enlargement of the spleen (splenomegaly), loss of motor skills, involuntary muscle spasms (spasticity), decreased muscle tone (hypotonia) and dysphagia. Gaucher type 2 usually results in fatality during the first to third year of life as a result of respiratory distress of suffocation from food entering the respiratory passage. Finally, patients with Gaucher type 3 (so-called chronic neuronopathic Gaucher disease) may experience the same blood and bone anomalies as Gaucher type 2, however, with the neurological complications usually progressing at a slower rate. Gaucher type 3 usually onsets during the first decade of life and patients may live into their teens, early 20s and some even longer. Some patients diagnosed with Gaucher type I develop neurological symptoms later in life.

Gaucher is the most common lysosomal storage disease. The prevalence of all Gaucher types is about 1 to 100,000 individuals or higher in the general population with an exceptionally high prevalence in the Ashkenazi Jewish population (1 per 850 individuals).[33] Neuronopathic Gaucher (Gaucher type 2 and 3), which is the focus of Orphazyme, is estimated to account for 10-30% of all patients with Gaucher in the Western world whereas it becomes the dominant form of the disease in North Africa and South-East Asia, including India, Pakistan, China and Japan.[34]

The diagnosis of Gaucher is made upon characteristic symptoms, as described above, especially in conjunction with enlargements of the spleen and/or liver and fractures. The diagnosis can be confirmed by clinical evaluations and numerous tests measuring the activity in e.g. the white blood cells or skin cells along with an analysis of potentially mutated genes.

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[31] Medical Marketing Economics for Orphazyme, 2016
[32] Medical Marketing Economics for Orphazyme, 2016 and Actelion Pharmaceuticals Annual Report 2016
[33] Siebert et al., Brain, 2014
[34] EMA and Orpha.net (http://www.orpha.net/consor/cgi-bin/index.php)

Treatment options for Gaucher and unmet need

Two types of treatment are currently available for patients with Gaucher; enzyme replacement therapy, such as Cerezyme (Sanofi), Elelyso (Pfizer) and Vpriv (Shire), and substrate reduction therapy using Zavesca or Cerdelga (Sanofi) that reduce the production of substrates. These treatments are all approved based on their ability to improve the peripheral features but not the neurological manifestations of Gaucher disease[35] and none of them are therefore approved for treatment of Gaucher disease type 2 and 3.

As enzyme replacement therapy and substrate reduction therapy does not treat the pathology of the central nervous system, a significant unmet need for treatment of Gaucher continues to exist, especially for Gaucher type 2 and type 3 and Gaucher type 1 patients developing neurological symptoms at a later stage.

Competition for treatment of Gaucher

In addition to the availability of enzyme replacement therapy and substrate reduction therapy, one study, in addition to Orphazyme's program, is currently being performed to test the safety and efficacy of another treatment option. A phase II trial on GZ/SAR 402671 (conducted by Sanofi), a substrate reduction therapy, is currently in progress.[36] The clinical efficacy in this study is unknown to the Company.

Market for treatment of Gaucher

Based on the prevalence above the total number of Gaucher patients in the United States and EU is conservatively estimated at between 10,000 and 15,000 individuals. Of the total market, Orphazyme focuses on the 10-30% with Gaucher type 2 or type 3, as well as type 1 patients with neurological symptoms as the treatment needs of these groups are not met by current treatment options.

A number of reference points exist for the market for treatment options for Gaucher type 1. In 2016, global sales of products for treatment of Gaucher disease were in excess of EUR 1.2 billion.

Drug development and, particularly, orphan drug development

The development of drugs is subject to strict regulation by various regulatory and ethical authorities. A drug is granted marketing authorisation by the regulatory authorities only when the authorities assess that sufficient documentation concerning its safety and efficacy has been obtained. To obtain such documentation, biotech and pharmaceutical companies undergo several pre-specified phases to cover all aspects of a pharmaceutical compound. The different phases are: the discovery phase, the pre-clinical phase and the clinical phase. Only once safety and efficacy have been documented in these phases, can a marketing authorization be granted by the authorities. The drug development process, from discovery to authorization, often takes 10-20 years and requires significant financial investments.

As part of the drug development process, drug developers must engage in extensive chemistry and manufacturing controls (CMC) activities. Such activities are necessary as a part of the drug development and commercialization processes. CMC activities include activities performed to establish the properties of product candidates: their chemical composition, stability, and solubility as well as activities relating to subsequent scaling of manufacturing processes to ultimately industrial scale, as typically only small amounts are manufactured for use in pre-clinical studies followed by larger size production of the drug for clinical studies.

Manufacturing activities must take place in accordance with good manufacturing practices ("**GMP**") which is a regulatory framework for ensuring that medicinal products are consistently produced and controlled according to quality standards and which covers all aspects of production; from the starting materials, premises and equipment to the training and personal hygiene of staff. GMP requires that there are systems in place to provide documented proof that correct procedures are consistently followed at each step in the manufacturing process.

Finally, in order to manufacture pharmaceuticals, both for clinical trials and sale in the market, the relevant manufacturer must hold a valid manufacturing permit from the relevant authorities. For a permit to be granted, manufacturing must take place in suitable premises and be performed by use of suitable equipment, and also be performed in accordance with cGMP. The activity of manufacturing pharmaceutical products as well as the related process validation, -development and -optimisation is lengthy and resource consuming. It is common practice for biopharmaceutical development stage companies to rely heavily on outsourcing CMC-activities to CDMOs that have the required level of expertise while maintaining at the same time a limited in-house CMC-capability devoted to organising and checking the outsourcing activity and making sure that all steps of the drug development process are being co-ordinated.

[35] Bennett et al, Ann Pharmacother, 2013
[36] Company analysis and clinicaltrials.gov

Discovery phase and pre-clinical phase

Drug development begins with a screening of pharmaceutical compounds to identify lead molecules that may target the pathology of diseases of interest. Laboratory tests are performed to characterize the effects of the lead molecule on the disease in question. At this stage, the first patent applications are usually filed.

When a potential drug candidate has been identified, pre-clinical studies are performed to study the effect of the pharmaceutical compound in living cells ('in vitro') and animals ('in vivo'). Usually, the in vitro phase is used to establish and characterize the mechanism of action, the potential efficacy and toxicity of the lead molecule. Different cell types can be used including cells originating from patients with diseases of interest. The 'in vivo' phase includes pre-clinical studies in animals to further characterize the lead molecule in terms of efficacy and toxicity/safety and its pharmacokinetics. Studies of potential side effects in animals are a prerequisite imposed by regulators to maximize patient safety in subsequent studies. The pre-clinical studies investigate the compound's safety and mechanism of action, and evaluate whether the compound has sufficient pharmacological activity. Finally, a secure starting dose is calculated for administration to humans in the subsequent clinical phase.

Upon completion of the pre-clinical studies, the results are presented to the regulatory authorities and ethical committees. Based on the pre-clinical results and a clinical study protocol an application for the initiation of clinical studies is submitted to the relevant regulatory authorities. When approved, clinical trials may be initiated.

Clinical phase

The next phase of the drug development process is to test the drug in humans in what is referred to as clinical trials. Clinical trials are investigations to evaluate new ways to prevent, diagnose or treat diseases in human patients. They test medical products or techniques, or they explore new ways of using existing treatments. Based on the results of clinical trials, scientists can evaluate treatments in relation to both safety and effectiveness. Clinical trials to evaluate potential new therapies are launched when there is good evidence from pre-clinical studies that the evaluated potential new therapy has an acceptable benefit/risk ratio. Clinical trials are sometimes referred to as 'clinical studies', 'medical research trials' or simply 'trials'. As described below, the clinical phase is typically comprised of three different sub-phases; phase I, II and III.

Phase I

In phase I trial, focus is primarily on testing the safety and pharmacokinetics of a drug candidate. Phase I trials are usually performed on a small group of healthy volunteers (except where the drug may have severe toxicity, such that testing in healthy people would be unethical). Different doses of the drug are administered to the group of patients being studied to monitor how well the drug is tolerated. If a drug demonstrates a satisfactory safety and pharmacokinetic profile, the phase I trial concludes with a dose selection for continued human trials.

Phase I/II

A trial that tests the safety, side effects, and best dose of a new treatment. Phase I/II clinical trials also test how well certain patient responds to a new treatment. In the phase II part of the clinical trial, patients usually receive the highest dose of treatment that did not cause harmful side effects in the phase I part of the clinical trial. Combining phase I and II may allow research questions to be answered more quickly or with fewer patients.

Phase II

In phase II trials, the drug candidate's safety and efficacy are studied within a specified group of patients who suffer from the disease for which the drug candidate is intended. Trials are usually based on a limited number of patients yet large enough to allow dose finding and enable appreciation of efficacy (albeit usually not statistically fully powered) and safety of the drug candidate in the targeted patient population. The purpose of phase II is to evaluate the drug candidate's effects on the targeted disease and its symptoms. In the event of tolerability problems or other adverse effects, the dosage for later trials is determined.

Phase III

If a drug candidate successfully completes phases I and II, phase III (sometimes called confirmatory) trials are initiated to generate data on the drug candidate's safety and efficacy profiles required for achieving the necessary regulatory approvals and marketing authorizations. Usually, the drug candidate is compared to placebo (a formulation without an active component) or to a different treatment for the same disease. The aim is to confirm the findings from Phase II by studying a larger patient population which is necessary for obtaining a sound basis for statistical analysis, in particular of the efficacy endpoints. In order to achieve a marketing authorization, the applicant must be able to document that the treatment induces an improvement for the patient in terms of effect and/or adverse effects.

Phase II/III

In certain cases, the objectives of the phase II and III trials can be accomplished by performing a single phase II/III trial.

Design of clinical trials

For a treatment to be introduced (or changed), trials showing improvements for the patients in terms of effect and/or adverse effects, are needed. In order to do so, two or more patient groups are compared in the same trial. Each patient group in a clinical trial is called a "study arm" or simply an "arm". Patients are usually allocated into each of the arms on a random basis after which each arm receives a distinct treatment. The most common comparative trial is called a placebo-controlled trial and, in the case of two arms, involves one arm receiving the new treatment option and the other a placebo. Any differences between the arms in terms of effects and/or adverse effects can then be attributed to the new treatment option. However, a placebo-controlled trial does not address whether the new treatment option is better, just as good as or poorer than existing treatment options available in the market. To do this, a direct comparative trial can be performed by treating one group with an existing drug and another with the new treatment option (which typically requires a very large sample size). Upon completion of the trial, any differences between the groups in terms of effects and/or adverse effects can be attributed to the new treatment option.

Clinical trials can be performed in several ways. "Open label" trials imply that both patients and physicians are aware of the treatment option. On the contrary, "double-blind" trials imply that the patients and the physician (as well as the sponsor) are not aware of the specific treatment a patient has been given to remove any biases.

Regulatory process

The drug development process is governed by an extensive set of rules and laws that developers must adhere to at all times. The rules and laws vary in different countries. However, the following information focuses on orphan drugs and primarily addresses regulations in the United States, with elements of EU rules.

The regulatory process during pre-clinical and clinical trials

Prior to the initiation of pre-clinical trials on animals, specific permissions must be obtained from the relevant authorities in the country where the trial is to be conducted. In order for any clinical trials to be initiated, the drug candidate must fulfill an extensive set of regulatory requirements and approvals must have been obtained beforehand.

If the drug being developed targets a disease which only affects a small number of people, and if certain other criteria are met, the regulatory bodies can grant a potential drug an orphan drug designation. An orphan drug designation entails multiple benefits during the development phase including protocol assistance from the FDA and EMA and financial incentives. For more information on the orphan drug designation, please see the section on *"Orphan Drugs"* above.

In the United States, Investigational New Drug ("**IND**") applications are submitted to the FDA and in the EU Clinical Trial Applications ("**CTA**") are submitted in each of the countries where the trial of the drug is to be conducted. Furthermore, the applicant must also seek and receive approvals from relevant local ethical committees. During the drug development phase, a pharmaceutical company can request a scientific advice meeting with the regulatory authorities or authorized bodies such as the FDA, the Danish Medicines Agency ("**DMA**"), the English Medicines and Healthcare Products Regulatory Agency ("**MHRA**") or EMA. The purpose of these meetings is to receive regulatory guidance on the company's development plans and to improve the quality of the application for a marketing authorization once the clinical trials are concluded. Companies seeking advice for a drug with an orphan drug designation can request scientific advice meetings at any time during the development phase. One important meeting is usually the End of Phase II-meeting with the FDA held before the phase III trials commence. During this meeting, the company presents results from the phase II trials along with information on the planned phase III trials and other studies supporting the new drug application ("**NDA**") or the marketing authorisation application ("**MAA**"). The purpose of the meeting is to receive guidance on the requirements and design of the phase III trials and the intended package of clinical and non-clinical studies as well as CMC to support the NDA or MAA. In addition to the aforementioned scientific advice meetings, orphan drug developers may also receive answers on questions related to the retention of the orphan drug designation and approval of orphan drugs.

The regulatory process upon completion of clinical trials

Based on a successful completion of the phase III trials an application for marketing authorization (NDA in the US; MAA in the EU) is submitted to the relevant regulatory authority or authorized body, e.g. the FDA in the United States and EMA in the EU. For these applications an electronic Common Technical Document ("**eCTD**") is submitted to the agencies that consists of five modules covering administrative and prescribing information, information on quality (pharmaceutical documentation), non-clinical information (pharmacology/toxicology) and clinical trial information and overview and summaries of non-clinical, clinical and pharmaceutical quality documentation. Typically, a large proportion of the information included in the dossier for the application of one indication will be relevant for applications for other indications as well. This includes for example information on drug quality, non-clinical toxicology studies, phase I studies and clinical pharmacology studies and safety information across studies conducted thus far. The FDA's review process usually takes around

twelve months. However, drugs that address an unmet medical need, and where the data generated during the pre-clinical and clinical trials is particularly strong, may have a considerably shorter review period with the FDA. The FDA has several options of expediting the review process, including fast track status, priority review designation, accelerated approval and breakthrough therapy designations.

In the EU, the PRIME scheme was introduced in March 2016 to shorten the time to market of drugs that target an unmet medical need. Through earlier and greater degree of interaction between the developers of drugs and the EMA, the purpose of the PRIME scheme is to accelerate the process of launching drugs to the market. Orphan drugs may qualify for such schemes although that is not always the case, as the authorities make this determination on an individual basis.

In the United States, the FDA may review drug applications in a Fast Track process if the FDA assesses that the drug in question targets a serious disease for which there is a significant medical need and the results obtained to date indicate clinical benefits. A Fast Track process entails ongoing reviews by the FDA throughout the development process to ensure that questions and issues are resolved quickly. Furthermore, the Fast Track process allows for modules of the NDA to be submitted and reviewed on a "rolling basis" to ensure they are completed by the time the clinical trials are completed and reviewed.

The review process of the FDA is currently performed on one of two levels; standard and priority. A standard review is the default review granted to most drug developers and targets a review period of 10 months. In 2007 the US Congress created the priority review voucher program to encourage development of drugs for neglected tropical diseases. The program was in 2012 expanded to include rare pediatric diseases that meet certain criteria. Under the program, a sponsor who receives an approval for a new medicinal product for a "rare pediatric disease" may qualify for a voucher that can be redeemed to receive a priority review of a subsequent marketing application for a different product that would otherwise be ineligible for priority review. The priority review voucher system for pediatric diseases was reauthorised by the US Congress in December 2016 extending the program to 1 October 2020. Under a priority review process, the FDA seeks to take action on the application generally within six months (although it does not mean that the application will necessarily be approved within this timeframe). Any rare pediatric disease priority review voucher may be revoked by the FDA, if the product for which such voucher was awarded is not marketed in the US within one year from the approval of the drug. Further, the sponsor of an approved rare pediatric disease product shall report to FDA not later than five years after the approval of the applicable application of: the estimated population in the United States suffering from the rare pediatric disease, the estimated demand in the United States for such rare pediatric disease product and the actual amount of such rare pediatric disease product distributed in the United States. Vouchers may be freely sold. A voucher's sale price may be freely determined and depends on supply and demand. Reported sales prices have been significant. By way of example only, in February 2017, Sarepta Therapeutics Inc. reported that it has sold to Gilead Sciences, Inc. a Rare Pediatric Disease Priority Review Voucher awarded to it by FDA in connection with the approval of Exondys 51™ (eteplirsen). As of 1 October 2017, the range of disclosed sale prices for priority review vouchers is $67,500,000 to $350,000,000.

Orphan drug developers can in some cases also be eligible for an accelerated approval process. An accelerated approval enables the approval of drugs for treatment of severe conditions with a great medical need on the grounds of a surrogate endpoint (i.e. non-clinical endpoint), given that reasonably likelihood exists of clinical benefits being documented at a later stage. The approval process thus reduces the time required to conduct the clinical trials as surrogate endpoints can be used to infer efficacy earlier than if clinical endpoints, obtained at the end of the clinical trials, were to be obtained prior to approval. However, the FDA usually requires additional post-approval studies to strengthen the evidence of clinical benefits upon which the approval was granted initially. Drugs intended to treat a serious or life threatening disease, and whose preliminary clinical data indicates considerable improvements compared to existing treatments, may be granted a Breakthrough Therapy Designation. Such a designation provides eligibility for priority review and other benefits, such as access to FDA senior management and ongoing regulatory communications and often leads to a shorter approval process with the FDA.

Business

Investors should read this section in conjunction with the more detailed information contained in this document, including the financial and other information appearing in "Operating and Financial Review". Where stated, financial information in this section has been extracted from Orphazyme's financial information and related notes.

Overview of the business

Orphazyme is a biotech company with a late stage orphan drug pipeline based in Copenhagen, developing new treatment options for orphan protein misfolding diseases. The Company focuses on severe and mostly fatal diseases with high unmet need, and with a particular 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 Europe and the United States after which launch and commercialisation may be undertaken by the Company alone or through a partnership with a pharma or biotech company.

The Company's lead candidate, arimoclomol, stimulates the body's own production of heat shock proteins ("**HSPs**") in cells experiencing stress or toxicity. HSPs form the core of the cells' protein rescue system and guard against the toxicity arising from misfolded proteins by supporting correct folding of mutated proteins and the cell's recycling system.

Arimoclomol is in development as a potential treatment for four orphan diseases; two neuromuscular diseases, sIBM and ALS, and two lysosomal storage diseases, NPC and Gaucher disease. Across phase I trials and phase II trials in sIBM and SOD1-ALS, arimoclomol was well tolerated. Phase II trials have shown trends of treatment benefit. The clinical trials in NPC and Gaucher disease are pursuant to phase I trials in healthy volunteers and pre-clinical studies showing benefit of arimoclomol in the respective lysosomal storage diseases. Orphazyme is currently conducting, or in late stage planning of, three trials that are expected to serve as pivotal/registration trials if completed with a successful outcome.

As of the date hereof, Orphazyme employs 30 people with relevant experience and expertise in the research and clinical development of orphan drugs. The Company conducts research in the field of HSPs with the aim of identifying diseases and further characterising pathologies that may be targeted with HSP-based treatments. The Company has developed assays and expertise to assess new leads and have identified a number of new molecular entities ("**NMEs**"), which may generate clinical candidates. Clinical development is managed by a clinical development team with experience in rare diseases. A number of activities are outsourced to contract research organisations ("**CROs**") and contract development and manufacturing organisations ("**CDMOs**"). This helps ensure a cost-effective development model that allows internalisation of the relevant expertise at the appropriate time during the development process.

History and development of the Company

Orphazyme was founded in 2009 with the objective to develop new therapies for patients suffering from protein misfolding diseases with no or limited treatment options available. At inception, the Company was based on a scientific discovery on the function of HSPs by Thomas Kirkegaard Jensen (current CSO) and Professor Marja Jäättelä that was published in the scientific journal Nature.[37] Since inception, the Company has translated the scientific discovery into a late stage clinical development programme.

A brief historical overview of the key milestones in Orphazyme's development is presented below:

2009	• Orphazyme is founded to pursue the opportunity of developing new therapies based on the cell protective function of HSPs
2010	• Orphazyme's current CSO and co-authors publish scientific foundation of the Company in Nature • Orphazyme raises DKK 22 million in a seed financing round (share issue). Novo Holdings and Sunstone Capital become shareholders
2011	• Orphazyme acquires arimoclomol and a portfolio of other molecules from the United States based biopharmaceutical company CytRx • Orphazyme completes a series A financing round with cash proceeds of DKK 104 million. Aescap Venture becomes shareholder
2013	• EMA grants orphan drug designation to HSP70 for the treatment of NPC
2014	• EMA grants orphan drug designation to arimoclomol for the treatment of NPC • Wellcome Trust Pathfinder Award is awarded to Orphazyme in collaboration with Oxford University for the project 'Regulation of the Heat Shock Response as a Treatment for Niemann-Pick Type C disease' • Orphazyme receives the EY Entrepreneur of the Year award in the Life Science category

[37] Kirkegaard et al., Nature, 2010

2015	• Orphazyme raises DKK 150 million in a series B financing round (share issue). Kurma Partners and Idinvest Partners become shareholders • FDA grants orphan drug designation to arimoclomol for the treatment of NPC • Orphazyme initiates observational trial in NPC
2016	• University College London and University of Kansas invest DKK 1.3 million as part of the sIBM collaboration • Pre-clinical and phase II clinical data with arimoclomol as a potential for the treatment for sIBM is published[38] • EMA grants orphan drug designation to arimoclomol for the treatment of sIBM • Orphazyme and University of Miami announce successful phase II trial of arimoclomol in SOD1-ALS patients.[39] Data presented at the 27th International Symposium on ALS/MND in Dublin • Orphazyme's current CSO and co-authors publish pre-clinical data demonstrating the potential of HSP70 and arimoclomol as a treatment for multiple LSDs, including NPC and Gaucher disease[40], in Science Translational Medicine • FDA grants a fast track designation for the phase II/III clinical trial with arimoclomol for the treatment of NPC • Dosing begins in Orphazyme's phase II/III clinical trial with arimoclomol for the treatment of NPC
2017	• Orphazyme raises DKK 109 million as an extension to the series B financing round (share issue). LSP and the ALS Investment Fund (through ALS Invest 2 B.V. ("**ALS Invest**")) become shareholders • Orphazyme completes enrolment into the clinical phase II/III trial with arimoclomol for the treatment of NPC • Dosing begins in Orphazyme's phase II/III clinical trial with arimoclomol for the treatment of sIBM • End of phase II meeting with FDA regarding arimoclomol for ALS

Orphazyme's key strengths

Orphazyme believes that it has a number of strengths, which will contribute to a successful implementation of its strategy, including:

- *Late stage project pipeline with several potential registration studies ongoing or in planning*
- *Careful choice of target indications, based on insight into the function of HSPs and optimisation for clinical success by focusing on diseases with well-defined patient populations, disease pathology and/or genetics*
- *Technology platform that shows potential with clinical proof-of-concept of the lead molecule*[41]
- *Technology protection with active patenting strategy aimed at maintaining Orphazyme's technology position*
- *Experienced management team with relevant and complementary skill set*

Late stage project pipeline with several potential registration studies ongoing or in planning

Orphazyme is developing arimoclomol in four indications. Orphazyme has fully recruited a phase II/III trial in NPC (trial results expected Q3 2018), initiated a phase II/III trial in sIBM in August 2017 and plans to start a phase II/III trial in ALS in H2 2018. In addition, a phase II proof-of-concept trial in Gaucher disease will be initiated in 2018. If positive, all three phase II/III trials are intended to form basis for a single study filing in each respective indication. The Company expects to have completed all three phase II/III trials by the end of 2020 with the first potential marketing authorisation in 2020.

The ongoing phase II/III trial to assess the safety and efficacy of arimoclomol in NPC was initiated following pre-clinical data supporting the advancement of arimoclomol in NPC and favourable tolerability in phase I trials. While the phase II/III trial is a small trial including 50 patients and the first trial of arimoclomol in NPC, it has the potential to form the basis for a single study approval.

Following clinical proof-of-concept for arimoclomol (based on trend effect across independent efficacy endpoints) in a phase II trial in sIBM, a phase II/III trial has been initiated in August 2017 to assess the safety and efficacy of arimoclomol for the treatment of sIBM. The trial was started as an investigator initiated trial under a FDA grant and the Company is expected to assume full control of the trial by Q1 2018. Similarly, following the clinical proof-of-concept (based on trend effect across independent efficacy endpoints) achieved in a phase II trial of arimoclomol in ALS, a phase II/III trial is planned to start in H2 2018. Subject to a positive outcome, Orphazyme intends to file for registration based on a single trial in each of these indications.

Orphazyme plans to conduct a phase II trial in arimoclomol for Gaucher disease in Q2 2018 with results expected in 2019. The Company expects to start a phase III registration study, if the phase II trial demonstrates clinical proof-of-concept (based on trend effect across independent efficacy endpoints).

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[38] Ahmed M et al, Science Translational Medicine, 2016
[39] Company announcement *"University of Miami and Orphazyme ApS Announce Successful Phase II Trial of Arimoclomol in ALS Patients"*, 9 December 2016
[40] Kirkegaard et al, Nature, 2017
[41] Benefit across efficacy endpoints and assessments as compiled evidence that constitutes proof-of-concept, also in absence of statistical significance.

Three phase II/III trials will provide results within three years. If successful, each trial has the potential to form the basis of filing for approval.

Careful choice of target indications, based on insight into the function of HSPs and optimisation for clinical success by focusing on diseases with well-defined patient populations, disease pathology and/or genetics
Orphazyme integrates insight into the function of HSPs with information about the genetics and molecular pathology of diseases to identify good fits for a HSP-based treatment approach. The Company makes use of recent advances in the characterisation of the genetics and molecular pathologies of protein misfolding diseases, which provide a large source of data to identify relevant diseases. The Company selects candidate diseases for HSP-based treatment by performing pre-clinical experiments in-house and in collaboration with leading researchers around the world.

Orphazyme aims to enhance development success rates by choosing target patient populations with well-defined pathology and/or genetics that are expected to respond more uniformly to treatment and by using clinical and biochemical parameters to select and stratify patients for the clinical trials. A tightly defined patient population increases the likelihood of robust signal detection in a clinical trial and to optimise chances of clinical success, Orphazyme therefore specifically pursues rare disease indications instead of more common diseases.

The principle of Orphazyme's development approach was demonstrated in the recently completed a first of its kind phase II clinical trial of arimoclomol in ALS patients with pathogenic SOD1 mutations and rapid disease progression. SOD1-ALS is a genetically defined sub-type of ALS that accounts for approximately 2% of ALS patients. The trial results paved the way for a phase II/III trial in ALS that is expected to commence in H2 2018. Orphazyme believes that ALS is a good fit for a HSP-based treatment approach since it is a protein misfolding disease involving the formation of toxic protein aggregates. Arimoclomol has shown promising pre-clinical data in the best characterised animal model for ALS – a mouse model with a pathogenic SOD1 mutation.

Technology platform that shows potential with clinical proof-of-concept of the lead molecule[42]
Orphazyme possesses extensive insight into the function of HSPs and the convergence of HSP function with protein misfolding and cellular recycling pathways. Orphazyme believes that the ability to apply these insights into the disease pathology in protein misfolding diseases is key to successful development of HSP-based therapies.

Orphazyme's lead program, arimoclomol, has been studied in a number of pre-clinical experiments allowing it to be characterised in a variety of different disease models and pathologies, many of which are published in leading peer-reviewed scientific journals including Nature Medicine and Science Translational Medicine. The ability of arimoclomol to mobilise HSPs and provide benefit have been validated across several protein misfolding diseases including relevant animal models for NPC, ALS and sIBM. Moreover, the benefit of arimoclomol has been established and characterised in cells from patients diagnosed with NPC and Gaucher disease across a range of mutations.

Arimoclomol has been studied in double blinded, placebo-controlled, clinical phase II trials in both sIBM and ALS including 24 and 36 patients, respectively. The primary endpoint of these trials was safety and tolerability. Efficacy endpoints were secondary exploratory endpoints, implying that neither trial was powered to show efficacy. However, both trials showed consistent trends of clinically relevant efficacy. Further, arimoclomol has been well-tolerated with no significant safety risks identified as of the date hereof.

Technology protection with active patenting strategy aimed at maintaining Orphazyme's technology position
Since its founding Orphazyme has strategically and actively pursued patent protection of its inventions. Orphazyme now holds the rights to a comprehensive patent portfolio and continues to actively pursue further patent protection and exclusivity opportunities. Orphazyme focuses on protecting small molecule inducers of HSPs, including the main candidate arimoclomol, as well as NMEs.

Protection is sought for a relevant scope to obtain commercial exclusivity in the main areas, and covering geographically at least the United States and Europe (via the European Patent Convention), as well as further geographic areas of interest. Patents directed to arimoclomol and its use in treating NPC, Gaucher disease and ALS are issued and in force in the United States and a range of European countries.

Experienced management team with relevant and complementary skill set
Orphazyme's senior management team has experience from the biotech and pharmaceutical industries with a broad range of skills to succeed in the industry, including experience with research and development, finance and intellectual property rights. The Company's CEO and CSO have been with the Company since its inception.

[42] Benefit across efficacy endpoints and assessments as compiled evidence that constitutes proof-of-concept, also in absence of statistical significance.

The current and new Board of Directors comprises industry experts with experience from companies focused on rare diseases such as Genzyme and Swedish Orphan Biovitrum.

Strategy

Orphazyme's strategy is to develop treatments for orphan diseases with protein misfolding where it can apply its specialised know-how in HSPs. Important elements of Orphazyme's strategy are the following:

- *Advance the development of arimoclomol for the treatment of sIBM, ALS, NPC and Gaucher disease by completion of clinical development programmes*
- *Design the commercialisation and go-to-market strategy for arimoclomol*
- *Develop new molecular entities for other protein misfolding diseases based on current technology platform*

Advance the development of arimoclomol for the treatment of sIBM, ALS, NPC and Gaucher disease by completion of clinical trials

Orphazyme's objective is to successfully conduct and complete the planned and ongoing trials of arimoclomol for the treatment of the neuromuscular diseases, sIBM and ALS, and the lysosomal storage diseases, NPC and Gaucher disease. Orphazyme develops new therapies for orphan diseases where few products, if any, have made it through to regulatory approval and the Company maintains frequent interactions with the regulatory bodies in the United States and Europe to advance its programme toward approval in the most expedient manner.

Design the commercialisation and go-to-market strategy for arimoclomol

As the clinical development programme for arimoclomol progresses, Orphazyme intends to refine and finalise its commercialisation strategy and build its commercial structure and operations. The Company intends to build its own sales force in key markets. In markets outside the United States and Europe, Orphazyme currently intends to partner with local/regional distributors or license partners in certain other geographical areas.

Develop new molecular entities (NMEs) for other protein misfolding diseases based on current technology platform

Protein misfolding is the hallmark of a broad range of diseases and Orphazyme's strategy is to use its expertise, including proprietary know-how to select and develop new leads for suitable diseases. In line with this, the Company is developing a proprietary suite of NMEs with improved characteristics. Orphazyme intends to select diseases suitable for the NMEs based on genetic and mechanistic insights into selected protein misfolding diseases.

Arimoclomol

Introduction

Orphazyme develops treatments for protein misfolding diseases based on expertise in heat shock proteins, which form the core of the cells' rescue system. Orphazyme and academic collaborators have developed and used assays and models to characterise the benefit of HSP-based therapy in a number of pre-clinical disease models of lysosomal storage disorders ("**LSDs**") and protein misfolding and/or aggregation diseases such as NPC, Gaucher disease, sIBM, and ALS.[43] Based on these insights, Orphazyme is developing a broad pipeline of drugs targeted at amplifying the body's own production of cytoprotective HSPs, spearheaded by arimoclomol.

Pipeline

Orphazyme's current clinical programmes investigate arimoclomol as a treatment for four indications: the neurodegenerative and -muscular diseases sIBM and ALS, and in the LSDs NPC and Gaucher disease. Orphazyme's development pipeline for the four target indications is shown below, with the important next steps highlighted. The NME programme focuses on the development of new molecules as treatment for relevant protein misfolding diseases.

[43] Kirkegaard et al., Nature, 2010; Kirkegaard, Gray et al., Science Translational Medicine, 2016, Kieran et al., Nature Med, 2004; Ahmed et al., Science Translational Medicine, 2016.

Area	Key milestones achieved	Next steps
sIBM	• Phase II trial (24 patients), arimoclomol was well-tolerated and clinical proof-of-concept[44] achieved	• Phase II/III trial (12 months treatment duration), initiated in August 2017
ALS	• Phase II SOD1-ALS trial (36 patients), arimoclomol was well-tolerated and clinical proof-of-concept[45] achieved	• Initiation of phase II/III trial (12 months treatment duration), start H2 2018 • Pursue broad ALS indication
NPC	• Pre-clinical data in animal models and patients' cells supporting the advancement of arimoclomol in NPC • Enrolment of phase II/III clinical trial (50 patients) completed • Fast track designation granted by the FDA • Paediatric investigation plan agreed with the EMA	• Results of phase II/III trial in Q3 2018 • If positive: filing for NDA (US) / MAA (EU) in H2 2019
Gaucher disease	• Pre-clinical data in patients' cells supporting the advancement of arimoclomol in Gaucher disease: arimoclomol demonstrate activity across all major genotypes	• Initiation of phase II trial, start anticipated Q2 2018
NMEs	• Pre-clinical studies on new molecules	• Additional early phase studies to establish leads

Arimoclomol's mechanism of action ("MOA")

Arimoclomol works by increasing the production of HSPs inside the cells and thereby enhancing the natural biological mechanisms that reduce protein misfolding and aggregation.

Protein misfolding diseases

Proteins are our cells' molecular machines, making certain that each cell in the body fulfils its function. To do their job, they must attain the correct shape. In many diseases protein misfolding may cause toxicity either as a consequence of protein aggregation or the loss of protein function. Protein aggregates, a hallmark of neuromuscular diseases such as sIBM and ALS, leads to toxicity as aggregates perturb the cell functions. Loss of function, a classical feature of the LSDs such as NPC and Gaucher, leads to an accumulation of toxic substances as a result of the absence of functional proteins that would otherwise have dealt with such waste products.

Heat shock proteins

The HSPs constitute a natural system that makes other proteins work correctly and guard against the toxicity arising from misfolded proteins and dysfunctional cellular recycling systems. In particular, HSPs are molecular chaperones that promote the survival of stressed cells by re-folding misfolded proteins into their correct conformation, or by directing 'terminally' misfolded protein to be broken down. They also protect cells by inhibiting lysosomal membrane permeabilisation; stabilising lysosomes (cellular structures where waste products are broken down), allowing cells to clear away waste and return to their healthy status.

There are several different types of HSPs which work in conjunction – a cardinal member is HSP70, which Orphazyme uses as the key parameter to measure activity of its drug candidates ("**HSP70**"). HSP70 has been shown to protect against the formation of protein aggregates which are the defining characteristic of a number of neurodegenerative diseases including ALS and sIBM. In addition, HSP70 has been identified as a co-factor for lysosomal sphingolipid breakdown, a necessary step in the metabolism of stored lipids which cause toxicity if accumulated in the lysosome. In both NPC and Gaucher disease, as well as other LSDs, mutations lead to misfolding and loss of enzyme functions involved in the breakdown and recycling of critical cellular components within the cells recycling centres, the lysosomes. By amplifying the production of HSPs, this pathological cascade can be addressed by rescuing the function of the recycling enzymes and helping them perform better in the lysosomes.

Arimoclomol's MOA

Arimoclomol stimulates the production of HSPs.

The production of HSPs is regulated by a transcription factor, heat shock factor 1 ("**HSF1**"). A transcription factor is a protein that regulates production of other proteins in the cell. In the case of HSF1, the proteins being regulated are HSPs. Activation of HSF1 starts the production of the major stress-inducible HSP70-chaperone along with other HSP-chaperones, which help reshape the cells' misfolded proteins and take care of the recycling systems. Under normal cellular conditions, HSF1 is inactive. However, the transcription factor can be activated by an initial cellular stress, such as protein misfolding, and becomes fully activated under a sustained stress signal.

Arimoclomol amplifies and prolongs the activated, HSP-producing state of HSF1. This leads to an amplification in the production of cell protective HSPs, but only in physiologically stressed cells.

[44] Clinical proof-of-concept in sIBM achieved by demonstrating benefit across efficacy endpoints and assessments as compiled evidence, also in absence of statistical significance.
[45] Clinical proof-of-concept in ALS achieved by demonstrating benefit across efficacy endpoints and assessments as compiled evidence, also in absence of statistical significance.

Clinical profile and uses of arimoclomol

The clinical experience with arimoclomol supports its continued development. Highlights from clinical trials performed to date are summarised below.

Safety

As of the date hereof, no safety risks from taking arimoclomol have been identified in seven phase I trials and three phase II trials, and, overall, tolerability of arimoclomol (i.e. the degree to which an overt adverse effect can be tolerated by the subject/patient) was comparable to a placebo.

A comprehensive non-clinical program covering pharmacology, pharmacokinetics, single and repeat dose toxicity, genotoxicity, reproductive toxicity, and interaction studies has been conducted. In these non-clinical studies, oral administration of arimoclomol was found to be safe and well tolerated. Mild gastrointestinal effects were reported in a few animals after administration of arimoclomol.

To date, a total of 112 healthy subjects have received between 50 mg and 1800 mg arimoclomol daily. 82 ALS patients received arimoclomol across the 12-week phase II dose ranging trial (AALS-001) and the six-month open-label extension trial (AALS-001-OL). In an investigator-initiated trial in SOD1-ALS, 19 patients were treated with 300-600 mg arimoclomol daily for 12 months. In another investigator-initiated trial in sIBM 16 patients were treated with 300 mg arimoclomol daily for four months. Potential mild tolerability side effects identified were diarrhea, nausea and dry mouth. In the trials these side effects did not result in treatment discontinuation. Arimoclomol is an inhibitor of the organic cation transporter 2 (OCT2), and thus inhibits OCT2 dependent transport of creatinine. Thereby arimoclomol can lead to a transient decrease in creatinine clearance and increase in serum creatinine that is not considered a safety risk. In the trials, serum creatinine increases were still within normal limits and the values went back to baseline levels while subjects continued treatment with arimoclomol.

In the AALS-001 trial, nine serious adverse events (SAEs), including three deaths, were reported in the placebo-controlled main part of the study; two incidences of pulmonary embolus; two incidences of myasthenia/ALS progression; one incidence of apnea; one incidence of bone fracture; one incidence of dysphagia; and one incidence of respiratory disorder; one incidence of thrombophlebitis. All SAEs were determined to be either unlikely or not related to trial medication. There were no treatment related deaths. In the AALS-001-OL, the open label extension of the trial, 13 patients had SAEs. There was one SAE, a pulmonary embolism, which was deemed possibly related to trial medication. All other SAEs were deemed not related or unlikely related to the trial medication. In the phase II SOD1-ALS trial adverse events, occurred with similar frequency in the two treatment groups, and were largely considered unrelated to trial drug by the site investigators. 22 SAEs were reported (15 in the placebo group and seven in the arimoclomol-treated group), none of which were considered related to trial drug. A single participant stopped arimoclomol treatment because of a skin rash that was deemed probably related to trial drug. In the sIBM trial involving 16 patients on active treatment and eight patients on placebo there was no difference in the rate, type and severity of adverse events between the arimoclomol and the placebo group. One SAE of high blood pressure was noted in the arimoclomol group. In the ongoing phase II/III trial in NPC, two patients developed urticarial and angioedema, respectively, the treatment allocation remains blinded until the end of the pivotal part of the trial.

While none of the above events has led to the identification of a safety risk that would be considered associated with the treatment with arimoclomol, given that still a limited number of patients have been exposed to date for a limited time and daily dose, rare safety risks or risks associated with higher doses cannot be excluded. Often, uncommon risks are only identified when thousands of patients have been treated long term, not uncommonly after years on the market.

Pharmacokinetics

Arimoclomol is orally administered and has a bioavailability of 80-90%. High bioavailability reduces the amount of drug administered while achieving the desired pharmacological effect.

In addition, arimoclomol readily crosses the blood-brain barrier. The blood-brain barrier is a selectively permeable membrane which prevents large compounds from entering the cerebrospinal fluid ("**CSF**") (liquid surrounding the brain and spinal column). Arimoclomol reaches CSF drug concentrations comparable to the concentration found in circulating blood. Good brain penetrance is key to treatment of many central neurological diseases.

Efficacy

Arimoclomol has achieved a clinical proof-of-concept in ALS[46] and sIBM[47]. Phase II trials in both indications have shown consistent trends of clinically relevant efficacy in pre-defined analyses.

[46] Clinical proof-of-concept in ALS achieved by demonstrating benefit across efficacy endpoints and assessments as compiled evidence, also in absence of statistical significance.

[47] Clinical proof-of-concept in sIBM achieved by demonstrating benefit across efficacy endpoints and assessments as compiled evidence, also in absence of statistical significance.

Arimoclomol for the treatment of sIBM

sIBM is a rare and relatively slowly progressing muscle wasting disorder with onset typically after 50 years of age. The prognosis for patients with sIBM is poor, with the majority progressing relentlessly towards disability over a period of 10 to15 years. Pathophysiologically, sIBM is characterised by abnormal processing of proteins leading to the formation of protein aggregates or plaques, known as inclusion bodies, in muscle and immune cells. These inclusion bodies can be seen using a microscope following a muscle biopsy, and are the hallmark of the disease.

With the exception of very rare familial forms of inclusion body myositis ("**IBM**"), IBM is predominantly a sporadic (spontaneous) disease. The size of the patient population in Europe and the United States is not fully elucidated but has been conservatively estimated to be between 7,000 and 15,000 individuals.[48] A recent meta-analysis of nine sIBM prevalence studies concludes that prevalence is approximately 24.8 per million or 17,000 individuals in the United States and the major European countries.[49] Many clinicians believe that sIBM is underdiagnosed and thus the number of patients is likely higher than current estimates. No effective treatment is currently available for sIBM, resulting in a high unmet medical need.

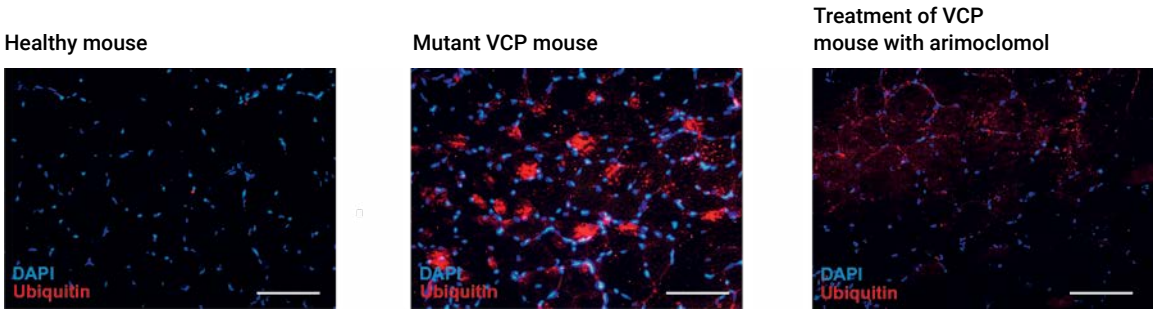
Results from a randomised, double blinded, placebo-controlled phase II clinical trial in sIBM was published in 2016.[50] The primary endpoint was safety but trends of efficacy were shown on clinically relevant endpoints at all assessment time points. Post-hoc responder analysis indicated a very consistent treatment response (please see *"Business—Arimoclomol for the treatment of sIBM—Clinical trials of arimoclomol for sIBM"*).

The next step for Orphazyme is to conduct a clinical phase II/III trial that, if successful, has the potential to form the basis for registration. The first patient was enrolled in August 2017.

Pre-clinical studies of arimoclomol in sIBM

Arimoclomol has been tested in both in vitro and in vivo models of sIBM. In in vitro models of sIBM in primary rat myocyte cultures, administration of arimoclomol was found to induce HSP70 and provided improvement on the molecular pathological hallmarks of the disease; the inclusion bodies and the processes leading to their formation. This was demonstrated by the reduction of the accumulation of ubiquitinated inclusion bodies and other molecular markers of protein aggregation.

The cellular effects of arimoclomol were recapitulated in a mouse model of the disease, where arimoclomol provided the same benefit in the mouse muscles, which was accompanied by a significant functional improvement in clinically relevant manifestations of the disease such as muscle force. In these studies, treatment with arimoclomol was initiated after onset of clinical symptoms in the mice.[51] The figure below illustrates the test of arimoclomol's effect on IBM-like pathology in a mouse model of the disease, illustrating a consistent reduction of disease pathology upon treatment with arimoclomol.



Mutated mice with IBM hallmarks were treated with arimoclomol from disease onset (four months) plus 10 months. A cross-section of muscle cells from healthy mouse before the disease is illustrated to the left. No inclusion bodies are present (ubiquitin, red). In the middle picture, inclusion bodies (red) are evident in a mutant VCP mouse throughout the muscle cells. The mouse has a significant decrease in muscle force. In the picture to the right, treatment of the VCP mouse with arimoclomol results in upregulated HSP70, inclusion bodies are reduced and the loss of muscle force is prevented. Source: Ahmed et. al. Targeting protein homeostasis in sporadic inclusion body myositis, Science Translational Medicine, page 7, 23 March 2016, vol. 8, issue 331.

[48] Defined Health for Orphazyme, 2017
[49] Aoife C et al, Journal of Neuromuscular Diseases, 2017
[50] Ahmed et al, Science Translational Medicine, 2016
[51] Ahmed et al, Translational Medicine, 2016

Clinical trials of arimoclomol for sIBM

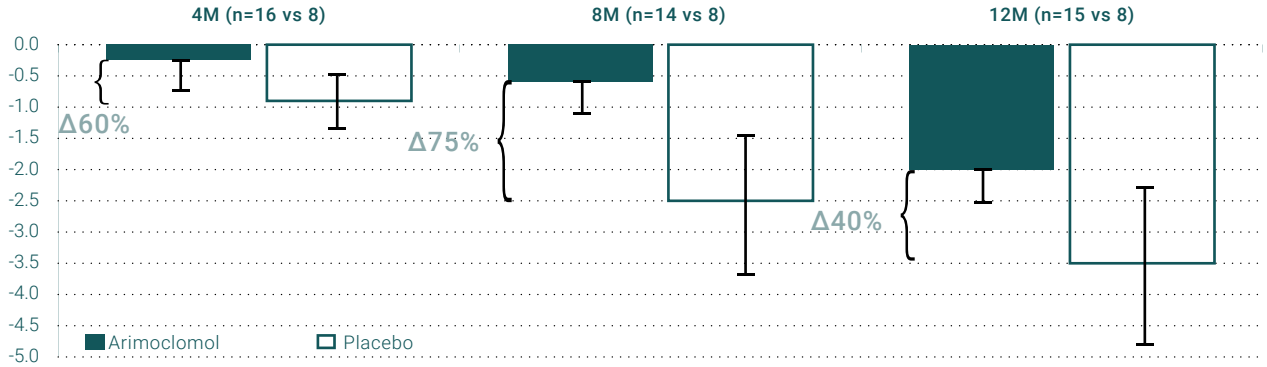
The arimoclomol phase II trial in sIBM indicated consistent trends of benefit in favour of arimoclomol across pre-defined independent efficacy endpoints and assessment time points. Following the in vitro and in vivo pre-clinical studies, arimoclomol was assessed in a phase II investigator initiated trial. The trial was a double-blinded and placebo-controlled with oral administration at 100 mg three times a day for four months with assessments up to 12 months after initiation of randomised treatment.

Trial design. Patients were randomised 2:1 to arimoclomol versus placebo treatment, respectively, and treated for four months. Baseline clinical and demographic characteristics were similar between the groups. Efficacy assessments continued beyond the randomised treatment period for eight months and efficacy was thus assessed for a total of 12 months. The primary endpoint was safety and tolerability. Efficacy endpoints were secondary and statistically not powered for. These secondary endpoints[52] included the IBM functional rating scale ("**IBMFRS**"), manual muscle testing and the maximum voluntary isometric contraction testing. IBMFRS is a ten-domain functional rating scale with a total score range 0-40 (within each domain, 0 ascribed to no function and 4 ascribed to normal function) which assesses a patient's ability to complete a range of tasks relevant in daily living, including writing, swallowing and climbing stairs.

Patient number	24
Randomisation	2:1 (arimoclomol: placebo)
Dosage	100 mg three times per day
Treatment length	4 months
Follow-up assessment length	8 months
Primary endpoint	Safety and tolerability
Secondary endpoints	IBMFRS and muscle strength

Results. Arimoclomol was well tolerated. After four months of treatment the arimoclomol group demonstrated 60% reduction in progression on the IBMFRS sum score from the baseline compared to placebo. The effect of arimoclomol on the change in IBMFRS sum score was maintained beyond the four-months treatment period. Compared to the placebo group, the decline in IBMFRS sum score was reduced by 75% and 40% at eight months and at 12 months[53], respectively. Although not powered for efficacy, there was a trend in favour of arimoclomol on the IBMFRS reaching almost statistical significance (p-value of 0.055) at eight months. Similar effects were observed for the two other efficacy endpoints, manual muscle testing and the maximum voluntary isometric contraction testing.

Change in IBERMRS sum score

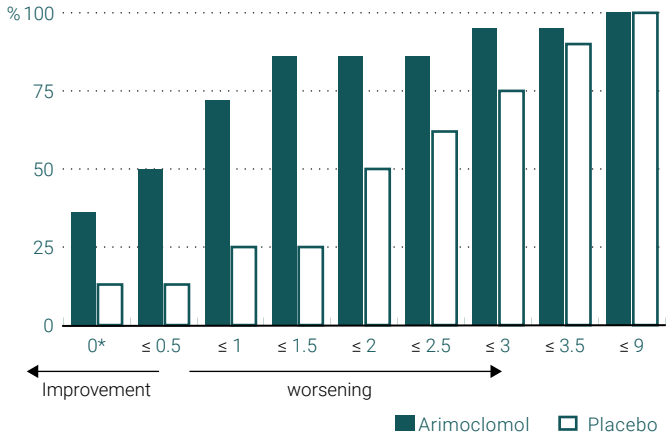


Change in IBMFRS score (disease progression) among trial participants receiving either arimoclomol (filled bar) or placebo (white bar). The figure illustrates that the progression was faster among patients receiving placebo compared to patients receiving arimoclomol. N value indicates the number of patients receiving arimoclomol who attended each clinic visit (not all patients were able to attend the eight and 12 month follow up visits). The trial was not powered for efficacy, and results were not statistically significant. Source: Ahmed et. al. Targeting protein homeostasis in sporadic inclusion body myositis, Science Translational Medicine, page 8, 23 March 2016, vol. 8, issue 331.

[52] Clinical endpoints are the target outcomes of a clinical trial. Primary endpoints provide the most relevant evidence which the trial is designed to investigate, e.g. whether one treatment is more effective in treating a disease than another. Clinical trials are designed so that there is sufficient statistical power to indicate if the primary endpoint(s) has (have) been met. Secondary endpoints are outcomes of interest, but ones which the trial is not specifically powered to measure.

[53] Ahmed, M.; Machado, P. M.; Miller, A.; Spicer, C.; Herbelin, L.; He, J.; Noel, J.; Wang, Y.; McVey, A. L.; Pasnoor, M.; Gallagher, P.; Statland, J.; Lu, C.-H.; Kalmar, B.; Brady, S.; Sethi, H.; Samandouras, G.; Parton, M.; Holton, J. L.; Weston, A.; Collinson, L.; Paul Taylor, J.; Schiavo, G.; Hanna, M. G.; Barohn, R. J.; Dimachkie, M. M.; Greensmith, L. Targeting protein homeostasis in sporadic inclusion body myositis. Sci. Transl. Med. 8 (331), 331ra41 (2016) – DOI: 10.1126/scitranslmed.aad4583.

In a post-hoc analysis, the assessment of disease progression showed that, irrespective of responder definition, patients on arimoclomol treatment fared better than those on placebo treatment, as illustrated in the figure below. Typically, sIBM patients progress by losing 2.0 to 2.5 points per 8 months[54]. In the arimoclomol trial, 86% of patients in the arimoclomol treatment group lost 1.5 points or less on the IBMFRS score over eight months compared to 25% in the placebo group. Equally noteworthy, 36% of the patients did not progress at all or improved during the eight months (0 points lost).



Response Criteria	Ari (%)	Pbo (%) (%)
Stable or improvement	36	13
≤ 0.5 pt progression	50	13
≤ 1.5 pt progression	86	25
≤ 9 pt progression (all patients)	100	100

Cumulative distribution analysis of disease progression in arimoclomol-treated versus placebo patients at eight months. The cumulative percentage of patients in the treatment and placebo arms for each disease progression rank in the left chart shows that there is a higher proportion of patients with no or slow disease progression in the arimoclomol arm than the placebo arm. The right chart illustrates the share of patients in each of the treatment groups with a disease progression less than or equal to four pre-specified thresholds and shows a higher proportion of patients with no or slow disease progression when treated with arimoclomol compared to placebo.

Conclusion. The trial results demonstrate that 100 mg arimoclomol administered three times per day for four months was well tolerated and associated with clinically meaningful benefits when comparing with placebo treatment and these benefits persisted for several months beyond treatment period. The trial was not powered to show efficacy and did not reach statistical significance. However, a responder analysis revealed that the trends of efficacy were very consistent in the arimoclomol treated group.

Next steps. The next step in the development of arimoclomol for the treatment of sIBM is the conduct of a phase II/III trial in the United States and Europe to establish efficacy. The trial was initiated in August 2017 and is intended to form the basis for registration. Orphazyme conducts the trial in collaboration with the University of Kansas and the University College London. The trial is a randomised, double blinded, placebo-controlled phase II/III trial assessing efficacy and safety of arimoclomol 400 mg three times a day in patients with sIBM. The primary endpoint analysis is after 12 months, while the trial duration will continue for up to 20 months. 150 patients (75 per arm) will be enrolled, randomised 1:1 to receive arimoclomol or placebo. The primary efficacy endpoint will be the change from baseline to 12 months in the IBMFRS total score. Secondary endpoints include the change from baseline to month 20 in the IBMFRS sum score (durability of treatment) and changes from baseline to month 12 and 20 in different measures of strength and function. The average changes in these outcomes will be compared between placebo and arimoclomol at 12 and 20 months, respectively. While the trial started as an investigator initiated trial led by the University of Kansas, and supported by a FDA grant, the trial sponsorship has been and the IND is currently in the process of being transferred to Orphazyme. Orphazyme expects to assume full control of the trial by Q1 2018. Results from the phase II/III trial are expected in H1 2020.

Arimoclomol for the treatment of ALS

ALS is an aggressive and fatal neuromuscular degenerative disease with onset typically around 40-60 years of age. The expected time from onset to mortality is generally two to five years. The disease is rapidly progressive and invariably fatal. It attacks neurons responsible for controlling voluntary muscles (muscle action we are able to control, such as those in the arms, legs, and face).

The incidence of ALS is estimated at between 1-3 per 100,000 individuals per year. The disease occurs more frequently in men than women, whereas prevalence is roughly the same throughout the world (4-5.4 per 100,000 individuals) with no ethnic, racial or socioeco-

[54] Cortese et al., Neuromuscul Disord, 2013; Morrow et al., Lancet Neurol., 2016.

nomic boundaries. The patient population in Europe and the United States is estimated to be approximately 50,000 patients[55] and about 10 percent of ALS cases are inherited.[56]

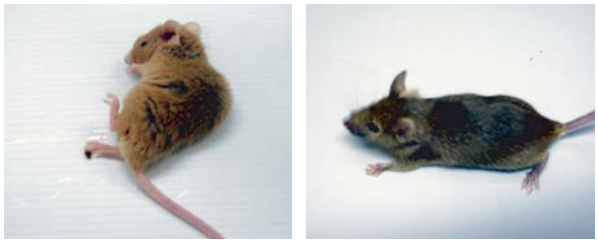
Until recently, the only available therapy was Riluzole, which has a limited efficacy demonstrating approximately three months prolonged survival. In May 2017, the FDA approved the use of edaravone for the treatment of ALS, based on a phase III trial in 137 patients. However, there is still an urgent need for new therapies. For further information, please see "*Industry—ALS—Treatment options for ALS and unmet need*".

A randomised, double-blinded, placebo-controlled phase II clinical trial in SOD1 ALS was concluded in 2016. Consistent trends of efficacy were observed across all endpoints and arimoclomol was well-tolerated with no risks identified. The Company met with the FDA at an end-of-phase II meeting in May 2017 to discuss the results.

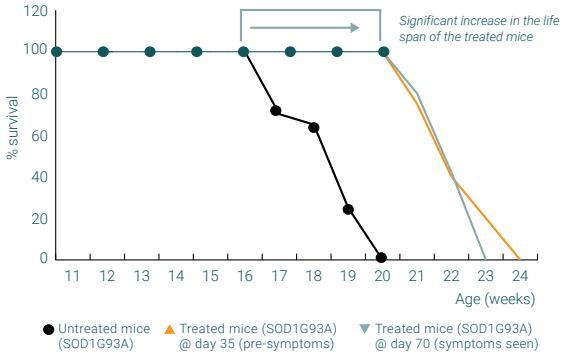
The next step for Orphazyme is to initiate a clinical phase II/III trial in the broad ALS population expected to commence in H2 2018. If the outcome is positive, the Company intends to file for registration for ALS.

Pre-clinical studies of arimoclomol in ALS

Arimoclomol has been extensively tested in the best characterised pre-clinical model of ALS – a transgenic mouse model overexpressing human mutant SOD1. This model has a phenotype and pathology similar to the human disease, including loss of motor neurons in the spinal cord and resulting loss of muscle function. In this model, arimoclomol treatment led to amplified production of HSPs with concomitant reduction of protein aggregates in affected motor neurons in the spinal cord. The arimoclomol-treated mice also showed improvement in motor neuron survival, functional benefit and extension of life by up to 22%, even when arimoclomol was administered after symptom onset[57] as illustrated in the figure below. An expert review when the findings were published stated that more than 70 drugs have been tested in the mouse model, but only few compounds prolong survival by more than 10%, even when started pre-symptomatically.[58]



Untreated Arimoclomol treated



Results of the SOD1-ALS mouse study. The pictures of mice to the left depict untreated and arimoclomol-treated mice. The untreated SOD1 mouse shows significant signs of hind limb muscle wasting, no toe-spreading reflex, marked kyphosis and is unable to right itself. This mouse was judged to have reached the disease endpoint. The arimoclomol treated, age-matched SOD1 littermate mouse shows definite toe-spreading reflex, no signs of hind limb muscle wasting or kyphosis, and is able to perform a righting reflex test with no delay. The panel on the right depicts Kaplan-Meier survival plots of SOD1 mice over time. A decline in the curve represents a death, so the higher the line, the more extended the survival. Mice treated with arimoclomol (red and green triangles) survived significantly longer than untreated mice (black circles), even when treatment was started after disease onset (at 70 days, green triangles). Source: Kieran et al. Treatment with arimoclomol, a coinducer of heat shock proteins, delays disease progression in ALS mice, Nature Medicine, page 404, April 2004, vol. 10, no. 4.

Clinical trials of arimoclomol in ALS

A 12 weeks dose-ranging trial in ALS patients was performed with the objective to assess the safety, tolerability, and pharmacokinetics of arimoclomol in ALS. 84 participants with ALS received arimoclomol at one of three oral doses (25, 50, or 100 mg three times daily) or placebo. The primary outcome measure was safety and tolerability. Arimoclomol at doses up to 300 mg/day was well tolerated and safe. Arimoclomol resulted in dose-linear pharmacologic exposures and the half-life did not change with continued treatment. Importantly, arimoclomol CSF levels increased with dose and reached a CSF-to-serum ratio of 1 at steady state and equilibrium indicating that arimoclomol is fully CNS penetrant. 69 participants who completed 12 weeks of treatment enrolled in a six-month open-label trial at the highest dose (100 mg three times daily). In the open label trial, the rate of decline of ALSFRS-R was slower in the arimoclomol treated group than in a historical placebo control group, controlling for baseline ALSFRS-R (P=0.034).

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[55] Defined Health for Orphazyme, Mehta, P.; Kaye, W.; Bryan, L.; Larson, T.; Copeland, T.; Wu, J.; Muravov, O.; Horton, K. Prevalence of Amyotrophic Lateral Sclerosis - United States, 2012-2013. MMWR Surveill Summ, 2016;65(8):1–12 and Chiò, A.; Logroscino, G.; Traynor, BJ.; Collins, J.; Simeone, JC.; Goldstein, LA.; White, LA. Global epidemiology of amyotrophic lateral sclerosis: a systematic review of the published literature. Neuroepidemiology. 2013; 41(2): 118–130. DOI:10.1159/000351153.

[56] Taylor et al., Nature, 2016

[57] Kieran et al., Nature Medicine, 2004; Kalmar et al., J Neurochemistry, 2008

[58] Robert H Brown Jr, News and Views, Nature Medicine, 2004

Consistent trends of benefit across aldl pre-defined independent efficacy endpoints has been achieved in an investigator led randomised, double-blinded, placebo-controlled phase II clinical trial in SOD1 ALS. The purpose of the trial was to investigate the safety and efficacy of arimoclomol in ALS patients with pathogenic mutations in the SOD1 gene. Although the trial was not powered for efficacy, the results revealed a consistent trend of clinically meaningful efficacy across all efficacy endpoints. A key aspect of this trial was the selection of a trial patient population with pathogenic SOD1 mutation and rapid disease progression which allowed for efficacy signal detection despite the small size of the trial.

Trial design. Patients were randomised 1:1 into two treatment arms, arimoclomol and placebo, and assessed for a total of 12 months. The primary endpoints were safety and tolerability. The secondary efficacy endpoints included survival, the revised ALSFRS-R and forced expiratory volume in six seconds ("**FEV6%**"), a measurement of lung function. In addition, the combined assessment of survival and function ("**CAFS**"), a composite score of survival and ALSFRS-R, was included.

Patient number	36 (all SOD1 mutations) of which 26 with A4V mutation
Geography	United States and Canada
Primary investigator	Professor Michael Benatar, University of Miami (United States)
Randomisation	1:1 (arimoclomol : placebo)
Dosage	200 mg three times per day
Treatment length	12 months
Follow-up assessment length	None
Primary endpoint	Safety and tolerability
Secondary endpoints	Survival, ALSFRS-R, FEV6%, CAFS

Baseline clinical and demographic characteristics were similar between groups with the exception of pulmonary function and SOD1 mutations other than the A4V mutation. Of the SOD1-ALS subpopulation, the mutation known as A4V is the most commonly detected mutation in the SOD1 gene in the United States, and it is associated with a very aggressive disease course. The most frequent mutation was the A4V mutation, accounting for 72% of patients enrolled. A4V patients were well balanced across the two treatment arms (given the large proportion, sub-analyses were pre-defined for patients with this mutation).

Results. The primary endpoints of safety and tolerability were met. While not powered to show statistically significant therapeutic effect, the trial results indicated a consistent benefit of arimoclomol over placebo on all pre-defined clinical endpoints as exemplified by a hazard ratio of death 0.67 in favour of arimoclomol. Patients with the common A4V mutation were balanced between the treatment and placebo group. Other mutations were either only in the treated group or in the placebo group. An efficacy analysis of the A4V subpopulation was therefore predefined in the protocol.

In the A4V patient population, the trial demonstrated a clinically meaningful reduction of the progression rate on the ALSFRS-R scale by up to 39%. The treatment difference increased when correcting for Riluzole use and baseline pulmonary function. On the survival endpoint, hazard ratios of 0.59 were in favour of arimoclomol. In line with a functional and survival benefit there was also a reduction in the decline of pulmonary function by up to 33%. CAFS, a score combining functionality and mortality, also indicated that arimoclomol was superior to placebo (20.5 for arimoclomol versus 14.5 for placebo, where a lower rank score indicates a worse outcome). Sensitivity analyses correcting for baseline imbalances confirmed the effects observed in the primary analyses.

Conclusion. The trial results demonstrate that 200 mg arimoclomol administered three times per day for 12 months was well tolerated. The trial also demonstrated that patients treated with arimoclomol showed trends of clinically meaningful efficacy across all clinical endpoints when compared to placebo treatment. The trial was not powered to show efficacy and thus did not reach statistical significance. However, a pre-defined analysis of patients revealed that the trends of efficacy were consistent in the arimoclomol treated group across all pre-defined clinical endpoints.

Next steps. Based on the mechanism of action of arimoclomol that is expected to be relevant across the broad ALS patient population, the results in the phase II SOD1 ALS trial, trend effect observed in the open label portion of the phase II trial in sporadic ALS and encouraging feedback from FDA, Orphazyme is working on a phase II/III trial design intended to support a marketing authorisation in the broad ALS population. The Company is currently evaluating a design that will include clinical and genetic enrichment strategies to ensure homogeneous disease progression in the trial. The next step is to finalise the protocol for submission to the EMA for scientific advice prior to enrolling patients in the trial. Expectedly, the trial will be a randomised, double blinded, placebo-controlled phase II/III trial assessing efficacy and safety of arimoclomol 400 mg three times a day. The trial is expected to enrol approximately 200-300 patients.

The plans have been discussed with the FDA at an end-of-phase-II meeting. The agency was supportive of the approach. The trial is planned to start in H2 2018 and results are expected in H2 2020.

Arimoclomol for the treatment of NPC

NPC disease is a rare, genetic and progressive disease that is usually fatal. The onset of NPC varies from fatality during the first months after birth to a progressive disorder not diagnosed until adulthood and the majority of individuals with the disease die before the age of 20. NPC is caused by an inherited mutation in the NPC1 gene, which prevents cells from properly processing waste lipids and leads to an accumulation of lipids in the lysosomes. These lysosomes become overwhelmed and the toxicity causes progressive loss of cell function. The CNS involvement results in progressive motor and brain impairment. Symptom onset and disease progression are highly variable and most patients do not expect to live beyond their late teens.[59]

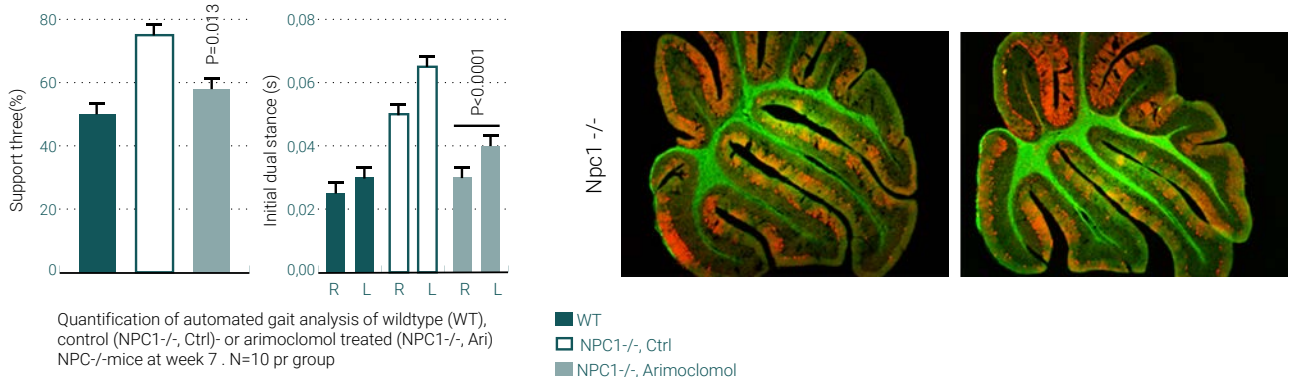
The incidence of NPC is approximately 1 in 120,000 new-borns, meaning there is a conservatively estimated total patient population in Europe and the United States of 1,000-2,000 patients.[60] In line with the higher end of this range, the United Kingdom Niemann-Pick disease patient organisation reports registration of 101 patients (as of August 2017). There is currently one compound, miglustat, approved in Europe for treatment of NPC.

Orphazyme published its pre-clinical studies in 2016 and the results demonstrated that arimoclomol may correct the underlying pathology of NPC. Following a pre-IND meeting with the FDA, Orphazyme moved directly from pre-clinical studies and clinical phase I data in healthy volunteers to a phase II/III registration trial. The trial was fully enrolled in Q2 2017 and is expected to have trial results in Q3 2018.

Pre-clinical studies of arimoclomol in NPC

Arimoclomol has been extensively tested in pre-clinical models of NPC. Recent scientific publications underscore that the induction of HSPs is an attractive therapeutic target in LSDs.[61] This is particularly the case for NPC, where HSP70 has been demonstrated to be critical for the proper folding and activity of the NPC1 protein.[62] In addition, it has been demonstrated that the majority of NPC patients have mutations that are responsive to therapies aimed at rescuing and improving the folding of the NPC1 protein.[63]

The pre-clinical studies published in 2016 demonstrated a clear potential therapeutic benefit of arimoclomol in NPC.[64] These studies demonstrated that arimoclomol increased the activation of HSF1 and production of HSP70 in the brains of the NPC mouse model. The studies further demonstrated that HSP70 reduces CNS white matter (myelin) loss and cerebellar atrophy in the NPC mouse model. Importantly, treatment with arimoclomol improved all measurable manifestations of loss of motor coordination/ataxia in the animal model and extended life. This is important as loss of coordination is identified to be the most important factor for quality of life in patients.



Quantification of automated gait analysis of wildtype (WT), control (NPC1-/-, Ctrl)- or arimoclomol treated (NPC1-/-, Ari) NPC-/-mice at week 7 . N=10 pr group

Rescue of brain white matter and quantification of improved coordination. The figure on the right illustrates a brain cross section of NPC mice without (left) and following (right) treatment to increase brain levels of HSP70. The increased green area demonstrates the rescue of white matter. The figure on the left illustrates the loss of coordination quantitatively measured by gait analyses. Treatment with arimoclomol demonstrated a response on all (13 out of 13) quantitative ataxic manifestations of the disease, two of the most important which are depicted here.

[59] Vanier MT., Orphanet Journal of Rare Diseases, 2010
[60] Medical Marketing Economics for Orphazyme, 2016
[61] Ingemann & Kirkegaard, J Lipid Research, 2014
[62] Nakasone et al., J Biol Chem, 2014
[63] Macias-Vidal et al., FEBS J, 2014
[64] Kirkegaard, Gray et al., Science Translational Medicine, 2016

Clinical trials of arimoclomol in NPC

A phase II/III, randomised placebo-controlled trial was initiated after regulatory advice from the authorities. Interaction with the European Paediatric Committee took place in 2015 and resulted in an agreed paediatric investigation plan. A scientific advice meeting was held with EMA on refined clinical endpoints and statistical analysis plan is in preparation for Q1 2018. FDA granted Orphazyme a Fast Track designation for the investigation of arimoclomol intended for the treatment of NPC in June 2016. Based on regulatory feedback from the regulatory authorities, the Company expects that this single trial may be sufficient to form the basis for approval in NPC. The aim of the trial is to investigate the safety and efficacy of arimoclomol. Prior to the interventional trial, patients were enrolled in a prospective observational trial allowing for the assessment of natural progression in the very same patients who participate in the phase II/III trial. Information obtained from this observational trial offered the opportunity to adjust the phase II/III statistical analysis plan. The multi-centre trial completed enrolment in May 2017, with patients recruited at sites across Europe and the United States.

Trial design. Patients are randomised 2:1 to arimoclomol and placebo, respectively, and assessed for a total of 12 months of randomised treatment, followed by open-label treatment of up to 24 months. The purpose of the trial is to assess the efficacy and safety of arimoclomol when administered in addition to patient's current prescribed best standard of care. The primary endpoint is disease severity as measured by the NPC clinical severity score (NPCCSS).

Patient number	50
Geography	Europe and the United States
Randomisation	2:1 (arimoclomol : placebo)
Dosage	Weight-adjusted 200 mg three times per day
Randomised treatment length	12 months
Follow-up assessment length	24 months open-label extension
Primary endpoint	NPCCSS
Secondary endpoints	CGI, other clinical endpoints,biomarkers, liver / spleen pathology assessments and quality of life

Next steps. Orphazyme's phase II/III clinical trial is ongoing as of the date hereof. The trial was fully enrolled in Q2 2017 and trial completion is expected in H2 2018. Contingent on positive results of the trial, Orphazyme expects to file an NDA (US) and MAA (EU) in H2 2019 with potential approval in H1 2020.

Arimoclomol for the treatment of Gaucher disease

Gaucher disease is an inherited disorder affecting many of the body's organs and tissues. Patients may live into their teens, early 20's and some even longer. The disease is caused by mutations in the beta-glucosidase gene ("**GBA**") which causes waste lipid to accumulate in lysosomes of cells in the brain, spleen, liver, bones and the immune system. Gaucher disease is classified into three subtypes; types II and III are known as 'neuronopathic' because of the involvement of the nervous system, resulting in progressive brain damage. While splenomegaly with anemia and thrombocytopenia, hepatosplenomegaly and bone disease usually are the prominent symptoms in Gaucher disease type I, treated patients living longer have a high risk of developing parkinsonism later in their disease course. In spite of occurrence of neurological symptoms, these patients are usually not reclassified as type III.

Gaucher disease is the most common LSD, affecting a conservatively estimated patient population of 10,000-15,000 in Europe and the United States combined, of which neuronopathic Gaucher disease accounts for 10-30%.[65] Orphazyme focuses on Gaucher disease type II and III with a focus on neuronopathic symptoms. Type II occurs in new-borns and infants and usually progresses to life-threatening complications between the first to third year of life.

Two primary therapies exist for patients with Gaucher disease; ERT and a substrate reduction therapy (Cerdelga® and Zavesca) that reduce the production of substrates. These treatments improve the peripheral features of the disease, but not the neuropathic symptoms as none of the treatments are capable of effectively crossing the blood-brain barrier.

In pre-clinical studies arimoclomol has been demonstrated to amplify HSP70 production leading to an increase in refolding, maturation and correct localisation of GBA, meaning that the enzyme was correctly built and located in the part of the cell where it is needed. The increase in GBA activity was confirmed in a complimentary neurological Gaucher disease model system.

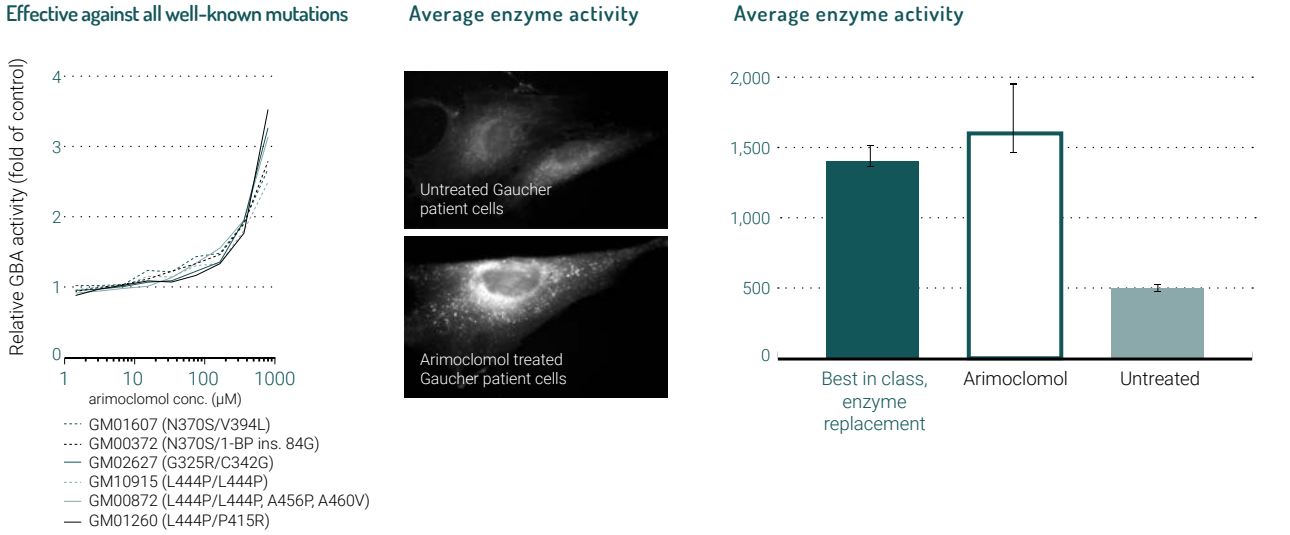
The next step for Orphazyme is to conduct a phase II clinical trial in approximately 40 patients. The Company expects the first patients to be enrolled in Q2 2018 with a primary analysis to take place after six months of intervention.

[65] EMA and Orpha.net (http://www.orpha.net/consor/cgi-bin/index.php)

Pre-clinical studies of arimoclomol in Gaucher disease

The potential efficacy of arimoclomol in Gaucher disease has been extensively tested in pre-clinical *in vitro* assays. In accordance with its mechanism of action, arimoclomol induced HSP70 and facilitated the proper folding, maturation and lysosomal localisation of GBA leading to a marked effect on GBA activity across a number of different mutations, including the most abundant L444P and N370S mutations, as illustrated in the figures below. The results in primary patient fibroblasts were corroborated in a neurological model system, employing neuron-like cells (so-called neuronally differentiated multipotent adult stem cells ("**MASCs**")) from neuronopathic Gaucher patients. Consistent with the studies on primary fibroblasts, neuronal differentiated MASCs treated with arimoclomol had increased GBA activity across genotypes.

Additionally, in vitro studies comparing the effect of the gold standard treatment (i.e. ERT) for non-neurological Gaucher disease with arimoclomol demonstrated that arimoclomol provides a similar increase to GBA activity in cells from Gaucher disease patients.



Highlight of pre-clinical data on arimoclomol for the treatment of Gaucher disease. The left chart illustrates that arimoclomol increases the activity of GBA across several different mutations in cells from Gaucher patients. The microscopy pictures in the middle illustrate arimoclomol's ability to increase the GBA enzyme activity and its correct cellular localisation. Increase in active GBA and correct localisation (brighter fluorescence, cellular distribution) were visible following treatment with arimoclomol. The chart to the right illustrates that treatment with arimoclomol increases the GBA enzyme activity in Gaucher disease patient cells to levels comparable to treatment with the current, best in class ERT therapy.

Clinical trials of arimoclomol in Gaucher disease

A randomised double blinded dose-ranging phase II trial is planned to start in Q2 2018. The trial will include approximately 40 patients, who are naive to any treatment for Gaucher. The trial will take place at clinical sites in India where access to ERT is limited, allowing for assessment of the effect of arimoclomol treatment on pharmakodynamic biomarkers in blood and other tissues, as ERT treatment may obscure the effect of arimoclomol on peripheral symptoms and biomarkers. Peripheral markers of effect are validated biomarkers for treatment effectiveness in Gaucher disease. In addition, performing the trial in India will provide children suffering from this rare disease with the opportunity to receive a potential treatment.

Trial design. Patients are to be randomised 1:1:1:1 into four treatment arms – active treatment at three different doses and placebo, and assessed for six months. Following the placebo-controlled period, the placebo group will be re-randomised into one of the three active treatment groups for a six-month extension. The primary endpoint in this trial is chitotriosidase levels in CSF and blood. Chitotriosidase is a known marker of Gaucher disease, because it is found in high levels in patients whose immune cells have accumulated an excess lipid burden.

Patient number	Approximately 40
Geography	India
Randomisation	1:1:1:1 (1200 mg/day: 600 mg/day: 300 mg/ day: placebo)
Dosage	1200/600/300 mg per day
Treatment length	Six months followed by long-term open label extension
Follow-up assessment length	Ongoing assessment every six months until market authorisation or cessation of the development program in Gaucher disease
Primary endpoint	Chitotriosidase serum and CSF levels
Secondary endpoints	Additional biomarkers

Next steps. The next steps for arimoclomol in Gaucher disease will be decided based on the outcome of the phase II clinical trial.

Manufacturing of arimoclomol

Orphazyme has contracted current Good Manufacturing Practices ("**cGMPs**") for manufacturing of the active ingredient to a Contract Development and Manufacturing Organisation ("**CDMO**"). The manufacturing is done at an FDA inspected American facility. The CDMO also manages quality control, release and warehousing. In 2017, the manufacturing process went through a process risk assessment which ended in a non-public Quality by Design report. In addition, a manufacturing contract is made on a quote-by-quote basis with another partner for packaging, storage and distribution of clinical trial supplies to Orphazyme's clinical trials.

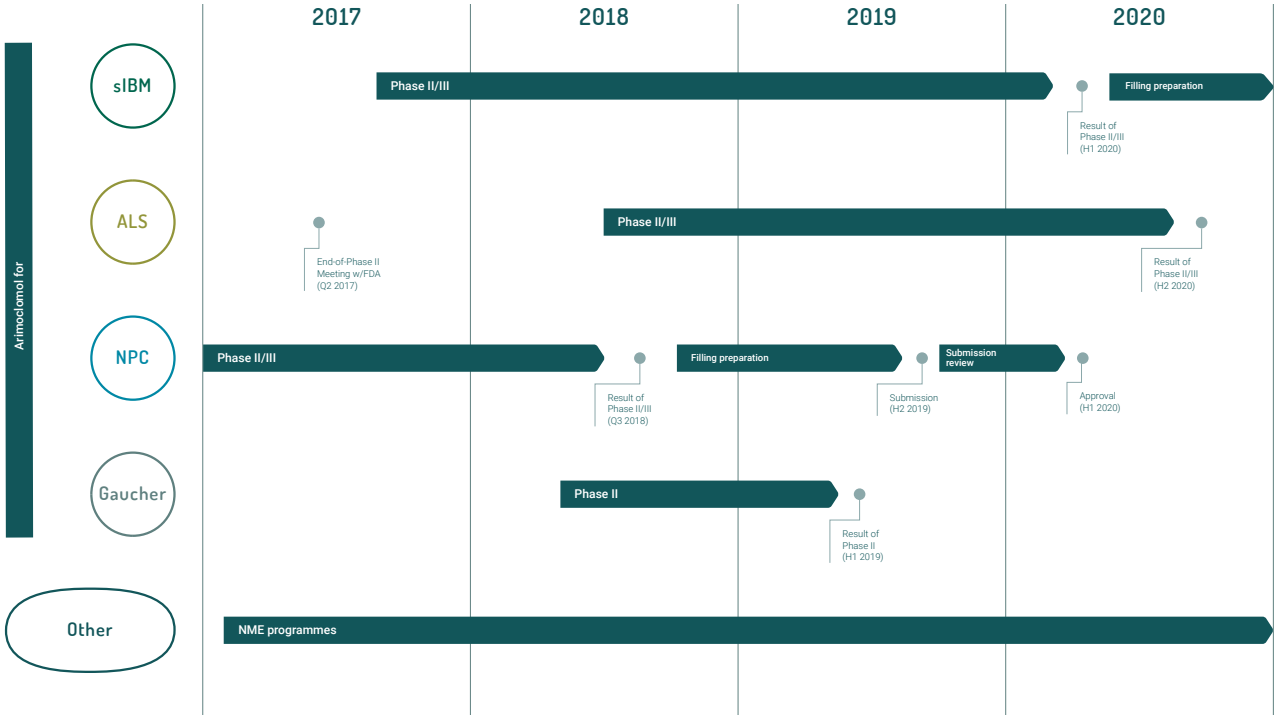
Go-to-market strategy

Orphazyme believes that a small sales force and medical and market access field teams specialised in rare diseases should be sufficient to launch the drug in both the United States and major European countries. The Company believes that such sales, medical and market access field teams can be supported by core rare disease commercial and medical operational functions based in the United States and Denmark under a unified global strategy, and with oversight by global commercial and medical leadership. The commercial build-out in Europe is expected to be more gradual and aligned with the progress and expected timelines for obtaining acceptable pricing and funding for patient access in each country.

As additional rare disease indications are approved, Orphazyme believes that this strategy allows for optimal leveraging of its core commercial capabilities and expansion when necessary to support the rare disease launches in each country. In case of successful outcomes within all four indications, the Company sees notable synergy potential and will consider expanding and organising two separate sales, market access and medical field teams based on the designated therapeutic areas of LSDs and neuromuscular diseases, supported by a central commercial and medical operations function in each country under global leadership and strategy.

Summary of expected news flow

Orphazyme has potential registration studies in several indications ongoing or in the planning. The figure below provides an overview of the expected news flow until 2020.



Timeline subject to success of trials. The expected net proceeds from the Offering will finance clinical studies and certain parts of Orphazyme's NME programmes. Funding for filing preparation, registration and potential commercialisation are not included in the use of net proceeds from the Offering. In the event of positive results from the phase II/III trial with arimoclomol for the treatment of NPC, Orphazyme may choose to reallocate funds to facilitate filing and registration activities in arimoclomol for NPC, thus accelerating the use of proceeds from the Offering.

The first indication for arimoclomol is expected to be the treatment of NPC. The results of the clinical trial in NPC are expected in Q3 2018 and regulatory approvals are expected in H1 2020. Phase II/III trials have been initated in sIBM and the first patient was enrolled in August 2017 with results expected in H1 2020. Additionally, Orphazyme expects to initiate clinical trials in the ALS and Gaucher disease in 2018.

Technology protection

Patents

Orphazyme's patent strategy aims at ensuring the protection and enforcement of inventions covering its development programmes. This has resulted in a patent portfolio with a wide scope of protection and geographic coverage. Orphazyme's main product candidate, arimoclomol, is covered by composition-of-matter patents acquired from CytRx. In addition, second medical use patents and pending patent applications cover the medical use of arimoclomol and other inducers of HSPs in the treatment of relevant medical indications. These include the LSDs, including specifically NPC and Gaucher disease, and neuromuscular/neurodegenerative disorders including ALS and Parkinson's disease. The Company is covered by patents for treatment of LSDs until 2029, with potential for up to three and five year extensions in the United States and Europe, respectively. Within the protein aggregation diseases, the Company continuously explores patent coverage for the use of arimoclomol for treatment of specific protein aggregation diseases.

Orphazyme has launched a NME programme to develop novel small molecule inducers of heat shock proteins. Compounds are continuously evaluated to obtain patent protection for the NME programme. Additionally, a new formulation of arimoclomol is tested and covered by the current patent portfolio.

Orphazyme holds the rights to 11 patent families, each with a number of issued patents (>130 total) and pending patent applications (>25 total). Patents generally expire 20 years from the filing date, with the possibility to extend the exclusivity period for pharmaceutical products of up to five years via Supplementary Protection Certificates ("**SPC**") available in most European countries (an additional six months can be added as paediatric extension if the clinical data includes children), and via the United States Analogue Patent Term Extension ("**PTE**"). The Company considers it realistic that at least one SPC/PTE can be obtained per product.

Patent (title)	Type	Expiration[66]	Regions	Status
Use of hsp70 as a regulator of enzymatic activity	Second medical use – directed to hydroxylamine derivative type small molecule inducers of the heat shock proteins, including arimoclomol, and HSP70 protein, for treatment of lysosomal storage diseases, including NPC and Gaucher	Projected patent expiry: 26.06.2029	AU, BR, CA, CN, EP, HK, IL, JP, RU, US	Granted and maintained in: AU, CN, EP*, HK, IL, JP, RU, US Pending in: BR, CA, EP, IL, US
Methods for increasing intracellular activity of hsp70	Second medical use – directed to hydroxylamine derivative type small molecule inducers of the heat shock proteins, including arimoclomol, and HSP70 protein, for treatment of additional lysosomal storage diseases	Projected patent expiry: 22.11.2031	US and EP	Granted and maintained in: US Pending in: EP and US
Arimoclomol formulation	Formulation: Extended-release formulation of arimoclomol	Projected patent expiry: 15.09.2035	AU, BR, CA, CN, EP, IL, IN, JP, KR, RU, US	Pending in: AU, BR, CA, CN, EP, IL, IN, JP, KR, RU, US
Heat Shock Proteins and Cholesterol Homeostasis	Second medical use – directed to hydroxylamine derivative type small molecule inducers of the heat shock proteins, including arimoclomol, and HSP70 protein, for treatment of diseases associated with dysregulation of cholesterol homeostasis	Projected patent expiry: 10.04.2037	International patent application (PCT)	Pending - PCT
Arimoclomol for treating glucocerebrosidase associated disorders	Second medical use – directed to arimoclomol for treatment of glucocerebrosidase GBA-associated disorders, including GBA-associated Parkinson's disease	Projected patent expiry: 28.04.2037	International patent application (PCT)	Pending - PCT

[66] Excluding potential patent term adjustments and extensions

Heat shock protein inducers and frontotemporal disorders[67]	Second medical use – directed to hydroxylamine derivative type small molecule inducers of the heat shock proteins, including arimoclomol, and HSP70 protein, for treatment of frontotemporal disorders	Projected patent expiry: 24.05.2038	Priority founding patent application (unpublished)	A PCT-application will be filed no later than 24.05.2018
Hydroxylamine derivatives useful for enhancing molecular chaperon production and the preparation thereof	Directed to hydroxylamine derivative compounds including bimoclomol in methods of increasing molecular chaperone expression	Basic patent expiry: 01.11.2016 – maintained in the US with added patent term adjustment (2018 and 2020, respectively)	US	Granted and maintained in: US
N-[2-hydroxy-3-(1-piperidinyl)propoxy]pyridine-1-oxide-3-carboximidoyl chloride and its use in the treatment of insulin resistance	Composition-of-matter - directed to the hydroxylamine derivative compound arimoclomol, its stereoisomers and salts thereof, and the use of said compounds for treatment of insulin resistance	Projected patent expiry: 24.02.2020	AU, CA, EP, IL, JP, RU, US, ZA	Granted and maintained in: AU, CA, EP*, IL, JP, RU, US, ZA
A pyridine-1-oxide derivative, and process for its transformation into pharmaceutically effective compounds	Manufacture - directed to an arimoclomol intermediate compound and its use in the preparation of arimoclomol	Projected patent expiry: 17.04.2021	AU, BR, CA, CN, EP, IL, JP, US	Granted and maintained in: AU, BR, CA, CN, EP*, IL, JP, US
Use of a hydroximic acid halide derivative in the treatment of neurodegenerative diseases	Second medical use – directed to arimoclomol for treatment of neurodegenerative diseases, including ALS and Parkinson's disease	Projected patent expiry: 25.10.2024	AU, BR, CA, EP, JP, RU, US, ZA	Granted and maintained in: AU, CA, EP*, JP, RU, US, ZA Pending in BR, US
Optically active pyridyl-4H-1,2,4-oxadiazine derivative and its use in the treatment of vascular diseases	Composition-of-matter - directed to the hydroxylamine derivative compound iroxanadine, salts thereof, and the use of said compounds for treatment of vascular diseases	Projected patent expiry: 07.12.2019	AU, CA, EP, JP, US	Granted and maintained in: AU, CA, EP*, JP, US

* Validated in select countries

Orphan drug designation

Arimoclomol has been granted orphan drug designations from the FDA and EMA for the treatment of ALS and NPC, and from the EMA for the treatment of sIBM. The Company has applied for an orphan drug designation for the treatment of sIBM with the FDA in September 2017 and expects to apply for Gaucher disease at a later stage.

Generally, the orphan drug designations entail market exclusivity for seven years in the United States and ten years in Europe, should arimoclomol receive a marketing authorisation within the targeted indications. Paediatric extensions to the orphan drug designation are available for indications with clinical data including children for an additional six months in the United States and two years in Europe.

Orphan drug designation	Authority	Regions	Market exclusivity
Treatment of sporadic Inclusion Body Myositis	EMA*	Europe	Ten years
Treatment of Amyotrophic Lateral Sclerosis	EMA and FDA	United States and Europe	Seven years in the United States Ten years in Europe
Treatment of Niemann-Pick Type C	EMA and FDA	United States and Europe	Seven years in the United States (+ 6 m paediatric extension) Ten years in Europe (+ 2 years paediatric extension)

*An application was submitted to the FDA in September 2017.

[67] Filed in the name of Orphazyme ApS and UCL Business PLC (University College London); governed by a Collaborative Research Agreement and Material Transfer Agreement signed in April 2015.

Trademarks
Orphazyme has registered "ORPHAZYME" as word mark in relevant trademark classes and jurisdictions (EU and International Madrid Protocol designating US, CN, IL, IN, JP and RU). Registration of the word mark in Israel and Japan is still pending.

Partnerships and academic collaborations

Orphazyme strives to develop its expertise within its therapeutic areas of interest through close collaborations with academic experts and patient organisations. Through these partnerships, Orphazyme supports the advancement of molecular and clinical understandings and performs pre-clinical evaluations in biological models of relevant diseases. Orphazyme's academic partners include academic professors and clinicians from institutions such as the University of Oxford, University Hospital of Udine, University College London, University of Miami, University of Cambridge and University of Kansas. As an example of such partnership structures, Orphazyme collaborates with Professor Frances Platt at the University of Oxford, a partnership which has lasted almost from the inception of the Company. Professor Platt is an expert in LSD research and pioneered the first pharmacological approach to treat these disorders. Through this collaboration, Orphazyme has conducted a number of pre-clinical studies that have provided insights into the potential of HSP amplifying therapeutic strategies for the LSDs. Further, partnerships with the patient community has ensured that patient-relevant outcomes have been assessed in scientific models and subsequently published in peer-reviewed scientific journals.

New molecular entity (NME) programmes

Orphazyme is developing a new series of HSP amplifying drugs based on its expertise and know-how about the convergences of HSPs, protein aggregation and cellular recycling systems, and how these can be targeted for therapeutic benefit. As of the date hereof, Orphazyme has several leads that constitute potentially new intellectual property opportunities. These molecules are currently being vetted and will be prioritised based on their suitability for specific diseases.

The US priority review voucher program

Orphazyme targets rare paediatric diseases and it intends to make voucher requests in connection with its FDA approval processes. The Company believes that it may be possible to satisfy the applicable voucher requirements. However, the Company's efforts and prospects in this respect are not regarded as an essential component of the Company's business strategy. Rather, the Company's efforts will be opportunistically driven only.

Orphazyme considers it possible that a rare paediatric disease voucher in connection with the NDA approval for arimoclomol for either NPC or, if the programme is still available, for Gaucher disease when neuronopathic symptoms are present. A preparation for the rare paediatric disease designation is currently ongoing.

Organisation

Orphazyme is located in Copenhagen, Denmark with address at Copenhagen Bio Science Park, Ole Maaløes Vej 3, 2200 Copenhagen N. The office in Copenhagen serves as headquarter and focuses on coordination and execution of the drug development process and on the conduct of pre-clinical and clinical trials and administration. As of the date hereof, the organisation comprises:

- Senior Management (4 persons comprising the Executive Management and Key Employees, see "*Board of Directors, Executive Management and Key Employees*"
- Administration (2 persons)
- Clinical development (9 persons)
- CMC/QA (as defined below) (2 persons)
- Regulatory (2 persons)
- Research (9 persons)

Administration is responsible for supporting the Senior Management, Human Resources, Finance and Public Relations. Clinical development comprises clinical trial and central laboratory oversight. Chemistry, Manufacturing and Controls ("**CMC**") and Quality Assurance ("**QA**") comprise internal and external audits. Regulatory comprise interactions with regulators and related strategic oversight. Research comprises internal and external efforts primarily to support ongoing clinical development and to a lesser extent identify new lead compounds with potential to enter clinical development.

As of the date hereof, Orphazyme has a total of 30 employees. Most of the staff is engaged in the development of strategies, design, planning, procurement and project management and execution of clinical trials and pre-clinical studies and the required regulatory interaction. Orphazyme intends to retain a relatively limited staff to manage its clinical and pre-clinical development programmes. An overview of Orphazyme's staff is presented below.

Period	H1'17	FY'16	FY'15
Employees (End-of-period)	28	21	14

Any expansion or recruitment as a consequence of the commercial strategy is expected to be announced during 2018.

Material Contracts

Asset Purchase Agreement with CytRx

In May 2011, Orphazyme entered into an Asset Purchase Agreement with the US biopharmaceutical company CytRx. Pursuant to this agreement, CytRx irrevocably sold and transferred certain pre-clinical and clinical data, intellectual property rights and other assets, including contractual rights and obligations relating to a portfolio of chemical compounds, including arimoclomol, to Orphazyme.

Under the terms of the Asset Purchase Agreement, Orphazyme made an up-front cash payment of USD 150,000 and further agreed to make future payments to CytRx contingent upon the achievement of specified clinical/regulatory and sales milestones as well as royalty payments based on a specified percentage of any eventual net sales of products containing one of the compounds purchased as summarised further below.

Clinical/Regulatory Milestone payment obligations (non-ALS or stroke products)

Orphazyme has agreed to pay CytRx clinical and regulatory milestone payments for the first two products being developed for indications other than for the treatment or prevention of amyotrophic lateral sclerosis or stroke (non-ALS or stroke products). Payments are triggered upon achieving certain key clinical or regulatory milestones. The aggregate amount of milestone payments that may be triggered is USD 12.10 million for the first non-ALS or stroke product and USD 10.25 million for the second non-ALS or stroke product developed assuming (for both products) approval in the EU (or a major European market), US and Japan. A second non-ALS or stroke product is not calculated as a second product (and hence does not trigger milestone payments) unless is contains a different compound than the first non-ALS or stroke product. In 2016, Orphazyme paid CytRx USD 0.1 million for achievement of a clinical milestone for the first product.

Clinical/Regulatory Milestone payment obligations (ALS or stroke products)

Orphazyme has also agreed to pay CytRx clinical and regulatory milestone payments (payable one time only) for each product developed that is being developed or labelled for the treatment or prevention of amyotrophic lateral sclerosis or stroke (ALS or stroke products). Payments are triggered upon achieving certain key clinical or regulatory milestones. The aggregate amount of milestone payments that may be triggered per ALS or stroke product is USD 23.750 million assuming approval in the EU (or a major European market), US and Japan. The milestone obligations are payable only once per ALS or stroke product. A subsequent ALS or stroke product may only achieve a milestone and trigger a payment obligation, if it contains a different compound than an ALS or stroke product previously achieving the same milestone or if it contains the same compound as another ALS or stroke product previously achieving same milestone but is for a different indication. Under the terms of the Asset Purchase Agreement, Orphazyme was assigned and hence became the party to a royalty agreement with ALS Charitable Remainder Trust and must pay 1% royalty to ALS Charitable Remainder Trust on world-wide net sales of products to treat ALS.

Sales milestones. Orphazyme also agreed to pay CytRx milestone payments upon reaching certain aggregated world-wide net sales of all products developed by Orphazyme and containing any of the compounds purchased from CytRx. The first milestone payment is triggered on aggregate net sales exceeding USD 100 million. The aggregate milestone payment obligations may be up to USD 50 million assuming world-wide net sales in excess of USD 1 billion.

Royalties. Orphazyme has agreed to pay CytRx a low double-digit royalty on net sales of ALS or stroke products and a mid-single digit royalty on net sales of all other products developed by Orphazyme or its licensees containing any of the compounds purchased from CytRx. Royalties accrue on a country-by-country and product-by-product basis until the latest of expiration of relevant patent claims in the country, expiry of regulatory exclusivity in the country or 10 years from the date of the approval of the product in the country; provided, however, that the royalty rate may be reduced on a country-by-country and product-by-product basis by 20% for the remainder of the royalty term on expiration of the relevant patent claims and the expiration of regulatory exclusivity; and by 40% in the event that there are no valid patent claims or any regulatory exclusivity at the time of first commercial sale in the country.

Orphazyme has no contractual obligations towards CytRx to develop or commercialise any products under the terms of the asset purchase agreement and Orphazyme cannot be held liable towards CytRx for failure to do so.

Option Agreement with University of Miami

In May 2017, Orphazyme entered into an option agreement with the University of Miami. Pursuant to the option agreement, Orphazyme is during an initial period granted a first option to negotiate a world-wide royalty bearing, exclusive license to data, know-how and patent rights generated by the University of Miami in a phase II clinical trial of arimoclomol to treat ALS with the a4V SOD1 mutation to use or apply the study data. Orphazyme has also been granted internal development use rights to the data, know-how and patent rights. If Orphazyme elects to exercise the option, the option agreement provides that the license may involve payment of certain license fees, milestone payments and annual fees as well as a royalty of 1.5% of net sales of products sold within ALS linked to mutations in the SOD1 gene. Any annual fees will be creditable against royalty and milestone payments. If Orphazyme elects not to exercise the option, or if the parties do not execute a license, the University of Miami may license data, know-how and patent rights to a third party, provided that certain conditions are met.

License agreement with University of Kansas and UCL Business PLC

In October 2017, Orphazyme entered into a license agreement with the University of Kansas and UCL Business PLC (a wholly-owned subsidiary of University College London). The license agreement grants Orphazyme the world-wide, royalty bearing exclusive license to develop and commercialize products under all data generated in the course of the ongoing phase II/III clinical trial on arimoclomol for the treatment of sIBM. Orphazyme's license includes any inventions and know-how included in such data. The trial was initiated in August 2017 with the University of Kansas as sponsor, but the license agreement provides that the IND and trial sponsorship shall be transferred to Orphazyme on Orphazyme's request.

Under the terms of the license agreement, Orphazyme shall pay an aggregate royalty of 2% of net sales of products sold for the treatment of sIBM. Orphazyme is required to use commercially diligent efforts to develop and commercialize such products. The license agreement also provides that Orphazyme in consideration of the license shall issue bonus Shares in favour of the University of Kansas and UCL Business PLC, for up to an aggregated value of USD 2.5 million (around DKK 15.8 million) in total depending on the size of the grants awarded to the universities under the trial (with a price per Share calculated based on the average closing price of the Shares on Nasdaq Copenhagen for the 30 days immediately prior to the date of issuance). The Shares shall be issued or delivered on a yearly basis subject to certain reporting requirements.

Agreements related to manufacture and clinical studies of arimoclomol

In order to support a cost-effective development model that allows internalisation of the relevant expertise at the appropriate time during the development process, Orphazyme engages with third party CDMOs to manufacture, store and distribute products for clinical trials. These third party CDMOs include, in particular, a cGMP manufacturer of the active ingredient and a provider of packaging, storage and distribution services in relation to Orphazyme's clinical trials. The Company also engages with third party CROs to facilitate and assist in its clinical trials.

In 2013, Orphazyme entered into a Master Service Agreement with a UK-based CRO. Under the terms of the Master Service Agreement, the CRO agreed to provide various services to support Orphazyme's clinical testing of arimoclomol to treat NPC – currently Orphazyme's II/III clinical trial of arimoclomol to treat NPC. Contracted services comprise trial set-up and monitoring, trial and data management, statistical analytical work as well as preparation of final study report. As part of the study set-up Orphazyme is co-signing as sponsor on the clinical trial agreements entered into with the individual trial sites. Under the terms of the Master Service Agreement, all intellectual property rights pertaining to arimoclomol including possible patents based on data provided from inventions made during the trial are owned by Orphazyme. Orphazyme has undertaken to indemnify the CRO for third party claims arising from the performance of the trial or from the use of arimoclomol.

Following completion of the Offering, Orphazyme expects to enter into similar type service agreements for clinical trial services with two additional CROs, to support Orphazyme's dose-ranging phase II trial of arimoclomol for Gaucher disease and its phase II/III clinical trial in sIBM.

Material agreements entered into outside the ordinary course

Save as disclosed above, there are no contracts (other than entered into in the ordinary course of business) to which Orphazyme is a party which (i) are, or may be, material to Orphazyme and which have been entered into in the two years immediately preceding the date of this Offering Circular; or (ii) contain any obligations or entitlements which are, or may be, material to Orphazyme as of the date of this Offering Circular.

Legal Proceedings and Investigations

From time to time, Orphazyme may be involved in litigation matters arising in the ordinary course of business. Orphazyme does not believe that any of the liabilities arising from the outcome of such matters, individually or in the aggregate, will have a significant effect on its financial position or profitability. Orphazyme has not within the last twelve months from the date of this Offering Circular been, and is not currently, party to any governmental, litigation, administrative, arbitration, or dispute proceedings, that could have, or have had in the recent past, a material adverse effect on Orphazyme's business, results of operations or financial condition. Orphazyme is not aware of any threatened or potential dispute or governmental proceeding that could have a material adverse effect on Orphazyme's business, results of operations or financial condition in the future.

Research and Development Policy

Orphazyme's core technology stimulates the body's own production of HSPs. The Company employs this technology to develop a potential treatment for four orphan diseases; two neuromuscular diseases. The related research and development cost for Orphazyme during 2015, 2016 and H1 2017 amounted to DKK 45.9 million, DKK 55.8 million and DKK 46.9 million, respectively.

Property

Orphazyme leases office and laboratory space from COBIS A/S on Ole Maaløes Vej 3, 2200 Copenhagen N, with a current total gross area of around 720 square metres. The lease agreement expires automatically on 31 January 2023. Notice of termination is six months for the Company. COBIS A/S is entitled to relocate the Company to another leasehold in the same building with the same size and standard and on the same terms, against payment of reasonable costs associated with the relocation.

Selected Historical Financial and Operating Information

The selected financial information set forth below have been extracted from the Company's financial statements for the indicated periods as included in the F-pages of this Offering Circular.

The Company prepared its statutory Audited Financial Statements for the period 1 January – 31 December 2016 (12 months) with comparative figures for the period 1 July 2015 – 31 December 2015 (6-month conversion period) and the period 1 January 2014 – 30 June 2015 (12 months) in accordance with IFRS, issued by the International Accounting Standards Board (IASB) and adopted by the European Union and additional requirements in the Danish Financial Statements Act. As the Company's previously published statutory financial statements reflect uneven financial periods, due to the financial period being converted from 1 July – 30 June to 1 January – 31 December in 2015, the Company has in addition to the indicated published financial statements prepared a set of reviewed unaudited 2015 Comparative Financial Statements for the period 1 January – 31 December 2015, which have been prepared based on the Company's accounting policies for recognition and measurement and reviewed by the Company's auditors, as included on pages F-37 - F-46. Such financial statements for the period 1 January – 31 December 2015 together with the Audited Financial Statements for the period 1 January – 31 December 2016 form the basis for the operational and financial review below.

Further, the Company has prepared reviewed unaudited condensed Interim Financial Statements for the period 1 January – 30 June 2017 with comparative figures for the period 1 January – 30 June 2016 prepared in accordance with the International Accounting Standard 34 "Interim Financial Reporting" (IAS 34) as adopted by the European Union and additional requirements in the Danish Financial Statements Act, as included on pages F-3 - F-13, and which in respect of the period 1 January – 30 June 2017 have been reviewed by the Company's auditors, EY, whereas the comparative figures for the period 1 January – 30 June 2016 has not been subject to any review.

Investors should read the selected historical financials set forth below together with the Audited Financial Statements and Interim Financial Statements, including the notes thereto, and the sections "*Presentation of Financial and Certain Other Information*" and "*Operating and Financial Review*".

Statement of profit or loss and other comprehensive income

	Six months period ended 30 June		Twelve months ended 31 December	
(TDKK)	**H1 2017**	**H1 2016**	**2016**	**2015**
Research and development expenses	(46,870)	(27,875)	(55,817)	(45,865)
Administrative expenses	(7,972)	(2,696)	(7,703)	(7,220)
Operating loss	**(54,842)**	**(30,571)**	**(63,520)**	**(53,085)**
Net financials	(122)	35	85	(317)
Loss before tax	**(54,964)**	**(30,536)**	**(63,435)**	**(53,402)**
Income tax benefit	2,841	2,750	5,500	5,688
Net loss for the period	**(52,123)**	**(27,786)**	**(57,935)**	**(47,714)**
Other comprehensive income/(loss	-	-	-	-
Total comprehensive loss	**(52,123)**	**(27,786)**	**(57,935)**	**(47,714)**
Loss per share, basic and diluted				
Class C preferred shares			(16.71)	(14.56)
Class B preferred shares			(17.12)	(15.05)
Class A ordinary shares	5.09	2.83	(26.08)	(23.19)

Statement of financial position

(TDKK)	As of 30 June 2017	As of 31 December 2016	As of 31 December 2015
Assets			
Non-current assets			
Property, plant and equipment	1,007	987	1,487
Corporation tax receivable	5,591	2,750	2,750
Deposits	357	310	211
Total non-current assets	**6,955**	**4,047**	**4,448**
Current assets			
Corporation tax receivable	5,500	5,500	5,875
Receivable capital increase	91,319	-	-
Other receivables	5,104	3,421	644
Prepayments	3,245	4,624	5,970
Cash and cash equivalents	33,589	14,349	68,015
Total current assets	**138,757**	**27,894**	**80,504**
Total assets	**145,712**	**31,941**	**84,952**
Equity and liabilities			
Equity			
Share capital	5,103	3,361	3,346
Share premium	380,454	226,285	224,999
Accumulated deficit	(264,260)	(212,137)	(154,202)
Total equity	**121,297**	**17,509**	**74,143**
Current liabilities			
Trade payables	7,614	4,718	2,447
Payables to shareholders	223	-	-
Other payables	16,578	9,714	8,362
Total current liabilities	**24,415**	**14,432**	**10,809**
Total equity and liabilities	**145,712**	**31,941**	**84,952**

Statement of cash flows

	Six months period ended 30 June		Twelve months ended 31 December	
(TDKK)	**H1 2017**	**H1 2016**	**2016**	**2015**
Operating activities				
Net loss before tax	(54,842)	(30,571)	(63,435)	(53,402)
Adjustments to reconcile loss before tax to cash flows from operating activities				
Share-based expense	-	-	-	-
Depreciation and write-down	291	287	706	554
Gain/loss on sale and disposal of assets	-	-	33	-
Change in other receivables	(1,730)	(1,032)	(2,876)	(277)
Change in prepayments	1,379	1,296	1,347	(4,969)
Change in trade payables	2,896	1,134	2,271	858
Change in other payables	6,821	2,284	1,352	4,330
Cash flows from taxes	-	-	5,875	6,250
Interest paid, net	(122)	35	-	241
Net cash used in operating activities	**(45,307)**	**(26,567)**	**(54,727)**	**(46,414)**
Investing activities				
Investment in property, plant and equipment	(311)	-	(238)	(495)
Net cash used in investing activities	**(311)**	**-**	**(238)**	**(495)**
Financing activities				
Capital contributions from shareholders	65,431	-	1,330	85,970
Cash from convertible loan	-	-	-	-
Bank loans	-	463	-	-
Expenses related to capital contributions	(573)	-	(30)	(215)
Net cash provided by financing activities	**64,858**	**463**	**1,300**	**85,755**
Net change in cash and cash equivalents	**19,240**	**(26,104)**	**(53,665)**	**38,846**
Net foreign exchange differences	-	-	-	-
Cash and cash equivalents at the beginning of the period	14,349	68,014	68,014	29,169
Cash and cash equivalents at the end of the period	**33,589**	**41,910**	**14,349**	**68,015**

Operating and Financial Review

The following is a discussion of Orphazyme's financial condition and results of operations and cash flows as at and for the six months period ended 30 June 2017 and 2016 and as at and for the years ended 31 December 2016 and 2015. This discussion should be read in conjunction with the selected historical financial information included under "Selected Historical Financial and Operating Information", the Audited Financial Statements and Interim Financial Statements and related notes included under "Financial Information". For information on the basis of preparation of the financial statements, see "Presentation of Financial and Certain Other Information".

Some of the information contained in the following discussion contains forward-looking statements that are based on assumptions and estimates and are subject to risks and uncertainties. Investors should read the section entitled "Special Notice Regarding Forward-Looking Statements" for a discussion of the risks and uncertainties related to those statements. Investors should also read the section entitled "Risk Factors" for a discussion of certain factors that may affect Orphazyme's business, results of operations, financial condition and prospects.

Overview

Orphazyme is a Danish biotech company developing medicines for the treatment of orphan protein misfolding diseases. The Company targets diseases with a high unmet need and well-defined patient populations with known disease pathology and/or genetics. Orphazyme's expertise in protein misfolding diseases has resulted in a pipeline of new drug candidates.

Orphazyme uses its lead candidate, arimoclomol, to develop new drug candidates within neuromuscular and lysosomal storage diseases.

The Company has a late stage orphan drug project pipeline with two completed phase II programs in arimoclomol for sIBM and ALS and pre-clinical data supporting the advancement of arimoclomol in NPC and Gaucher disease. Following successful trial outcomes, Orphazyme has initiated a phase II/III program in NPC, phase II/III program in sIBM and expects to initiate a phase II program for Gaucher in Q2 2018 as well as a phase II/III trial for ALS in H2 2018.

Since its inception, Orphazyme has financed its operations through capital increases as well as funding for research from governmental grants. Most of Orphazyme's expenditures to date have been incurred to discover and develop its pipeline of drug candidates, and to seek or obtain patents for its intellectual property.

Being a biotech company with a developed pipeline but no products ready for marketing and distribution, Orphazyme has never generated any revenue and has an accumulated deficit of DKK 264.3 million.

Principal factors affecting Orphazyme's results of current operations

Research and development expenses

Orphazyme focuses on development of novel treatments addressing medical needs of patients with rare diseases. As such, the successful development of new programs will greatly affect the Company's long-term performance and ability to deliver shareholder returns.

Research and development expenses include salaries including share-based compensation and costs arising from research activities, clinical development, legal expenses related to the protection, defense and enforcement of the Company's intellectual property and rent associated with facilities used for research purposes.

The Company's research and development costs vary from period to period depending on the phase of development of its product candidates. For the periods presented, research and development costs were mainly affected by the pre-clinical studies and clinical trials performed by the Company.

Governmental grants received regarding research activities are deducted from research expenses. Orphazyme has received government grants from the Innovation Fund Denmark and from the Danish Ministry of Science for employment of Ph.D. students. Government grants are recognised at the time when a final and firm right to the grant has been obtained and to the extent that the entity has obtained reasonable assurance to comply with the conditions attaching to them and the grants will be received. Grants related to expenses incurred are set off against the related expenses for which the grants are intended to compensate. Government grants based on cost reimbursement are recognised under research and development expenses.

Administrative expenses
Administrative expenses include salaries for administrative staff and management, costs of share-based payments and rent associated with facilities not used for research purposes and fluctuate mainly based on changes in the number of employees.

Licensing and cooperation agreements
Orphazyme is due to pay milestone payments and royalty payments relating to the development of arimoclomol. These payments relate to Orphazyme's acquisition of a portfolio of molecules, including arimoclomol, from CytRx. Due to the nature of the agreements, the exact timing of the potential milestone and royalty payments cannot be precisely estimated. For more information, refer to "*Business—Arimoclomol—Acquisition from CytRx and the key terms*".

Other initiatives affecting results of operations
Orphazyme operates in a highly regulated industry and is, as are other pharmaceutical and biotech companies, generally affected by governmental, economic, fiscal, monetary and political policies. Historically, such policies have not materially affected Orphazyme's results of operations. For risks relating to changes in the regulatory environment, see "*Risk Factors*".

Accounting policies
A full description of the Company's accounting policies is provided in the Audited Financial Statements for 2016, as included in this Offering Circular on pages F-21 – F-24.

In 2015, the Company changed its financial year to follow the calendar year, having previously used a financial year spanning 1 July to 30 June.

Critical accounting estimates and judgments
In preparing financial statements under IFRS, certain rules and standards require Executive Management's judgments, estimates and assumptions. Such judgments, estimates and assumptions are considered important in order to understand the accounting policies and Orphazyme's compliance with the standards. The following summarises the areas involving a higher degree of judgment or complexity, or areas where assumptions and estimates are significant to the financial statements.

Accrued expenses
Orphazyme expenses research and development costs as incurred. Research and development expenses include, among other things, clinical trial costs.

Accounting for clinical trials relating to activities performed by clinical research organizations (CROs) and other external vendors requires management to exercise significant estimates in regards to the timing and accounting for these costs. The diverse nature of services being provided by CROs and other arrangements, the different compensation arrangements that exist for each type of service and the limitations in respect of information related to certain clinical activities add complexity to the estimation of accruals for services rendered by CROs and other vendors in connection with clinical trials. Furthermore, certain CROs and vendors are paid upfront in connection with clinical activities. In estimating the relevant periods etc., the Company evaluates the start-up, treatment and wrap-up periods, compensation arrangements and services rendered attributable to each clinical trial. Estimated costs are regularly tested against payment plans and trial completion assumptions. Accrued expenses are recognised as other payables under current liabilities.

The Company has recognised accruals related to clinical trial development costs presented under trade payables of DKK 7.7 million, DKK 1.4 million and DKK 4.1 million for the six months period ended 30 June 2017, twelve months period ended 31 December 2016 and the twelve months period ended 31 December 2015, respectively.

The Company has recognised prepaid costs related to clinical trial development costs of DKK 1.5 million, DKK 4.5 million and DKK 5.8 million for the six months period ended 30 June 2017, twelve months period ended 31 December 2016 and the twelve months period ended 31 December 2015, respectively.

Deferred tax
The Company recognises deferred income tax assets if it is deemed probable that sufficient taxable income will be available in the future against which the temporary differences and unused tax losses can be utilised. Management has considered future taxable income in assessing whether deferred income tax assets should be recognised and has concluded that the deferred income tax assets related to taxable losses to be carried forward do not meet the criteria for being recognised as assets in the statement of financial position.

The Company has net tax loss carry-forwards that are not recognized of DKK 135 million and DKK 97 million as of 31 December 2016 and 2015, respectively. The Company has net tax loss carry-forwards that are not recognized of DKK 187 million and DKK 125 million as of 30 June 2017 and 2016, respectively.

Income tax benefit includes the current benefit due from the current period's taxable loss and deferred tax adjustments. The benefit is comprised primarily of refundable tax credits for costs incurred in connection with research and development activities under the Danish Tax Credit Regime.

The tax loss carry forwards have no expiry date. The Company's ability to use tax loss carry forwards in any one year is limited to 100% of the first DKK7.5 million of taxable income plus 60% of taxable income above DKK 7.5 million.

Six months period ended 30 June 2017 and 30 June 2016

Results of operations
Research and development costs totaled DKK 46.9 million in the six months period ended 30 June 2017 compared to DKK 27.9 million in the six months period ended 30 June 2016. The increase was mainly due to initiation of the NPC Phase II/III trial in mid-2016 and the costs for the preparation of a phase II trial in India for Gaucher disease. Furthermore, the period is impacted by cost of manufacturing arimoclomol for the IBM phase III trial.

Administrative expenses were DKK 8.0 million in the six months period ended 30 June 2017 compared to DKK 2.7 million in the six months period ended 30 June 2016. The increase is mainly due to costs for lawyers, auditors and consultants for preparing the Initial Public Offering (IPO), as well as the hiring of new members of management and additional staff. Part of the costs directly associated with the IPO and the expected issuance of new Shares is recognised within prepayments until the Offering takes place. As at 30 June 2017, the Company has in total recognised prepayments related to the IPO of DKK 1.4 million.

Net financials totaled an expense of DKK 0.1 million in the six months period ended 30 June 2017 compared to an income of DKK 0.0 million in the six months period ended 30 June 2016. Financial income primarily reflects exchange rate gains. Financial expenses primarily cover interest expenses on bank accounts and bank fees.

Income tax benefit totaled DKK 2.8 million in the six months period ended 30 June 2017 compared to DKK 2.8 million in the six months period ended 30 June 2016. Income tax benefits for the two periods include a tax credit for research and development expenses at the applicable tax rate under the Danish Corporate Income Tax Act.

Liquidity and capital resources
As of 30 June 2017, Orphazyme had cash and cash equivalents of DKK 33.6 million compared with cash and cash equivalents of DKK 14.3 million as of 31 December 2016. The increase reflects the capital increase of DKK 65.4 million in the six months period ended 30 June 2017 and is partly offset by the costs associated with Orphazyme's business activities, including costs of current and planned clinical trials. In the six months period ended 30 June 2017, operating activities has net cash outflow of DKK 45.3 million, compared to DKK 26.6 million in the six months period ended 30 June 2016. The Company has increased cash flow from financing activities through capital increases from both current and new investors. For additional information regarding liquidity and capital resources, see section "*Description of the shares and share capital*". In the six months period ended 30 June 2017, the Company carried out a capital increase by issuing 534,007 C class shares to existing shareholders with net proceeds of DKK 48.1 million. On 8 March 2017, the Company completed a DKK 108.7 million financing round by issuing new C class shares to LSP and ALS. Of the total financing round, DKK 17.4 million have been received by the Company in the six month period ended 30 June 2017 and the investors have subscribed capital for remaining DKK 91.3 million. As at 30 June 2017, the Company has therefore recognized a receivable from capital increase of DKK 91.3 million. The receivable has been received after the balance sheet date.

Cash flows
Net cash flow from operating activities amounted to an outflow of DKK 45.3 million in the six months period ended 30 June 2017 compared to DKK 26.6 million in the six months period ended 30 June 2016. Net cash flow from operating activities is attributable primarily to the initiation and progression of clinical development activities, as well as administrative expenses.

Net cash outflow from investing activities amounted to an outflow of DKK 0.3 million in the six months period ended 30 June 2017 compared to DKK 0.0 million in the six months period ended 30 June 2016. Investing activities comprise investment in equipment for research and development purposes as well as refurbishment of the new facility. The increase is due to a higher activity level as well as the Company has moved to new facilities in the six months period ended 30 June 2017. Net cash flow from financing activities amounted to

an inflow DKK 64.9 million in the six months period ended 30 June 2017 compared to DKK 0.5 million in the six months period ended 30 June 2016. The increase is due to capital increases from both existing and new investors in the six months period ended 30 June 2016.

Capital expenditure
Capital expenditure amounted to DKK 0.3 million in the six months period ended 30 June 2017 compared to DKK 0.0 million in the six months period ended 30 June 2016. The increase was primarily due to investments in equipment for research and development purposes as well as refurbishment of the new facility. All investments in equipment have been financed by the Company using cash.

Years ended 31 December 2016 and 2015

Results of operations
Research and development costs totaled DKK 55.8 million in 2016 compared to DKK 45.9 million in 2015. The increase was mainly due to the initiation of the NPC phase II/III trial in mid-2016 and cost of new employees to support the increase in the level of activity.

Administrative expenses were DKK 7.7 million in 2016 compared to DKK 7.2 million in 2015. The increase from 2015 to 2016 was due to the hiring of new members of management at the end of 2016.

Net financials total an income of DKK 0.1 million in 2016 compared to an expense of DKK 0.3 million. Financial income primarily reflects exchange rate gains. Financial expenses primarily covers interest expenses on bank accounts and bank fees. Furthermore, in 2015 financial expenses also covers interest on convertible debt, which was converted to equity in 2015.

Income tax benefit totaled DKK 5.5 million in 2016 compared to DKK 5.7 million in 2015. Income tax benefits for the two years include a tax credit for research and development expenses at the applicable tax rate under the Danish Corporate Income Tax Act.

Liquidity and capital resources
Since its inception, Orphazyme has financed its operations through capital increases as well as funding for research from governmental grants.

As of 31 December 2016, Orphazyme had cash and cash equivalents of DKK 14.3 million compared with cash and cash equivalents of DKK 68.0 million as of 31 December 2015. The decrease reflects the costs associated with Orphazyme's business activities, including costs of current and planned clinical trials.

Cash flows
Net cash flow from operating activities amounted to an outflow of DKK 54.7 million in 2016 compared to DKK 46.4 million in 2015. Net cash flow from operating activities is attributable primarily to the initiation and progression of clinical development activities, as well as administrative expenses.

Net cash outflow from investing activities amounted to DKK 0.2 million in 2016 compared to DKK 0.5 million in 2015. Investing activities primarily comprise investments in leasehold improvements. The decrease was due to limited investments in leasehold improvements before the planned move to a new office location.

Net cash flow from financing activities amounted to an inflow of DKK 1.3 million in 2016 compared to DKK 85.8 million in 2015. Net cash from financing activities was primarily attributable to net proceeds in connection with the issue of Shares. In 2015, Orphazyme completed a funding round in two tranches with current and new investors of which the first tranche was paid in 2015 resulting in net proceeds to the Company of DKK 85.8 million. In 2016, as part of a research collaboration two universities invested DKK 1.3 million in the Company.

Capital expenditure
Capital expenditure amounted to DKK 0.2 million in 2016 compared to DKK 0.5 million in 2015. The decrease was due to limited investments in leasehold improvements before the planned move to a new office location. All investments in equipment have been financed by the Company using cash.

The Company has no material current investments and has made no commitment to material future investments.

Contractual obligations

The following table summarises Orphazyme's contractual obligations and commercial commitments as of 30 June 2017.

Contractual obligations (TDKK)

	Payment due by period				
	Less than 1 year	**1-3 years**	**3-5 years**	**More than 5 years**	**Total**
Operating lease obligations	659	-	-	-	659
Other contractual obligations	46,645	11,279	602	-	58,526
Total	47,304	11,279	602	-	59,185

Other contractual obligations primarily include committed costs relating to agreements with CROs used for pre-clinical studies, stability studies and clinical trials as well as funding of Ph.D.-students with collaboration partners.

Pensions

Orphazyme has a defined contribution pension scheme for its employees.

Financial and market risk

Foreign currency exchange

The Company maintains operations in Denmark and uses DKK as its functional currency. The Company conducts cross border transactions where the functional currency is not always used. Accordingly, future changes in the exchange rates of DKK, EUR, USD and/or GBP will expose the Company to currency gains or losses that will impact the reported amounts of assets, liabilities, income and expenses and the impact could be material.

Interest rates

Orphazyme generally maintain its cash in a cash account in order to preserve capital and liquidity funding. Orphazyme had no floating rate borrowings as of 30 June 2017. Orphazyme's only direct exposure to interest rate fluctuations is to the interest rates paid or charged on its cash balances.

Current trading

As of the date hereof, there have been no significant changes in the business or financial condition of the Company since 30 June 2017, other than changes resulting from the ordinary course of business; the payment of DKK 91.3 million receivable from previous capital increases; and the 2017 Capital Structure Adjustment (as described in "*Ownership Structure and Main Shareholders*").

The phase II/III program in sIBM was initiated in August. During October 2017, Orphazyme entered into a license agreement with the University of Kansas and UCL Business PLC. Under the terms of the license agreement, Orphazyme is entitled to take over the IND and trial sponsorship from the University of Kansas. The sponsorship will require Orphazyme to fund the trial until completion. For further details on the costs related to the sponsorship, please see "*Background to the Offering and Use of Proceeds*" and for a further description of the license agreement, please see *"Business—Material Contracts—License Agreement with University of Kansas and UCL*".

Off balance sheet items

As part of the Asset Purchase Agreement with CytRx, Orphazyme may be required to make royalty payments based on potential future sale and milestone payments based on development milestones. No minimum unconditional royalties have been committed. Orphazyme has no royalty liabilities prior to the occurrence of a potential future sale. Accordingly, no such liabilities have been recognized. Reference is made to "*Business—Material Contracts—Asset Purchase Agreement with CytRx* ".

Prospective Financial Information for the Financial Year ending 31 December 2017

Statement by the Board of Directors and Executive Management

The Company has prepared and presented the prospective financial information for the financial year ending 31 December 2017, including the principal assumptions stated under "*—Methodology and Assumptions*". The accounting policies applied are in accordance with the accounting policies set out in the notes to the Company's financial statements for the financial year 2016 included in this Offering Circular. The prospective financial information for the financial year ending 31 December 2017 is prepared for the purpose of this Offering Circular.

The prospective financial information for the financial year ending 31 December 2017 is based on a number of factors, including certain estimates and assumptions. The principal assumptions upon which the Company has based the prospective financial information for the financial year ending 31 December 2017 are described under "*—Methodology and Assumptions*". The prospective financial information for the financial year ending 2017 is based on a number of assumptions, and many of the significant assumptions the Company has used in preparing this information are outside of the Company's control or influence.

The prospective financial information for the financial year ending 2017 represents the best estimates of the Board of Directors and Executive Management at the date of publication of this Offering Circular. Actual results are likely to be different from the prospective financial information for the financial year ending 31 December 2017, since anticipated events may not occur as expected and the variation may be material. You should read the financial information for the financial year ending 31 December 2017 in this section in conjunction with "*Risk Factors*" included elsewhere in this Offering Circular. See also "*Special Notice Regarding Forward-Looking Statements*".

Copenhagen, 6 November 2017

Orphazyme A/S

Board of Directors

Georges Gemayel
Chairman

Bo Jesper Hansen
Deputy Chairman

Martijn Kleijwegt
Board Member

Martin Bonde
Board Member

Martin Rahbek Kornum
Board Member

Nanna Lüneborg
Board Member

Patrick J.H. Krol
Board Member

Rémi Droller
Board Member

Sten Verland
Board Member

Executive Management

Anders Mørkeberg Hinsby
CEO

Anders Vadsholt
CFO

Report from Orphazyme's Independent Auditors regarding the Prospective Financial Information for the Financial Year ending 31 December 2017

To shareholders and potential shareholders

We have evaluated whether the prospective financial information for 2017 of Orphazyme A/S (the "Company"), in all material respects, has been properly compiled on the basis stated and whether the basis of accounting used for the prospective financial information is consistent with the accounting policies of Orphazyme A/S.

The prospective financial information for 2017 is stated on pages 113 to 114 of this Offering Circular. The basis is stated in the section "*Methodology and Assumptions*".

We will express reasonable assurance in our conclusion.

The purpose of the prospective financial information is to reflect the expected financial effect of the planned actions by management of the Company for 2017.

The Company's actual results of operations for 2017 are likely to deviate from the prospective financial information for 2017, since anticipated events frequently do not occur as expected. Such deviations may be material.

The prospective financial information with appertaining statement has been prepared for the purpose of this Offering Circular, which is prepared in accordance with Commission Regulation (EC) No 809/2004, as subsequently amended, and may therefore not be used for another purpose. Our report is issued in accordance with Commission Regulation (EC) No 809/2004, as subsequently amended, and has been prepared in accordance with generally accepted Danish practice for such reports and only in connection with the contemplated public offering of new Shares in the Company and admission for trading and official listing on Nasdaq Copenhagen of the Shares in the Company.

Management's Responsibility

The Company's management is responsible for the proper compilation of the prospective financial information on the basis stated and for the basis of accounting used for the prospective financial information being consistent with the accounting policies of Orphazyme A/S and for such internal control as the Company's management determines is necessary to enable the preparation of prospective financial information on the basis stated.

Furthermore, the Company's management is responsible for the assumptions underlying the prospective financial information.

Auditors' Responsibility

Our responsibility is, in accordance with the Commission Regulation (EC) No 809/2004, as subsequently amended, to express a conclusion as to whether the prospective financial information has been properly compiled on the basis stated and whether the basis of accounting used for the prospective financial information is consistent with the accounting policies of Orphazyme A/S.

We have performed our work in accordance with ISAE 3000 (revised), Assurance Engagements Other than Audits or Reviews of Historical Financial Information, and additional requirements under Danish audit regulation.

We are subject to the International Standard on Quality Control, ISQC 1, and thus apply a comprehensive quality control system, including documented policies and procedures concerning compliance with ethical requirements, professional standards and current statutory requirements and other regulation.

We complied with independence requirements and other ethical standards under FSR - Danish Auditors' Code of Ethics for Professional Accountants, which rely on general principles regarding integrity, objectivity, professional competence and due care, confidentiality and professional conduct.

As part of our work, we have examined whether the prospective financial information has been properly compiled on the basis of the assumptions stated and according to the accounting policies stated in the audited financial statements for 2016 of the Company as included in the F-pages F-21 to F-24, including examination of the numerical consistency of the prospective financial information for 2017.

Our work did not comprise an assessment of whether the assumptions applied are documented, well-founded, realistic and complete or whether the prospective financial information for 2017 can be realised, and therefore we express no conclusion thereon.

Conclusion
In our opinion, the prospective financial information for 2017 has, in all material respect, been properly compiled on the basis stated and the basis of accounting used for the prospective financial information is consistent with the accounting policies of Orphazyme A/S.

6 November 2017, Copenhagen

ERNST & YOUNG
Godkendt Revisionspartnerselskab
CVR no. 30 70 02 28

Christian Schwenn Johansen
State Authorised Public Accountant

Lars Hansen
State Authorised Public Accountant

Introduction

The Company has prepared the prospective financial information for the year ending 31 December 2017 for use in this Offering Circular in accordance with applicable laws and regulations. Such information is the responsibility of the Board of Directors and Executive Management.

The prospective financial information was not prepared with a view towards compliance with published guidelines of the U.S. Securities and Exchange Commission and the American Institute of Certified Public Accountants (the "**AICPA**"), for preparation and presentation of prospective financial information. Accordingly, this information does not include disclosure of all information required by the AICPA guidelines on prospective financial information. The prospective financial information is necessarily based upon a number of assumptions and estimates that, while presented with numerical specificity and considered reasonable by the Company, are inherently subject to significant business, operational, economic and competitive uncertainties and contingencies, and upon assumptions with respect to future business decisions that are subject to change.

Orphazyme's expectations as to future developments may deviate substantially from actual developments, and Orphazyme's actual results of operations are likely to deviate, and may deviate materially, from the forecast provided. Accordingly, potential investors should treat this information with caution and not place undue reliance on the expectations set forth below.

Methodology and assumptions

The prospective financial information for the financial year ending 31 December 2017 has been prepared in accordance with the accounting policies presented in the Company's Audited Financial Statements for 2016 which have been prepared in accordance with International Financial Reporting Standards (IFRS) as adopted by the European Union.

The prospective financial information for the financial year ending 31 December 2017 is prepared for the purpose of this Offering Circular.

The prospective financial information for 2017 has been based on management's updated budget for 2017 prepared in accordance with Orphazyme's forecasting and budgeting procedures and on a basis comparable to the historical financial information included elsewhere in this Offering Circular.

The prospective financial information for the financial year ending 31 December 2017 is based on a number of factors, including certain estimates and assumptions. The key assumptions concerning the future, and other key sources of estimation uncertainty at the date of the prospective financial information that have a significant risk of causing a material adjustment to the prospective amounts of expenses, assets and liabilities within the period until 31 December 2017, are listed below. The Company based its assumptions and estimates on information available when the prospective financial information was prepared.

Certain assumptions, uncertainties and contingencies relating to the prospective financial information are wholly or partly within the control of Orphazyme, while others are outside or substantially outside the control of the Company.

While Orphazyme has presented the key assumptions on which the prospective financial information is based in the following, it is likely that one or more of the assumptions that Orphazyme has relied upon will not prove to be accurate in whole or in part.

Orphazyme's result of operations could deviate materially from its forecasts as a result of other factors, including but not limited to those described in *"Special Notice Regarding Forward-Looking Statements"* and *"Risk Factors"*. For more information regarding principal factors affecting the Company's results of operations, see *"Operating and Financial Review—Principal Factors Affecting Orphazyme's Results of Current Operations"*.

For the purpose of preparing the prospective financial information for the financial year ending 31 December 2017, Orphazyme has applied the key assumptions below:

Currency

The prospective financial information for the financial year ending 31 December 2017 is presented in the Company's reporting currency DKK. Multiple expected costs are denominated in foreign currencies, especially costs from clinical research organizations (CROs). Accordingly, future changes in the exchange rates of the DKK, the EUR, the USD and/or the GBP will impact the Company's actual expenses and expose the Company to currency gains or losses that will impact the expected amounts of assets and liabilities, income and expenses and the impact could be material. The currency assumptions applied for purposes of the prospective financial information are outside the control of the Company.

For the time being, the Company has decided not to utilise foreign currency forward contracts or other derivative instruments to mitigate cash flow or market value risks associated with foreign-currency-denominated transactions.

Research and development expenses
Research and development expenses include salaries, including share-based compensation and costs arising from research activities, clinical development, legal expenses related to the protection, defense and enforcement of the Company's intellectual property and rent associated with facilities used for research purposes.

The Company's research and development expenses vary from period to period depending on the phase of development of its product candidates. For the financial year ending 31 December 2017, research and development expenses were mainly affected by the pre-clinical studies and clinical trials performed by the Company.

For the financial year ending 31 December 2017, the Company expects to incur substantial costs associated with clinical trials. The objective of the development programs is to develop a pharmaceutical drug for treatment of the following diseases: sIBM, ALS, NPC and Gaucher disease.

Budgeting for clinical trials relating to activities performed by CROs and other external vendors requires management to exercise significant estimates in regard to the timing and accounting for these costs. The diverse nature of services being provided by CROs and other arrangements, the different compensation arrangements that exists for each type of service and the limitations in respect of information related to certain clinical activities add complexity to the estimation of accruals for services rendered by CROs and other vendors in connection with clinical trials. Furthermore, certain CROs and vendors are paid upfront in connection with clinical activities. In the estimation for the financial year ending 31 December 2017, the Company evaluates the start-up, treatment and wrap-up periods, compensation arrangements and services rendered attributable to each clinical trial. The majority of the assumptions associated with estimating research and development costs are considered substantially outside the Company's control. The prospective financial information for the financial year ending 31 December 2017 takes into consideration the trial designs for the respective product candidates as described in "*Business*" as to the activities planned for 2017.

The prospective financial information reflects the initiation of the NPC phase II/III in mid-2016, costs for preparations of a phase II trial in India for Gaucher disease and costs related to manufacturing of arimoclomol for the IBM phase II/III trial. Furthermore, management expects increasing staff due to the expected increasing level of activity. Income from public grants has been set off as a reduction of research and developments costs.

Administrative expenses
Administrative expenses include salaries for administrative staff and management, costs of share-based payments and rent associated with facilities not used for research purposes. The assumptions associated with estimating administrative expenses are substantially within the Company's control.

Orphazyme's administrative expenses are expected to increase as the business expands and the Company is expected to incur additional costs associated with being a publicly listed company. This will include costs related to retaining personnel to strengthen the finance department and develop the internal controls and financial reporting processes, as well as engaging investor relations companies. The impact of the Company becoming a publicly listed company also includes increased costs related to new personnel that needs to be retained in connection with both administrative and operational activities, legal compliance fees, accounting and audit fees, D&O insurance premiums, and costs related to general investor relations.

Income tax benefit
The prospective financial information for the financial year ending 31 December 2017 contains estimated income tax benefit and includes tax credit for research and development expenses at the applicable tax rate under the Danish Corporate Income Tax Act. No capitalization of deferred tax assets is included.

Expectations for the net result for the financial year ending 31 December 2017

Based on the assumptions above, the Company expects a net loss for 2017 of DKK 125 million - DKK 135 million. Orphazyme has no revenue and costs are primarily related to research and development. Income tax benefit and income from public grants have a positive effect on the net result for the year.

Orphazyme's result of operations for 2017 could deviate materially from this forecast as a result of other factors, including, but not limited to, those described in *"Special Notice Regarding Forward-Looking Statements"* and *"Risk Factors"*.

Board of Directors, Executive Management and Key Employees

Overview

The Company has a two-tier governance structure consisting of the Board of Directors and the Executive Management. The two bodies are separate and have no overlapping members. The Executive Management is supported by Orphazyme's key employees (the "**Key Employees**"). The business address of the Board of Directors, Executive Management and the Key Employees is Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark.

Board of Directors

The Board of Directors is responsible for the overall and strategic management and proper organisation of the Company's business and operations and it supervises the Company's activities, management and organisation. The Board of Directors appoints and dismisses the members of the Executive Management, who are responsible for the day-to-day management of the Company.

In accordance with article 9.1 of the Articles of Association, the general meeting of the Company shall elect not less than six and not more than nine members to the Board of Directors. The Board of Directors elects a chairman (the "**Chairman**") and, if so decided, a deputy chairman ("**Deputy Chairman**") of the Board of Directors among its members. See article 9.3 of the Articles of Association.

The members of the Board of Directors elected by the general meeting are elected for a term of one year. Members of the Board of Directors may be re-elected.

The existing Board of Directors is currently comprised of nine members elected by the general meeting comprising the Chairman and eight board members (the "**Existing Board of Directors**"). An extraordinary general meeting is expected to be held on 17 November 2017 no later than at 8:00 a.m. (CET) after expiry of the Offer Period but before Admission. The Company's shareholders have confirmed that if the vote is held as expected, they will vote in favour of electing Anders Hedegaard and Catherine Moukheibir as a new independent members of the Board of Directors (the "**New Board Members**") and Martin Rahbek Kornum, Nanna Lüneborg and Patrick J.H. Krol will resign from the Board of Directors, thus constituting the Company's Board of Directors as of Admission (the "**New Board of Directors**"). If the Offering is closed before 16 November 2017, the day of the extraordinary general meeting may be moved forward accordingly. The result of the extraordinary general meeting regarding election of the New Board Members and the composition of the New Board of Directors will be published through Nasdaq Copenhagen and made available on the Company's website. Information on the Company's website does not form part of, and is not incorporated by reference into, this Offering Circular.

The Company believes that the members of the New Board of Directors possess the professional skills and experience required to serve as board members of the Company and to supervise and manage a company with shares admitted to trading and official listing on Nasdaq Copenhagen.

The following table presents an overview of the Existing Board of Directors and the New Board of Directors:

Name	Position	Independent[1]	Year of first appointment	Expiration of term
Existing Board of Directors				
Georges Gemayel	Chairman	Independent	2012	2018
Bo Jesper Hansen	Deputy Chairman	Independent	2010	2018
Martijn Kleijwegt	Member	Independent	2017	2018
Martin Bonde	Member	Independent	2010	2018
Martin Rahbek Kornum	Member	Independent	2009	2018
Nanna Lüneborg	Member	Independent	2016	2018
Patrick J.H. Krol	Member	Independent	2013	2018
Rémi Droller	Member	Independent	2015	2018
Sten Verland	Member	Independent	2010	2018

New Board of Directors				
Georges Gemayel	Chairman	Independent	2012	2018
Bo Jesper Hansen	Deputy Chairman	Independent	2010	2018
Anders Hedegaard	Member	Independent	2017	2018
Catherine Moukheibir	Member	Independent	2017	2018
Martijn Kleijwegt	Member	Independent	2017	2018
Martin Bonde	Member	Independent	2010	2018
Rémi Droller	Member	Independent	2015	2018
Sten Verland	Member	Independent	2010	2018

(1) The Company has based its assessment of independence on the basis of the criteria set out in the current Corporate Governance Recommendations (as defined below).

All members of the New Board of Directors will be considered independent under the current Corporate Governance Recommendations.

Biographies

Other than as presented below, none of the members of the Board of Directors have been a member of the administrative, management or supervisory bodies of a company or a partnership or been a partner in a partnership outside Orphazyme within the past five years.

Biographies—Existing Board Members

Georges Gemayel (born 1960, American nationality) has been a member of the Board of Directors since November 2012 and Chairman since September 2014. Georges Gemayel is currently chairman of the board of directors of Enterome SA, OxThera AB and Dimension Therapeutics Inc. (publ), as well as a member of the board of directors of Momenta Pharmaceuticals Inc. (publ) and Supernus Pharmaceuticals Inc.(publ). Georges Gemayel is currently a consultant for Noveome Biotherapeutics Inc. and partner in Gemayel Investment LLC as well as a director of the non-governmental organization, St. Andrew's School in Ngong Inc. and a trustee of the Gemayel Family Foundation. In the past five years, Georges Gemayel has previously been chairman of the board of directors of Epitherapeutics ApS, Vascular Magnetics Inc. and Syndexa Pharmaceuticals Inc. as well as a member of the board of directors of NPS Pharmaceuticals Inc. (publ), Raptor Pharmaceuticals Corp. (publ), Prosensa N.V. (publ) and Adolor Corp. (publ), a consultant for Novo Ventures 1 A/S and Fidelity Ventures as well as a director of the non-governmental organization, International Institute of New England. Georges Gemayel holds a master's degree and a PhD degree in Pharmacology from Paris-Sud University and a Docteur d' Exercice en Pharmacie from the St. Joseph University.

Bo Jesper Hansen (born 1958, Danish nationality) has been a member of the Board of Directors since December 2010 and Deputy Chairman since 24 October 2017. Bo Jesper Hansen is currently chairman of the board of directors of Laborie Inc. and Innoventa Medica ApS as well as a member of the board of directors of Azanta A/S, Ablynx NV, Newron Pharmaceuticals SpA and CMC Sweden AB. Bo Jesper Hansen is also an advisory consultant for Wellington Partners Life Science Fund LP, Aescap 2.0, Nordic Capital, EQT AB, Broad Street Principal Investments Europe Ltd. and senior business advisor for HBM Ventures Ltd. In the past five years, Bo Jesper Hansen has previously been chairman of the board of directors and a member of the executive management of Swedish Orphan Biovitrum AB (publ), chairman of the board of directors of Reapplix ApS, Topotarget A/S (publ) (dissolved by merger), Karolinska Development AB (publ) as well as a member of the board of directors of Hyperion Therapeutics Inc. (publ) (dissolved following acquisition), Gambro AB, Inspyr Inc. (publ), Zymenex Holding A/S, Zymenex A/S, ACE Bioscience A/S, Mipsalus Holding ApS and MipSalus ApS. In the past five years, Bo Jesper Hansen has also served as an expert evaluator for FP7-Health-2012 and in 2013 for the European Commission and has been a thesis supervisor for the Faculty of Pharmaceutical Services. Bo Jesper Hansen holds a MD and PhD degree in Medicine from the University of Copenhagen.

Martijn Kleijwegt (born 1955, Dutch nationality) has been a member of the Board of Directors since January 2017. Martijn Kleijwegt is currently founder and manging partner at LSP Management Group BV. and a member of the board of directors of Kiadis Pharma N.V. (publ), OxThera AB, Eloxx Pharmaceuticals Ltd. and Pharvaris BV. In the past five years, Martijn Kleijwegt has previously been a member of the board of directors of Prosensa N.V. (publ). Martijn Kleijwegt holds a master's degree from the University of Amsterdam.

Martin Bonde (born 1963, Danish nationality) has been a member of the Board of Directors since June 2010 when he was chairman of the Board of Directors until September 2014 . Martin Bonde is currently a member of the executive management of Vaccibody AS and Bohrs Towers IVS as well as a member of the board of directors and the executive management of Biotopix ApS. Martin Bonde is also chairman of the board of directors of the trade organization Dansk Biotek. In the past five years, Martin Bonde has previously been a member of the board of directors of Visiopharm A/S and a member of the executive management of Epitherapeutics ApS. Martin Bonde holds a Graduate Diploma in Business Administration (HD i Udenrigshandel) from Copenhagen Business School, a Master of Science and a PhD in Chemical Engineering from the Technical University of Denmark.

Martin Rahbek Kornum (born 1976, Danish nationality) has been a member of the Board of Directors since June 2009. Martin Rahbek Kornum is currently a senior patent counsel at Coloplast A/S (publ) and a member of the executive management of Rahbek Kornum IVS. In the past five years, Martin Rahbek Kornum has been a member of the board of directors of OZ Holding ApS. Martin Rahbek Kornum holds a Master of Science in human biology from the University of Copenhagen, is a registered European Patent Attorney and has passed the U.S. Patent Bar Exam.

Nanna Lüneborg (full name: Nanna Liebach Lüneborg, born 1975, Danish nationality) has been a member of the Board of Directors since April 2016. Nanna Lüneborg is currently a principal at Novo Holdings A/S and a member of the board of directors of Epsilon-3 Bio Ltd, Inventiva Pharma and Glionova AB. In the past five years, Nanna Lüneborg has previously been chairman of the board of directors of Affinicon ApS as well as a member of the board of directors of Inthera Bioscience AG, Obseva SA, Pcovery ApS, MinervaX ApS and IO Biotech ApS and a member of the executive management of Avilex Pharma ApS. Nanna Lüneborg holds a BA Honours in Psychology, Philosophy and Physiology from the University of Oxford, a PhD degree in Neuroscience from University College London and an MBA from the University of Cambridge.

Patrick J.H. Krol (full name: Patrick Johan Hendrik Krol, born 1963, Dutch nationality) has been a member of the Board of Directors since May 2013. Patrick J.H. Krol is currently a managing partner of AESCAP Venture I and Inspirational Visions BV as well as a fund manager of AESCAP 2.0 and is currently a member of the board of directors of Shire International Licensing BV (publ), Cassini BV, Easyscan BV, F-Star Alpha Ltd, F-Star Beta Ltd, F-Star Delta Ltd, F-Star Gamma Ltd, f-star Biotechnologische Forschungs- und Entwickelungsges m.b.H., STAK to-BBB and STAK i-Optics. In the past five years, Patrick J.H. Krol has previously been a member of the board of directors in i-Optics BV, to-BBB BV and Aquapharm Biodiversity Ltd. Patrick J.H. Krol holds a bachelor's degree in physical therapy from Hogeschool Utrecht, a master's degree in Entrepreneurship from Hogeschool Inholland and a master's degree in Executive Management and Consultancy from Lemniscaat Management School.

Rémi Droller (full name: Rémi Pascal Louis Droller, born 1975, French nationality) has been a member of the Board of Directors since January 2015. Rémi Droller is currently managing partner of Kurma Partners SA and a chairman of the board of directors in Dyncaure SAS and ImCheck SAS as well as a member of the board of directors of OxThera AB, AM Pharma BV, STAT Dx S.L. and Pharvaris BV. In the past five years, Rémi Droller has previously been chairman of the board of directors of Step Pharma SAS and a member of the board of directors of Prosensa N.V. (publ) and Onxeo SA (publ). Rémi Droller holds a master's degree in molecular biology from Université Pierre et Marie Curie and a master's degree in finance and management of innovation from Masternova.

Sten Verland (born 1957, Danish nationality) has been a member of the Board of Directors since December 2010. Sten Verland is currently co-founder of Sunstone Capital A/S, senior partner at Sunstone Life Science Ventures A/S and a member of the board of directors of Anergis SA, Vaximm AG, F2G Ltd., MinervaX ApS, OxThera AB, the Danish Venture Capital and Private Equity Association (DVCA) as well as a member of the board of directors in certain companies in or associated with the Sunstone group. Sten Verland is also currently a member of the executive management of Verland Capital ApS, Verland Holding ApS, Verland Holding II ApS and Genobiotix ApS as well as a member of the executive management in certain companies in or associated with the Sunstone group. In the past five years, Sten Verland has previously been chairman of the board of directors of Zymenex Holding A/S and Zymenex A/S as well as a member of the board of directors of Rigontec GmbH, ACE Biosciences A/S, Scanbur A/S, Selskabet af 9. september 2015 A/S, Selskabet af 23. September 2015 ApS, Tera Holding A/S and VetVerland ApS. In the past five years, Sten Verland has previously been a member of the executive management of VetVerland ApS as well as a partner and a member of the executive management in certain companies in or associated with the Sunstone group. Sten Verland holds a master's degree in Biology and Mathematics and a PhD in Immunology, both from the University of Copenhagen.

Biography—New Board Members

Anders Hedegaard (born 1960, Danish nationality). Anders Hedegaard is expected to be elected as a member of the Board of Directors at an extraordinary general meeting expected to be held on 17 November 2017. Anders Hedegaard is currently chief executive officer of GN Store Nord A/S and GN Hearing A/S as well as a member of the board of directors of the Confederation of Danish Enterprise, Hearing Instrument Manufacturers Software Association A/S and HIMSA II A/S. In the past five years, Anders Hedegaard has previously been chairman of the board of directors of GN Otometrics A/S, Aktieselskabet af 1. juni 2011 I and BN Washington D.C. Holding A/S as well as a member of the board of directors in of Origio A/S and certain companies in or associated with the Bavarian Nordic group. Previously, Anders Hedegaard has been chief executive officer of Bavarian Nordic A/S and a member of the executive management of ALK-Abelló A/S and FOSS A/S as well as international marketing director at Novo Nordisk A/S. Anders Hedegaard holds a Master of Science in Chemical Engineering and Biochemistry from the Technical University of Denmark.

Catherine Moukheibir (born 1959, American, Lebanese and British nationality) is expected to be elected as a member of the Board of Directors at an extraordinary general meeting expected to be held on 17 November 2017. Catherine Moukheibir is currently chairman of the board of directors of MedDay Pharmaceuticals SA, member of the board of directors of Zealand Pharma A/S (publ) and Genkyotex SA (publ), as well as a member of the board of directors of Ablynx NV (publ) and Cerenis Therapeutics SA (publ). In the past five years,

Catherine Moukheibir has previously been a member of the executive management and a consultant for Innate Pharma Inc (publ), chairman of the board of directors of Creabilis, a member of the board of directors in Octoplus NV. Catherine Moukheibir is also a member of the advisory board of Imperial College Business School and the international advisory board of the Yale School of Management. Catherine Moukheibir holds a Master in Economics and an MBA degree, both from Yale University.

Board practices and committees

The Board of Directors has resolved that the following board practices and committees shall take effect from the day of Admission.

The Board of Directors normally holds at least five regular meetings annually, including a strategy review, plus ad hoc meetings as required. Extraordinary board meetings are convened by the Chairman when necessary or when requested by a member of the Board of Directors, a member of the Executive Management or by the Company's auditor. The Board of Directors forms a quorum when more than half of its members are represented, including the Chairman or the Deputy Chairman. Resolutions of the Board of Directors are passed by a simple majority of the votes present at the meeting. In the event of equal votes, the Chairman or, in his/her absence, the Deputy Chairman shall have the casting vote. The Board of Directors conducts an annual evaluation of the effectiveness, performance, achievements and competencies of the Board of Directors and of the individual members as well as the collaboration with the Executive Management.

The following board committees have been established by the Board of Directors, each of which has a charter setting forth its purpose and responsibilities. All the committees report and make recommendations to the Board of Directors.

Audit Committee

The Company's audit committee (the "**Audit Committee**") shall review accounting and audit matters that by decision of the Board of Directors or the Audit Committee require a more thorough evaluation, and assess the internal controls and risk management systems of the Company. Its duties also include supervision of the Company's auditors and review of the audit process.

In accordance with the Recommendations on Corporate Governance of the Danish Committee on Corporate Governance issued in May 2013 and updated in November 2014 (the "**Corporate Governance Recommendations**"), the Company has decided that the Chairman of the Board of Directors may not also be the chairman of the Audit Committee and that a majority of the members of the Audit Committee are required to meet the independence requirements set out in the Corporate Governance Recommendations. In addition, at least one member shall have accounting or audit qualifications and between them, the members shall possess such expertise and experience as to provide an updated insight into, and experience in, the financial, accounting and audit aspects of companies with shares admitted to trading and official listing on a regulated market. The Audit Committee shall consist of no less than three members appointed by and among the Board of Directors, including the chairman of the Audit Committee, and is expected to consist of Catherine Moukheibir as chairman, Martijn Kleijwegt and Sten Verland. All of the members of the Audit Committee will meet the independence requirement set out in the Corporate Governance Recommendations. The CFO and the Company's external auditor shall participate in meetings of the Audit Committee if so requested by the Audit Committee and the external auditor shall attend at least one meeting per year or the relevant part hereof where the Executive Management is not present.

Remuneration Committee

The Company's remuneration committee (the "**Remuneration Committee**") shall ensure that the Company maintains a Remuneration Policy for the members of the Board of Directors and the Executive Management which includes the overall guidelines on incentive pay for the Board of Directors and Executive Management in accordance with Section 139 of the Danish Companies Act, and to evaluate and make recommendations for the remuneration of the members of the Board of Directors and the Executive Management.

The Remuneration Committee shall consist of no less than three members appointed by and among the Board of Directors, including Bo Jesper Hansen as chairman, Anders Hedegaard and Rémi Droller. All of the members of the Remuneration Committee will meet the independence requirements set out in the Corporate Governance Recommendations.

Nomination Committee

The Company's nomination committee (the "**Nomination Committee**") shall assist the Board of Directors with ensuring that appropriate plans and processes are in place for nomination of candidates to the Board of Directors, the Executive Management and the board committees. Moreover, the Nomination Committee shall evaluate the composition of the Board of Directors and the Executive Management. This includes making recommendations for nomination or appointment of members of (a) the Board of Directors, (b) the Executive Management and (c) the board committees established by the Board of Directors.

The Nomination Committee shall consist of no less than three members appointed by and among the Board of Directors, including Georges Gemayel as chairman, Martin Bonde and Sten Verland. All of the members of the Nomination Committee will meet the independence requirements set out in the Corporate Governance Recommendations.

Compensation of the Board of Directors

Members of the Board of Directors receive fixed annual fees. Additionally, Orphazyme is open to the practice of share-based remuneration for the members of the Board of Directors as is common among international biotech companies and the Company believes that share-based incentives may be beneficial to the shareholders' long-term interests. The remuneration paid to the Board of Directors will be presented for approval by the Company's shareholders at the annual general meeting. Certain members of the Board of Directors have participated in the Company's pre-IPO warrant programme (for more information, please see "*—Incentive Programmes—Pre-IPO warrant programme*").

In respect of the financial year ending 31 December 2016, the Board of Directors received compensation in the total amount of tDKK 524 for their services. Nanna Lüneborg, Martijn Kleijwegt, Patrick J.H. Krol , Sten Verland and Rémi Droller waived their right to compensation in respect of the financial year 2016.

The Company's extraordinary general meeting has approved a resolution that, subject to completion of the Offering, the members of the Board of Directors for the remainder of the financial year 2017 would receive a prorated share of a fixed annual base fee of EUR 31,500, while the Chairman and Deputy Chairman receive an additional fee of EUR 24,900 and EUR 12,450, respectively. Members of the Audit Committee, the Remuneration Committee and the Nomination Committee will receive a supplementary fee of EUR 6,700, EUR 4,700 and EUR 3,100, respectively, and the chairman of the Audit Committee, the Remuneration Committee and the Nomination Committee will receive a supplementary fee of EUR 13,500, EUR 9,000 and EUR 6,500, respectively. These board fees have been determined based on a benchmark analysis of similar Scandinavian and European biotech companies of comparable size and development stage.

The Company has not granted any loans, issued any guarantees or undertaken any other similar obligations to or on behalf of the Company's Board of Directors or any of its members. No member of the Board of Directors is entitled to any kind of compensation upon resignation as a member of the Board of Directors. The Company has not allocated funds or made provisions for any pension benefits, severance scheme or the like for the Board of Directors and has no obligation to do so.

Executive Management

According to article 10.1 of the Company's Articles of Association, the Company's Board of Directors appoints an Executive Management consisting of one to three members. The primary task of the Executive Management is to carry out the day-to-day management of Orphazyme with the support of the Key Employees.

The following table presents an overview of the current members of the Executive Management:

Name	Position	Year of first employment	Year of appointment to current position
Anders Hinsby	CEO	2009[1]	2010
Anders Vadsholt	CFO	2016	2016

[1] Anders Hinsby initially joined the Company in connection with its foundation in 2009 as a member of the Board of Directors.

The Company believes that both members of the Executive Management possess the professional skills and international experience required for their positions in Orphazyme and to manage a company with shares admitted to trading and official listing on Nasdaq Copenhagen.

Biographies

Other than as presented below, none of the members of the Executive Management have been members of the administrative, management or supervisory bodies of a company or a partnership or a partner in a partnership outside Orphazyme within the past five years.

Anders Hinsby (full name: Anders Mørkeberg Hinsby, born 1973, Danish nationality) has been CEO since he joined Orphazyme in March 2010. Anders Hinsby is currently a member of the executive management of WoB Holding IVS as well as a member of the board of directors of the trade organization Dansk Biotek and a panel member for Innovationsfonden. In the past five years, Anders Hinsby has been a member of the board of directors of OZ Holding ApS. Anders Hinsby holds a Master of Science in Human Biology and a PhD in Medicine from the University of Copenhagen.

Anders Vadsholt (full name: Anders Fink Vadsholt, born 1969, Danish nationality) has been CFO since he joined Orphazyme in May 2016. Anders Vadsholt is currently owner and a member of the executive management of Alpha Healthcare Investments ApS and Lakeside Invest ApS as well as a partner at Obton Solenergi Sinope Komplementaranpartsselskab. In the past five years, Anders Vadsholt has previously been

a member of the executive management of Copenhagen Innovation Capital Management ApS and TopoTarget A/S (publ) (dissolved by merger), Topotarget UK Limited, TopoTarget Gmbh and Topotarget Inc. Anders Vadsholt holds a Bachelor of Science in Corporate Law from the University of Aalborg, an MBA in Finance and Strategy from the University of Melbourne, a Master of Science in Corporate Law and Economics from Copenhagen Business School as well as a Diploma in Basic Pharmaceutical Medicine, Pharmacology and Pathology from LIF.

Compensation of Executive Management

For the financial year ending 31 December 2017, the compensation of the Executive Management consists of a combination of fixed salary, a cash bonus as well as customary benefits in accordance with market standards.

Additionally, the Executive Management have been offered to participate in Orphazyme's share-based long-term incentive programme (for more information, please see "*—Incentive Programmes—Long-term incentive programme (the "LTIP")*". The Executive Management have participated in the Company's pre-IPO warrant programme (for more information, please see "*—Incentive Programmes—Pre-IPO warrant programme*").

The Executive Management may receive a performance-based cash bonus of up to 33% of their respective fixed annual salaries for the financial year 2017.

The Executive Management is at all times required to hold a minimum amount of Shares in the Company equal to their respective annual fixed salaries.

The Executive Management are, subject to certain conditions, entitled to an extraordinary takeover retention bonus amounting to 12 months' fixed salary, provided that they remain employed by the Company and are not under notice on the first anniversary following completion of a public takeover bid for the Shares of the Company resulting in a change of control.

The following table presents an overview of the compensation booked by Orphazyme to the Executive Management in respect of the financial year 2016:

tDKK	Anders Hinsby	Anders Vadsholt[1]
Salaries and other remuneration	1,776	670
Cash bonus	335	50
Share-based payments	—	—

[1] Anders Vadsholt joined the Company as interim CFO in May 2016 and was promoted to CFO in October 2016.

The fixed salaries of the members of the Executive Management are scheduled to increase in order to be competitive and comparable with similar publicly listed biotech companies. As of Admission, the fixed annual salaries of the CEO and CFO will be tDKK 2,126 and tDKK 1,603, respectively. The fixed salaries and cash bonuses of the Executive Management have been determined based on a benchmark analysis of similar Scandinavian and European biotech companies of comparable size and development stage.

Orphazyme has not granted any loans, issued any guarantees or undertaken any other similar obligations to or on behalf of the Executive Management.

The CEO and CFO are, under their respective service contracts, currently entitled to a notice period of 12 months if the employment is terminated by the Company. Subject to certain conditions, the Company may terminate the employment of the CEO and CFO with three months' notice in case of long-term illness. The CEO and the CFO may terminate the employment with six months' notice. The CEO and CFO are not entitled to any agreed severance pay. The CEO and CFO are not covered by a pension scheme. The Company has not allocated funds or made provisions for any pension benefits, severance scheme or the like for the Executive Management and has no obligation to do so.

The CEO and CFO are subject to a non-competition clause and a non-solicitation clause for a period of 12 months after expiry of the notice period. The CEO and the CFO are entitled to compensation for undertaking the clauses corresponding to 100 % and 60% of the fixed salary, respectively.

Key Employees

The Key Employees are employed by Orphazyme with responsibility for their functional areas.

The following table presents an overview of Orphazyme's current Key Employees:

Name	Position	Year of first employment	Year of appointment to current position
Thomas Blaettler	Chief Medical Officer	2016	2016
Thomas Kirkegaard Jensen	Chief Scientific Officer	2009[1]	2010

[1] Thomas Kirkegaard Jensen joined the Company as CEO in 2009 and was made CSO in 2010.

Biographies

Other than as presented below, none of the Key Employees have been members of the administrative, management or supervisory bodies of a company or a partnership or a partner in a partnership outside Orphazyme within the past five years.

Thomas Blaettler (born 1967, Danish and Swiss nationalities) has been CMO, since he joined Orphazyme in November 2016. Thomas Blaettler has previously been PD Neuroscience Group Medical Director at F. Hoffmann-La Roche Ltd. Thomas Blaettler holds a Doctorate in Medicine from the University of Zürich and a Medical School Certificate Swiss State Examinination from the Medical School of the University of Zürich and is a board certified neurologist by the Swiss Medical Association (the Foederation Medicorum Helveticorum).

Thomas Kirkegaard Jensen (born 1977, Danish nationality) has been CSO, since he joined Orphazyme in June 2009. Thomas Kirkegaard Jensen is currently a member of the executive management of Dare to Dream IVS and is an expert reviewer for the European Research Council and is a member of the advisory board for the Rare Disease Report. In the past five years, Thomas Kirkegaard Jensen has been a member of the board of directors and executive management of OZ Holding ApS and vice-chairman of the national Orphan Disease Council. Thomas Kirkegaard holds a Bachelor of Science in Biochemistry, a Master of Science in Human Biology and a PhD in Medicine from the University of Copenhagen.

Compensation of Key Employees

For the financial year ending 31 December 2017, the compensation of the Key Employees consists of a combination of fixed salary, cash bonus as well as customary benefits in accordance with market standards. Additionally, the Key Employees have been offered to participate in Orphazyme's share-based long-term incentive programme (for more information, please see "*—Incentive Programmes—Long-term incentive programme (the "LTIP")*". The Key Employees have participated in the Company's pre-IPO warrant programme (for more information, please see "*—Incentive Programmes—Pre-IPO warrant programme*").

The Key Employees may receive a performance-based cash bonus of up to 33% of their respective fixed annual salaries for the financial year 2017.

The Key Employees are at all times required to hold a minimum amount of Shares in the Company equal to their respective annual fixed salaries.

The Key Employees are, subject to certain conditions, entitled to an extraordinary takeover retention bonus amounting to 12 months' fixed salary, provided that they remain employed by the Company and are not under notice on the first anniversary following completion of a public takeover bid for the Shares of the Company resulting in a change of control.

For the financial year ending 31 December 2016, the Key Employees received compensation, which consisted of a fixed salary and pension contributions in the aggregate amount of tDKK 1,902, a cash bonus in the aggregate amount of tDKK 357 as well as customary benefits in accordance with market standards.

The fixed salaries of the Key Employees are scheduled to increase in order to be competitive and comparable with similar publicly listed biotech companies. As of Admission, the aggregate fixed annual salaries of the CSO and CMO will be a total of tDKK 3,581. The fixed salaries and cash bonuses of the Key Employees have been determined based on a benchmark analysis of similar Scandinavian and European biotech companies of comparable size and development stage.

Orphazyme has not granted any loan, issued any guarantees or undertaken any other similar obligations to or on behalf of the Key Employees.

The Company may currently terminate the employment of its Key Employees with notices of six and eight months, respectively, and the Key Employees may terminate their respective positions with the Company with three to four months' notice. Subject to certain conditions,

the Company may terminate the employment of the Key Employees with one month's notice in case of long-term illness. None of the Key Employees are entitled to agreed severance pay upon termination of employment. However, after having been employed for 12 or 17 years, respectively, the Key Employees are entitled to statutory severance pay of one or three months' salary pursuant to the Danish Consolidated Act no. 81 of 3 February 2009 on salaried employees, as amended. The Company has not allocated funds or made provisions for any pension benefits, severance scheme or the like for the Key Employees and has no obligation to do so.

The CMO is subject to a combined non-competition and non-solicitation clause for a period of 6 months after expiry of the notice period. As compensation for undertaking the clauses, the CMO is entitled to a monthly compensation amounting to 60 % of the salary. Subject to certain conditions, the compensation will be reduced, e.g. if the CMO takes up other suitable employment. The Company may terminate the clauses with one month's notice.

The CSO is subject to a non-competition clause and a non-solicitation clause for a period of 12 months after expiry of the notice period. As compensation for undertaking the clauses, the CSO is entitled to an agreed monthly compensation amounting to 75 % of the salary. Subject to certain conditions, the compensation will be reduced, e.g. if the CSO takes up other suitable employment. The Company may terminate the clauses with one month's notice.

Under Danish mandatory law, non-competition clauses cannot be enforced after expiry of the notice period, if termination is initiated by the Company without the Key Employee in question having given reasonable cause for the dismissal.

Incentive Programmes

Orphazyme's current incentive programmes comprise a short-term performance-based cash bonus programme and a long-term share-based incentive programme to be implemented subsequent to Admission. As of the date hereof, Orphazyme has a pre-IPO warrant programme in place, which will be discontinued in connection with the Offering (subject to completion thereof).

In accordance with Section 139 of the Danish Companies Act, the Company has adopted a remuneration policy, including overall guidelines on incentive pay, for the Board of Directors and Executive Management, which has been approved by the general meeting. Any future amendments to the remuneration policy will be presented to the general meeting for approval. The remuneration policy is available on the Company's website.

Short-term incentive programme (the "STIP")

The Executive Management and Key Employees are eligible to receive an annual performance-based cash bonus subject to certain predefined corporate and individual goals as determined by the Board of Directors on an annual basis. These predefined goals may include financial and/or operational targets, e.g. related to financing, working capital needs, organizational development as well as pre-clinical and clinical development. A cash bonus received under the STIP may not exceed 100% of the annual fixed salary of the participants. For the financial year 2016, Orphazyme expensed tDKK 743 on cash bonuses for the Executive Management and Key Employees.

Long-term incentive programme (the "LTIP")

In connection with and subject to completion of the Offering, the Executive Management and Key Employees have been offered to subscribe for Offer Shares ("**Investment Shares**") at the Offer Price for a maximum amount corresponding to approximately 15% (CMO) and 20% (CEO, CFO and CSO) of their respective current annual base salaries. The Board of Directors may decide to offer other current or new employees of Orphazyme to participate in the LTIP. A loan facility with Danske Bank A/S is being made available in order to facilitate the Executive Management and Key Employees' subscription for Investment Shares as part of the LTIP.]

The participants may be allocated a number of Shares ("**Performance Shares**") at a price per Performance Share of DKK 1 at the end of a vesting period of four years from the date of Admission. The number of Performance Shares shall be proportional to the potential increase in the price of the Shares at the time of vesting compared to the Offer Price. The potential increase in the price of the Shares will be calculated as the volume weighted average share price as quoted on Nasdaq Copenhagen during the 10 trading days preceding the vesting date (being the fourth anniversary following Admission to trading and official listing on Nasdaq Copenhagen). The maximum allocation of Performance Shares will be six (CEO) and four (CFO, CMO and CSO) times the number of Investment Shares subscribed for in connection with the Offering. Performance Shares will be allocated on a linear scale with maximum allocation triggered by an 80% increase in the price of the Shares at the time of vesting, whereas no Performance Shares will be allocated, if the price of the Shares has increased 20% or less at the time of vesting. Additionally, vesting is inter alia subject to the participants having maintained ownership of their Investment Shares and continued employment at the time of vesting.

In order to also promote a short-term incentive for the participants to retain their employment with Orphazyme, they may be allocated a number of Shares ("**Matching Shares**") at a price per Matching Share of DKK 1 in connection with the first anniversary following Admission. The number of Matching Shares shall be equal to the number of Investment Shares subscribed for in connection with the Offering and vesting will be subject to the participants having maintained ownership of their Investment Shares and continued employment at the time of vesting.

The Matching Shares and Performance Shares may vest on an accelerated basis in connection with a public takeover bid for the Shares (subject to the vesting conditions of the LTIP being satisfied at such time). Furthermore, the Board of Directors may at its sole discretion decide to accelerate vesting under other special circumstances, e.g. in connection with a merger, demerger or delisting.

The number of Matching Shares and Performance Shares may be adjusted in connection with certain changes to the capital structure of Orphazyme, e.g. capital increases below market value (subject to certain exceptions, e.g. capital increases made in connection with share-based incentive programmes).

The aggregate value of the LTIP is dependent on a number of factors, e.g. the level of investment by the participants, the Offer Price and the price of the Shares at the time of vesting. Assuming maximum investment by the participants and maximum allocation of Matching Shares and Performance Shares at the respective vesting dates, a maximum of 18,956 Matching Shares and 87,628 Performance Shares would be allocated to the current participants in the LTIP (assuming an Offer Price at the mid-point of the Offer Price Range).

Orphazyme's obligation to deliver Matching Shares and Performance Shares under the LTIP may be covered by a variety of means, e.g. Shares held in treasury by the Company accumulated through share buy-backs, or directed issues of Shares and/or bonus Shares.

The LTIP will be treated as an equity-settled share-based incentive programme and expensed over the four-year vesting period.

IPO cash bonus

Subject to completion of the Offering, all employees of Orphazyme (including the Executive Management and Key Employees) may be eligible to receive a cash bonus corresponding to 10% of their respective annual base salaries (excluding pension contributions). The cash bonus is subject to the success of the IPO as determined by the Board of Directors. The aggregate value of the cash bonus is expected to be approximately DKK 2.9 million (excluding social costs).

Pre-IPO warrant programme

A warrant programme was introduced in September 2010 with the purpose of aligning the interests of the participants and the shareholders (the "**2010 Warrant Programme**"). As of the date hereof, a total of 462,701 warrants have been issued and made available for allocation under the 2010 Warrant Programme to the Executive Management, Key Employees, certain other of Orphazyme's employees, certain members of the Board of Directors and certain external consultants from September 2010 to September 2015. These warrants have been allocated according to a specific allocation scheme subject to continued employment or other relevant affiliation with Orphazyme, as the case may be. As of the date hereof, all warrants have been allocated under the 2010 Warrant Programme. Each warrant entitles the holder of the warrant to subscribe for one Share at a price of DKK 44 per Share.

In March 2017, another warrant programme was introduced on materially the same terms as the 2010 Warrant Programme, however, with an adjusted exercise price in order to adjust the warrant programme to the changes in Orphazyme's capital structure (the "**2017 Warrant Programme**"). As of the date hereof, a total of 700,051 warrants have been issued and made available for allocation under 2017 Warrant Programme to the Executive Management, Key Employees, certain other of Orphazyme's employees and certain members of the Board of Directors during 2017. As of the date hereof, all warrants have been allocated under the 2017 Warrant Programme. In connection with the Offering, an additional 130,541 warrants have been issued and made available for allocation on an accelerated basis to certain members of the Board of Directors, Executive Management and Key Employees as an IPO bonus (the "**IPO Warrant Programme**"). The IPO Warrant Programme has been introduced on materially the same terms as the 2017 Warrant Programme with allocation of the IPO Warrant Programme being subject to completion of the Offering and, thus, not subject to a specific allocation scheme. Each warrant entitles the warrant holder to subscribe for one Share at a price of DKK 1 per Share.

According to the terms and conditions of the 2010, 2017 and IPO Warrant Programmes, an exercise window of 10 working days has been triggered in connection with the Offering during which all allocated warrants may be exercised amounting to a total of 1,293,293 warrants ("**Pre-IPO Warrants**"). Pre-IPO warrants that are not exercised within the exercise period will lapse without further compensation. Upon exercise of Pre-IPO Warrants allocated under the 2017 Warrant Programme, the warrant holder's right to exercise warrants allocated under the 2010 Warrant Programme, if any, will be reduced accordingly and vice versa. For this reason and as 86,181 Pre-IPO Warrants (of which 36,373 warrants are still deemed exercisable pursuant to their terms) under the 2010 Warrant Programme are exempt from the aforementioned reduced right of exercise, the maximum nominal capital increase resulting from the exercise of the Pre-IPO Warrants is limited to

DKK 866,965. The capital increase related to exercise of Pre-IPO Warrants, if any, is expected to take place in connection with settlement of the Offering, i.e. on or around the Settlement Date being 21 November 2017.

The terms governing the 2010, 2017 and IPO Warrant Programmes are attached to the Articles of Association and available on the Company's website.

Statement on Past Records

During the past five years, none of the members of the Board of Directors, the Executive Management or any of the Key Employees have been (i) convicted of fraudulent offenses; (ii) directors or officers of companies that have entered into bankruptcy, receivership or liquidation, except as set out immediately below; or (iii) subject to any public incrimination and/or sanctions by statutory regulatory authorities (including designated professional bodies), and have not been disqualified by a court from acting as a member of an issuer's board of directors, executive board or supervisory body or being in charge of an issuer's management or other affairs.

Georges Gemayel was chairman of Vascular Magnetics Inc., when it was dissolved in 2014 and chairman of Epitherapeutics ApS until 2015, when the company was dissolved following merger and Syndexa Pharmaceuticals Inc. until 2012 (dissolved in 2013). Georges Gemayel was a member of the board of directors of NPS Pharmaceuticals Inc. until 2015, when the company was dissolved following merger; Raptor Pharmaceuticals Corp. until 2016, when the company was dissolved following merger; and Prosensa N.V. until 2015, when it was dissolved following merger.

Anders Hedegaard was chairman of the board of directors of Scanning Technology A/S, when it was dissolved following merger in August 2015.

Anders Vadsholt was a member of the executive management of Topotarget A/S (publ), when it was dissolved following merger in July 2014.

Bo Jesper Hansen was chairman of the board of directors of Topotarget A/S (publ), when it was dissolved by merger in July 2014 and a member of the board of directors of Hyperion Therapeutics Inc. until 2015 (dissolved following acquisition) and Gambro AB until December 2013 (dissolved following acquisition).

Martijn Kleijwegt was a member of the board of directors of Prosensa N.V. until 2015, when the company was dissolved following merger.

Martin Bonde was a member of the executive management of Epitherapeutics ApS, when it was dissolved following merger in 2015.

Nanna Lüneborg was a member of the board of directors of Pcovery ApS until February 2016. The company is currently undergoing voluntary liquidation.

Patrick J.H. Krol was a member of the board of directors of Aquapharm Biodiversity Ltd, until 2013 (dissolved following merger in 2014) as well as a member of the board of directors of T-BBB Technologies BV until 2015 (currently undergoing liquidation).

Sten Verland was chairman of the board of directors of Action Pharma af 22. Februar 2012 A/S, when it was dissolved by voluntary liquidation in May 2013; a member of the board of directors of P/S Sunstone Biomedicinsk Venture III, which is currently undergoing voluntary liquidation in January 2001; Sunstone LSV & Co. Special Limited Partner III Holding ApS, when it was dissolved following merger in December 2014; Biovision A/S, when it was dissolved following declaration of payments in November 2014; Scanbur Corporation ApS, when it was dissolved following merger in December 2014; Scanbur Technology ApS, when it was dissolved following merger in December 2014; Scanbur Research ApS, when it was dissolved following merger in December 2014; NsGene A/S, when it was dissolved by voluntary liquidation in May 2017; member of the executive management of Sunstone LSV Partners Holding III ApS, when it was dissolved following declaration of payments in January 2015;Sunstone LSV Partners & Co. Holding III ApS, when it was dissolved following declaration of payments in January 2015; Sunstone LSV Special Limited Partner III Holding ApS, when it was dissolved following merger in December 2014; Sunstone LSV Invest III Holding ApS, when it was dissolved following merger in December 2014; Sunstone LSV & Co. Invest III Holding ApS, when it was dissolved following merger in December 2014 and Sunstone LSV Partners Holding III ApS, when it was dissolved by voluntary liquidation in January 2015.

Statement on Conflicts of Interest

There are no family ties among the members of the Board of Directors, the Executive Management or any of the Key Employees.

With the exception of the members of the Board of Directors, Martin Kleijwegt, Nanna Lüneborg, Patrick J.H. Krol, Rémi Droller and Sten

Verland, the Company is not aware of any member of the Board of Directors, or the Executive Management or any of the Key Employees having been appointed to their current position pursuant to an agreement or understanding with the major shareholders, customers, suppliers or other parties.

None of the members of the Board of Directors, or the Executive Management or any other Key Employees have conflicts of interest with respect to their duties as members of the Board of Directors, or the Executive Management or as Key Employees except for the members of the Board of Directors, Martin Kleijwegt, Nanna Lüneborg, Patrick J.H. Krol, Rémi Droller and Sten Verland, for the reasons set out in the paragraph above.

None of the members of the Board of Directors, the Executive Management or the Key Employees have positions in other companies which could result in a conflict of interest vis-à-vis such companies, either because Orphazyme has an equity interest in such company or because Orphazyme and the company concerned have an ongoing business relationship, except as disclosed under "*Related Party Transactions*". However, Orphazyme may do business in the ordinary course with companies in which members of the Board of Directors, or the Executive Management, or the Key Employees may hold positions as directors or officers.

It follows from the Rules of Procedure of the Company's Board of Directors and the Danish Companies Act that a member of the Board of Directors or the Executive Management shall not participate in the preparation, discussions or the decision-making process concerning an agreement between the Company and the member in question or concerning legal proceedings between the member in question and the Company or an agreement between the Company and any third party or legal proceedings brought against any third party if the member in question has a significant interest therein that may conflict with its interests.

Description of Internal Control and Financial Reporting Procedures

The Board of Directors, the Audit Committee and the Executive Management are ultimately responsible for Orphazyme's risk management and internal controls in relation to its financial reporting, and approve Orphazyme's general policies in that regard. The Audit Committee assists the Board of Directors in overseeing the reporting process and the most important risks involved in this respect. The Executive Management are responsible for the effectiveness of the internal controls and risk management and for the implementation of such controls aimed at mitigating the risk associated with the financial reporting.

Orphazyme has internal control and financial reporting procedures aimed at enabling it to monitor its performance, operations, funding and risk. While Orphazyme continues to improve its procedures and internal control, including documentation of the internal control systems, Orphazyme believes that its reporting and internal control systems enable it to be compliant with disclosure obligations applying to issuers of shares admitted to trading and official listing on Nasdaq Copenhagen. Orphazyme's internal control and financial reporting procedures include, among other things:

- Monthly financial information, including income statement, balance sheet, cash flow results and actual amounts compared with budgeted performance, latest forecast and explanations of any material deviations. The monthly financials are reported to the Executive Management;
- Monthly highlight reports, including key performance indicators for each project in the pipeline and general corporate activities on actual performance compared with budgeted performance and previous year's performance and explanations of any material deviations. The monthly highlights are reported to the Executive Management and discussed at monthly review sessions with project management supervising the progress of each project in the pipeline;
- Monthly, clinical development meetings are held between project leaders and CMO and CFO where significant changes to projects are discussed. Significant changes are approved in accordance with defined level-of-authority.
- Quarterly detailed review of accruals for clinical trials relating to activities performed by CROs and other external vendors, especially in respect of the sIBM, ALS, NPC and Gaucher disease programs by project management and the CFO.
- Liquidity management is executed on a daily basis, with a view to securing the Company's required liquidity through appropriate cash management, and maintaining adequate liquidity reserves at any time. As part of the liquidity management, the Company applies controls regarding cash disbursements based on a defined level-of-authority; and
- Centralised planning processes including a centrally driven budget process with bottom-up input from all project managers responsible for the individual product programs and from Executive Management in respect of corporate activities, updated "full year estimates" and, from 2017, the introduction of a "36 months rolling forecast process"; and
- On a quarterly basis, a detailed reporting of financial information and project development is reported to the Board of Directors.

External audit

Orphazyme's independent auditors are appointed for a term of one year by the shareholders at the Company's annual general meeting upon recommendation from the Audit Committee. The Board of Directors assesses the independence and competencies and other

matters pertaining to the auditors. The framework for the auditors' compensation and duties, including audit and non-audit tasks, is agreed annually between the Board of Directors and Orphazyme's auditors based on recommendation from the Audit Committee. Orphazyme has regular dialogue and exchange of information with its auditors.

Corporate Governance

The Company is committed to exercising good corporate governance at all times and the Board of Directors will regularly assess rules, policies and practices according to the Corporate Governance Recommendations. Nasdaq Copenhagen has incorporated the Corporate Governance Recommendations in the Rules for issuers of shares of 3 July 2016 (the "**Issuer Rules of Nasdaq Copenhagen**"). Accordingly, as a company with shares admitted to trading and official listing on Nasdaq Copenhagen, the Company will be required to comply with or explain deviations from the Corporate Governance Recommendations as also required pursuant to Section 107b of the Danish Consolidated Act no. 1580 of 10 December 2015 on financial statements.

In connection with the Offering and with effect from the Admission, the Board of Directors has prepared a statutory statement on corporate governance that reflects the compliance of the Company with each of the Corporate Governance Recommendations. The Company intends to comply with the Corporate Governance Recommendations in all material respects except that the Company has opted to deviate from the Corporate Governance Recommendations in the following areas:

- For the time being, the Company has decided only to publish half-yearly financial reports, as the Company is still in a developmental phase until potential commercialisation of its product candidates. Accordingly, the Company does not find it necessary at present to communicate its limited financial performance to investors and other stakeholders on more than a half-yearly basis.
- The Company does not regard age as a deciding factor with respect to the selection and nomination of candidates for the Board of Directors or the performance of the individual board members. The Company believes that the annual performance evaluation of the Board of Directors and the board members' one-year terms are adequate in this respect. Additionally, the Company takes note that this recommendation appears to have been abolished in the most recent proposal for revised Corporate Governance Recommendations expected to enter into force in 2018.
- Share-based compensation, e.g. shares, share options, performance shares or warrants, constitutes a common part of the board remuneration in competing international biotech companies. To remain competitive in the international market and to be able to attract and retain qualified members of the Board of Directors, Orphazyme is open this practice and believes that it may be beneficial to the shareholders' long-term interests.
- The LTIP is currently structured as a one-off four-year incentive programme, i.e. it is not a roll-over programme and does not involve annual grants. The Board of Directors wishes to have flexibility with respect to designing and implementing new share-based incentive schemes adjusted to the needs of the business. However, the Board of Directors is open to introducing a share-based incentive programme involving annual grants at a future point in time.
- As members of the Board of Directors are elected for a term of one year, the Company finds that any share-based instruments that are granted to members of the Board of Directors should have a maturity of one year from the date of allocation.
- Under LTIP, the participants may be granted a number of Matching Shares on the first anniversary following the IPO, subject to continued employment and a specific minimum shareholding at such time. Accordingly, the Matching Shares have a maturity of less than three years. The Matching Shares are intended to promote short-term retention of the participants and act as a bridge until potential vesting of the Performance Shares after a four-year performance period.

The Company's corporate governance practices are also accounted for in the statutory statement on corporate governance, which is available on the Company's website.

Ownership Structure and Main Shareholders

2017 Capital Structure Adjustment

In preparation of the Offering, the capital structure of Orphazyme has been adjusted by way of a merger of the three previous share classes into one combined with an issue of bonus Shares in order to account for the now abolished preference shares (the "**2017 Capital Structure Adjustment**"). In connection with the 2017 Capital Structure Adjustment, the class B and C preference shares of the Company were converted into Shares on a 1:1 ratio. In order to account for the preferential rights attached to the preference shares, a directed issue of bonus Shares using free reserves of the Company was carried out at par value in favour of the preference shareholders, assuming an Offer Price in the top end of the Offer Price Range. In case the Offer Price is set below the top end of the Offer Price Range, an additional number of bonus Shares will be issued shortly before Admission in favour of the former preference shareholders in order to account for the Offer Price. Assuming an Offer Price at the bottom end of the Offer Price Range, up to 1,621,968 bonus Shares with a nominal value of DKK 1 each will be issued, corresponding to 14.0% of the total share capital and voting rights of the Company as of the date hereof.

Ownership Structure

As of the date hereof, Novo Holdings A/S ("**Novo Holdings**") holds 33.1%; Coöperative Aescap Venture I U.A. ("**Aescap Venture**") holds 15.2%; Sunstone Life Science Ventures Fund II K/S ("**Sunstone Capital**") holds 15.6%; Orpha Pooling B.V. (a joint venture between LSP V Coöperatieve U.A. ("**LSP**") and ALS Invest) holds 21.0%; FCPI Idinvest Patrimoine n°3, FCPI Idinvest Patrimoine n°4, FCPI Objectif Innovation Patrimoine n°6 and FCPI Objectif Innovation Patrimoine n°7 (together "**Idinvest**") collectively hold 7.2%; and Kurma Biofund II holds 6.6% of the Company's Shares, respectively (together referred to as the "**Main Shareholders**") and UCL Business PLC holds 0.1% and Kansas Life Sciences Development Inc. holds 0.1%. The remaining Shares (1.1%) are indirectly held by the founders of the Company (CSO Thomas Kirkegaard Jensen; CEO Anders Hinsby; Marja Helena Jaattela; and member of the Existing Board of Directors, Martin Rahbek Kornum) through their respective wholly-owned holding companies. As of the date of hereof, the Company's share capital amounts to a nominal value of DKK 11,590,092 divided into 11,590,092 Shares of DKK 1 each, each of which is fully paid. Prior to Admission, up to 866,965 new Shares with a nominal value of DKK 1 each may be issued upon exercise of Pre-IPO Warrants vesting in connection with the Offering. Additionally, upon determination of the Offer Price, but prior to Admission, up to 1,621,968 bonus Shares with a nominal value of DKK 1 each may be issued as part of the 2017 Capital Structure Adjustment.

A total of 1,293,293 Pre-IPO Warrants may be exercised (or lapse if not exercised) in connection with the Offering. As further described in "*Board of Directors, Executive Management and Key Employees—Incentive Programmes—Pre-IPO warrant programme*", the maximum nominal capital increase resulting from the exercise of the Pre-IPO Warrants is limited to DKK 866,965. The capital increase related to exercise of Pre-IPO Warrants, if any, is expected to take place in connection with settlement of the Offering, i.e. on or around the Settlement Date being 21 November 2017.

At the time of completion of the Offering, the Company will issue a number of new Shares, the number of which will be determined based on gross proceeds of approximately DKK 600 million (excluding the Overallotment Option). Assuming completion of the Offering, the Company's registered share capital will increase by a nominal value of up to DKK 9,375,000 as a result of the issue of Offer Shares (excluding the Overallotment Option). The exact number of Offer Shares will be determined based on a book-building process.

Skandinaviska Enskilda Banken, Danmark, branch of Skandinaviska Enskilda Banken AB (Publ.), Sweden; Vækstfonden; BI Asset Management Fondsmæglerselskab A/S on behalf of certain clients; Handelsbanken, branch of Svenska Handelsbanken AB (Publ.), Sweden; and Spar Nord Bank A/S have in connection with the Offering subject to certain conditions undertaken to subscribe for Offer Shares as Cornerstone Investors for a total subscription amount of DKK 230 million, corresponding to approximately 38.3% of the Offering (excluding the Overallotment Option).

The existing Shares issued as of the date hereof will be diluted in connection with the Offering by the issuance of up to 10,781,250 new Shares, corresponding to a nominal value of DKK 10,781,250, assuming full exercise of the Overallotment Option. Following completion of the Offering, the existing Shares issued and outstanding will make up 57.9% of the Company's share capital, assuming full subscription for all new Shares issued in connection with the Offering (including full exercise of the Overallotment Option) as well as exercise of all Pre-IPO Warrants vesting in connection with the Offering, and adjusting for the 2017 Capital Structure Adjustment (assuming an Offer Price at the midpoint of the Offer Price Range and, thereby, an issue of 720,875 bonus Shares prior to Admission as part of the 2017 Capital Structure Adjustment). The Company's net capital as of 30 September 2017 was DKK 94,500,000, corresponding to a net book value per Share of DKK 18.5, corresponding to DKK 8.2 per Share based on the Company's capital structure as of the date hereof. The net book value per Share is calculated by dividing the total net capital by the total number of Shares issued. Based on the Company's net capital as of 30 September 2017 and the same assumptions as described above as well as adjusting for the estimated costs related to the Offering, the Company's net book value per Share as of completion of the Offering would be DKK 32.1.

Table of shareholders

The following table sets out the information regarding the Company's ownership structure as at the date hereof; and immediately following the completion of the Offering, assuming exercise of all Pre-IPO Warrants vesting in connection with the Offering.

At an Offer Price at the top end of the Offer Price Range, no additional bonus Shares will be issued as part of the 2017 Capital Structure Adjustment. At an Offer Price at the bottom end of the Offer Price Range, 1,621,968 additional bonus Shares will be issued to the former preference shareholders prior to Admission as part of the 2017 Capital Structure Adjustment. Any such issue of additional bonus Shares has been reflected in the following table to the extent relevant.

The information in the table below includes indirect holdings through legal entities (e.g. private holding companies) to the extent relevant. In case the shareholding percentages do not sum to 100% in the following table, this is due to rounding.

			Shares held after completion of the Offering, assuming full exercise of the Overallotment Option					
	Shares held as at the date hereof[1]		At an Offer Price at the bottom end of the Offer Price Range		At an Offer Price at the midpoint of the Offer Price Range		At an Offer Price at the top end of the Offer Price Range	
Shareholders	**Number of Shares**	**%**	**Number of Shares**	**%**	**Number of Shares**	**%**	**Number of Shares**	**%**
Novo Holdings	3,830,832	33.1%	4,377,085	17.6%	4,073,611	17.9%	3,830,832	18.2%
Aescap Ventures	1,765,605	15.2%	2,017,802	8.1%	1,877,692	8.2%	1,765,605	8.4%
Sunstone Capital	1,804,405	15.6%	2,062,228	8.3%	1,918,993	8.4%	1,804,405	8.6%
Orpha Pooling B.V. (LSP and ALS Invest)	2,431,672	21.0%	2,758,354	11.1%	2,576,865	11.3%	2,431,672	11.5%
Idinvest	833,362	7.2%	953,815	3.8%	886,897	3.9%	833,362	4.0%
Kurma Biofund II	765,947	6.6%	879,886	3.5%	816,587	3.6%	765,947	3.6%
Kansas Life Sciences Development Inc.	16,691	0.1%	19,016	0.1%	17,724	0.1%	16,691	0.1%
UCL Business PLC	16,578	0.1%	18,874	0.1%	17,598	0.1%	16,578	0.1%
Founders[2]	125,000	1.1%	125,000	0.5%	125,000	0.5%	125,000	0.6%
Warrant holders	-	-	866,965	3.5%	866,965	3.8%	866,965	4.1%
New shareholders[3]	-	-	10,781,250	43.4%	9,583,333	42.1%	8,625,000	40.9%
Total[4]	**11,590,092**	**100%**	**24,860,275**	**100%**	**22,761,265**	**100%**	**21,082,057**	**100%**
Existing Board of Directors[5]								
Georges Gemayel[6]	-	-	87,758	0.4%	87,758	0.4%	87,758	0.4%
Bo Jesper Hansen[7]	-	-	91,945	0.4%	85,000	0.4%	79,445	0.4%
Martijn Kleijwegt	-	-	-	-	-	-	-	-
Martin Bonde[8]	-	-	46,790	0.2%	46,356	0.2%	46,009	0.2%
Martin Rahbek Kornum[9]	18,750	0.2%	41,476	0.2%	41,476	0.2%	41,476	0.2%
Nanna Lüneborg	-	-	-	-	-	-	-	-
Patrick J.H. Krol	-	-	-	-	-	-	-	-
Rémi Droller	-	-	-	-	-	-	-	-
Sten Verland	-	-	-	-	-	-	-	-
New Board Members[5]								
Anders Hedegaard	-	-	7,812	0.0%	6,944	0.0%	6,250	0.0%
Catherine Moukheibir	-	-	10,000	0.0%	8,888	0.0%	8,000	0.0%
Executive Management								
Anders Hinsby[10]	18,750	0.2%	206,236	0.8%	205,498	0.9%	204,908	1.0%
Anders Vadsholt[11]	-	-	126,556	0.5%	126,000	0.6%	125,556	0.6%
Key Employees								
Thomas Blaettler[12]	-	-	113,189	0.5%	112,668	0.5%	112,252	0.5%
Thomas Kirkegaard Jensen[13]	68,750	0.6%	230,285	0.9%	229,729	1.0%	229,285	1.1%

(1) Subsequent to the initial steps of the 2017 Capital Structure Adjustment (including an issue of 6,487,882 bonus Shares, assuming an Offer Price at the top end of the Offer Price Range).
(2) Excluding any Pre-IPO Warrants held or exercised by the founders (as otherwise set out in foot-notes 9, 10 and 13 below) and/or any Investment Shares subscribed for as part of the LTIP (as otherwise set out in foot-notes 10 and 13 below).
(3) Assuming no exercise of the Overallotment Option, the new shareholders will hold 9,375,000 Shares (40.0%) at the bottom end of the Offer Price Range; 8,333,333 Shares (38.7%) at the midpoint of the Offer Price Range; and 7,500,000 Shares (37.6%) at an Offer Price at top end of the Offer Price Range, respectively.
(4) Assuming no exercise of the Overallotment Option, the Company's total share capital will consist of 23,454,025 Shares at the bottom end of the Offer Price Range; 21,511,265 Shares at the midpoint of the Offer Price Range; and 19,957,057 Shares at an Offer Price at top end of the Offer Price Range, respectively.
(5) It is assumed in the table above that certain members of the Board of Directors participate in the Offering by subscribing for Offer Shares for an amount equal to the interest indications provided by the relevant board members prior to the Offering.
(6) Under the 2010 and 2017 Warrant Programmes, Georges Gemayel may exercise a maximum of 87,758 Pre-IPO Warrants in connection with the Offering.
(7) Under the 2010 and 2017 Warrant Programmes, Bo Jesper Hansen may exercise a maximum of 29,445 Pre-IPO Warrants in connection with the Offering.
(8) Under the 2010 and 2017 Warrant Programmes, Martin Bonde may exercise a maximum of 42,884 Pre-IPO Warrants in connection with the Offering.
(9) Under the 2010 and 2017 Warrant Programmes, Martin Rahbek Kornum may exercise a maximum of 22,726 Pre-IPO Warrants in connection with the Offering. Martin Rahbek Kornum also has an indirect ownership stake in the Company through a wholly-owned holding company.
(10) Assuming maximum subscription of Investment Shares as part of the LTIP (i.e. 20% of his current annual base salary). Under the 2010 and 2017 Warrant Programmes, Anders Hinsby may exercise a maximum of 180,846 Pre-IPO Warrants in connection with the Offering. Anders Hinsby also has an indirect ownership stake in the Company through a wholly-owned holding company.
(11) Assuming maximum subscription of Investment Shares as part of the LTIP (i.e. 20% of his current annual base salary). Under the 2010 and 2017 Warrant Programmes, Anders Vadsholt may exercise a maximum of 121,556 Pre-IPO Warrants in connection with the Offering.
(12) Assuming maximum subscription of Investment Shares as part of the LTIP (i.e. 15% of his current annual base salary). Under the 2010 and 2017 Warrant Programmes, Thomas Blaettler may exercise a maximum of 108,502 Pre-IPO Warrants in connection with the Offering.
(13) Assuming maximum subscription of Investment Shares as part of the LTIP (i.e. 20% of his current annual base salary). Under the 2010 and 2017 Warrant Programmes, Thomas Kirkegaard Jensen may exercise a maximum of 156,535 Pre-IPO Warrants in connection with the Offering. Thomas Kirkegaard Jensen also has an indirect ownership stake in the Company through a wholly-owned holding company.

The Company is not aware of being owned or controlled, directly or indirectly, by others, and the Company is not aware of any agreement that could later result in others taking control over it.

The Main Shareholders

Novo Holdings
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 oversees, invests and manages the assets of the Novo Nordisk Foundation. Besides being the controlling shareholder in the Novo Group companies, including Novo Nordisk A/S and Novozymes A/S, Novo Holdings A/S provides seed and venture capital to development stage companies and takes significant ownership positions in well-established companies within life science, as well as manages a broad portfolio of financial assets.

Aescap Venture
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 is managed by Aescap Venture Management B.V.

Aescap Venture is an investor across the life science sector in Europe.

Sunstone Capital
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, CVR. no. 30575245.

Sunstone Life Sciences Ventures is an early-stage investor across the life science sector in Europe.

Orpha Pooling B.V.
Orpha Pooling B.V. is a liability liability company organised under the laws of the Netherlands under registration number 67827055 with its registered address at Johannes Vermeer, Plein 9, 1071 DV Amsterdam, the Netherlands. The shareholders of Orpha Pooling B.V. are (i) LSP V Coöperatieve U.A., a cooperative with excluded liability, having its official seat in Amsterdam, the Netherlands, and its registered office address at Johannes Vermeerplein 9, 1071 DV Amsterdam, the Netherlands, registered with the Dutch trade register under number 61888575, and (ii) ALS Invest 2 B.V., 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 Dutch closed-end investment fund, which is a subsidiary of LSP Management Group B.V., an experienced healthcare investment firm with offices in Amsterdam, Munich and Boston. ALS Invest 2 B.V. is a special purpose vehicle incorporated for the investment in Orphazyme. ALS Invest 2 B.V. is managed by Sunu Ventures B.V., a Dutch venture capital firm dedicated to support and finance biotech companies that develop drugs and diagnostics for the disease ALS.

Idinvest

Idinvest Partners manages its investment 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. Idinvest Partners is a French société anonyme, governed by the laws of France, having its registered office at 117 Avenue des Champs-Elysées, 75 008 Paris, France, registered with the Registry of Trade and Companies of Paris under number 414 735 175With EUR 8 billion under management, Idinvest Partners is a leading pan-European private equity firm focused on the mid-market segment. Idinvest Partners has developed several complementary areas of expertise including investments in innovative European start-ups, primary, secondary and mezzanine investments in unlisted European companies, and private equity consulting. Founded under the name AGF Private Equity in 1997, Idinvest Partners was formerly part of Allianz until 2010 when it became an independent firm.

Kurma Biofund II

Kurma Biofund II is a fund organised under the laws of France under registration number 510043136 with its registered address at 5-7 rue de Monttessuy, 75007 Paris, France. Kurma Biofund II is managed by Kurma Partners, registration number 510 043 136.

Kurma Partners was founded in July 2009 and is a key European venture capital fund in the life sciences sector, notably through its Kurma Biofund I fund, Kurma Biofund II fund and Kurma Diagnostics acceleration fund. Managing funds totalling over EUR 250 million, Kurma's investment strategy is based both on a selection of the best European companies and on the creation of a portfolio of "proprietary" companies in which Kurma intervenes from their very creation.

Agreements related to the Ownership of Orphazyme

The Main Shareholders have entered into a shareholders' agreement dated 6 January 2015, as amended and supplemented (the "**Shareholders' Agreement**"). This Shareholders' Agreement, other than for certain customary terms inter alia related to confidentiality, will in accordance to its terms cease to have effect upon completion of the Offering.

Related Party Transactions

The Board of Directors, the Executive Management and the Key Employees are considered related parties of the Company as they exercise a significant influence on the Company's operations. Related parties also include such persons' relatives as well as undertakings in which such persons have significant interests. As of the date hereof, the Company is not ultimately controlled by any of its existing shareholders.

Except as set out below, the Company has not undertaken any significant transactions with its Board of Directors, Executive Management, Key Employees, or undertakings outside of Orphazyme in which related parties have significant interests.

During the calendar years 2015 and 2016 and to the date hereof, the Company has made the following transactions with related parties which were all carried out on arm's length terms:

Transactions with Board of Directors, Executive Management and Key Employees

The Company has not had any significant transactions with the members of the Board of Directors, Executive Management or the Key Employees apart from the wages/salaries, pensions and other social security and staff costs in respect of the Executive Management and Key Employees, and board fees and warrants received with respect to the Board of Directors. Reference is also made to notes 5, 6 and 14 of Orphazyme's Audited Financial Statements, as included in this Offering Circular on F-26, F-27 and F-35 - F36, and note 6 of the Interim Financial Statements, as included in this Offering Circular on F-12.

Transactions with the Shareholders and their affiliates

The Company has not had any significant transactions with its existing shareholders or their affiliates apart from the capital increases set out in "*Description of the Shares and share capital—Movement in the share capital*" and the issue of bonus Shares as part of the 2017 Capital Structure Adjustment (see "*Ownership Structure and Main Shareholders*"). Reference is also made to note 10 of Orphazyme's Audited Financial Statements, as included in this Offering Circular on F-30, and notes 3 and 6 of the Interim Financial Statements, as included in this Offering Circular on F-10 and F-12.

Since 30 June 2017, Aescap Venture and Orpha Pooling B.V. (LSP and ALS Invest) have paid DKK 91.3 million receivable from previous capital increases.

In connection with and subject to completion of the Offering, certain members of the Board of Directors, Executive Management and Key Employees have been granted a number of warrants under the 2017 Warrant Programme and a number of warrants under the IPO Warrant Programme on an accelerated basis as an IPO bonus. For further information, please see "*Board of Directors, Executive Management and Key Employees—Incentive Programmes—Pre-IPO warrant programme*".

Description of the Shares and Share Capital

The following is a summary of material information relating to the Company's share capital, including a summary of certain provisions of the Company's Articles of Association dated 2 november 2017, as well as a brief description of certain provisions of the Danish Companies Act. This summary does not purport to be exhaustive and should be read in conjunction with the full text of the Company's Articles of Association as well as in the context of applicable Danish law. See "*Annex A—Excerpt of the Articles of Association of the Company*".

The Company is a public limited liability company incorporated on 19 June 2009 and later converted into a Danish public limited company in 20 October 2017 and is organised under the laws of Denmark under the name Orphazyme A/S with its registered office at Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark. The Company is registered with the Danish Business Authority under company registration (CVR) no. 32266355.

Registered Share Capital

As of the date of this Offering Circular, the Company's share capital had a nominal value of DKK 11,590,092, divided into 11,590,092 Shares of DKK 1 each or multiples thereof. All Shares are issued and fully paid up. The Shares are not divided into share classes, and all Shares have the same rights and rank *pari passu* in respect of voting rights, preemption rights, redemption, conversion and restrictions or limitations according to the Articles of Association or eligibility to receive dividend or proceeds in the event of dissolution and liquidation. No Shares shall carry special rights, restrictions or limitations pursuant to the Company's Articles of Association.

Each Share of the nominal value DKK 1 gives the holder the right to one vote at the Company's general meetings.

As of the date hereof, the Company has issued a number of indemnification warrants to certain of its current shareholders (Novo Holdings, Aescap Venture, Sunstone Capital, Idinvest and Kurma Biofund II) giving the warrant holders a right to subscribe for the Company's former class C preference shares in case certain contractual warranty claims under the Shareholders' Agreement materialise. According to their terms and conditions, the indemnification warrants will lapse without compensation in connection with Admission. Other than the indemnification warrants and as described in "*Board of Directors, Executive Management and Key Employees—Pre-IPO Warrant Programme*", the Company has not issued any securities that are convertible, exchangeable nor have warrants attached other.

Before completion of the Offering, the Company's share capital will have a nominal value of DKK 12,310,967, divided into 12,310,967 Shares of DKK 1 each or multiples thereof, assuming an Offer Price at the midpoint of the Offer Price Range (and thereby an issue of 720,875 bonus Shares prior to Admission as part of the 2017 Capital Structure Adjustment), which will all be issued and fully paid up.

Immediately after payment of the new Shares to be issued by the Company pursuant to the Offering (excluding the Overallotment Option) and registration of the related capital increase, the Company's registered share capital will have a nominal value of DKK 21,511,265, divided into 21,511,265 Shares of nominal value DKK 1 each, assuming exercise of all Pre-IPO Warrants vesting in connection with the Offering and an Offer Price at the midpoint of the Offer Price Range (and thereby an issue of 720,875 bonus Shares prior to Admission as part of the 2017 Capital Structure Adjustment).

Movement in the Share Capital

The table set forth below presents the development of the Company's share capital from its incorporation until the date hereof.

Date of approval	Transaction type	Share capital before change (DKK)	Share capital change (DKK)	Share capital after change (DKK)	Number of shares after change
19 June 2009	Incorporation of the Company as a private limited company	—	—	125,000	A-shares: 125,000
15 March 2010	Capital increase by cash contribution	125,000	100,000	225,000	A-shares: 125,000 B-shares: 100,000
9 December 2010	Capital increase by cash contribution and debt conversion (50/50)	225,000	204,082	429,082	A-shares: 125,000 B-shares: 304,082
12 May 2011	Capital increase by cash contribution	429,082	136,054	565,136	A-shares: 125,000 B-shares: 440,136
12 July 2011[1]	Capital increase by cash contribution	565,136	760,204	1,325,340	A-shares: 125,000 B-shares: 1,200,340

29 August 2011[1]	Capital increase by cash contribution	1,325,340	304,081	1,629,421	A-shares: 125,000 B-shares: 1,504,421
3 April 2013	Capital increase by cash contribution	1,629,421	545,787	2,175,208	A-shares: 125,000 B-shares: 2,050,208
6 January 2015[2]	Capital increase by cash contribution and debt conversion	2,175,208	1,042,823	3,218,031	A-shares: 125,000 B-shares: 2,050,208 C-shares: 1,042,823
12 November 2015	Capital increase by cash contribution	3,218,031	127,724	3,345,755	A-shares: 125,000 B-shares: 2,050,208 C-shares: 1,170,547
16 September 2016	Capital increase by cash contribution	3,345,755	14,786	3,360,541	A-shares: 125,000 B-shares: 2,050,208 C-shares: 1,185,333
26 January 2017	Capital increase by cash contribution	3,360,541	534,007	3,894,548	A-shares: 125,000 B-shares: 2,050,208 C-shares: 1,719,340
26 January 2017	Capital increase by cash contribution	3,894,548	772,022	4,666,570	A-shares: 125,000 B-shares: 2,050,208 C-shares: 2,491,362
29 June 2017	Capital increase by cash contribution	4,666,570	435,640	5,102,210	A-shares: 125,000 B-shares: 2,050,208 C-shares: 2,927,002
20 October 2017	Conversion into a public limited liability company	5,102,210	—	5,102,210	A-shares: 125,000 B-shares: 2,050,208 C-shares: 2,927,002
2 November 2017	Consolidation of share classes[3]	5,102,210	—	5,102,210	5,102,210
2 November 2017	Issue of bonus Shares[3]	5,102,210	6,487,882	11,590,092	11,590,092

[1] As corrected with the Danish Business Authority on 9 April 2013.
[2] DKK 827,165 of the capital increase was paid for in cash and DKK 215,658 was paid for by debt conversion.
[3] In connection with the 2017 Capital Structure Adjustment involving a 1:1 consolidation of the then existing share classes, a number of bonus Shares were issued in order to account for the preferential rights attached thereto. For a further description of the 2017 Capital Structure Adjustment, please see "*Ownership Structure and Main Shareholders—2017 Capital Structure Adjustment*".

Authorisation to Increase the Share Capital

The Board of Directors has pursuant to the Articles of Association been granted authorisation to increase the Company's share capital.

In accordance with article 3.1 of the Articles of Association, the Board of Directors is, until 31 December 2017, authorised to increase the share capital of the Company in one or more issues of new Shares without preemption rights for the existing shareholders of the Company by up to a nominal amount of DKK 9,400,000. The capital increase shall take place at market price as determined through a book-building process and shall be effected by cash payment. See also "*The Offering—Authorisation*".

In accordance with article 3.2 of the Articles of Association, the Board of Directors is, until 31 December 2017, authorised to increase the share capital of the Company in one or more issues of new Shares without preemption rights for the existing shareholders of the Company by up to a nominal amount of DKK 1,500,000 for purposes of the Overallotment Option. The capital increase shall take place at the Offer Price and shall be effected by cash payment. See also "*The Offering—Authorisation*".

In accordance with article 3.3 of the Articles of Association, the Board of Directors is, until 2 November 2017, authorised to increase the share capital of the Company in one or more issues without preemption rights for the existing shareholders of the Company by up to a nominal amount of DKK 1,700,000 in connection with a directed issue of bonus Shares to one or more shareholders. The capital increase shall take place at par value. See also "*Ownership Structure and Main Shareholders—2017 Capital Structure Adjustment*" regarding the reorganisation of Orphazyme's previous share capital structure.

In accordance with article 3.4 of the Articles of Association, the Board of Directors is, until 2 November 2022, authorised to increase the share capital of the Company in one or more issues of new Shares without preemption rights for the existing shareholders of the Company by up to a nominal amount of DKK 5,000,000. The capital increase shall take place at market price and may be effected by cash payment, conversion of debt or by contribution of other assets than cash.

In accordance with article 3.5 of the Articles of Association, the Board of Directors is, until 2 November 2022, authorised to increase the share capital of the Company in one or more issues without preemption rights for the existing shareholders of the Company by up to a nominal amount of DKK 1,300,000 in connection with the issue of new Shares to members of the Board of Directors, executives and/or employees of the Company. The new Shares shall be issued against cash payment at a subscription price to be determined by the Board of Directors, which may be below the market price.

In accordance with article 3.6 of the Articles of Association, the Board of Directors is, until 2 November 2022, authorised to increase the share capital of the Company in one or more issues without preemption rights for the existing shareholders of the Company by up to a nominal amount of DKK 15,750,000 in connection with issues of bonus Shares, and/or directed issues of new Shares effected by cash payment, to Kansas Life Sciences Development Inc. and UCL Business PLC (or entities designated by them), respectively. The capital increase shall take place at par value. The value of such new Shares to be issued can in any case not exceed a maximum of USD 2.5 million with a fixed exchange rate of DKK 6.30 per 1 USD based on the average closing price of the Company's Shares on Nasdaq Copenhagen for the 30 days immediately prior to the date of issuance.

The capital increase shall take place at market price and may be effected by cash payment, conversion of debt or by contribution of other assets than cash.Shares issued pursuant to the Board of Directors' authorisations shall be fully paid up, shall be issued in the name of the holder and shall be recorded in the holder's name in the Company's register of shareholders, shall be negotiable instruments and shall in every respect carry the same rights as the existing Shares. The Board of Directors is authorised to lay down the terms and conditions for capital increases pursuant to the above authorisations.

Authorisation to Acquire Treasury Shares

The Board of Directors is authorised in the period until 2 November 2022 to approve the acquisition of treasury shares, on one or more occasions, with a total nominal value of up to 10% of the share capital of the Company from time to time after completion of the Offering, subject to the Company's holding of treasury shares after such acquisition does not exceed 10% of the Company's share capital. The consideration may not deviate more than 10% from the official price quoted on Nasdaq Copenhagen at the time of acquisition.

The Company does not hold any treasury shares as of the date hereof.

General Meetings and Voting Rights

The Company's general meetings shall be held in the Capital Region of Denmark.

The Company's annual general meeting shall be held each year in due time for the audited and approved annual report to be received by the relevant authorities before the applicable statutory time limit. Not later than eight weeks before the contemplated date of the annual general meeting, the Company shall publish the date of the general meeting and the deadline for submitting requests for specific proposals to be included in the agenda.

Extraordinary general meetings shall be held when determined by the Board of Directors or requested by the Company's auditor. Furthermore, the Board of Directors shall convene an extraordinary general meeting within two weeks of receipt of a written request from shareholders representing no less than 5% of the share capital containing specific proposals for the business to be transacted at such extraordinary general meeting.

General meetings shall be convened by the Board of Directors with at least three weeks' and not more than five weeks' notice. The notice shall be published on the Company's website. Furthermore, a notice of the general meeting shall be sent electronically to all shareholders recorded in the Company's register of shareholders who have requested such notice.

In accordance with Danish law, the notice shall specify the time and place of the general meeting and the agenda containing the business to be transacted at the general meeting. If a proposal to amend the Articles of Association is to be considered at the general meeting, the main contents of the proposal shall be specified in the notice.

The Company's general meetings shall be held in English. The Board of Directors may decide to offer simultaneous interpretation into Danish. Documents prepared in connection with or following a general meeting shall be in English and, to the extent required by law or if decided by the Board of Directors, in Danish.

Annual reports shall be prepared in English and, if decided by the Board of Directors, in Danish.

Every shareholder is entitled to have specific business transacted at the general meeting, provided that the shareholder submits a written request to that effect to the Board of Directors not later than six weeks before the date of the general meeting.

The right of a shareholder to attend a general meeting and to vote is determined by the Shares held by the shareholder at the record date. The record date is one week before the general meeting. The Shares held by each shareholder are determined at the record date based on the number of Shares held by that shareholder as registered in the Company's register of shareholders and any notification of ownership received by the Company for the purpose of registration in its register of shareholders, but which have not yet been registered.

At the general meeting each Share of the nominal value of DKK 1 shall carry one vote.

A shareholder who is entitled to attend the general meeting pursuant to the Articles of Association and who wants to attend the general meeting shall request an admission card no later than three days prior to the date of the general meeting. A shareholder may, subject to having requested an admission card in accordance with the Company's Articles of Association, attend in person or by proxy, and the shareholder or the proxy may attend together with an adviser.

The right to vote may be exercised by a written and dated instrument of proxy in accordance with applicable laws. The Board of Directors may be appointed as proxy. A shareholder who is entitled to participate in the general meeting according to the Articles of Association may vote by postal vote in accordance with the Danish Companies Act. Such postal votes shall be received by the Company no later than the business day before the general meeting. Postal votes cannot be withdrawn.

In accordance with Danish law, the notice shall specify the time and place of the general meeting and the agenda containing the business to be transacted at the general meeting. If a proposal to amend the Articles of Association is to be considered at the general meeting, the main contents of the proposal shall be specified in the notice.

Resolutions by the General Meetings and Amendments to the Articles of Association

Resolutions at general meetings shall be passed by a simple majority of votes cast, unless otherwise prescribed by law or by the Articles of Association.

Adoption of changes to the Articles of Association, dissolution of the Company, merger or demerger requires that the resolution is adopted by at least 2/3 of the votes cast as well as the share capital represented at the general meeting, unless applicable laws prescribe stricter or less strict adoption requirements or applicable laws confer specific authority to the Board of Directors or other bodies.

The provisions in the Articles of Association relating to a change of the rights of shareholders or a change to the capital are not more stringent than required by the Danish Companies Act.

Registration of Shares

The Shares will be delivered in book-entry form through allocation to accounts with VP Securities through a Danish bank or other institution authorised as custodian.

The Shares are issued in dematerialised form through VP Securities. The address of VP Securities is Weidekampsgade 14, P.O. Box 4040, 2300 Copenhagen S, Denmark.

The Shares shall be fully paid up, issued in the name of the holder and recorded in the holder's name, in the Company's register of shareholders through the holder's custodian bank. The Company's register of shareholders is kept by VP Services A/S, registration (CVR) no. 30201183.

Transfer of Shares

The Shares are negotiable instruments, and no restrictions under Danish law apply to the transferability of the Shares. See "*Selling Restrictions*" and "*Transfer Restrictions*" for certain restrictions applicable to the Offer Shares.

Preemption Rights

Under Danish law, the Company's shareholders generally have preemption rights if the general meeting of the Company resolves to increase the share capital by cash payment. However, the preemption rights of the shareholders may be derogated by a majority compris-

ing at least 2/3 of the votes cast and of the share capital represented at the general meeting if the share capital increase is made at market price. The Board of Directors is authorised to increase the Company's share capital in one or more issues at market price without preemption rights to the Company's shareholders. See "*—Authorisation to Increase the Share Capital*".

The exercise of preemption rights may be restricted for shareholders resident in certain jurisdictions, including but not limited to the United States, Canada, Japan and Australia, unless the Company decides to comply with applicable local requirements.

The Company intends to evaluate at the time of any issue of Shares subject to preemption rights or in a rights offer, as the case may be, the cost and potential liabilities associated with complying with any local requirements, as well as the indirect benefits to the Company of enabling the exercise of non-Danish shareholders of their preemption rights to Shares or participation in any rights offer, as the case may be, and any other factors considered appropriate at the time, and then to make a decision as to whether to comply with any local requirements. No assurances are given that local requirements will be complied with or that any registration statement would be filed in the U.S. or elsewhere so as to enable the exercise of such holders' preemption rights or participation in any rights offer.

Redemption and Conversion Provisions

Except as provided for in the Danish Companies Act, see "*The Danish Securities Market—Mandatory Redemption of Shares*", no shareholder is under an obligation to have his Shares redeemed in whole or in part by the Company or by any third party, and none of the Shares carry any redemption or conversion rights or any other special rights.

Dissolution and Liquidation

In the event of dissolution and liquidation, the Company's shareholders are entitled to participate in the distribution of assets in proportion to their nominal shareholdings after payment of the Company's creditors.

Indication of Takeover Bids

No takeover offers have been made by any third party in respect of the Shares during the past or current financial year.

The Articles of Association do not contain provisions that are likely to have the effect of delaying, deferring or preventing a change in control of the Company. Consistent with the Corporate Governance Recommendations, the Board of Directors has adopted a set of guidelines for handling of takeover bids.

Disclosure of Information

The Board of Directors has adopted a set of internal rules aiming, inter alia, to ensure that the disclosure of information complies with the applicable stock exchange regulations and rules applicable to the Company's securities listed on regulated markets. All company announcements are published via Nasdaq Copenhagen and can subsequently be accessed from the Company's website. All company announcements will be published in English and, if decided by the Board of Directors, in Danish. The annual report and any interim reports will be prepared in English and, if decided by the Board of Directors, in Danish. Investor presentations and/or telephone conferences are expected to be held following the publication of each interim and annual report to provide participants the opportunity to ask questions to the Executive Management. Audiocasts of such presentations will subsequently be available on the Company's website. Investors may also contact the Company's investor relations department to obtain additional information subject to any restrictions under applicable law.

Taxation

Danish Tax Considerations

The following is a summary of certain Danish income tax considerations relating to an investment in the Shares. The Temporary Purchase Certificates are from a Danish tax perspective evidence of a corresponding Share and each Temporary Purchase Certificate will automatically be exchanged into a Share when the Shares have been listed in the permanent ISIN in VP Securities. The Danish National Tax Board (in Danish: Skatterådet) has in a binding tax ruling of 30 August, 2016, confirmed that a temporary purchase certificate from a tax perspective shall be regarded as a share (and not as a financial instrument). Consequently, the exchange of the Temporary Purchase Certificates into Shares is not regarded as a disposal of shares. Accordingly, the following description of the taxation of Shares is also a description of the taxation of the Temporary Purchase Certificates.

The summary is for general information only and does not purport to constitute exhaustive tax or legal advice. It is specifically noted that the summary does not address all possible tax consequences relating to an investment in the Shares. The summary is based solely upon the tax laws of Denmark in effect on the date of this Offering Circular. Danish tax laws may be subject to change, possibly with retroactive effect. The summary does not cover investors to whom special tax rules apply and, therefore, may not be relevant, for example, to investors subject to the Danish Act on Pension Investment Return Taxation (i.e., pension savings), professional investors, certain institutional investors, insurance companies, pension companies, banks, stockbrokers and investors with tax liability on return on pension investments. The summary does not cover taxation of individuals and companies who carry on a business of purchasing and selling shares. Sales are assumed to be sales to a third party.

Potential investors in the Shares are advised to consult their tax advisers regarding the applicable tax consequences of acquiring, holding and disposing of the Shares based on their particular circumstances. Investors who may be affected by the tax laws of other jurisdictions should consult their tax advisers with respect to the tax consequences applicable to their particular circumstances, as such consequences may differ significantly from those described herein.

Taxation of Danish tax resident shareholders

Sales of shares—individuals

Gains from the sale of shares are taxed as share income at a rate of 27% on the first DKK 51,700 in 2017 (for cohabiting spouses, a total of DKK 103,400) and at a rate of 42% on share income exceeding DKK 51,700 (for cohabiting spouses over DKK 103,400). Such amounts are subject to annual adjustments and include all share income (i.e., all capital gains and dividends derived by the individual or cohabiting spouses, respectively).

Gains and losses on the sale of shares admitted to trading on a regulated market are calculated as the difference between the purchase price and the sale price. The purchase price is generally determined using the average method which means that each share is considered acquired at a price equivalent to the average acquisition price of all the shareholder's shares in the issuing company.

Losses on the sale of shares admitted to trading on a regulated market can only be offset against other share income deriving from shares admitted to trading on a regulated market (i.e., received dividends and capital gains on the sale of shares admitted to trading on a regulated market). Unused losses will be offset against a cohabiting spouse's share income deriving from shares admitted to trading on a regulated market. Any remaining losses after the above deduction can be carried forward indefinitely and offset against future share income deriving from shares admitted to trading on a regulated market.

Losses on shares admitted to trading on a regulated market may only be set off against gains and dividends on other shares admitted to trading on a regulated market if the Danish Tax Authorities have received certain information concerning the ownership of the shares. This information is normally provided to the Danish Tax Authorities by the securities dealer.

Sale of shares—companies

Tax on the sale of shares by companies is subject to different regimes depending on whether the shares are considered as Subsidiary Shares, Group Shares, Tax-Exempt Portfolio Shares or Taxable Portfolio Shares.

"**Subsidiary Shares**" are generally defined as shares owned by a shareholder holding at least 10% of the nominal share capital of the issuing company.

"**Group Shares**" are generally defined as shares in a company in which the shareholder of the company and the issuing company are subject to Danish tax consolidation or fulfil the requirements for international tax consolidation under Danish law.

"**Tax-Exempt Portfolio Shares**" are generally defined as shares not admitted to trading on a regulated market owned by a shareholder holding less than 10% of the nominal share capital in the issuing company. Tax-Exempt Portfolio Shares are not relevant in respect of this Offering and will not be described in further detail.

"**Taxable Portfolio Shares**" are shares that do not qualify as Subsidiary Shares, Group Shares or Tax-Exempt Portfolio Shares.

Gains or losses on disposal of Subsidiary Shares and Group Shares are not included in the taxable income of the shareholder.

Special rules apply in order to prevent avoidance of the 10% ownership requirement through pooling of shareholdings in a holding company. These rules will not be described in further detail.

Capital gains from the sale of Taxable Portfolio Shares are taxable at the corporate income tax rate of 22%. Losses on such shares are generally deductible. Gains and losses on Taxable Portfolio Shares are, as a general rule, calculated in accordance with the mark-to-market principle. According to the mark-to-market principle, each year's taxable gain or loss is calculated as the difference between the market value of the shares at the beginning and end of the tax year. Thus, taxation will take place on an accrual basis even if no shares have been disposed of and no gains or losses have been realised. If the Taxable Portfolio Shares are sold or otherwise disposed of before the end of the income year, the taxable income of that income year equals the difference between the value of the Taxable Portfolio Shares at the beginning of the income year and the value of the Taxable Portfolio Shares at realisation. If the Taxable Portfolio Shares have been acquired and realised in the same income year, the taxable income equals the difference between the acquisition sum and the realisation sum. If the Taxable Portfolio Shares are acquired in the income year and not realised in the same income year, the taxable income equals the difference between the acquisition sum and the value of the Shares at the end of the income year.

A change of status from Subsidiary Shares/Group Shares to Taxable Portfolio Shares (or vice versa) is for tax purposes deemed to be a disposal of the shares and a reacquisition of the shares at market value at the time of change of status.

Dividends—individuals
Dividends received by individuals who are tax residents of Denmark are taxed as share income. Share income is taxed at a rate of 27% on the first DKK 51,700 in 2017 (for cohabiting spouses, a total of DKK 103,400) and at a rate of 42% on share income exceeding DKK 51,700 (for cohabiting spouses over DKK 103,400). Such amounts are subject to annual adjustments and include all share income (i.e., all capital gains and dividends derived by the individual or cohabiting spouses, respectively).

Dividends paid to individuals are generally subject to 27% withholding tax.

Dividends—companies
Dividends received on Taxable Portfolio Shares are subject to the standard corporate tax rate of 22% irrespective of ownership period.

The withholding tax rate is 22%. If the distributing company withholds a higher amount, the shareholder can claim a refund of the excess tax. A claim for repayment must be filed within two months; otherwise the excess tax will instead be credited in the corporate income tax for the year.

Dividends received on Subsidiary Shares and Group Shares will not be subject to taxation irrespective of ownership period.

Taxation of shareholders residing outside Denmark

Sales of shares—individuals and companies
Shareholders not resident in Denmark will normally not be subject to Danish tax on any gains realised on the sale of shares, irrespective of the ownership period. Where a non-resident of Denmark holds Taxable Portfolio Shares which can be attributed to a permanent establishment in Denmark, such gains are taxable pursuant to the rules applicable to Danish tax residents as described above.

Dividends—individuals
Under Danish law, dividends paid in respect of shares are generally subject to Danish withholding tax at a rate of 27%. A request for a refund of Danish withholding tax can however be made by the shareholder in the following situations:

1) Double Taxation Treaty

In the event that the dividend receiving individual is a resident of a state having a double taxation treaty with Denmark, the shareholder may claim a refund of the tax amount exceeding the treaty rate, through certain application procedures, from the Danish tax authorities.

Denmark has executed double taxation treaties with approximately 75 countries, including almost all members of the EU. The double taxation treaties generally provide for a 15% withholding tax rate. The refund is sought by filling out an online form on The Danish Tax Authorities' website. The shareholder must be able to document that the Danish dividend has been received, that the Danish dividend tax has been withheld and the Danish dividend tax exceeds the final taxation in accordance with the double taxation treaty with the shareholder's resident country or under current Danish tax law.

Note that refund of withholding tax as described in this section requires a certification by the applicable local tax authority.

2) Credit under Danish law

In addition, if the shareholder holds less than 10% of the nominal share capital of the company and the shareholder is a tax resident in a state which has a double taxation treaty or an international agreement, convention or other administrative agreement on assistance in tax matters according to which the competent authority in the state of the shareholder is obliged to exchange information with Denmark, dividends are subject to tax at a reduced rate of 15%. If the shareholder is a tax resident outside the EU, it is an additional requirement for eligibility for the 15% tax rate that the shareholder together with related shareholders holds less than 10% of the nominal share capital of the company. Note that the reduced tax rate does not affect the withholding rate. Thus, the shareholder must also in this situation claim a refund as described above in order to benefit from the reduced rate.

Note that refund of withholding tax as described in this section requires a certification by the applicable local tax authority.

Where a non-resident of Denmark holds shares which can be attributed to a permanent establishment in Denmark, dividends are taxable pursuant to the rules applicable to Danish tax residents described above. See "*—Taxation of Danish tax resident shareholders*".

Dividends—companies

Dividends from Subsidiary Shares are exempt from Danish withholding tax provided the taxation of the dividends is to be waived or reduced in accordance with the Parent Subsidiary Directive (2011/96/EU as amended 2015/121/EU) or in accordance with a tax treaty with the jurisdiction in which the company investor is resident.

Dividends from Group Shares are exempt from Danish withholding tax provided the company investor is a resident of the EU or the EEA and the taxation of dividends should have been waived or reduced in accordance with the Parent Subsidiary Directive (2011/96/EU as amended 2015/121/EU) or in accordance with a tax treaty with the country in which the company investor is resident had the shares been Subsidiary Shares.

Dividend payments on Taxable Portfolio Shares are subject to a Danish withholding tax rate of 27% irrespective of ownership period. A request for a refund of Danish withholding tax can however be made by the shareholder in the following situations:

1) All foreign corporate shareholders

All foreign corporate shareholders can claim a refund from the Danish tax authorities of the tax amount exceeding 22%.

2) Double Taxation Treaty

In the event that the dividend receiving company is a resident of a state with which Denmark has entered into a double taxation treaty, the shareholder may claim a refund from the Danish tax authorities of the tax amount exceeding the treaty rate, through certain certification procedures. Denmark has executed double taxation treaties with approximately 75 countries, including the United States and almost all members of the EU. The double taxation treaties generally provide for a 15% withholding tax rate. The refund is sought by filling out an online form on The Danish Tax Authorities' website. The shareholder must be able to document that the Danish dividend has been received, that the Danish dividend tax has been withheld and the Danish dividend tax exceeds the final taxation in accordance with the double taxation treaty with the shareholder's resident country or under current Danish tax law.

Note that the refund of withholding tax as described in this section requires a certification by the applicable local tax authority.

3) Credit under Danish law

In addition, if the shareholder holds less than 10% of the nominal share capital of the company and the shareholder is a tax resident in a state which has a double taxation treaty or an international agreement, convention or other administrative agreement on assistance in tax

matters according to which the competent authority in the state of the shareholder is obliged to exchange information with Denmark, dividends are subject to tax at a reduced rate of 15%. If the shareholder is a tax resident outside the EU, it is an additional requirement for eligibility for the 15% tax rate that the shareholder together with related shareholders holds less than 10% of the nominal share capital of the company. Note that the reduced tax rate does not affect the withholding rate. Thus, the shareholder must also in this situation claim a refund as described above in order to benefit from the reduced rate.

Note that refund of withholding tax as described in this section requires a certification by the applicable local tax authority. Where a non-resident of Denmark holds shares which can be attributed to a permanent establishment in Denmark, dividends are taxable pursuant to the rules applicable to Danish tax residents described above. (see "*—Taxation of Danish tax resident shareholders*").

Share transfer tax and stamp duties
No Danish share transfer tax or stamp duties are payable on transfer of the shares.

Withholding tax obligations
An issuer of shares is subject to Danish withholding tax obligations in accordance with applicable Danish laws.

The Offering

Joint Global Coordinators

The Offering is being arranged by Carnegie Investment Bank, filial af Carnegie Investment Bank AB (publ), Sverige and Danske Bank A/S in their capacity as Joint Global Coordinators and Joint Bookrunners and Oddo as Co-Lead Manager.

The Offering

The Offering consists of: (i) an initial public offering to retail and institutional investors in Denmark and (ii) private placements to institutional investors in in certain other jurisdictions (excluding the United States). The Offering outside the United States will be made in compliance with Regulation S under the U.S. Securities Act.

The Company is offering up to 10,781,250 new Shares in order to raise gross proceeds of DKK 690 million (hereof DKK 90 million pursuant to the Overallotment Option). Assuming completion of the Offering, the Company's registered share capital will increase by a nominal value of DKK 9,375,000 as a result of the issue of Offer Shares (exluding the Overallotment Option). The exact number of Offer Shares will be determined based on a book-building process. The existing shareholders of Orphazyme will not be offering any existing Shares as part of the Offering.

Skandinaviska Enskilda Banken, Danmark, branch of Skandinaviska Enskilda Banken AB (Publ.), Sweden; Vækstfonden; BI Asset Management Fondsmæglerselskab A/S on behalf of certain clients; Handelsbanken, branch of Svenska Handelsbanken AB (Publ.), Sweden; and Spar Nord Bank A/S have in connection with the Offering subject to certain conditions undertaken to subscribe for Offer Shares as Cornerstone Investors for a total subscription amount of DKK 230 million, corresponding to approximately 38.3% of the Offering (excluding the Overallotment Option). The commitments undertaken by the Cornerstone Investors are subject to certain conditions, e.g. that the Offer Price is set within the Offer Price Range and there being no significant new factor, material mistake or inaccuracy in the information contained in this Offering Circular capable of affecting the Cornerstone Investors' assessment of the Shares and which would be required to be mentioned in a supplement to the Offering Circular under the Danish Securities Trading Act.

The Company has granted the Joint Global Coordinators, on behalf of the Managers, an Overallotment Option to subscribe for up to 1,406,250 Overallotment Shares (new Shares) at the Offer Price, exercisable, in whole or in part, from the date of Admission until 30 calendar days thereafter, solely to cover overallotments or short positions, if any, incurred in connection with the Offering. The number of Overallotment Shares will be adjusted if less than the maximum number of Offer Shares (other than the Overallotment Shares) are subscribed for in the Offering, such that the number of Overallotment Shares will not exceed 15% of the number of Offer Shares (other than Overallotment Shares). See "*Plan of Distribution*".

Offer Price

The Offer Price will be determined through a book-building process. Book-building is a process in which the Joint Global Coordinators, prior to the final pricing of the Offering, determine the Offer Price by collecting expressions of interest in the Offer Shares from potential institutional investors. Following the book-building process, the Offer Price will be determined by the Company in consultation with the Joint Global Coordinators. The Offer Price is expected to be announced through Nasdaq Copenhagen no later than 8:00 a.m. (CET) on 17 November 2017.

The Offer Price is free of brokerage charges and is expected to be between DKK 64 and DKK 80 per Offer Share. This indicative Offer Price Range has been set by the Company following consultation with the Joint Global Coordinators, taking into account, among other things, the Company's historic and projected revenue and earnings, the Company's objective to establish an orderly after market in the Offer Shares and prevailing market conditions.

It is currently expected that the Offer Price will be set within the Offer Price Range. If the Offer Price Range is adjusted, the Company will make an announcement through Nasdaq Copenhagen and publish a supplement to this Offering Circular. Following publication of such supplement, investors who have submitted orders to purchase Offer Shares in the Offering will have two trading days to withdraw their purchase offer. In such an event, the announcement of the Offer Price will not be published until the period for exercising such withdrawal rights has ended. See also "*—Investors' Withdrawal Rights*".

Offer Period

The Offer Period will commence on 6 November 2017 and will close no later than 16 November at 12:00 p.m. (noon) (CET). The Offer

Period may be closed prior to 16 November 2017; however, the Offer Period will not be closed in whole or in part before 15 November 2017 at 00:01 a.m. (CET). If the Offering is closed before 16 November 2017, the announcement of the Offer Price, allocation and the Admission may be moved forward accordingly. The Offer Period in respect of applications for purchases of amounts up to, and including, DKK 3 million may be closed before the remainder of the Offering is closed. Any such earlier closing, in whole or in part, will be announced through Nasdaq Copenhagen.

Submission of Bids

Applications to purchase or subscribe for amounts of up to and including DKK 3 million
Applications by Danish investors to purchase or subscribe for amounts of up to and including DKK 3 million should be made by submitting the application form enclosed in the Offering Circular to the investor's own account-holding bank during the Offer Period or such shorter period as may be announced through Nasdaq Copenhagen. Applications are binding and cannot be altered or cancelled. Bids may be made at a maximum price per Offer Share in Danish kroner. If the Offer Price exceeds the maximum price per Offer Share stated in the application form, then no Temporary Purchase Certificates or Offer Shares will be allocated to the investor. Where no maximum price per share has been indicated, applications will be deemed to be made at the Offer Price. All applications made at a price equivalent to the Offer Price, or a higher price, will be settled at the Offer Price following allotment, if any. Applications should be made for a number of Temporary Purchase Certificates representing Offer Shares or for an aggregate amount rounded to the nearest Danish kroner amount. Only one application will be accepted from each account in VP Securities. For binding orders, the application form must be submitted to the investor's own account-holding bank in complete and executed form in due time to allow the investor's own account-holding bank to process and forward the application to ensure that it is in the possession of Danske Bank A/S, no later than 12:00 p.m. (noon) (CET) on 16 November 2017, or such earlier time at which the Offering is closed.

Applications to purchase or subscribe for amounts of more than DKK 3 million
Investors who wish to apply to purchase or subscribe for amounts of more than DKK 3 million can indicate their interest to one or more of the Managers during the Offer Period. During the Offer Period, such investors can continuously change or withdraw their declarations of interest, but these declarations of interest become binding applications at the end of the Offer Period. Immediately following the determination of the Offer Price, investors will be allocated a number of Temporary Purchase Certificates representing Offer Shares at the Offer Price within the limits of the investor's most recently submitted or adjusted declaration of interest. All applications made at a price equivalent to the Offer Price, or a higher price, will be settled at the Offer Price following allotment, if any.

Minimum and Maximum Subscription Amounts

The minimum subscription/purchase amount is one Offer Share. No maximum subscription amount applies to the Offering. However, the number of shares is limited to the number of Offer Shares in the Offering (including the Offer Shares pursuant to the Overallotment Option, if exercised).

Allocation and Reduction

In the event that the total amount of shares applied for in the Offering exceeds the number of Offer Shares, reductions will be made as follows:

- With respect to applications for amounts of up to and including DKK 3 million, reductions will be made mathematically.
- With respect to applications for amounts of more than DKK 3 million, individual allocations will be made. The Joint Global Coordinators will allocate the Offer Shares after agreement upon such allocations with the Company.
- Up to 3,593,750 Offer Shares will be reserved for the Cornerstone Investors to subscribe for at the Offer Price in connection with the Offering.
- Up to 93,750 Offer Shares will be reserved for certain members of the Board of Directors to subscribe for in connection with the Offering at the Offer Price.
- Up to 21,875 Offer Shares will be reserved for the Executive Management and Key Employees to subscribe for at the Offer Price as an investment in connection with Orphazyme's long-term incentive programme.
- Up to 93,750 Offer Shares will be reserved for Orphazyme's employees to subscribe for in connection with the Offering at the Offer Price.

It is expected that the result of the Offering, the Offer Price and the basis of the allocation will be announced through Nasdaq Copenhagen no later than 8:00 a.m. (CET) on 17 November 2017. If the Offer Period is closed before 16 November 2017, announcement of the Offer Price and allocation will be brought forward accordingly.

Following the expiration of the Offer Period, investors will receive a statement indicating the number of Temporary Purchase Certificates representing Offer Shares allocated, if any, and the equivalent value at the Offer Price unless otherwise agreed between the investor and the relevant account-holding bank.

Orders as well as indications of interest may not result in an allocation of Offer Shares.

If the total applications in the Offering exceed the number of Offer Shares, a reduction will be made. In such event, the Joint Global Coordinators reserve the right to require documentation to verify that each application relates to a single account in VP Securities. Further, the Joint Global Coordinators reserve the right to require documentation to verify the authenticity of all orders, to demand the name of each purchaser, to pass on such information to the Company and the Main Shareholders, and to make individual allocations if there are several orders that are determined to have originated from the same investor or group of investors.

Authorisation

The Board of Directors passed a resolution on 6 November 2017 pursuant to the authorisation given to the Board of Directors at an extraordinary general meeting (see "*Description of the Shares and Share Capital—Authorisation to Increase the Share Capital*"), to increase the Company's share capital by a minimum of 7,500,000 new Shares with a total nominal value of DKK 7,500,000 and a maximum of up to 9,375,000 new Shares with a total nominal value of DKK 9,375,000. The capital increase will be made by cash payment and without preemption rights to the Company's existing shareholders.

On the same date, the Board of Directors passed a resolution pursuant to the authorisation given to the Board of Directors at an extraordinary general meeting to increase the Company's share capital by a maximum of up to 1,406,250 new Shares with a total nominal value of DKK 1,406,250 (in case the Overallotment Option is exercised in full). The capital increase will be made by cash payment and without preemption rights to the Company's existing shareholders.

For a description of the dilution effects on existing Shares in connection with the Offering, see "*Ownership Structure and Main Shareholders*".

Trading and Official Listing on Nasdaq Copenhagen

Application has been made for the Temporary Purchase Certificates to be admitted to trading on Nasdaq Copenhagen under the symbol "ORPHA TEMP" and for the Shares to be admitted to trading and official listing under the symbol "ORPHA" on Nasdaq Copenhagen. The Admission is subject to, among other things, Nasdaq Copenhagen's approval of the distribution of the Offer Shares, the Offering not being withdrawn prior to the settlement of the Offering and the Company making an announcement to that effect.

The first day of trading and official listing on Nasdaq Copenhagen is expected to be 17 November 2017 subject to the Offering not being withdrawn prior to settlement and completion of the Offering. The first day of trading of the Temporary Purchase Certificates on Nasdaq Copenhagen is expected to be 17 November 2017 under the temporary ISIN, and the last day of trading of the Temporary Purchase Certificates on Nasdaq Copenhagen is expected to be 21 November 2017. The Shares are expected to be admitted to trading and official listing on Nasdaq Copenhagen under the permanent ISIN on 22 November 2017. If the Offering is closed before 16 November 2017, the Admission, the Settlement Date, the delivery of Temporary Purchase Certificates, the automatic exchange of Temporary Purchase Certificates for Shares and the first day of trading and official listing of the Shares on Nasdaq Copenhagen may be moved forward accordingly.

Payment for and settlement of the Offer Shares are expected to take place on or around 21 November 2017 by way of delivery of Temporary Purchase Certificates. Upon completion of the Offering and after payment for the Temporary Purchase Certificates representing the Offer Shares, the capital increase relating to the new Shares to be issued by the Company pursuant to the Offering will be registered with the Danish Business Authority, which is expected to take place on the Settlement Date.

Subject to completion of the Offering and registration of the new Offer Shares with the Danish Business Authority, the Temporary Purchase Certificates will automatically be exchanged in VP Securities for a corresponding number of Shares, which are expected to be delivered on 23 November 2017. In connection with the Temporary Purchase Certificates being automatically exchanged for Shares, the Temporary Purchase Certificates will cease to exist. The existing shareholders have undertaken an obligation not to trade in the existing Shares until the Temporary Purchase Certificates have been exchanged for Shares and the Shares have been admitted to trading on Nasdaq Copenhagen.

If the Offering is not completed, no Temporary Purchase Certificates or Offer Shares will be delivered to investors. Consequently, any trades in the Temporary Purchase Certificates or the Shares effected on or off the market before settlement of the Offering may subject investors to liability for not being able to deliver the Temporary Purchase Certificates or the Shares sold and investors who have sold or acquired Temporary

Purchase Certificates or Shares on or off the market may incur a loss. Any such dealings will be at the sole risk of the parties concerned.

If the Offering is terminated or withdrawn, the Offering and any associated arrangements will lapse, all submitted orders will be automatically cancelled, any monies received in respect of the Offering will be returned to the investors without interest (less any transaction costs) and admission to trading and/or official listing of the Temporary Purchase Certificates or the Shares on Nasdaq Copenhagen will be cancelled. Consequently, any trades in the Temporary Purchase Certificates and/or Shares effected on or off the market before settlement of the Offering may subject investors to liability for not being able to deliver the Temporary Purchase Certificates and/or Shares sold, and investors who have sold or acquired Temporary Purchase Certificates and/or Shares on or off the market may incur a loss. All dealings in the Temporary Purchase Certificates and/or Offer Shares prior to settlement of the Offering are for the account of, and at the sole risk of, the parties concerned.

Identification

Permanent ISIN for the Shares: DK0060910917

Temporary ISIN for the Temporary Purchase Certificates: DK0060911055

The temporary ISIN code will be used for the settlement of Temporary Purchase Certificates representing the Offer Shares in VP Securities and on Clearstream and Euroclear in connection with the Offering.

Nasdaq Copenhagen Symbol for the Shares: "ORPHA"

Nasdaq Copenhagen Symbol for the Temporary Purchase Certificates: "ORPHA TEMP"

Share Lending Agreement

Novo Holdings has agreed with the Joint Global Coordinators that Novo Holdings will make available up to 1,406,250 existing Shares for purposes of delivery of the Offer Shares to investors in connection with the Overallotment Option. The Company has agreed to issue up to a corresponding number of new Shares that the Joint Global Coordinators will subscribe for and redeliver to Novo Holdings.

Registration and Settlement

The Temporary Purchase Certificates and Offer Shares will be registered in book-entry form electronically with VP Securities, Weidekampsgade 14, P.O. Box 4040, 2300 Copenhagen S, Denmark. All Temporary Purchase Certificates and Shares are registered on accounts with account-holding banks in VP Securities. Investors that are not residents of Denmark may use a Danish bank directly or their own bank's Danish correspondent bank as their account-holding bank or arrange for registration and settlement through Clearstream, 42 Avenue JF Kennedy, L-1855 Luxembourg, Luxembourg, or Euroclear, 1, Boulevard du Roi Albert II, B-1210 Brussels, Belgium.

Payment for and settlement of the Offer Shares are expected to take place on 21 November 2017 (i.e., the Settlement Date), by way of delivery of Temporary Purchase Certificates against payment in immediately available funds in Danish kroner in book-entry form to investors' accounts with VP Securities and through the facilities of Euroclear and Clearstream.

Subject to completion of the Offering and registration of the new Offer Shares with the Danish Business Authority, the Temporary Purchase Certificates will automatically be exchanged for a corresponding number of Shares, which are expected to be delivered two business days after the Settlement Date in book-entry form to the holder of the Temporary Purchase Certificates' account with VP Securities and through the facilities of Euroclear and Clearstream. If the Offering is closed before 21 November 2017 (i.e. the Settlement Date), the delivery of Temporary Purchase Certificates, the automatic exchange of Temporary Purchase Certificates for Shares and the first day of trading and official listing of the Shares on Nasdaq Copenhagen may be moved forward accordingly.

The account-holding bank will normally send a statement to the name and address registered in VP Securities showing the number of Temporary Purchase Certificates representing Offer Shares purchased or subscribed for by the investor unless otherwise agreed between the investor and the relevant account-holding bank. This statement also constitutes evidence of the investor's holding.

All dealings in the Temporary Purchase Certificates and/or the Offer Shares prior to settlement of the Offering will be for the account of, and at the sole risk of, the parties involved.

Withdrawal of the Offering

Completion of the Offering is conditional upon the Offering not being withdrawn. The Offering may be withdrawn by the Company and the Joint Global Coordinators at any time before pricing and allocation of the Offering take place. The Offering may also be withdrawn if Nasdaq Copenhagen is not satisfied that there will be a sufficiently broad distribution of the Shares to investors or if, for other reasons, the Temporary Purchase Certificates or the Shares cannot be admitted for trading and/or official listing on Nasdaq Copenhagen.

The Underwriting Agreement (as defined in "*Glossary*") contains a provision entitling the Joint Global Coordinators to terminate the Offering (and the arrangements associated with it) at any time prior to settlement of the Offering by delivery, and payment for the Temporary Purchase Certificates representing the Offer Shares expected on or around 21 November 2017 (including after Admission) in certain circumstances, including force majeure and material changes in the financial condition of the Company's business.

The termination rights of the parties to the Underwriting Agreement will lapse upon settlement of the Offering, currently expected to take place on 21 November 2017, except in respect of the Overallotment Shares. The termination rights of the parties to the Underwriting Agreement shall lapse, in respect of the Overallotment Shares, upon settlement of the sale of the Overallotment Shares, if the Overallotment Option is exercised.

Nasdaq Copenhagen's approval of the Admission on Nasdaq Copenhagen is subject to such termination rights not being exercised after pricing and prior to settlement of the Offering (excluding any termination rights in respect of the Overallotment Option).

The Underwriting Agreement contains closing conditions which the Company believes are customary for offerings such as the Offering. In addition, the Company has given usual representations and warranties to the Joint Global Coordinators and the Joint Bookrunners. The completion of the Offering is dependent on compliance with all of the closing conditions set forth in the Underwriting Agreement. If one or more closing conditions are not met, the Joint Global Coordinators and the Joint Bookrunners may, at their discretion, withdraw the Offering. If the Offering is terminated or withdrawn: the Offering and any associated arrangements will lapse, all submitted orders will be automatically cancelled, any monies received in respect of the Offering will be returned to the investors without interest (less any transaction costs) and admission to trading and/or official listing of the Temporary Purchase Certificates or the Shares on Nasdaq Copenhagen will be cancelled. All dealings in the Temporary Purchase Certificates and/or Offer Shares prior to settlement of the Offering are for the account of, and at the sole risk of, the parties concerned.

Any withdrawal of the Offering will be announced immediately through Nasdaq Copenhagen.

Investors' Withdrawal Rights

In the event that the Company is required to publish a supplement to this Offering Circular, between the date of publication of this Offering Circular and Admission, investors who have submitted orders to purchase Offer Shares in the Offering shall have two trading days following the publication of the relevant supplement within which the investors can withdraw their offer to subscribe for or purchase Offer Shares in the Offering in its entirety. The right to withdraw an application to subscribe for or purchase Offer Shares in the Offering in these circumstances will be available to all investors in the Offering, provided the obligation to publish a supplement to this Offering Circular was triggered before the Admission and provided no Offer Shares have been delivered. If the order is not withdrawn within the stipulated period any order to purchase Offer Shares in the Offering will remain valid and binding.

Costs of the Offering

The total expenses in relation to the Offering, including commissions and fees (fixed and discretionary) payable by the Company to the Managers, other advisor fees and expenses, are estimated to be approximately DKK 55 million (assuming full exercise of the Overallotment Option).

Further, the Company has agreed to pay a selling commission to account-holding banks (unless such account-holding bank is a Manager) equivalent to 0.25% of the Offer Price of the Offer Shares that are allocated in respect of orders of up to and including DKK 3 million submitted through the account-holding banks (except for the Managers) to be paid by the Company based on the number of Offer Shares that are sold).

None of the Company or the Managers will charge expenses to investors. Investors will have to bear customary transaction and handling fees charged by their account-holding banks.

Selling Agents for the Offering

Carnegie Investment Bank, filial af Carnegie Investment Bank AB (publ), Sverige
Overgaden neden Vandet 9B
DK-1414 Copenhagen K
Denmark

and

Danske Bank A/S
CVR no. 61126228
Holmens Kanal 2-12
DK-1092 Copenhagen K
Denmark

A request for copies of the Offering Circular may be submitted by persons who satisfy the requirements of the applicable selling restrictions from:

Carnegie Investment Bank, filial af Carnegie Investment Bank AB (publ), Sverige
Overgaden neden Vandet 9B
DK-1414 Copenhagen K
Denmark
e-mail: prospekter@carnegie.dk

and

Danske Bank A/S
CVR no. 61126228
Holmens Kanal 2-12
DK-1092 Copenhagen K
Denmark
e-mail: prospekter@danskebank.dk

In addition, the Offering Circular is available, subject to certain restrictions, on the Company's website (www.orphazyme.com). Information included on the Company's website does not form part of and is not incorporated into this Offering Circular.

The distribution of this Offering Circular and the offer or sale of the Offer Shares in certain jurisdictions is restricted by law. Persons possessing this Offering Circular are required by the Company, the Main Shareholders and the Managers to inform themselves about and to observe any restrictions. This Offering Circular does not constitute an offer to sell or a solicitation of an offer to buy or subscribe for any of the Offer Shares in any jurisdiction to any person to whom it would be unlawful to make such an offer in such jurisdiction.

Interests of Natural and Legal Persons Involved in the Offering

As described in "*Board of Directors, Executive Management and Key Employees—Statement on Conflicts of Interest*" and in "*Ownership Structure and Main Shareholders*", certain members of the Company's Board of Directors as well as the Executive Management and Key Employees are, or will upon exercise of Pre-IPO Warrants become, direct or indirect shareholders in the Company or hold economic interests therein and therefore have an interest in the Offering. No member of the Board of Directors or Executive Management or any of the Key Employees, directly or indirectly, hold more than 5% of the Company's share capital.

The Executive Management, Key Employees and certain members of the Board of Directors hold Pre-IPO Warrants granted by the Company as part of its pre-IPO warrant programme. These Pre-IPO Warrants may be exercised in connection with the Offering and, accordingly, the Executive Management, Key Employees and certain members of the Board of Directors have an interest in the Offering.

Skandinaviska Enskilda Banken, Danmark, branch of Skandinaviska Enskilda Banken AB (Publ.), Sweden; Vækstfonden; BI Asset Management Fondsmæglerselskab A/S on behalf of certain clients; Handelsbanken, branch of Svenska Handelsbanken AB (Publ.), Sweden; and Spar Nord Bank A/S have in connection with the Offering subject to certain conditions undertaken to subscribe for Offer Shares as Cornerstone Investors and, accordingly, will have a direct economic interest in the Offering.

The Managers and their respective affiliates have engaged in transactions with and performed various commercial banking, investment banking, financial advisory and other services for Orphazyme, and the Managers and their respective affiliates are currently providing and may in the future provide such services for Orphazyme. With respect to certain of these transactions and services, the sharing of information is restricted for reasons of confidentiality, internal procedures or applicable rules and regulations. The Managers have received and will receive customary fees and commissions for these transactions and services and may come to have interests that may not be aligned or could potentially conflict with potential investors' and the Company's interests. Danske Bank A/S is expected to also be a lender under a loan facility in favour of the Executive Management and Key Employees with respect to their subscription for Investment Shares (as defined herein) in connection with Orphazyme's long-term incentive programme.

See also "*Plan of Distribution*" for a description of certain interests of the Managers in the Offering.

The Company is not aware of any other potential interest of natural or legal persons involved in the Offering who may have a material interest in the Offering. See also "*Plan of Distribution*" for a description of certain interests of the Managers in the Offering.

Governing Law

The Shares are issued in accordance with Danish law.

The Danish Securities Market

Set forth below is a summary of certain information concerning the Danish securities market including information on certain provisions of Danish law and Danish securities market regulations in effect on the date of the Offering Circular. Such summary is qualified in its entirety by reference to the applicable Danish law and securities market regulations.

Nasdaq Copenhagen

Nasdaq Copenhagen is a company incorporated and organised under the laws of Denmark. Trading on Nasdaq Copenhagen is conducted by authorised firms, which include major Danish banks and other securities brokers, as well as certain mortgage credit institutions and the Danish Central Bank (Danmarks Nationalbank).

The trading system for equities trading in Denmark on Nasdaq Copenhagen operates between 9:00 a.m. and 4:55 p.m. (CET) on weekdays. After the end of the continuous trading there is a pre-closing call between 4:55 p.m. to 5:00 p.m. (CET). An after trade "post trade" session exists from 5:00 p.m. to 5:20 p.m. (CET). Before the continuous trading begins, there is a second after trade "pre-open" session from 8:00 a.m. to 9:00 a.m. and a morning call session from 8:45 a.m. to 9:00 a.m. (CET) for the purpose of establishing fair opening prices. After the opening prices have been presented, the continuous trading begins.

Registration Process

In connection with an initial public offering, a company's shares are registered in book-entry form on accounts maintained in the computer system of VP Securities, which acts as an electronic central record of ownership and as the clearing centre for all transactions in Denmark. The address of VP Securities is Weidekampsgade 14, P.O. Box 4040, DK-2300 Copenhagen S, Denmark.

Danish financial institutions, such as banks, are authorised to keep accounts for each specific investor with VP Securities, including for Euroclear and Clearstream. All Danish shares listed on Nasdaq Copenhagen are dematerialised, "non-certificated" and registered at VP Securities. The account is maintained through an account-holding bank.

The account-holding bank has the exclusive right to make transactions and registrations on these accounts on behalf of its customers.

Shares shall be registered in the name of the holder through the account-holding bank.

Nominees

An account may be kept on behalf of one or more owners, meaning that a shareholder may appoint a nominee.

A nominee shareholder is entitled to receive dividends and to exercise all subscription and other financial and administrative rights attached to the shares held in its name with VP Securities. The relationship between the nominee shareholder and the beneficial owner is regulated solely by an agreement between the parties, and the beneficial owner must disclose its identity if any of the aforementioned rights are to be exercised directly by the beneficial owner.

The right to appoint a nominee does not eliminate a shareholder's obligation to notify the Company and the Danish Financial Supervisory Authority (the "**Danish FSA**") of a major shareholding. See "*—Disclosure of Major Shareholdings*" below.

Settlement Process

Settlement in connection with trading on Nasdaq Copenhagen normally takes place on the second business day after effecting a sale or purchase transaction. On behalf of VP Securities, the account-holding bank sends a statement to the name and address recorded in VP Securities, showing the amount of shares held in that name, which provides the holder with evidence of its rights. Settlement can also take place through the clearing facilities of Euroclear and Clearstream.

Disclosure of Major Shareholdings

Holders of shares in Danish companies with shares admitted to trading and official listing on Nasdaq Copenhagen are, pursuant to Section 29 of the Danish Securities Trading Act, required to give simultaneous notice to the company and the Danish FSA of the sharehold-

ing in the company, immediately, when the shareholding reaches, exceeds or falls below thresholds of 5%, 10%, 15%, 20%, 25%, 50% or 90% and limits of one-third or two-thirds of the voting rights or nominal value of the total share capital.

Holders of shares in a company mean a natural or legal person who, directly or indirectly, holds: (i) shares in the company on behalf of himself and for his own account; (ii) shares in the company on behalf of himself, but for the account of another natural or legal person; or (iii) share certificates, where such holder is considered a shareholder in relation to the underlying securities represented by the certificate.

The duty to notify set forth above further applies to natural and legal persons who are entitled to acquire, sell or exercise voting rights which are:

(i) held by a third party with whom that natural or legal person has concluded an agreement, which obliges them to adopt, by concerted exercise of the voting rights they hold, a lasting common policy towards the management of the issuer in question (common duty to inform for all parties to the agreement);
(ii) held by a third party under an agreement concluded with that natural or legal person providing for the temporary transfer of the voting rights in question in return for consideration;
(iii) attached to shares which are lodged as collateral for that natural or legal person, provided the person controls the voting rights and declares an intention of exercising them;
(iv) attached to shares in which that natural or legal person has a lifelong right of disposal;
(v) held, or may be exercised within the meaning of (i) to (iv), by an undertaking controlled by that person or entity;
(vi) attached to shares deposited with that natural or legal person and which the person can exercise at his own discretion in the absence of specific instructions from the shareholders;
(vii) held by a third party in its own name on behalf of that person; or
(viii) exercisable by that person through a proxy where that person may exercise the voting rights at his discretion in the absence of specific instructions of the shareholder.

The duty to notify set forth above also applies to anyone, who directly or indirectly holds (a) financial instruments that afford the holder a right to purchase existing shares (e.g., share options); and/or (b) financial instruments based on existing shares and with an economic effect equal to that of the financial instruments mentioned in (a), regardless of them not affording the right to purchase existing shares (e.g., cash-settled derivatives linked to the value of the shares in question). Holding these kinds of financial instruments counts towards the thresholds mentioned above and may thus trigger a duty to notify by themselves or when accumulated with a shareholding.

The notification shall be made immediately and within the same trading day (before midnight) as the transaction, and in accordance with the provisions of Danish Executive Order no. 1256 of 4 November 2015 and disclose the number of voting rights and shares held directly or indirectly following the transaction. The notification shall further state the transaction date on which the threshold was reached or no longer reached and the identity of the shareholder as well as the identity of any natural or legal person with the right to vote on behalf of the shareholder and in the case of a group structure, the chain of controlled undertakings through which voting rights are effectively held. The information shall be notified to the company and simultaneously submitted electronically to the Danish FSA. Failure to comply with the notification requirements is punishable by fine or suspension of voting rights in instances of gross or repeated non-compliance. The Danish Securities Trading Act will be replaced by a new Act no. 650 of 8 June 2017 on capital markets (the "**Capital Markets Act**"), which enters into force in January 2018. In the context of major shareholder reporting, the Capital Markets Act will change the latest time for making major shareholder notifications from "within the same trading day as the transaction" to "no later than four trading days after the transaction".

When an obligation to notify rests on more than one natural or legal person the notification may be made through a joint notification. However, use of a joint notification does not exempt the individual shareholders or natural or legal persons from their responsibilities in connection with the obligation to notify or the contents of the notification.

After receipt of the notification, the company shall publish the contents of the notification.

A similar duty, as set forth above, to publish major shareholdings and notify the Danish FSA hereof also applies to a company's holding of treasury shares, voting rights and financial instruments (as described above), regardless of the company holding these on behalf of itself or through a natural or legal person acting on behalf of himself, but for the account of the company. Subsequent to the entry into force of the Capital Markets Act, issuers will only be required to make such disclosures when the holding of treasury shares reaches, exceeds or falls below thresholds of 5% and 10% of the voting rights or nominal value of the total share capital.

Furthermore, the general duty of notification under Section 55 of the Danish Companies Act in respect of notification of significant holdings (similar to the thresholds set out in the Danish Securities Trading Act Section 29) applies, including when the limit of 100% of the share capital's voting rights or nominal value of the company is reached or are no longer reached. Section 58 of the Danish Companies Act provides that a company shall publish information related to major shareholdings received pursuant to Section 55 of the Danish Companies Act in an electronic public register of shareholders which is kept by the Danish Business Authority. Moreover, Section 58a of the Danish Companies Act requires companies to seek out and publish information related to any natural legal persons that directly or indirectly owns a considerable amount of the shares or votes in the company in question (generally, 25 % or more of the total number of shares or votes), or who controls the company by any other means, in an electronic public register of beneficial shareholders, which is also kept by the Danish Business Authority. However, companies, whose shares are admitted to trading on a regulated market, are exempt from this registration requirement.

Short Selling

The Short Selling Regulation (236/2012/EU) includes certain notification requirements in connection with short selling and imposes restrictions on uncovered short selling of shares admitted to trading on a trading venue (including Nasdaq Copenhagen).

When a natural or legal person reaches or falls below a net short position of 0.2% of the issued share capital of a company that has shares admitted to trading on a trading venue, such person shall notify the relevant competent authority, which in Denmark is the Danish FSA. The obligation to notify, moreover, applies in each case where the net short position reaches or falls below each 0.1% threshold above the 0.2% threshold. In addition, when a natural or legal person reaches or falls below a net short position of 0.5% of the issued share capital of a company that has shares admitted to trading on a trading venue and each 0.1% threshold above that, such person shall make a public announcement of its net short position.

A natural or legal person is prohibited from entering into a short sale of shares admitted to trading on a trading venue unless one of the following conditions is satisfied: (i) the natural or legal person has borrowed the share or has made alternative provisions resulting in a similar legal effect; (ii) the natural or legal person has entered into an agreement to borrow the share or has another absolutely enforceable claim under contract or property law to be transferred ownership of a corresponding number of securities of the same class so that settlement can be effected when it is due; or (iii) the natural or legal person has an arrangement with third party under which that third party has confirmed that the share has been located and has taken measures vis-à-vis third parties necessary for the natural or legal person to have a reasonable expectation that settlement can be effected when it is due. Certain exemptions apply to the prohibition, such as in the case of market-makers or in connection with stabilisation in accordance with the Commission Delegated Regulation (EU) 2016/1052 (the "**Safe Harbor Regulation**").

Mandatory Tender Offers

The Danish Securities Trading Act (Part 8) and the Danish Executive Order no. 562 of 2 June 2014 on Takeover Bids includes rules concerning public offers for the acquisition of shares admitted to trading on a regulated market (including Nasdaq Copenhagen) or an alternative marketplace.

If a shareholding is transferred, directly or indirectly, in a company with one or more share classes admitted to trading on a regulated market or an alternative market place, to an acquirer or to persons acting in concert with such acquirer, the acquirer shall give all shareholders of the company the option to dispose of their shares on identical terms, if the acquirer gains a controlling interest as a result of the transfer.

Controlling interest exists if the acquirer, directly or indirectly, holds at least one-third of the voting rights in the company, unless it can be clearly proven in special cases that such ownership does not constitute a controlling interest. An acquirer who does not hold at least one-third of the voting rights in a company, nevertheless has a controlling interest when the acquirer has:

(i) the right to control at least one-third of the voting rights in the company according to an agreement with other investors;
(ii) the right to control the financial and operational affairs of the company according to the articles of association or agreement; or
(iii) the right to appoint or dismiss a majority of the members of the supervisory body, and this body has controlling influence over the company.

Warrants, call options and other potential voting rights, which may currently be exercised or converted, shall be taken into account in the assessment of whether the acquirer holds a controlling interest. Voting rights attached to treasury shares shall be included in the calculation of voting rights.

The Danish Takeover Order specifically exempts transfers of shares by inheritance or transfer within the same group and as a result of a creditor's debt enforcement proceedings from the obligation to submit a mandatory takeover offer. Exemptions from the mandatory tender offer rules may be granted under special circumstances by the Danish FSA.

Mandatory Redemption of Shares

Where a shareholder holds more than 90% of the shares in a company and a corresponding proportion of the voting rights, such shareholder may, pursuant to the Danish Companies Act, Section 70, decide that the other shareholders have their shares redeemed by that shareholder. In this case, the other shareholders must be requested, under the rules governing notices for general meeting, to transfer their shares to the shareholder within four weeks after the request to transfer their shares. In addition, the other shareholders shall through the Danish Business Authority's IT system be requested to transfer their shares within the same four-week period. Specific requirements apply to the contents of the notices to the other shareholders regarding the redemption. If the redemption price cannot be agreed upon, the redemption price must be determined by an independent expert appointed by the court in the jurisdiction of the company's registered office in accordance with the provisions of the Danish Companies Act. However, the redemption price will be deemed fair under any circumstances, provided that (i) the redemption takes place in continuation of a voluntary tender offer by which the bidder obtained at least 90% of the voting rights or (ii) the redemption takes place after a mandatory tender offer. To the extent any minority shareholders have not transferred their shares to the acquiring shareholder before the expiry of the four-week period, the redeeming shareholder shall, as soon as possible thereafter, deposit the amount required for redemption for the benefit of such minority shareholders. Upon the deposit, such minority shareholders will have been redeemed and the minority shareholders shall in such case through the Danish Business Authority's IT system be notified that the right to require determination of the redemption price by the independent expert expires at the end of a period, which cannot be less than three months pursuant to the Danish Companies Act, Section 72.

Furthermore, where a shareholder holds more than 90% of the shares in a company and a corresponding proportion of the voting rights, the other shareholders may require such shareholder to acquire their shares pursuant to Section 73 of the Danish Companies Act. If the redemption price cannot be agreed upon, the redemption price must be determined by an independent expert appointed by the court in the jurisdiction of the company's registered office in accordance with the provisions of the Danish Companies Act. Expenses relating to the determination of the redemption price must be paid by the shareholder requesting such determination. If the valuation is higher than that offered by the redeeming shareholder, the court may order the redeeming shareholder to pay the expenses relating to determination of the redemption price in full or in part.

Disclosure Requirements for Companies Admitted to Trading and Official Listing on Nasdaq Copenhagen

As a company with its securities admitted to trading on a regulated market, the Company will under Regulation (EU) no. 596/2014 on Market Abuse (the "**Market Abuse Regulation**") and the Issuer Rules of Nasdaq Copenhagen be obliged to inform the public and the Danish FSA of inside information, as defined in Article 7 of the Market Abuse Regulation, as soon as possible if such information directly concerns the Company. Inside information must be disclosed as soon as possible unless the Company is in a position to delay such disclosure to the public with reference to Article 17(4) of the Market Abuse Regulation.

In addition, the Company will be obliged to disclose certain other information to the public pursuant to the Danish Securities Trading Act, the Danish Executive Order no. 1258 of 9 November 2015 on an Issuer's Duty to Provide Information and the Issuer Rules of Nasdaq Copenhagen, regardless of whether this information amounts to inside information. Information which would have to be disclosed under these rules includes, for example: (i) changes to the Company's Board of Directors, Executive Management and auditors; (ii) decisions to introduce incentive schemes; (iii) substantial changes in business activities; (iv) material acquisitions and divestments; (v) unexpected and significant deviations in the Company's financial result or position; (vi) proposed changes in the capital structure; and (vii) annual and interim reports and accounts. Furthermore, the Company will be required to make sure that no unauthorised person gains access to inside information prior to its publication to the market.

Plan of Distribution

The Offering

As of the date hereof, the Company and the Managers named below have entered into an underwriting agreement (the "**Underwriting Agreement**") with respect to the Offer Shares. Subject to certain conditions set forth in the Underwriting Agreement and the execution of a pricing agreement, the Company will agree to sell to the subscribers procured by the Managers or, failing which, to the Managers themselves; and each of the Managers, severally but not jointly, will agree to procure subscribers for, or failing such procurement, to subscribe from the Company the percentage of total number of Offer Shares offered listed opposite such Manager's name below.

Managers	**Percentage of Offer Shares**
Carnegie Investment Bank, filial af Carnegie Investment Bank AB (publ), Sverige	42.5%
Danske Bank A/S	42.5%
Oddo BHF SCA	15.0%
Total	**100.00%**

The Underwriting Agreement provides that the obligations of the Managers are subject to: (i) entry into the pricing agreement between the Company and the Managers, which will contain the Offer Price and the exact number of Offer Shares; (ii) receipt of opinions on certain legal matters from counsel; and (iii) certain other conditions, including receipt of auditor letters and reports and officer certificates. The Company has agreed to indemnify the Managers against certain losses and liabilities arising out of or in connection with the Offering.

The Underwriting Agreement provides that, upon the occurrence of certain events, such as the general suspension of all trading on Nasdaq Copenhagen, a material adverse change in the Company's business, results of operations or financial condition or in the financial markets and under certain other conditions, the Managers may elect to terminate their several commitments and have the right to withdraw from the Offering before settlement of the Offering (i.e., payment for and settlement of the Offer Shares by way of delivery of Temporary Purchase Certificates). If the Managers elect to terminate their several commitments, the Offering may be cancelled, and if it is cancelled, no Temporary Purchase Certificates or Offer Shares will be delivered. All dealings in the Temporary Purchase Certificates and/or the Offer Shares prior to settlement of the Offering are at the sole risk of the parties concerned.

Pursuant to the Underwriting Agreement, the Joint Global Coordinators have, on behalf of the Managers, been granted an option to subscribe for an aggregate of up to 1,406,250 additional new Shares, solely to cover overallotments or short positions, if any, exercisable for a period of 30 calendar days after Admission. The number of Overallotment Shares will be adjusted if less than the maximum number of Offer Shares (other than the Overallotment Shares) is subscribed for in the Offering, such that the number of Overallotment Shares will equal 15% of the number of Offer Shares (other than Overallotment Shares). If any Overallotment Shares are agreed to be subscribed for under this option, each Manager will be obligated, subject to certain conditions contained in the Underwriting Agreement, to subscribe for a number of additional Overallotment Shares proportionate to that Manager's initial percentage of Offer Shares reflected in the table above, and the Company will be obligated to issue a number of Shares proportionate to the additional Overallotment Shares over which they have granted this option. Novo Holdings has agreed with the Joint Global Coordinators that Novo Holdings will make available a number of existing Shares equal to the number of Overallotment Shares for purposes of delivery of the Offer Shares to investors in connection with the Overallotment Option.

Purchasers of the Offer Shares may be required to pay stamp taxes and other charges in accordance with the laws and practices of the country of purchase in addition to the Offer Price.

Application has been made for the Temporary Purchase Certificates to be admitted to trading and for the Shares to be admitted to trading and official listing on Nasdaq Copenhagen. The Admission is subject to, among other things, Nasdaq Copenhagen's approval of the distribution of the Offer Shares, the election of the New Board Members], the Offering not being withdrawn prior to settlement of the Offering (including registration of the capital increase with respect to the new Offer Shares with the Danish Business Authority) and the Company making an announcement to that effect.

The Offer Shares are expected to be delivered on or around 21 November 2017 by way of delivery of Temporary Purchase Certificates against payment in immediately available funds in Danish kroner to investors' accounts with VP Securities and through the facilities of Euroclear and Clearstream. Subject to completion of the Offering and registration of the new Offer Shares with the Danish Business

Authority, the Temporary Purchase Certificates will automatically be exchanged for a corresponding number of Shares, which are expected to be delivered two business days after the Settlement Date in book-entry form to the holder of the Temporary Purchase Certificates' account with VP Securities and through the facilities of Euroclear and Clearstream. The first day of trading and official listing on Nasdaq Copenhagen is expected to be 17 November 2017 subject to the Offering not being withdrawn prior to settlement and completion of the Offering. Trading in the Temporary Purchase Certificates is expected to commence on 17 November 2017 provided that the announcement of the Offer Price and allocation has been published through Nasdaq Copenhagen no later than 8:00 a.m. (CET) on 17 November 2017. The last day of trading of the Temporary Purchase Certificates on Nasdaq Copenhagen is expected to be 21 November 2017. The first day of trading of the Shares on Nasdaq Copenhagen under the permanent ISIN is expected to be 22 November 2017. In connection with the Temporary Purchase Certificates being automatically exchanged for Shares, the Temporary Purchase Certificates will cease to exist. All dealings in the Temporary Purchase Certificates and/or the Offer Shares prior to settlement of the Offering will be for the account of and at the sole risk of the parties involved.

Up to 93,750 Offer Shares will be reserved for certain members of the Board of Directors to subscribe for in connection with the Offering at the Offer Price.

Up to 21,875 of the Offer Shares will be reserved for the participants in Orphazyme's LTIP to subscribe for at the Offer Price as an investment in connection with the Offering.

Orphazyme's employees (including the Executive Management and Key Employees) have been offered to subscribe for Offer Shares at the Offer Price for a maximum amount of DKK 200,000 per employee. Accordingly, up to 93,750 Offer Shares will be reserved for Orphazyme's employees to subscribe for in connection with the Offering at the Offer Price.

Up to 3,593,750 of the Offer Shares will be reserved for Skandinaviska Enskilda Banken, Danmark, branch of Skandinaviska Enskilda Banken AB (Publ.), Sweden; Vækstfonden; BI Asset Management Fondsmæglerselskab A/S on behalf of certain clients; Handelsbanken, branch of Svenska Handelsbanken AB (Publ.), Sweden; and Spar Nord Bank A/S, who have in connection with the Offering subject to certain conditions undertaken to subscribe for Offer Shares as Cornerstone Investors for a total subscription amount of DKK 230 million, corresponding to approximately 38.3% of the Offering (excluding the Overallotment Option). The commitments undertaken by the Cornerstone Investors are subject to certain conditions, e.g. that the Offer Price is set within the Offer Price Range and there being no significant new factor, material mistake or inaccuracy in to the information contained in this Offering Circular capable of affecting the Cornerstone Investors' assessment of the Shares and which would be required to be mentioned in a supplement to the Offering Circular under the Danish Securities Trading Act.

In connection with the Offering, the Managers and any affiliates acting as investors for their own account may take up the Shares and in that capacity may retain, purchase or sell the Shares, for their own account and may offer or sell such securities otherwise than in connection with the Offering, in each case, in accordance with applicable law. The Managers do not intend to disclose the extent of any such investment or transactions otherwise than in accordance with any legal or regulatory obligation to do so.

No action has been or will be taken in any jurisdiction other than Denmark that would permit a public offering of the Offer Shares, or the possession, circulation or distribution of this Offering Circular or any other material relating to the Company or the Offer Shares, in any jurisdiction where action for that purpose is required. Accordingly, the Offer Shares may not be offered or sold, directly or indirectly, and neither this Offering Circular nor any other offering material or advertisements in connection with the Offer Shares may be distributed or published, in or from any country or jurisdiction, except in compliance with any applicable rules and regulations of such country or jurisdiction.

Prior to the Offering, the Shares have never been listed, and there is currently no public market for the Shares. The Offer Price will be determined by the Company following consultation with the Joint Global Coordinators, on the basis of a number of factors, including the following:

- the orders, in terms of price and quantity, received from potential institutional and retail investors;
- prevailing market conditions;
- Orphazyme's historical, operational and financial performance;
- estimates of Orphazyme's business potential and earning prospects; and
- the market valuation of publicly traded common stock of comparable companies.

The Offer Price is expected to be announced no later than at 8:00 a.m. CET on 17 November 2017. The indicative Offer Price Range set forth on the cover page of this Offering Circular is subject to change as a result of market conditions and other factors. See also "*The Offering—Offer Price*". There can be no assurance that an active trading market will develop for the Shares or that the Shares will trade in the public market after the Offering at, or above, the Offer Price. See also "*Risk Factors—Risks Relating to the Offering*".

Lock-up Arrangements

The Company has agreed with the Managers that it will not, except as set forth below, for a period of 180 days from Admission, without the prior written consent of the Managers: (i) issue, offer, pledge, sell, contract to sell, sell any option or contract to purchase, purchase any option or contract to sell, grant any option, right or warrant to purchase, lend, or otherwise transfer or dispose of (or publicly announce such action), directly or indirectly, any Shares or any securities convertible into or exercisable or exchangeable for Shares; (ii) enter into any swap or other arrangement that transfers to another, in whole or in part, any of the economic consequences of ownership of Shares, whether any such transaction described in clause (i) or (ii) above is to be settled by delivery of Shares or such other securities, in cash or otherwise; or (iii) submit to the Company's shareholders a proposal to effect any of the foregoing. The foregoing shall not apply to issue(s) of Shares by the Company as a result of exercise of Pre-IPO Warrants, the grants by the Company to participants in its LTIP or any Shares issued or transferred as part of the 2017 Capital Structure Adjustment.

The Main Shareholders have agreed with the Managers that they will not, except as set forth below, for a period starting on the date hereof and ending on the earlier of (i) the Company's publication of the results of the ongoing NPC phase II/III trial, currently expected for Q3 2018 (however, not earlier than 180 days after Admission), or (ii) 360 days after Admission, without the prior written consent of the Managers: (i) offer, pledge, sell, contract to sell, sell any option or contract to purchase, purchase any option or contract to sell, grant any option, right or warrant to purchase, lend, cause the Company to issue Shares, or otherwise transfer or dispose of (or publicly announce such action), directly or indirectly, any of their Lock-Up Shares held as of Admission (excluding any Offer Shares subscribed for in connection with the Offering), or any securities convertible into or exercisable or exchangeable for such Lock-Up Shares; (ii) enter into any swap or other arrangement that transfers to another, in whole or in part, any of the economic consequences of ownership of any Lock-Up Shares, whether any such transactions described in clause (i) or (ii) above are to be settled by delivery of such Lock-Up Shares or such other securities, in cash or otherwise; or (iii) propose any general meeting of the Company, or convene or take action to convene any general meeting for the purpose of proposing, any resolution of the Company authorizing the issue of any Shares or warrants to subscribe for Shares in each case, without the prior written consent of the Joint Global Coordinators (such consent not to be unreasonably withheld or delayed). In case the Managers consent in writing to release Lock-Up Shares held by a Main Shareholder, Lock-up Shares held by other Main Shareholders shall be released on a pro rata basis.

The foregoing shall not apply to: (i) the disposal of Shares to the direct or indirect shareholders or wholly-owned subsidiaries of the relevant Main Shareholder in connection with or arising out of any dividend or other distributions, or any liquidation, dissolution, reorganisation or other similar event affecting such Main Shareholder or any of its affiliates; (ii) any disposal of Shares subscribed for in connection with the Offering or acquired on or after Admission; (iii) for Novo Holdings, the lending of Shares under the Stock Lending Agreement (see *"The Offering—Share Lending Agreement"*); (iv) taking up any Shares or other rights and any transfer of any rights granted in respect of a rights issue or other pre-emptive share offering by the Company; (v) the disposal of Shares in accordance with any order made by a court of competent jurisdiction or required by law or regulation; and (vi) any disposal of Shares pursuant to a general offer made to all holders of shares in the Company made in accordance with takeover regulations on terms which treat all such holders alike (and the execution and delivery of an irrevocable commitment or undertaking to accept such general offer), provided that, in case of any transfer comprised by the foregoing, as a condition to such disposal of Shares, , each such shareholder has agreed to assume the obligations of such Main Shareholders.

The members of the Company's Board of Directors at the time of Admission (other than the New Board Members), Executive Management and the Key Employees have agreed with the Managers that, for a period of 360 days from Admission, in respect of the Shares owned as of Admission (and any Shares subscribed for as a result of exercise of Pre-IPO Warrants, if applicable), they will be subject to substantially the same restrictions as those of the Company and Main Shareholders as set forth above. The foregoing shall not apply to: (i) the disposal to their respective (a) spouse, (b) child or (c) any legal entity over which they alone (or together with any other of their respective related parties) have a controlling influence, provided that such persons agree to adhere to similar restrictions, (ii) disposal made with a view to settle any tax liabilities incurred as a result of exercise of Pre-IPO Warrants, (iii) disposal in accordance with a court order or as required by law or regulation, (iv) any disposal of Shares pursuant to a general offer made to all holders of shares in the Company made in accordance with takeover regulations on terms which treat all such holders alike (and the execution and delivery of an irrevocable commitment or undertaking to accept such general offer), (v) any disposal of any rights granted in respect of a rights issue or other pre-emptive share offering by the Company whose proceeds will be used in their entirety to fund the subscription or purchase of the balance of any rights granted pursuant to any such rights issue or other pre-emptive share offering by the Company, (vi) disposal occurring after death, permanent disability or interruption in employment for a continuous period of not less than 16 weeks due to disability or illness, and (vii) disposal occurring after termination of employment by the Company or resignation from the Board of Directors.

Moreover, all existing shareholders of the Company as of the date hereof have undertaken an obligation not to trade in the existing Shares until the Temporary Purchase Certificates have been exchanged for Shares and the Shares have been admitted to trading and official listing on Nasdaq Copenhagen.

Price Stabilization and Short Positions

In connection with the Offering, Danske Bank, as the stabilising manager, or its agents, on behalf of the Managers, may engage in transactions that stabilise, maintain or otherwise affect the price of the Shares for up to 30 days from the Admission. Any profits from stabilization actions will be afforded to the Managers.

Specifically, the Managers, Novo Holdings and the Company have agreed that the stabilizing manager on behalf of the Managers may over allot Offer Shares by accepting offers to subscribe for a greater number of Offer Shares than for which they are obligated to procure subscribers under the Underwriting Agreement, creating a short position. A short sale is covered if the short position is no greater than the number of Offer Shares available for subscription by the stabilising manager on behalf of the Managers under the Overallotment Option.

The Managers can close out a covered short sale by exercising the Overallotment Option or purchasing Shares in the open market. In determining the source of Shares to close out a covered short sale, the Managers will consider, among other things, the open market price of Shares compared to the price available under the Overallotment Option.

As an additional means of facilitating the Offering, the stabilising manager or its agents may effect transactions to stabilise the price of the Shares. These activities may support the market price of the Offer Shares at a level higher than that which might otherwise prevail. Such transactions may be effected on Nasdaq Copenhagen, in the over-the-counter markets or otherwise.

The stabilising manager and its agents are not required to engage in any of these activities, and, as such, there is no assurance that these activities will be undertaken; if undertaken, the stabilising manager or its agents may end any of these activities at any time, and they must be brought to an end at the end of the 30-day period mentioned above. Save as required by law or regulation, the stabilising manager does not intend to disclose the extent of any Stabilisation transactions under the Offering.

Other Relationships

The Managers and their respective affiliates are full service financial institutions engaged in various activities, which may include securities trading, commercial and investment banking, financial advisory, investment management, investment research, principal investment, hedging, financing and brokerage activities. Certain of the Managers and their respective affiliates have from time to time engaged in, and may in the future engage in, commercial banking, investment banking and financial advisory transactions and services in the ordinary course of their business with the Company or the Main Shareholders or any of the Company's or their respective related parties. With respect to certain of these transactions and services, the sharing of information is generally restricted for reasons of confidentiality, internal procedures or applicable rules and regulations. The Managers have received and will receive customary fees and commissions for these transactions and services and may come to have interests that may not be aligned or could potentially conflict with potential investors' and the Company's interests. Danske Bank A/S is expected to also be a lender under a loan facility in favour of the Executive Management and Key Employees with respect to their subscription for Investment Shares (as defined herein) in connection with Orphazyme's long-term incentive programme.

In addition, in the ordinary course of business, the Managers and their respective affiliates may make or hold a broad array of investments including serving as counterparties to certain derivative and hedging arrangements and actively trade debt and equity securities (or related derivative securities) and financial instruments (including bank loans) for their own account and for the accounts of their customers, and such investment and securities activities may involve securities and/or instruments of the Company. The Managers and their respective affiliates may also make investment recommendations and/or publish or express independent research views in respect of such securities or instruments and may at any time hold, or recommend to clients that they acquire, long and/or short positions in such securities and instruments.

Selling Restrictions

United States

The Offer Shares have not been and will not be registered under the U.S. Securities Act or with any securities regulatory authority of any state of the United States for offer or sale as part of their distribution and may not be offered or sold within the United States except in certain transactions exempt from the registration requirements of the U.S. Securities Act. The Offer Shares may only be resold outside the United States of America in offshore transactions in compliance with Regulation S under the U.S. Securities Act and in accordance with applicable law. Terms used herein shall have the meanings given to them by Regulation S.

European Economic Area

In relation to each Relevant Member State of the EEA that has implemented the Prospectus Directive (with the exception of Denmark), no offer of the Offer Shares may be made to the public in that Relevant Member State, except that offers of the Offer Shares may be made under the following exemptions under the Prospectus Directive as implemented in that Relevant Member State:

- to any qualified investor as defined in the Prospectus Directive;
- to fewer than 150 natural or legal persons (other than qualified investors as defined in the Prospectus Directive) subject to obtaining the prior consent of the Joint Global Coordinators for any such offer; or
- in any other circumstances falling within Article 3(2) of the Prospectus Directive;

provided that no such offer of Offer Shares shall result in a requirement for the publication by the Company or any Manager of a prospectus pursuant to Article 3 of the Prospectus Directive or supplement a prospectus pursuant to Article 16 of the Prospectus Directive.

For the purposes of this paragraph, the expression an "offer of the Offer Shares may be made to the public" in relation to any of the Offer Shares in any Relevant Member State, means the communication in any form and by any means of sufficient information on the terms of the Offering and the Offer Shares to be offered so as to enable an investor to decide to purchase or subscribe for the Offer Shares, as the same may be varied in that Relevant Member State by any measure implementing the Prospectus Directive in that Relevant Member State.

United Kingdom

In the United Kingdom, this Offering Circular is being distributed only to, and is directed only at, persons who: (i) have professional experience in matters relating to investments falling within the definition of "investment professionals" in Article 19(5) of The Financial Services and Markets Act 2000 (Financial Promotion) Order 2005, as amended (the "Order 2005"); (ii) are high net worth bodies corporate, unincorporated associations and partnerships and the trustees of high value trusts, as described in Article 49(2) of the Order 2005; (iii) the Company believes on reasonable grounds to be persons to whom Article 43(2) of the Order 2005 applies for these purposes; or (iv) other persons to whom it may lawfully be communicated (all such persons being referred to in (i), (ii), (iii) and (iv) are defined as "Relevant Persons"). In the United Kingdom, any investment or investment activity to which this Prospectus relates is only available to and will only be engaged in with Relevant Persons. Any other persons who receive this Prospectus should not rely on or act upon it.

General

No action has been or will be taken in any country or jurisdiction other than Denmark that would, or is intended to, permit a public offering of the Offer Shares, or the possession or distribution of this Offering Circular or any other offering material, in any country or jurisdiction where action for that purpose is required.

Persons into whose hands this Offering Circular comes are required by the Company, the Main Shareholders and the Managers to comply with all applicable laws and regulations in each country or jurisdiction in or from which they purchase, offer, sell or deliver Offer Shares or have in their possession or distribute such offering material, in all cases at their own expense. Neither the Company, the Main Shareholders or the Managers accept any legal responsibility for any violation by any person, whether or not a prospective subscriber or purchaser of any of the Offer Shares, of any such restrictions.

Transfer Restrictions

The Offer Shares have not been, and will not be, registered under the U.S. Securities Act and may not be offered or sold within the United States except pursuant to an exemption from the registration requirements of the U.S. Securities Act and applicable state securities laws.

Each purchaser of the Offer Shares outside the United States in compliance with Regulation S will be deemed to have represented and agreed that it has received a copy of this Offering Circular and such other information as it deems necessary to make an informed investment decision and that:

(1) the purchaser is authorised to consummate the purchase of the Offer Shares in compliance with all applicable laws and regulations;
(2) the purchaser acknowledges that the Offer Shares have not been and will not be registered under the U.S. Securities Act, or with any securities regulatory authority of any state of the United States, and, subject to certain exceptions, may not be offered or sold within the United States;
(3) the purchaser and the person, if any, for whose account or benefit the purchaser is acquiring the Offer Shares, was located outside the United States at the time the buy order for the Offer Shares was originated and continues to be located outside the United States and has not purchased the Offer Shares for the account or benefit of any person in the United States or entered into any arrangement for the transfer of the Offer Shares or any economic interest therein to any person in the United States;
(4) the purchaser is not an affiliate of the Company or a person acting on behalf of such affiliate;
(5) the Offer Shares have not been offered to it by means of any "directed selling efforts" as defined in Regulation S;
(6) the purchaser is aware of the restrictions on the offer, sale and transfer of the Offer Shares pursuant to Regulation S and acknowledges that the Company shall not recognise any offer, sale, pledge or other transfer of the Offer Shares made other than in compliance with the above stated restrictions;
(7) if it is acquiring any of the Offer Shares as a fiduciary or agent for one or more accounts, the purchaser represents that it has sole investment discretion with respect to each such account and that it has full power to make the foregoing acknowledgements, representations and agreements on behalf of each such account; and
(8) the purchaser acknowledges that the Company, the Managers and their respective affiliates will rely upon the truth and accuracy of the foregoing acknowledgements, representations and agreements.

Each person in a Relevant Member State, other than persons receiving offers contemplated in the Offering Circular in Denmark, who receives any communication in respect of, or who acquires any Offer Shares under, the offers contemplated hereby will be deemed to have represented, warranted and agreed to and with each of the Managers, the Main Shareholders and the Company that:

(1) it is a qualified investor within the meaning of the law in that Relevant Member State implementing Article 2(1)(e) of the Prospectus Directive; and
(2) in the case of any Offer Shares acquired by it as a financial intermediary, as that term is used in Article 3(2) of the Prospectus Directive: (i) the Offer Shares acquired by it in the offer have not been acquired on behalf of, nor have they been acquired with a view to their offer or resale to, persons in any Relevant Member State other than qualified investors, as that term is defined in the Prospectus Directive, or in other circumstances falling within Article 3(2) of the Prospectus Directive and the prior consent of the Joint Global Coordinators has been given to the offer or resale; or (ii) where Offer Shares have been acquired by it on behalf of persons in any Relevant Member State other than qualified investors, the offer of those Offer Shares to it is not treated under the Prospectus Directive as having been made to such persons.

For the purposes of this provision, the expression an "offer" in relation to any of the Offer Shares in any Relevant Member States means the communication in any form and by any means of sufficient information on the terms of the offer and any Offer Shares to be offered so as to enable an investor to decide to purchase or subscribe for the Offer Shares, as the same may be varied in that Relevant Member State by any measure implementing the Prospectus Directive in that Relevant Member State.

Legal Matters

Certain legal matters in connection with the Offering will be passed upon for the Company by Gorrissen Federspiel Advokatpartnerselskab, legal counsel to the Company. Certain legal matters in connection with the Offering will be passed upon for the Managers by Plesner Advokatpartnerselskab, legal counsel to the Managers.

State authorised public accountants

The name and address of the Company's independent auditors are as follows:

Ernst & Young Godkendt Revisionspartnerselskab
Osvald Helmuths Vej 4
2000 Frederiksberg
Denmark

Ernst & Young Godkendt Revisionspartnerselskab ("**EY**") is represented by Christian Schwenn Johansen, State Authorised Public Accountant, and Lars Hansen, State Authorised Public Accountant, both members of FSR—Danish Auditors (*FSR – danske revisorer*).

Additional Information

Name, Registered Office and Date of Incorporation

Orphazyme A/S
Ole Maaløes Vej 3
DK-2200 Copenhagen N
Denmark
Telephone: +45 70 70 29 80
Website: www.orphazyme.com

The Company was incorporated in Denmark as a private limited liability company under the laws of Denmark on 19 June 2009 and later converted into a Danish public limited liability company in 20 October 2017. The Company does not have any secondary names.

The registered office is located in the municipality of Copenhagen at Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark.

The Company does not have any subsidiaries.

Registration

The Company is registered with the Danish Business Authority under registration (CVR) no. 32266355.

Objectives of the Company

According to article 1.2 of the Articles of Association the objective are research, development, production, marketing, sales and/or licensing of medicinal products for treatment of various disorders, including lysosomal storage diseases (LSD), neuromuscular disorders and other related diseases, as well as to carry out associated activities. Furthermore, the Company may, within its line of business, participate in partnerships or co-operate with other businesses.

Information Incorporated by Reference

The additional information explicitly listed in the table below has been incorporated by reference in the Offering Circular pursuant to Article 28 of the Prospectus Regulation as also set out in Section 19 of the Danish Executive Order on Prospectuses. Direct and indirect information references in the reports to other documents or websites are not incorporated by reference and do not form part of this Offering Circular. The reports speak only as at the date of their respective publications and have not been updated and in some cases have been made superfluous by the information in this Offering Circular. Potential investors should assume that the information in this Offering Circular as well as the information incorporated by reference is accurate as at the dates thereof only. Orphazyme's business, financial condition, cash flows and results of operations may have changed since those dates.

Potential investors are encouraged to read the information incorporated by reference in conjunction with the cautionary statements in "*Special Notice Regarding Forward-Looking Statements*" and in conjunction with the "*Risk Factors*" of this Offering Circular.

The additional information incorporated by reference in this Offering Circular is exclusively set out in the cross reference table below and is available for inspection at Orphazyme's address, Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark and on its website.

Financial statements for the period 1 July 2015 to 31 December 2015 prepared in accordance with Danish General Accepted Accounting Policies (Danish GAAP)

Information	Reference	Page(s)
Management statement	Annual report 2015	2
Independent auditor's report	Annual report 2015	3
Financial statements including notes	Annual report 2015	6-15

Financial statements for the period 1 July 2014 to 30 June 2015 prepared in accordance with Danish General Accepted Accounting Policies (Danish GAAP)

Information	Reference	Page(s)
Management statement	Annual report 2014/2015	1-2
Independent auditor's report	Annual report 2014/2015	3-5
Financial statements including notes	Annual report 2014/2015	20-26

Articles of Association (including appendices)

Information	Reference	Page(s)
Articles of Association	Articles of Association	1-14
Appendices	Articles of Association	15-221

General Meetings

The general meeting is the ultimate authority in all matters relating to the Company, subject to the limitations in Danish law and the Company's Articles of Association. See "*Description of the Shares and Share Capital—General Meetings and Voting Rights*".

Corporate structure

Orphazyme continuously seeks to optimise its corporate structure and, depending on the specific circumstances, a corporate reorganisation may be deemed appropriate. Such a corporate reorganisation may entail e.g. establishing subsidiaries possessing certain operations, products, intellectual property rights or know-how. However, no decision on any such corporate reorganisation has been made as of the date hereof and no plans of a specific nature are currently contemplated.

Principal Bankers

After completion of the Offering, Orphazyme's principal bankers will be Danske Bank A/S.

Share Issuing Agent

The Company's share issuing agent is:

Danske Bank A/S
CVR no. 61126228
Holmens Kanal 2-12
DK-1092 Copenhagen K
Denmark

Glossary

The following explanations are not intended as technical definitions and are provided purely for assistance in understanding certain terms as used in this Offering Circular.

2010 Warrant Programme	the warrant programme established in 2010 as described in "*Board of Directors, Executive Management and Key Employees—Incentive Programmes—Pre-IPO warrant programmes*"
2015 Comparative Financial Statements	reviewed unaudited financial statements for the period 1 January 2015 to 31 December 2015 comprising "*Statement of Profit or Loss and Other Comprehensive Income*", "*Statement of Financial Position*", "*Statement of Changes in Shareholders Equity*" and "*Statement of Cash Flows*" prepared based on the Company's accounting policies for recognition and measurement
2017 Capital Structure Adjustment	the adjustment of the Company's capital structure by way of a merger of the three previous share classes into one combined with an issue of bonus Shares in order to account for the now abolished preference shares, as further described in "*Ownership Structure and Main Shareholders*"
2017 Warrant Programme	the warrant programme established in 2017 as described in "*Board of Directors, Executive Management and Key Employees—Incentive Programmes—Pre-IPO warrant programmes*"
Admission	admission of the Company's Temporary Purchase Certificates to trading on Nasdaq Copenhagen
Aescap Venture	Coöperative Aescap Venture I U.A., company registration no. 34257886
ALS	Amyotrophic Lateral Sclerosis
AALS-001	a 12-week phase II dose ranging trial in ALS
AALS-001-OL	A six-month open-label extension trial in ALS
ALS Invest	ALS Invest 2 B.V., company registration no. 67804187
Articles of Association	the articles of association of the Company
Audit Committee	the audit committee of the Board of Directors, described in "*Board of Directors, Executive Management and Key Employees—Board of Directors—Board practices and committees*"
Audited Financial Statements	audited financial statements at and for the financial year 1 January 2016 – 31 December 2016 with comparative figures for the period 1 July 2015 – 31 December 2015 (six month conversion period) and the period 1 July 2014 – 30 June 2015 (12 months period), prepared in accordance with the IFRS and additional requirements of the Danish Financial Statements Act as included in the statutory annual report for 2016
Board of Directors	the board of directors of the Company at any given date
Brexit	British exit from the European Union
CAFS	combined assessment of survival and function
CAGR	compound annual growth rate
Capital Markets Act	Act no. 650 of 8 June 2017 on capital markets
CDMOs	contract development and manufacturing organisations
cGMPs	Good Manufacturing Practices
CHMP	Committee for Medicinal products for Human Use
Clearstream	Clearstream Banking S.A.
CMC	chemistry and manufacturing controls
Company	Orphazyme A/S, company registration (CVR) no. 32266355
Cornerstone Investors	Skandinaviska Enskilda Banken, Danmark, branch of Skandinaviska Enskilda Banken AB (Publ.), Sweden; Vækstfonden; BI Asset Management Fondsmæglerselskab A/S on behalf of certain clients; Handelsbanken, branch of Svenska Handelsbanken AB (Publ.), Sweden; and Spar Nord Bank A/S
Corporate Governance Recommendations	the Recommendations on Corporate Governance of the Danish Committee on Corporate Governance issued on 6 May 2013, as updated in November 2014
Co-Lead Manager	Oddo BHF SCA
CROs	contract research organisations
CSF	cerebrospinal fluid
CTA	clinical trial applications

CytRx	CytRx Corporation
Danish Central Bank	Danmarks Nationalbank
Danish Companies Act	the Danish Consolidated Act no. 1089 of 14 September 2015 on limited liability companies, as amended
Danish Executive Order on Issuer's Duty to Provide Information	Executive Order no. 1526 of 9 December 2016
Danish Executive Order on Major Shareholders	Executive Order no. 1256 of 4 November 2015
Danish Executive Order on Prospectuses	Executive Order no. 1257 of 6 November 2015 on prospectuses for securities admitted to trading in a regulated market and for offering to the public of securities of at least EUR 5,000,000, as amended
Danish FSA	Danish Financial Supervisory Authority
Danish Securities Trading Act	the Danish Consolidated Securities Trading Act no. 251 of 21 March 2017
Deputy Chairman	Deputy Chairman of the Board of Directors
"DKK" or "Danish kroner"	Danish kroner, the lawful currency of Denmark
DMA	Danish Medical Agency
EEA	European Economic Area
EMA	European Medicines Agency
EU	European Union
"euro", "EUR" or "€"	euro, the lawful currency of the participating member states in the Third Stage of the European and Monetary Union of the Treaty Establishing the European Community
Euroclear	Euroclear Bank S.A./N.A.
Executive Management	the executive management of the Company as registered with the Danish Business Authority
Existing Board Members	Georges Gemayel, Bo Jesper Hansen, Martijn Kleijwegt, Martin Bonde, Martin Rahbek Kornum, Nanna Lüneborg, Patrick J.H. Krol, Rémi Droller and Sten Verland
EY	Ernst & Young Godkendt Revisionspartnerselskab, company reg. (CVR) no. 30 70 02 28
FDA	US Food and Drug Administration
FEV6%	forced expiratory volume in six seconds
G&A activities	general and administrative activities
Gaucher	Gaucher disease
GBA	mutations in the beta-glucosidase gene
GCP	good clinical practices
GLP	good laboratory practices
GMP	good manufacturing practices
HSF1	heat shock factor 1
HSP70	heat shock protein 70
HSPs	heat shock proteins
IAS 34	International Accounting Standard 34 on "Interim Financial Reporting"
IASB	International Accounting Standards Board
IESBA Code	Ethics Standards Board for Accountants' Code of Ethics for Professional Accountants
IBMFRS	IBM functional rating scale
Idinvest	FCPI Idinvest Patrimoine n°3, company registration no. 414 735 175, FCPI Idinvest Patrimoine n°4 , company registration no. 414 735 175, FCPI Objectif Innovation Patrimoine n°6, company registration no. 414 735 175, FCPI Objectif Innovation Patrimoine n°7, company registration no. 414 735 175
IFRS	International Financial Reporting Standards as adopted by the EU
IND	investigational new drug
Interim Financial Statements	reviewed interim financial statements for the period 1 January 2017 to 30 June 2017 with unaudited and non-reviewed comparative figures for the periode 1 January 2016 to 30 June 2016 prepared in accordance with the IAS 34
Investment Shares	any investment shares related to the long-term incentive programme described in "*Board of Directors, Executive Management and Key Employees—Incentive Programmes—Long-term incentive programme (the "LTIP")*"
IPO	initial public offering
IPO Warrant Programme	the warrant programme established in connection with the Offering as described in "*Board of Directors, Executive Management and Key Employees—Incentive Programmes—Pre-IPO warrant programmes*"

Issuer Rules of Nasdaq Copenhagen	the rules for issuers of shares on Nasdaq Copenhagen of 3 July 2016
Joint Global Coordinators	Carnegie Investment Bank, filial af Carnegie Investment Bank AB (publ), Sverige and Danske Bank A/S
Key Employees	Thomas Kirkegaard Jensen and Thomas Blaettler
Lock-Up Shares	Shares subject to lock-up under the Underwriting Agreement
LTIP	the long-term incentive programme described in "*Board of Directors, Executive Management and Key Employees—Incentive Programmes—Long-term incentive programme (the "LTIP")*"
LSD	lysosomal storage disorders
MAA	marketing authorisation application
Main Shareholders	Novo Holdings; Aescap Ventures; Sunstone Capital; Orpha Pooling B.V.; Idinvest; and Kurma Biofund II
Managers	The Joint Global Coordinators and Co-Lead Manager
Market Abuse Regulation	Regulation (EU) no. 596/2014 on Market Abuse
MASCs	multipotent adult stem cells
Matching Shares	any matching shares related to the long-term incentive programme described in "*Board of Directors, Executive Management and Key Employees—Incentive Programmes—Long-term incentive programme (the "LTIP")*"
MHRA	Medicines and Healthcare Products Regulatory Agency
MOA	mechanism of action
Nasdaq Copenhagen	Nasdaq Copenhagen A/S, CVR no. 19 09 26 77
NDA	new drug application
New Board Members	Anders Hedegaard and Catherine Moukheibir
New Board of Directors	The Board of Directors as of Admission consisting of Georges Gemayel, Bo Jesper Hansen, Anders Hedegaard, Catherine Moukheibir, Martin Bonde, Martijn Kleijwegt, Rémi Droller and Sten Verland
NME	new molecular entities
Nomination Committee	the nomination committee of the Company's Board of Directors, described in "*Board of Directors, Executive Management and Key Employees—Board of Directors—Board practices and committees*"
Novo Holdings	Novo Holdings A/S, company registration (CVR) no. 24 25 76 30
NORD	The National Organization for Rare Disorders
NPC	Niemann Pick type C
OCT2	organic cation transporter 2
Offer Period	6 November 2017 to 16 November 2017 at 12:00 p.m. (noon) (CET), unless the Offering is closed earlier
Offer Price	the price per Offer Share at which the Offer Shares will be sold
Offer Price Range	the Offer Price is expected to be between DKK 64 and DKK 80 per Offer Share
Offer Shares	such number of new Shares as will raise gross proceeds of approximately DKK 690 million (hereof DKK 90 million pursuant to the Overallotment Option), unless the context indicates otherwise, all of which (except for the Overallotment Shares) will be delivered by way of Temporary Purchase Certificates representing the Offer Shares until being exchanged for a corresponding number of Shares
Offering	offering of up to 10,781,250 Offer Shares of DKK 1 nominal value each, including a number of Overallotment Shares pursuant to the Overallotment Option, if exercised
Orphan Drug Act	The Orphan Drug Act 1983
Orphazyme	Orphazyme A/S, company registration (CVR) no. 32266355
Overallotment Option	the option granted the Joint Global Coordinators by the Company to subscribe for additional new Shares at the Offer Price
Overallotment Shares	option granted by the Company to the Joint Global Coordinators to subscribe for up to 1,406,250 additional new Shares in the aggregate at the Offer Price
Performance Shares	any performance shares related to the long-term incentive programme described in "*Board of Directors, Executive Management and Key Employees—Incentive Programmes—Long-term incentive programme (the "LTIP")*"
Pre-IPO Warrants	a number of warrants granted under the 2010 and 2017 Warrant Programmes that may be exercised in connection with the Offering
Prospectus Directive	Directive 2003/71/EC of 4 November 2003 (together with any applicable implementing measures in any member state)

Prospectus Regulation	Commission Regulation (EC) no. 809/2004 of 29 April 2004, as amended
PTE	Analogue Patent Term Extension
QA	quality assurance
Regulation S	Regulation S under the U.S. Securities Act
Relevant Member State	any Member State of the European Economic Area that has implemented the Prospectus Directive, excluding Denmark
relevant persons	persons who: (i) are investment professionals falling within Article 19(5); or (ii) fall within Article 49(2)(a) to (d) ("high net worth companies, unincorporated associations, etc."), of the UK Financial Services and Markets Act 2000 (Financial Promotion) Order 2005 or other persons to whom such investment or investment activity may lawfully be made available
Remuneration Committee	the remuneration committee of the Company's Board of Directors, described in "*Board of Directors, Executive Management and Key Employees—Board of Directors—Board practices and committees*"
R&D	research and development
SAE	serious adverse event
Safe Harbour Regulation	Commission Delegated Regulation (EU) 2016/1052
Settlement Date	the date of payment for and settlement of the Offer Shares by way of delivery of Temporary Purchase Certificates expected to take place on or around 21 November 2017
Shares	the outstanding ordinary shares of the Company
Short Selling Regulation	Regulation (EU) 236/2012 of 14 March 2012 on short selling
sIBM	sporadic inclusion body myositis
SPC	Supplementary Protection Certificates
SOD1	copper-zinc superoxide dismutase
Sunstone Capital	Sunstone Life Science Ventures Fund II K/S, company registration (CVR) no. 30 58 22 68
STIP	the short-term incentive programme described in "*Board of Directors, Executive Management and Key Employees—Incentive Programmes—Short-term incentive programme (the "STIP")*"
Temporary Purchase Certificates	the temporary purchase certificates representing the Offer Shares from Admission until automatic exchange for a corresponding number of Shares
U.S. Securities Act	U.S. Securities Act of 1933, as amended
Underwriting Agreement	an agreement between the Company and the Managers entered into on 6 November 2017
VP Securities	VP SECURITIES A/S

Financial Information

Index of Financial Information

Reviewed unaudited condensed interim financial statements for the period 1 January - 30 June 2017 (the Interim Financial Statements):

Audited financial statements at 31 December 2016 and for the financial year 1 January 2016 - 31 December 2016 with comparative figures for the financial year 1 July 2015 - 31 December 2015 and 1 July 2014 - 30 June 2015 (the Audited Financial Statements):

Reviewed unaudited financial statements for the period 1 January - 31 December 2015 (the 2015 Comparative financial statements):

Introduction

The financial information set out on pages F-3 to F-46 comprises:

The Interim Financial Statements for the period 1 January - 30 June 2017

The reviewed unaudited condensed interim financial statements for the period 1 January 2017 to 30 June 2017 with unaudited and non-reviewed comparative figures for the period 1 January 2016 to 30 June 2016 prepared in accordance with the International Accounting Standard 34 on "Interim Financial Reporting" (IAS 34) as set out in pages F-3 - F-13. The Interim Financial Statements were prepared and approved by the Executive Management and the Board of Directors on xx October 2017. The Interim Financial Statements have been reviewed by Ernst & Young Godkendt Revisionspartnerselskab. Their report dated 6 November 2017 expressed an unmodified review opinion in respect of the period 1 January - 30 June 2017 and an other matter disclosure stating that the comparative figures for the period 1 January - 30 June 2016 have not been subject to review and, accordingly, Ernst & Young Godkendt Revisionspartnerselskab does not express an opinion or any other form of assurance on such comparative figures.

The Audited Financial Statements for 2016

The audited financial statements at 31 December 2016 and for the financial year 1 January 2016 - 31 December 2016 with comparative figures for the period 1 July 2015 - 31 December 2015 (6 month conversion period) and the period 1 July 2014 - 30 June 2015 (12 months period), prepared in accordance with the International Financial Reporting Standards as adopted by the European Union (IFRS) as included in the statutory annual report for 2016 and on pages F-14 - F-36. The Audited Financial Statements were prepared and approved by the Executive Management and the Board of Directors on 30 May 2017. The Audited Financial Statements have been audited by Ernst & Young Godkendt Revisionspartnerselskab. Their report dated 30 May 2017 expressed an unmodified opinion on the Audited Financial Statements.

In 2015, the Company changed its financial year to follow the calendar year, having previously used a financial year spanning 1 July to 30 June as the Company's peers also follows the calendar year. The current period in the audited financial statements at 31 December 2016 comprises the twelve-month period ended 31 December 2016 while the comparative figures comprise the six-month period ended 31 December 2015 and the twelve-month period ended 30 June 2015 and the figures are therefore not directly comparable. The Company has voluntarily decided to disclose the comparative figures for the financial year 2014/15.

The 2015 Comparative Financial Statements

The reviewed unaudited financial statements for the period 1 January 2015 to 31 December 2015 comprises "Statement of Profit or Loss and Other Comprehensive Income", "Statement of Financial Position", "Statement of Changes in Shareholders Equity" and "Statement of Cash Flows" and certain selected notes and as such these have not been prepared in accordance with International Financial Reporting Standards (IFRS) as a number of note disclosures are not included. However, the 2015 Comparative Financial Statements have been prepared based on the Company's accounting policies in respect of recognition and measurement applied in the financial statements at 31 December 2016 and for the period 1 January - 31 December 2016 with comparative figures for the financial year 1 July 2015 - 31 December 2015 and 1 July 2014 - 30 June 2015 as set out on pages F-37 to F-46, which were prepared in accordance with IFRS. The 2015 Comparative Financial Statements were prepared and approved by the Executive Management and the Board of Directors on 6 November 2017. The Audited Financial Statements have been reviewed by Ernst & Young Godkendt Revisionspartnerselskab. Their report dated 6 November 2017 expressed an unmodified review opinion in respect of the 2015 Comparative Financial Statements.

Statement by the Executive Management and the Board of Directors on the Condensed Interim Financial Statements for the period 1 January – 30 June 2017

The Board of Directors and the Executive Management have today considered and approved the Condensed Interim Financial Statements of Orphazyme A/S for the period 1 January - 30 June 2017.

The Condensed Interim Financial Statements for the period 1 January - 30 June 2017 have been prepared in accordance with IAS 34 "Interim Financial Reporting", as adopted by the European Union.

In our opinion, the Condensed Interim Financial Statements give a true and fair view of the Company's assets, liabilities and financial position at 30 June 2017 and of the results of the Company's operations and cash flows for the period 1 January - 30 June 2017.

Copenhagen, 6 November 2017

Board of Directors

Georges Gemayel
Chairman

Bo Jesper Hansen
Deputy Chairman

Martijn Kleijwegt
Board Member

Martin Bonde
Board Member

Martin Rahbek Kornum
Board Member

Nanna Lüneborg
Board Member

Patrick J. H. Krol
Board Member

Rémi Droller
Board Member

Sten Verland
Board Member

Executive Management

Anders Mørkeberg Hinsby
CEO

Anders Vadsholt
CFO

Independent auditor's review report on the unaudited Condensed Interim Financial Statements at and for the six months ended 30 June 2017 included on page F-5 to F-13

To the shareholders and potential shareholders

We have reviewed the condensed interim financial statements of Orphazyme A/S for the period 1 January – 30 June 2017, which comprise statement of profit or loss and comprehensive income, statement of financial position, statement of changes in shareholders' equity, statement of cash flow and notes. The condensed interim financial statements are prepared in accordance with IAS 34 "Interim Financial Reporting", as adopted by the European Union.

Management's responsibilities for the condensed interim financial statements

Management is responsible for the preparation of condensed interim financial statements in accordance with IAS 34 "Interim Financial Reporting", as adopted by the European Union and for such internal control as management determines is necessary to enable the preparation of condensed interim financial statements that are free from material misstatement, whether due to fraud or error.

Auditor's responsibilities

Our responsibility is to express a conclusion on the condensed interim financial statements. We conducted our review in accordance with the International Standard on Review of Interim Financial Information Performed by the Independent Auditor of the Entity and additional requirements applicable in Denmark.

This requires us to conclude whether anything has come to our attention that causes us to believe that the condensed interim financial statements, taken as a whole, are not prepared, in all material respects, in accordance with IAS 34 "Interim Financial Reporting", as adopted by the European Union. This standard also requires us to comply with ethical requirements.

A review of the condensed interim financial statements in accordance with the International Standard on Review of Interim Financial Information Performed by the Independent Auditor of the Entity is a limited assurance engagement. The auditor performs procedures primarily consisting of making enquiries of management and others within the Company, as appropriate, applying analytical procedures and evaluate the evidence obtained.

The procedures performed in a review are substantially less than those performed in an audit conducted in accordance with the International Standards on Auditing. Accordingly, we do not express an audit opinion on the condensed interim financial statements.

Other matter

The condensed interim financial statements of Orphazyme A/S for the period 1 January – 30 June 2017 contains comparative figures for the period 1 January 2016 – 30 June 2016. The comparative figures in the condensed interim financial statements have not been subject to review. Accordingly, we do not express an opinion or any other form of assurance on these comparative figures.

Conclusion

Based on our review, nothing has come to our attention that causes us to believe that these condensed interim financial statements are not prepared, in all material respects, in accordance with IAS 34 "Interim Financial Reporting", as adopted by the European Union.

Copenhagen, 6 November 2017

ERNST & YOUNG
Godkendt Revisionspartnerselskab
CVR no. 30 70 02 28

Christian Schwenn Johansen
State Authorised Public Accountant

Lars Hansen
State Authorised Public Accountant

Reviewed unaudited condensed interim financial statements for the period 1 January – 30 June 2017

Statement of Profit or Loss and Other Comprehensive Income

(TDKK)	Note	Six months ended 30 June 2017	Six months ended 30 June 2016
Research and development expenses	2	(46,870)	(27,875)
Administrative expenses	2	(7,972)	(2,696)
Operating loss		**(54,842)**	**(30,571)**
Net financials		(122)	35
Loss before tax		**(54,964)**	**(30,536)**
Income tax benefit	4	2,841	2,750
Net loss for the period		**(52,123)**	**(27,786)**
Other comprehensive income		-	-
Total comprehensive loss		**(52,123)**	**(27,786)**
Loss per share, basic and diluted	5	5.09	2.83

Statement of Financial Position

(TDKK)	Note	30 June 2017	31 December 2016
Assets			
Non-current assets			
Property, plant and equipment		1,007	987
Corporation tax receivable	4	5,591	2,750
Deposits		357	310
Total non-current assets		**6,955**	**4,047**
Current assets			
Corporation tax receivable	4	5,500	5,500
Receivable capital increase	3	91,319	-
Other receivables		5,104	3,421
Prepayments		3,245	4,624
Cash and cash equivalents		33,589	14,349
Total current assets		**138,757**	**27,894**
Total assets		**145,712**	**31,941**
Equity and liabilities			
Equity			
Share capital		5,103	3,361
Share premium		380,454	226,285
Accumulated deficit		(264,260)	(212,137)
Total equity	**3**	**121,297**	**17,509**
Current liabilities			
Trade payables		7,614	4,718
Payables to shareholders	6	223	0
Other payables		16,578	9,714
Total current liabilities		**24,415**	**14,432**
Total equity and liabilities		**145,712**	**31,941**

Statement of Changes in Shareholders' Equity

(TDKK)	Note	Share Capital	Share Premium	Accumulated deficit	Total
Balance as of 1 January 2016		**3,346**	**224,999**	**(154,202)**	**74,143**
Net loss for the period		-	-	(27,786)	(27,786)
Other comprehensive loss for the period		-	-	-	-
Balance as of 30 June 2016	**3**	**3,346**	**224,999**	**(181,988)**	**46,357**
Balance as of 1 January 2017		**3,361**	**226,285**	**(212,137)**	**17,509**
Net loss for the period		-	-	(52,123)	(52,123)
Other comprehensive loss for the period		-	-	-	-
Transactions with owners					
Capital increase decided, subscribed and paid		1,742	63,689	-	65,431
Capital increase decided, subscribed but not paid			91,319		91,319
Expenses, capital increase		-	(839)	-	(839)
Share-based payment expense		-	-	-	-
Balance as of 30 June 2017	**3**	**5,103**	**380,454**	**(264,260)**	**121,297**

Statement of Cash Flows

(TDKK)	Six months ended 30 June 2017	Six months ended 30 June 2016
Operating activities		
Operating loss	(54,842)	(30,571)
Adjustments to reconcile loss before tax to cash flows from operating activities		
Share-based payment expense	-	-
Depreciation and write-down	291	287
Change in other receivables	(1,730)	(1,032)
Change in prepayments	1,379	1,296
Change in trade payables	2,896	1,134
Change in other payables	6,821	2,284
Cash flows from taxes	-	-
Interest paid, net	(122)	35
Net cash flow used in operating activities	**(45,307)**	**(26,567)**
Investing activities		
Investment in property, plant and equipment	(311)	-
Net cash flow used in investing activities	**(311)**	**-**
Financing activities		
Capital contributions from shareholders	65,431	-
Bank loans	-	463
Expenses related to capital contributions	(573)	-
Net cash provided by financing activities	**64,858**	**463**
Net change in cash and cash equivalents	**19,240**	**(26,104)**
Net foreign exchange differences		-
Cash and cash equivalents at the beginning of the period	14,349	68,014
Cash and cash equivalents at the end of the period	**33,589**	**41,910**

Notes to the condensed interim financial statements

Note 1 – Accounting policies

Basis of Presentation
The condensed interim financial statements for the Company are prepared in accordance with International Accounting Standard 34 (IAS 34), "Interim Financial Reporting" .

The condensed interim financial statements included on pages F-5 to F-13 were prepared and approved by the Executive Management and the Board of Directors on 6 November 2017, as set out on page F-3.

Accounting Policies
The condensed interim financial statements have been prepared using the same accounting policies as outlined in accordance with the accounting policies set out in the Annual Report 2016 of Orphazyme A/S, discussed on pages F-21 to F-24 in the audited financial statements at and for the financial year 1 January 2016 - 31 December 2016 with comparative figures for the financial year 1 July 2015 - 31 December 2015 and 1 July 2014 - 30 June 2015.

Management Judgments and Estimates under IFRS
In preparing condensed interim financial statements, certain provisions under IFRS require management to make judgments (various accounting estimates and assumptions) which may significantly impact the Company's financial statements. The most significant judgments include, among other things, accrual and prepaid costs for clinical trial development costs and deferred tax assets. For additional descriptions of significant judgments and estimates, refer to note 2 in audited financial statements at and for the financial year 1 January 2016 - 31 December 2016 with comparative figures for the financial year 1 July 2015 - 31 December 2015 and 1 July 2014 - 30 June 2015 discussed on pages F-24 to F-25.

Note 2 – Warrants

The Company has issued warrants to employees, consultants providing similar services and key management. The warrants can be settled by subscribing for A-shares of the Company at an exercise price of DKK 44 per share. Due to the liquidation preference to B and C shares as discussed in note 10 in the annual report 2016, the exercise price for the warrants were significantly above the fair value of one A-share at the respective issuance dates.

Management has applied a Black Scholes option valuation model to determine fair value of the warrants. Fair value of the warrants granted in 2014/2015 and 2015 respectively amounts to TDKK 3 and TDKK 12 respectively.

The most significant assumption applied is the underlying share price. Fair value of one A-share has been determined on the basis of the share of fair value of Orphazyme attributable to A-shares. Fair value of Orphazyme has been determined as the implied fair value, which can be derived from the subscription price in the most recent capital increase round prior to granting the warrants. Fair value per A-share has been determined to be in the range DKK 1.87 – DKK 3.72.

In May 2017, Orphazyme, without cancelling or modifying former warrant programs, issued a new warrant program under which a mechanism was put in place ensuring that the respective warrant holders can only exercise warrants from either former programs or the new program. Orphazyme therefore has multiple warrant programs that run 'in parallel'.

The expense recognized by Orphazyme for warrant programs running in parallel and where management believes that both programs will vest, is determined based on

a) the grant date fair value of the old program under the original vesting terms, plus
b) the incremental fair value of the new warrant program, as at its grant date (being its fair value of the new programs less the fair value of the old programs at that date), over the vesting terms of the new program.

Management has applied a Black Scholes option valuation model to determine fair value of the warrants. Fair value of the warrants granted in the six month period ended 30 June 2017 amounts to TDKK 1.

The most significant assumption applied is the underlying share price. Fair value of one A-share has been determined on the basis of the share of fair value of Orphazyme attributable to A-shares. Fair value of Orphazyme has been determined as the implied fair value, which can be derived from the subscription price in the most recent capital increase round prior to granting the warrants. Fair value per A-share has been determined to be in the range DKK 1.87 – DKK 3.72.

The table below summarizes the activity related to the warrants for the six months ended 30 June 2017 and the six months ended 30 June 2016:

(TDKK)	Key management	Employees	Board of Directors	Consultants	Total Warrants	Warrants exercisable
Outstanding at 31 December 2015	211,879	76,176	124,122	9,700	421,877	266,621
Granted	-	-	-	-	-	
Exercised	-	-	-	-	-	
Expired	-	-	-	-	-	
Outstanding at 30 June 2016	**211,879**	**76,176**	**124,122**	**9,700**	**421,877**	**295,350**
Outstanding at 31 December 2016	211,879	76,176	124,122	9,700	421,877	324,078
Granted	211,879	215,573	124,122	-	551,574	
Exercised	-	-	-	-	-	
Expired	-	-	-	-	-	
Outstanding at 30 June 2017	**423,758**	**291,749**	**248,244**	**9,700**	**973,451**	**440,354**

The weighted average remaining contractual life of the warrants outstanding as of 30 June 2017 and 30 June 2016 was 4.1 years and 5.1 years, respectively.

According to the terms and conditions of the 2010, 2017 and IPO Warrant Programmes, an exercise window of 10 working days will be triggered in connection with an Initial Public Offering during which all allocated warrants may be exercised Warrants that are not exercised within the exercise period will lapse without further compensation. Upon exercise allocated under the 2017 Warrant Programme, the warrant holder's right to exercise warrants allocated under the 2010 Warrant Programme, if any, will be reduced accordingly and vice versa.

Note 3 – Equity

On 6 January 2015, the Company issued 1,704,554 indemnification warrants to the investors subscribing for C class shares at this date. The warrants entitle the holders to subscribe for additional C class shares at DKK 1 per share if certain liabilities of the Company prove higher than warranted by the Company in the Investment agreement. The warrants expire on 6 January 2018 and it is not expected to be realised. The warrants are considered as an adjustment mechanism to the subscription price and not as separate derivative liabilities.

In first quarter 2017, the Company has finished a capital increase by issuing 534,007 C class shares to existing shareholders for a net proceeds received of TDKK 48,061. In connection with the capital increase, the Company incurred expenses totaling TDKK 73.

On 8 March 2017 the Company completed a TDKK 108,690 financing round by issuing new shares to LSP V Coöperatieve U.A. and ALS Investment Fund. Of the total financing round, TDKK 17,371 has been invested in the six month period ended 30 June 2017 and the investors have subscribed capital of the remaining TDKK 91,319 but have not been paid yet the capital as at the balance sheet date.

Thus, as at 30 June 2017, the Company has recognized a receivable from capital increase of TDKK 91,319. The receivable from capital increases have been received after the balance sheet date. Reference is made to note 8 regarding subsequent events. In connection with the capital increase, the Company incurred expenses totaling TDKK 764.

Note 4 – Income tax and deferred tax

Income tax benefit for the period includes an estimated tax credit for research and development at the applicable tax rate under the Danish Corporate Income Tax Act.

The tax loss carry forwards have no expiry date. The Company's ability to use tax loss carry forwards in any one year is limited to 100% of the first MDKK 7.5 of taxable income plus 60% of taxable income above TDKK 7,500

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The Company recognizes deferred tax assets, including the tax base of tax loss carry forwards, if management assesses that these tax assets can be offset against positive taxable income within a foreseeable future. Significant management judgment is required to determine the amount of deferred tax assets that can be recognized, based upon the likely timing and the level of future taxable profits together with future tax planning strategies. This judgment is made periodically after considering current facts and circumstances, budgets and business plans as well as the risks and uncertainty associated with the Company's ability successfully commercialize and defend its intellectual property.

Note 5 – Loss per share

The following reflects the net loss attributable to shareholders and share data used in the basic and diluted loss per share computations for the six months ended 30 June 2017 and 2016:

(TDKK)	Six months ended 30 June 2017	Six months ended 30 June 2016
Loss for the period	(52,123)	(27,787)
Weighted average shares outstanding	10,248,928	9,833,637
Loss per share shares	**5.09**	**2.83**

Basic loss per share amounts are calculated by dividing the net loss for the period by the weighted average number of shares outstanding during each the period.

On 2 November 2017, the former share classes were rolled up in to common shares. In connection with the roll up in to common shares, the Company issued bonus shares to the holders of preference shares. The total number of bonus shares issued was 6,487,882, which has been included in the number of weigthed average shares outstanding for both the six months ended 30 June 2017 and the six months ended 30 June 2016.

Due to the fact that the Company has incurred losses for each period presented, the potential shares issuable related to outstanding warrants have been excluded from the calculation of diluted loss per share as the effect of such shares is anti dilutive. Therefore, basic and diluted loss per share are the same for each period presented.

Note 6 – Related party disclosures

The Company is not ultimately controlled by any of the investors. In addition to Novo Holdings A/S, Kurma Biofund II, Idinvest, Sunstone Life Science Ventures Fund II K/S and Cooperative Aescap Venture I U.A. all owns more than 5%.

As of 30 June 2017, the Company has a receivable from the financing round in March 2017 of TDKK 91,319 discussed in note 3. The receivable from capital increases has been received after the balance sheet date. Reference is made to note 8 regarding subsequent events. In connection with the March 2017 financing round, the Company is obliged to reimburse the investors for transactions cost incurred of TDKK 223. The Company has provided for these costs and recognized these costs under equity within the line expenses, capital increase.

There have been no transactions between related parties in the six months ended 30 June 2017 and the six months ended 30 June 2016 besides capital increases as described in note 3.

Terms and conditions of transactions with related parties

Amounts due to related parties are uncollateralized and interest free. There have been no guarantees provided or received for any related party receivables or payables. For the six months ended 30 June 2017 and the six months ended 30 June 2016, the Company has not recorded any impairment of receivables relating to amounts owed by related parties. There are no related party receivables at any of the balance sheet dates, besides the receivable from capital increase of TDKK 91,319 discussed in note 3. The receivable from capital increases have been received after the balance sheet date. Reference is made to note 8 regarding subsequent events.

Transactions with key management

The Company has not granted any loans, guarantees, or other commitments to or on behalf of any of the members of the Board of Directors or key management personnel.

Other than the remuneration, no other significant transactions have taken place with key management personnel during the period presented herein.

Compensation paid to members of the Board of Directors

Compensation paid to members of the Board of Directors are classified as administrative expense within the statement of loss. The following table lists compensation paid to members of the Board of Directors:

(TDKK)	Six months ended 30 June 2017	Six months ended 30 June 2016
Director fees	484	316
Warrants	0	0
Other fees	0	0
Total compensation paid to the Board of Directors	**484**	**316**

Certain members of the Board of Directors have received warrants in the Company (reference is made to note 2). Besides warrants and the director fees presented above, they have not received any other remuneration for their services.

Transactions with shareholders and affiliates

There have been no transactions with shareholders or affiliates of shareholders during the six months ending 30 June 2017 or the six months ending 30 June 2016, except for the capital increase disclosed in Note 3.

Note 7 – Contractual obligations

The following table summarises Orphazyme's contractual obligations and commercial commitments as of 30 June 2017.

Contractual obligations (TDKK)	**Payment due by period**				
	Less than 1 year	**1-3 years**	**3-5 years**	**More than 5 years**	**Total**
Operating lease obligations	659	-	-	-	659
Other contractual obligations	46,645	11,279	602	-	58,526
Total	**47,304**	**11,279**	**602**	**-**	**59,185**

Other contractual obligations primarily include committed costs relating to agreements with CROs used for pre-clinical studies, stability studies and clinical trials as well as funding of Ph.D.-students with collaboration partners.

Note 8 – Subsequent events

As at 30 June 2017, the Company has recognized a receivable from capital increase of TDKK 91,319. The receivable from capital increases have been received after the balance sheet date.

After the balance sheet date, the capital structure of Orphazyme has been adjusted by way of a merger of the three previous share classes into one combined with an issue of bonus Shares in order to account for the now abolished preference shares. The class B and C preference shares of the Company were converted into Shares on a 1:1 ratio.

In October 2017, the Company was converted from private limited liability company (ApS) into a public limited liability company (A/S).

Other than the events disclosed above, there were no other events that were required to be reported or disclosed that are not already included within these condensed interim financial statements.

Statement by the Executive Management and the Board of Directors on the financial statements at and for the financial year 1 January 2016 - 31 December 2016 with comparative figures for the financial year 1 July 2015 - 31 December 2015 and 1 July 2014 - 30 June 2015

The Executive Management and the Board of Directors have today discussed and approved the financial statements at 31 December 2016 and for the financial year 1 January 2016 - 31 December 2016 with comparative figures for the financial year 1 January - 31 December 2016 with comparative figures for the financial years 1 July - 31 December 2015 and 1 July 2014 - 30 June 2015.

The Financial Statements comprise statement of profit or loss and other comprehensive income, statement of financial position, statement of changes in shareholders' equity, statement of cash flows and notes, including a summary of significant accounting policies for Orphazyme ApS. The Financial Statements are prepared in accordance with International Financial Reporting Standards as adopted by the European Union.

In our opinion, the accounting policies applied are appropriate, and the Financial Statements give a true and fair view of Orphazyme ApS' financial position at 31 December 2016, 31 December 2015 and at 30 June 2015, respectively and of the results of the Company's operations and cash flows for the financial year 1 January - 31 December 2016, 1 July 2015 - 31 December 2015 and 1 July 2014 - 30 June 2015, respectively.

Copenhagen, 30 May 2017

Executive Management

Anders Mørkeberg Hinsby

Board of Directors

Georges Gemayel
Chairman

Bo Jesper Hansen
Deputy Chairman

Sten Verland
Board member

Patrick J.H. Krol
Board member

Nanna Lüneborg
Board members

Martin Bonde
Board member

Martijn Kleijwegt
Board member

Martin Rahbek Kornum
Board member

Rémi Droller
Board member

Independent auditor's report on the financial statements at 31 December 2016 and for the financial year 1 January 2016 - 31 December 2016 with comparative figures for the financial year 1 July 2015 - 31 December 2015 and 1 July 2014 - 30 June 2015

To the shareholders of Orphazyme ApS

Opinion
We have audited the financial statements of Orphazyme ApS (the "Company") for the financial year 1 January – 31 December 2016 with comparative figures for the financial years 1 July - 31 December 2015 and 1 July 2014 - 30 June 2015 (the "financial statements"), which comprise a statement of profit or loss and other comprehensive income, statement of financial position, statement of changes in shareholders' equity, statement of cash flows and notes, including accounting policies, as presented on pages F-17 to F-36. The financial statements are prepared in accordance with International Financial Reporting Standards as adopted by the European Union.

In our opinion, the financial statements as presented on pages F-17 to F-36 give a true and fair view of the financial position of Orphazyme ApS at 31 December, 2016 with comparative figures at 31 December, 2015 and 30 June 2015 and of the results of the Orphazyme's operations and cash flows for the financial year 1 January - 31 December 2016 with comparative figures for the financial years 1 July - 31 December 2015 and 1 July 2014 - 30 June 2015 in accordance with International Financial Reporting Standards as adopted by the European Union.

Basis for opinion
We conducted our audit in accordance with International Standards on Auditing ("**ISAs**") and additional requirements applicable in Denmark. Our responsibilities under those standards and requirements are further described in the "Auditor's responsibilities for the audit of the financial statements" section of our report. We are independent of the Company in accordance with the International Ethics Standards Board for Accountants' Code of Ethics for Professional Accountants ("**IESBA Code**") and additional requirements applicable in Denmark, and we have fulfilled our other ethical responsibilities in accordance with these rules and requirements. We believe that the audit evidence we have obtained is sufficient and appropriate to provide a basis for our opinion.

Management's responsibilities for the financial statements
Management is responsible for the preparation of financial statements that give a true and fair view in accordance with International Financial Reporting Standards as adopted by the European Union and additional requirements in the Danish Financial Statements Act and for such internal control as management determines is necessary to enable the preparation of financial statements that are free from material misstatement, whether due to fraud or error.

In preparing the financial statements, management is responsible for assessing the Company's ability to continue as a going concern, disclosing, as applicable, matters related to going concern and using the going concern basis of accounting in preparing the financial statements unless management either intends to liquidate the Company or to cease operations, or has no realistic alternative but to do so.

Auditor's responsibilities for the audit of the financial statements
Our objectives are to obtain reasonable assurance about whether the financial statements as a whole are free from material misstatement, whether due to fraud or error, and to issue an auditor's report that includes our opinion. Reasonable assurance is a high level of assurance, but is not a guarantee that an audit conducted in accordance with ISAs and additional requirements applicable in Denmark will always detect a material misstatement when it exists. Misstatements can arise from fraud or error and are considered material if, individually or in the aggregate, they could reasonably be expected to influence the economic decisions of users taken on the basis of the financial statements.

As part of an audit conducted in accordance with ISAs and additional requirements applicable in Denmark, we exercise professional judgment and maintain professional skepticism throughout the audit. We also:

- Identify and assess the risks of material misstatement of the financial statements, whether due to fraud or error, design and perform audit procedures responsive to those risks and obtain audit evidence that is sufficient and appropriate to provide a basis for our opinion. The risk of not detecting a material misstatement resulting from fraud is higher than for one resulting from error, as fraud may involve collusion, forgery, intentional omissions, misrepresentations or the override of internal control.

- Obtain an understanding of internal control relevant to the audit in order to design audit procedures that are appropriate in the circumstances, but not for the purpose of expressing an opinion on the effectiveness of the Company's internal control.

- Evaluate the appropriateness of accounting policies used and the reasonableness of accounting estimates and related disclosures made by management.
  Conclude on the appropriateness of management's use of the going concern basis of accounting in preparing the financial statements and, based on the audit evidence obtained, whether a material uncertainty exists related to events or conditions that may cast significant doubt on the Company's ability to continue as a going concern. If we conclude that a material uncertainty exists, we are required to draw attention in our auditor's report to the related disclosures in the financial statements or, if such disclosures are inadequate, to modify our opinion. Our conclusion is based on the audit evidence obtained up to the date of our auditor's report. However, future events or conditions may cause the Company to cease to continue as a going concern.

- Evaluate the overall presentation, structure and contents of the financial statements, including the note disclosures, and whether the financial statements represent the underlying transactions and events in a manner that gives a true and fair view.

We communicate with those charged with governance regarding, among other matters, the planned scope and timing of the audit and significant audit findings, including any significant deficiencies in internal control that we identify during our audit.

Copenhagen, 30 May 2017

ERNST & YOUNG
Godkendt Revisionspartnerselskab
CVR no. 30 70 02 28

Christian Schwenn Johansen
State Authorised Public Accountant

Lars Hansen
State Authorised Public Accountant

Statement of Profit or Loss and Other Comprehensive Income

(TDKK)	Note	Twelve months ended 31 December 2016	Six months ended 31 December 2015	Twelve months ended 30 June 2015
Research and development expenses	4, 5, 9	(55,817)	(25,478)	(31,604)
Administrative expenses	5, 9	(7,703)	(4,044)	(5,494)
Operating loss		**(63,520)**	**(29,522)**	**(37,098)**
Financial income	7	182	80	34
Financial expenses	8	(97)	(40)	(1,403)
Loss before tax		**(63,435)**	**(29,482)**	**(38,467)**
Income tax benefit	11	5,500	2,750	5,875
Net loss for the period		**(57,935)**	**(26,732)**	**(32,592)**
Other comprehensive income		-	-	-
Total comprehensive loss		**(57,935)**	**(26,732)**	**(32,592)**
Loss per share, basic and diluted	12			
Class C preferred shares		(16.71)	(7.98)	(11.15)
Class B preferred shares		(17.12)	(8.20)	(12.53)
Class A ordinary shares		(26.08)	(12.47)	(20.29)

Statement of Financial Position

(TDKK)	Note	31 December 2016	31 December 2015	30 June 2015	30 June 2014
Assets					
Non-current assets					
Property, plant and equipment	9	987	1,487	1,748	1,694
Corporation tax receivable	11	2,750	2,750	5,875	-
Deposits		310	211	184	179
Total non-current assets		**4,047**	**4,448**	**7,807**	**1,873**
Current assets					
Corporation tax receivable	11	5,500	5,875	6,250	7,500
Other receivables		3,421	644	408	269
Prepayments		4,624	5,971	2,721	190
Cash and cash equivalents		14,349	68,014	78,161	25,732
Total current assets		**27,894**	**80,504**	**87,540**	**33,691**
Total assets		**31,941**	**84,952**	**95,347**	**35,564**
Equity and liabilities					
Equity					
Share capital	10	3,361	3,346	3,218	2,175
Share premium	10	226,285	224,999	213,632	124,250
Accumulated deficit		(212,137)	(154,202)	(127,470)	(94,878)
Total equity		**17,509**	**74,143**	**89,380**	**31,547**
Current liabilities					
Bank debt		-	-	245	1
Trade payables		4,718	2,447	955	997
Other payables		9,714	8,362	4,767	3,019
Total current liabilities		**14,432**	**10,809**	**5,967**	**4,017**
Total equity and liabilities		**31,941**	**84,952**	**95,347**	**35,564**

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Statement of Changes in Shareholders' Equity

(TDKK)	Note	Share capital	Share premium	Accumulated deficit	Total
Balance as of 1 July 2014		**2,175**	**124,250**	**(94,878)**	**31,547**
Opening effects from conversion to IFRS	17	-	-	0	-
Net loss for the period		-	-	(32,592)	(32,592)
Other comprehensive loss for the period		-	-	-	-
Transactions with owners					
Capital increase		827	73,618	-	74,445
Conversion of debt		216	15,949	-	16,165
Expenses, capital increase		-	(185)	-	(185)
Share-based payment expense	6	-	-	-	-
Balance as of 30 June 2015		**3,218**	**213,632**	**(127,470)**	**89,380**
Net loss for the period		-	-	(26,732)	(26,732)
Other comprehensive loss for the period		-	-	-	-
Transactions with owners					
Capital increase		128	11,397	-	11,525
Expenses, capital increase		-	(30)	-	(30)
Share-based payment expense	6	-	-	-	-
Balance as of 31 December 2015		**3,346**	**224,999**	**(154,202)**	**74,143**
Net loss for the period		-	-	(57,935)	(57,935)
Other comprehensive loss for the period		-	-	-	-
Transactions with owners					
Capital increase		15	1,316	-	1,331
Expenses, capital increase		-	(30)	-	(30)
Share-based payment expense	6	-	-	-	-
Balance as of 31 December 2016		**3,361**	**226,285**	**(212,137)**	**17,509**

Statement of Cash Flows

(TDKK)	Note	Twelve months ended 31 December 2016	Six months ended 31 December 2015	Twelve months ended 30 June 2015
Operating activities				
Net loss before tax		(63,435)	(29,482)	(38,467)
Adjustments to reconcile loss before tax to cash flows from operating activities				
Share-based payment expense	6			0
Depreciation and write-down	9	706	286	504
Gain/loss on sale and disposal of assets		33	-	-
Change in other receivables		(2,876)	(263)	(145)
Change in prepayments		1,347	(3,250)	(2,531)
Change in trade payables		2,271	1,492	(42)
Change in other payables		1,352	3,595	1,748
Cash flows from taxes		5,875	6,250	1,250
Interest paid		-	-	1,245
Net cash flow used in operating activities		**(54,727)**	**(21,372)**	**(36,438)**
Investing activities				
Investment in property, plant and equipment	9	(238)	(25)	(558)
Net cash flow used in investing activities		**(238)**	**(25)**	**(558)**
Financing activities				
Capital contributions from shareholders	10	1,330	11,525	74,445
Cash from convertible loan		-	-	14,920
Bank loans		-	(245)	245
Expenses related to capital contributions		(30)	(30)	(185)
Net cash provided by financing activities		**1,300**	**11,250**	**89,425**
Net change in cash and cash equivalents		**(53,665)**	**(10,147)**	**52,429**
Net foreign exchange differences		-	-	-
Cash and cash equivalents at the beginning of the period		68,014	78,161	25,732
Cash and cash equivalents at the end of the period		**14,349**	**68,014**	**78,161**

Corporate information

Orphazyme ApS (the "Company") is a limited liability company incorporated and domiciled in Denmark. The registered office is located in Copenhagen, Denmark. The Company has no subsidiaries, and consequently, the financial statements are standalone financial statements. The financial statements for the year ended 31 December 2016 were authorized for approval at the Annual General Meeting to be held on 29 May 2017, with a resolution of the Board of Directors on 29 May 2017.

Note 1 – Accounting policies

Basis of preparation
The financial statements of the Company have been prepared in accordance with International Financial Reporting Standards, or IFRS, as adopted by the European Union. In prior years, the financial statements were prepared in accordance with the provisions applying to reporting class B enterprises under the Danish Financial Statements Act. Upon adoption of IFRS, standards and interpretations which are mandatory for reporting periods beginning on or after 1 January 2016 have been applied. The impact on reported financial position and financial performance is discussed in note 17.

The financial statements have been prepared on a historical cost basis except for share-based payment and convertible debt.
The financial statements are presented in Danish Kroner, or DKK, which is the functional currency of the Company based on facts and circumstances and the technical requirements of IFRS.

In 2015, the Company changed its financial year to follow the calendar year, having previously used a financial year spanning July 1 to 30 June as the Company's peers also follows the calendar year. The current period in the statement of loss comprises the twelve-month period ended 31 December 2016 while the comparative figures comprise the six-month period ended 31 December 2015 and the twelve-month period ended 30 June 2015 and the figures are therefore not directly comparable. The Company has voluntarily decided to disclose the comparative figures for the financial year 2014/15.

The opening balance as of 1 July 2014 has been prepared in applying the rules under IFRS. This has not resulted in any adjustments to the items in the opening balance.

Segment information
For management purposes, the Company is managed and operated as one business unit that is reflected in the organizational structure and internal reporting. No separate lines of business or separate business entities have been identified with respect to any product candidate or geographical market and no segment information is currently disclosed in the Company's internal reporting. Accordingly, it has been concluded that it is not relevant to include segment disclosures in the financial statements as the Company's business activities are not organized into business units, products or geographical areas.

Foreign currency transactions and balances
On initial recognition, transactions denominated in foreign currencies are translated at the foreign exchange spot rate at the transaction date. Differences arising between the foreign exchange spot rates at the transaction date and the date of payment are recognized in the income statement as financial income or financial expenses.

Receivables and payables and other monetary items denominated in foreign currencies are translated at the foreign exchange spot rates at the balance sheet date. The difference between the foreign exchange spot rates at the balance sheet date and the date at which the balance was recognized are recognized in the income statement as financial income or financial expenses.

Share-based payment
Employees and Management of the Company receive remuneration in the form of equity settled awards whereby services are rendered as consideration for warrants. The fair value of these equity settled awards is determined at the date of grant resulting in a fixed fair value at grant date that is not adjusted for future changes in the fair value of the equity awards that may occur over the service period. Fair value of warrants and options is determined using the Black Scholes model.

The cost of share based payments is recognized as an expense together with a corresponding increase in equity over the period in which the performance and/or service conditions are fulfilled. In the event that equity instruments are granted conditionally upon an equal number of equity instruments granted in prior periods not being exercised, they are treated as a new grant for the current period and a modification of the equity instruments granted in the prior period.

The fair value of equity settled awards is reported as compensation expense pro rata over the service period to the extent such awards are estimated to vest. No cost is recognized for awards that do not ultimately vest.

Employee benefits
Employee benefits are primarily made up of salaries, share-based payment and pension. The cost of these benefits is recognized as expense as services are delivered. The Group's contributions to the employee pension plan have not been material.

Leases
Leases that do not transfer substantially all the risks and rewards incident to the ownership to the Company are classified as operating leases. Payments relating to operating leases and any other leases are recognized in the income statement over the term of the lease.

The Company's aggregate liabilities relating to operating leases and other leases are disclosed under contingencies, etc.

Public grants
Public grants given to cover expenses are recognized in the income statement as a reduction to research and development expenses over the periods to which the costs relate. The terms of the grants do not obligate the Company to repay any of portion of the grant.

Financial instrument valuation hierarchy
Financial instruments recognized at fair value are allocated to one of the following valuation hierarchy levels:

- Level 1: Quoted (unadjusted) prices in active markets for identical assets or liabilities.
- Level 2: Other techniques for which all inputs that have a significant effect on the recorded fair value are observable, either directly or indirectly.
- Level 3: Techniques that use inputs that have a significant effect on the recorded fair value that are not based on observable market data.

No financial instrument is measured at fair value as of the balance sheet dates presented in the financial statements.

Property, plant and equipment
Property, plant and equipment includes fixtures, fittings, leasehold improvements and other plant and equipment, and are measured at cost less accumulated depreciation and impairment losses. Cost includes the acquisition price and costs directly related to the acquisition until the time at which the asset is ready for use. Depreciation is calculated on a straight-line basis over the expected useful lives of the underlying assets of five years. The residual values of equipment are not material. The useful life of and method of depreciation of equipment are reviewed by management at least each year-end or more often based on changes in facts and circumstances. Changes in useful lives or residual values are adjusted prospectively as changes in accounting estimates.

Property, plant and equipment are required to be tested for impairment when there are indications of impairment. Impairment tests are conducted at the individual asset level, or at the lowest level for which separately identifiable cash flows for groups of assets exist. Impaired assets or asset groups are written down to their recoverable amount, which is the higher of the value in use and the net realizable value of the asset or asset group, with impairment charges allocated proportionately to the assets within the impaired asset group.

Corporation tax receivable
Corporation tax receivable is recognized in the balance sheet as the tax benefit computed on the taxable loss for the year, adjusted for any changes to the prior year benefit due to changes in the taxable loss of prior years and for any taxes already paid or refunded. Management periodically evaluates positions taken in the tax returns with respect to situations in which applicable tax regulations are subject to interpretation or "uncertainty" and establishes provisions where appropriate. To date, there have been no provisions established for uncertain tax positions.

Deferred tax is measured using the balance sheet liability method on all temporary differences between the carrying amount and the tax value of assets and liabilities, with the exception of temporary differences occurring at the time of acquisition and liabilities neither effecting the result of operation nor the taxable income. The Company has no deferred tax balances as of 31 December 2016 or 2015 or as of 30 June 2015.

For further details please refer to note 2 and note 11.

Deposits
Deposits for property leased by the Company are measured at cost.

Prepayments
Prepayments include prepaid costs that will be incurred in subsequent financial reporting periods on a current basis.

Other receivables
Other receivables include prepaid costs that will be incurred on a noncurrent basis.

Financial assets
Initial recognition and measurement
Financial assets that meet certain criteria are classified at initial recognition as either financial assets at fair value through profit or loss, available for sale financial assets, held to maturity investments or receivables.

The Company's financial assets include other receivables and cash and cash equivalents. The Company does not hold assets that have been classified at fair value through profit or loss, available for sale or held to maturity. Generally, the Company's financial assets are available to support current operations; however, amounts expected to be realized within the next twelve months are classified within the statement of financial position as current assets.

The Company has no derivative financial assets nor has there been a change in classification of a financial asset after initial recognition and measurements as discussed herein. Financial assets are not acquired for trading or speculative purposes. The Company has not placed any assets as security for loans at either 31 December 2016 or 2015 or at 30 June 2015.

The Company's financial assets are recognized initially at fair value plus, in the case of financial assets not carried at fair value through profit and loss, transaction costs that are attributable to the acquisition of the financial asset, if any.

Subsequent measurement
Historically the Company's receivables are due within a short period and therefore the impact of using the effective interest rate method on the Company's financial statements has been immaterial. The same applies to cash and cash equivalents that comprise cash at banks available on demand.

Financial asset impairment
The Company assesses at the end of each reporting period whether there has been objective evidence that a financial asset or Company of financial assets may be impaired. Impairment losses are incurred if there is objective evidence of impairment and the evidence indicates that estimated future cash flows will be negatively impacted. For financial assets held at amortized costs, the amount of impairment loss to be recognized in the financial statements is measured as the difference between the carrying value of the financial asset and the present value of the expected cash flows of the financial asset using the original effective interest rate. The Company did not experience an impairment of a financial asset for either the twelve months ended at 30 June 2015, the six months ended 31 December 2015 or the twelve months ended 31 December 2016.

Other receivables
Receivables are measured at amortized cost. An impairment loss is recognized if there is an objective indication that a receivable or a group of receivables is impaired. Receivables that are concluded to be impaired are written down on an individual basis.

Cash and cash equivalents
Cash includes cash on hand and in banks, as well as short term marketable securities that are subject to an insignificant risk of changes in value.

Financial liabilities
The Company's financial liabilities historically have included bank debt, trade payables and other payables.

Bank debt
Bank debt of TDKK 245 as of 30 June 2015 had been repaid as of 31 December 2015. The Company has no bank debt as of 31 December 2016.

Trade payables
Trade payables relate to the Company's purchase of products and services from various vendors in the normal course of business.

Convertible debt
The Company in the past has issued convertible loans that meet certain technical requirements, including (but not limited to) settlement of the conversion option for a fixed number of the Company's ordinary shares, that are initially recognized at fair value, net of transaction costs incurred. Subsequently these convertible loans are measured at amortized cost and accounted for using the effective interest rate method. Gains and losses are recognized in the statement of profit or loss within other finance costs when the convertible loans are derecognized as well as through the effective interest rate amortization process. Amortized cost is calculated by taking into account any discount or premium from the face value of the convertible loan plus direct and incremental transaction costs incurred in connection with issuance of the convertible loan.

Other payables
Other payables are measured at net realizable value.

Statement of Profit or Loss and Other Comprehensive Income
Revenue
The Company does not have any revenue in any of the reporting periods.

Research and development costs
Research and development costs include salaries including share-based compensation and costs arising from research activities, clinical development, legal expenses related to the protection, defense and enforcement of the Company's intellectual property and rent associated with facilities used for research purposes. Given the uncertainty regarding the recoverability of clinical development costs, the Company has expensed all such expenses in the statement of loss and comprehensive loss for the periods presented.

Administrative expenses
Administrative expenses include salaries for administrative staff and management, costs of share-based payment and rent associated with facilities not used for research purposes.

Financial income and expense
Financial income and expense include interest income and expense, gains and losses due to changes in foreign exchange rates, interest expenses on convertible debt, allowances and surcharges related to the advance payment of tax scheme, and other miscellaneous items of financial income and expense.

Income tax benefit
Income tax benefit includes the current benefit due from the current period's taxable loss and deferred tax adjustments. The benefit is comprised primarily of refundable tax credits for costs incurred in connection with research and development activities under the Danish Tax Credit Regime.

Statement of cash flows
The statement of cash flows is presented using the indirect method and shows cash flows resulting from operating activities, investing activities, financing activities and the Company's cash and cash equivalents at the beginning and end of the year.

Cash flows used in operating activities primarily comprise the net loss for the year adjusted for non-cash items, such as foreign exchange gains and losses, depreciation, changes in working capital and cash received for interest and taxes.

Cash flows from investing activities are comprised primarily of investment in property, plant and equipment.

Cash flows from financing activities are comprised of repayment of bank debt and proceeds from capital increases net of transaction costs.

Note 2 - Significant accounting judgments, estimates and assumptions
The preparation of the financial statements requires management to make judgments, estimates and assumptions that affect the reported amounts of income, expenses, assets and liabilities as well as the accompanying disclosures. Uncertainty about these assumptions and estimates could result in outcomes that require a material adjustment to the carrying amount of the asset or liability affected in future periods.

Judgments made in applying accounting policies
In the process of applying the Company's accounting policies, management has made the following judgments and estimates that have the most significant effect on the amounts recognized in the financial statements. Refer to the following notes for more details:

- Estimation of accruals and prepaid costs for clinical research trials
- Judgment in respect of recognition of deferred taxes related to taxable losses to be carried forward (note 11)

Estimates and assumptions
The key assumptions concerning the future and other key sources of estimation uncertainty at the reporting date that have a significant risk of causing a material adjustment to the carrying amounts of assets and liabilities within the next financial year, are listed below. The Company based its assumptions and estimates on information available when the financial statements were prepared.

Estimation of accruals and prepaid costs for clinical trial development costs
The Company incurs substantial costs associated with clinical trials related to the AIDNPC program. The objective of the program is to develop a pharmaceutical drug for treatment of Niemann-Pick type C. Niemann-Pick type C (NPC) is a lysosomal storage disease affecting around 1 in 150,000 newborns and is caused by mutations in the NPC1 or NPC2 genes.

Accounting for clinical trials relating to activities performed by clinical research organizations (CROs) and other external vendors requires management to exercise significant estimates in regards to the timing and accounting for these costs. The diverse nature of services being provided by CROs and other arrangements, the different compensation arrangements that exists for each type of service and the limitations in respect of information related to certain clinical activities adds complexity to the estimation of accruals for services rendered by CROs and other vendors in connection with clinical trials. Furthermore, certain CROs and vendors are paid upfront in connection with clinical activities. In estimating the relevant periods etc., the Company evaluates the start-up, treatment and wrap-up periods, compensation arrangements and services rendered attributable to each clinical trial. Estimated costs are regularly tested against payment plans and trial completion assumptions.

The Company has recognized accruals related to clinical trial development costs of TDKK 0, TDKK 330 and TDKK 1,448 for the twelve months ended 31 December 2016, the six months ended 31 December 2015 and the twelve months ended 30 June 2015, respectively.

The Company has recognized prepaid costs related to clinical trial development costs of TDKK 3,794, TDKK 5,765 and TDKK 2,430 for the twelve months 31 December 2016, the six months ended 31 December 2015 and the twelve months ended 30 June 2015, respectively.

Judgment related to deferred taxes related to taxable losses to be carried forward
The Company recognizes deferred income tax assets if it is probable that sufficient taxable income will be available in the future against which the temporary differences and unused tax losses can be utilized. Management has considered future taxable income in assessing whether deferred income tax assets should be recognized and has concluded that the deferred income tax assets related to taxable losses to be carried forward do not meet the criteria for being recognized as assets in the statement of financial position.

The Company has net tax loss carry-forwards that are not recognized of MDKK 135, MDKK 97 and MDKK 81 for the twelve months 31 December 2016, the six months ended 31 December 2015 and the twelve months ended 30 June 2015, respectively.

The Company's tax losses can be carried forward infinitely subject to the general rules on limited deductibility due to ownership changes.

Reference is made to note 11.

Note 3 – Standards issued but not yet effective

The IASB has issued a number of new standards that become effective on or after 1 January 2017. Management's current expectation is that the new standards will be adopted by the Company at the effective date. Depending on the stage of development of the Company as of this point in time, the following new standards could have an impact of the financial statements:

IFRS 9 – Financial instruments
This standard addresses the accounting for financial assets and liabilities including their recognition, classification and measurement and hedge accounting. The Company does not anticipate adopting IFRS 9 before the mandatory effective date of 1 January 2018. The impact on the Company's financial statements of the future adoption of IFRS 9 will be determined based on facts and circumstances that exist at the time of adoption that cannot be predicted currently.

IFRS 16 – Leases

This standard introduces a single lessee accounting model and requires a lessee to recognize assets and liabilities for all leases with a term of more than twelve months, unless the underlying asset is of low value. A lessee is required to recognize a right of use asset representing its right to use the underlying leased asset and a lease liability representing its obligation to make lease payments. IFRS 16 has an effective date of 1 January 2019. The impact on the Group's financial statements of the future adoption of IFRS 16 will be determined based on facts and circumstances that exist at the time of adoption that cannot be predicted currently.

Note 4 – Government grant

Government grants comprise research funding from the Danish government and EU. Government grants are recognized in the period where the expenses funded by the grants have been incurred. Government grants are recognized as a reduction in research and development expenses as the grants are considered to be cost refunds. Income from government grants amount to TDKK 2,786, TDKK 1,003 and TDKK 0 respectively in 2016, 2015 and 2014/2015.

None of the government grants received are subject to repayment clauses.

Note 5 – Staff costs

(TDKK)	Twelve months ended 31 December 2016	Six months ended 31 December 2015	Twelve months ended 30 June 2015
Staff costs			
Wages/salaries	11,777	5,128	7,749
Share-based payment	-	-	-
Pensions	-	-	-
Other social security costs	117	30	57
Other staff costs	372	161	228
Total staff costs	**12,266**	**5,319**	**8,034**
Key management remuneration			
Wages/salaries	4,486	1,603	2,989
Share based payment	-	-	-
Pensions	60	-	-
Other social security costs	36	3	6
Other staff costs	-	-	-
Total key management remuneration	**4,582**	**1,606**	**2,995**
Total staff and key management costs	**16,848**	**6,925**	**11,029**
Administrative expenses	2,431	685	1,232
Research and development expenses	14,417	6,240	9,797
Total staff and key management costs	**16,848**	**6,925**	**11,029**

The amounts disclosed in the table above are the amounts recognized as an expense during the reporting periods. Key management consists of the Company's Chief Executive Officer, Chief Scientific Officer, Chief Financial Officer and Chief Medical Officer for the twelve months ended 31 December 2016. For the six months ended 31 December 2015 and the twelve months ended 30 June 2014 key management consisted of the Company's Chief Executive Officer and Chief Scientific Officer. See Note 14 for compensation paid to the members of the board of directors.

Note 6 – Warrants

The Company has issued warrants to employees, consultants providing similar services and key management. The warrants can be settled by subscribing for A-shares of the Company at an exercise price of DKK 44 per share. Due to the liquidation preference to B and C shares as discussed in note 10, the exercise price for the warrants were significantly above the fair value of one A-share at the respective issuance dates.

Management has applied a Black Scholes option valuation model to determine fair value of the warrants. Fair value of the warrants granted in 2014/2015 and 2015 respectively amounts to TDKK 3 and TDKK 12 respectively.

The most significant assumption applied is the underlying share price. Fair value of one A-share has been determined on the basis of the share of fair value of Orphazyme attributable to A-shares. Fair value of Orphazyme has been determined as the implied fair value, which can be derived from the subscription price in the most recent capital increase round prior to granting the warrants. Fair value per A-share has been determined to be in the range DKK 1.87 – DKK 3.72.

The table below summarizes the activity related to the warrants for the year ended 31 December 2016, the six-month period ended 31 December 2015 and the twelve months ended 30 June 2015:

(TDKK)	Key management	Employees	Board of Directors	Consultants	Total Warrants	Warrants exercisable
Outstanding at 30 June 2014	153,278	54,692	76,638	-	284,608	154,564
Granted	-	27,746	20,437	9,700	57,883	
Exercised	-	-	-	-	-	
Expired	-	-	-	-	-	
Forfeited	-	(12,773)	-	-	(12,773)	
Outstanding at 30 June 2015	**153,278**	**69,665**	**97,075**	**9,700**	**329,718**	**221,719**
Granted	58,601	21,789	27,047	-	107,437	
Exercised	-	-	-	-	-	
Expired	-	-	-	-	-	
Forfeited	-	(15,278)	-	-	(15,278)	
Outstanding at 31 December 2015	**211,879**	**76,176**	**124,122**	**9,700**	**421,877**	**266,621**
Granted	-	-	-	-	-	
Exercised	-	-	-	-	-	
Expired	-	-	-	-	-	
Outstanding at 31 December 2016	**211,879**	**76,176**	**124,122**	**9,700**	**421,877**	**324,078**

The weighted average remaining contractual life of the warrants outstanding as of 31 December 2016, 31 December 2015 and 30 June 2015 was 4.6 years, 5.6 years and 6.1 years, respectively.

Note 7 – Financial Income

(TDKK)	Twelve months ended 31 December 2016	Six months ended 31 December 2015	Twelve months ended 30 June 2015
Exchange gain	180	74	-
Other interest income	2	6	34
Total financial income	**182**	**80**	**34**

Note 8 – Financial expenses

(TDKK)	Twelve months ended 31 December 2016	Six months ended 31 December 2015	Twelve months ended 30 June 2015
Interest expense	71	26	14
Other financial expenses	26	14	15
Interest expense, convertible debt (note 15)	0	0	1,245
Exchange losses	0	0	129
Total financial expenses	**97**	**40**	**1,403**

Note 9 – Property, plant and equipment

(TDKK)	Fixtures and fittings, other plant and equipment	Leasehold improvements	Total
Cost at 30 June 2014	1,825	481	2,306
Additions	499	59	558
Cost at 30 June 2015	2,324	540	2,864
Additions	25	-	25
Cost at 31 December 2015	2,349	540	2,889
Additions	218	20	238
Disposals	0	(38)	(38)
Cost at 31 December 2016	**2,567**	**522**	**3,089**
Accum. depreciation at 30 June 2014	501	111	612
Depreciation expense	404	100	504
Accum. depreciation at 30 June 2015	905	211	1,116
Depreciation expense	232	54	286
Accum. depreciation at 31 December 2015	1,137	265	1,402
Depreciation expense	472	100	572
Write-down	0	134	134
Disposals	0	(6)	(6)
Accum. depreciation at 31 December 2016	**1,609**	**493**	**2,102**
Net book value at			
30 June 2014	1,324	370	1,694
30 June 2015	1,419	329	1,748
31 December 2015	1,212	275	1,487
31 December 2016	958	29	987

At the end of 2016 the Company started the process of moving to new premises. In connection with this the book value of the leasehold improvement has been impaired and written down to its expected net realizable value as of 31 December 2016. The write down has been recognized under research and development expenses in the statement of profit or loss and other comprehensive income. There has been no write down of property, plant and equipment in the six months ended 31 December 2015 and twelve months ended 30 June 2015.

Depreciation expense is included within operating loss as follows:

(TDKK)	Twelve months ended 31 December 2016	Six months ended 31 December 2015	Twelve months ended 30 June 2015
Administrative expenses	-	-	-
Research and development expenses	472	286	504
Total depreciation expense	**472**	**286**	**504**

Note 10 – Equity

The following table summarizes the Company's share activity:

	Class A ordinary shares	Class B preferred shares	Class C preferred shares
30 June 2014	**125,000**	**2,050,208**	**-**
Capital increase	-	-	827,165
Converted debt			215,658
30 June 2015	**125,000**	**2,050,208**	**1,042,823**
Capital increase	-		127,724
31 December 2015	**125,000**	**2,050,208**	**1,170,547**
Capital increase	-		14,786
31 December 2016	**125,000**	**2,050,208**	**1,185,333**

The share capital of the Company is divided into 3 classes, an A class, a B class and a C class. A class shares are ordinary shares. The B and C class shares both receive preference in all distributions for all amounts up to the amount paid in upon subscription plus an additional 10% compounded interest per year, with C class having the most senior preference. No distributions shall be made to the B class shares until the C class distributions have been satisfied, and likewise no distributions shall be made to the ordinary shares until the B class distributions have been satisfied. Once the B and C class distributions have been satisfied, any remaining distributions will be distributed on a pro rata basis amongst all issued shares of the Company.

Any B and C class share may at the request of its holder at any time be converted into an A class share (Conversion rate 1:1). The Board is authorized to adopt any changes to the Articles of Association required to implement any conversion request.

On January 6, 2015, the Company issued 1,704,554 indemnification warrants to the investors subscribing for C class shares at this date. The warrants entitle the holders to subscribe for additional C class shares at DKK 1 per share if certain liabilities of the Company prove higher than warranted by the Company in the Investment agreement. The warrants expiry on January 6, 2018 and it is not expected to be realised. The warrants are considered as an adjustment mechanism to the subscription price and not as separate derivative liabilities.

On February 5, 2015, the share capital was increased by 827,165 C class shares through issuance of shares for TDKK 74,445 in cash and issuance of 215,658 C class shares through conversion of convertible debt instruments with a principal amount of TDKK 15,527. The convertible bonds were issued on June 27, 2014 for proceeds of TDKK 14,920 that were received during July 2014. The conversion ratio was the lower of DKK 76.44 per share and the subscription price less 20% in a subsequent capital increase in excess of MEUR 10. The coupon rate was 8% p.a. Due to the variability in the conversion price, the conversion option was classified as a derivative financial instrument. Upon exercise, fair value of the conversion option TDKK 638 (for further details please refer to note 17) was transferred to equity and presented as part of the proceeds from the capital increase as they fulfill the requirements according to IFRS. In connection with the capital increase, the Company incurred expenses totaling TDKK 185.

As part of the subscription of C class shares in the six months ended 31 December 2015, the subscribers received 1,704,554 indemnification warrants to subscribe for additional C class shares at DKK 1 per share. The warrants may only be exercised in the event of the occurrence of one or more Warranty Claims as defined in the Investment Agreement dated December 30, 2014. Exercise of the warrants is further subject to the submission to the Company by the Owner of written notice of one or more Warranty Claims on or before January 6, 2016, or, in the event of a Warranty Claim which relates to tax issues, before January 6, 2018. The number of warrants exercisable is determined by the amount of the Claim. The warrants are accounted for as equity instruments.

On December 15, 2015, the share capital was increased by 127,724 C class shares though issuance of shares for TDKK 11,525 in cash. In connection with the capital increase, the Company incurred expenses totaling TDKK 30.

On December 9, 2016, share capital was increased by 14,786 C class shares through issuance of shares for TDKK 1,331 in cash. In connection with the capital increase, the Company incurred expenses totaling TDKK 30.

Until January 6, 2020, the Board of Directors are authorized to decide on one or more issues of warrants with the rights of subscription of 1 A class share per warrant in the Company in the total amount of up to TDKK 383 with addition of TDKK 245 without pre emptive subscription right for the Company's shareholders. The Board of Directors has issued a total of 462,701 warrants under the authorization with a right to subscribe for up to nominally DKK 462,701 A class shares. Consequently, warrants with a right to subscribe further A class shares in the nominal amount of DKK 165,299 may be issued under the authorization.

The Company has never paid a dividend on ordinary shares and does not expect to pay dividends for the foreseeable future.

Subsequent to 31 December 2016, the Company has finished a capital increase by issuing 534,007 C class shares to existing shareholders for a net proceeds received of MDKK 48 in January 2017. Furthermore, the Company completed a MDKK 104 financing round by issuing new C class shares to new shareholders in March 2017.

Note 11 – Income tax and deferred tax

(TDKK)	Twelve months ended 31 December 2016	Six months ended 31 December 2015	Twelve months ended 30 June 2015
Current tax on benefit on net loss	13,962	6,928	9,040
Adjustment to prior years	0	0	0
Tax credit research and development expenses	5,500	2,750	5,875
Change in unrecognized deferred tax before tax credit	(13,951)	(6,923)	(9,031)
Change in unrecognized deferred tax before tax credit	(11)	(5)	(9)
Total income tax benefit for the period	**5,500**	**2,750**	**5,875**

Reconciliation of effective tax rate to Danish statutory tax rate

(TDKK)	31 December 2016	31 December 2015	30 June 2015
Net loss before tax	(63,465)	(29,482)	(38,467)
Corporate income tax rate in Denmark	22%	23.5%	23.5%
Computed income tax benefit	**13,962**	**6,928**	**9,040**
Tax effect of:			
Adjustment to prior years	0	0	0
Other non-deductible expenses, including share-based compensation	(11)	(5)	(9)
Deferred tax asset not recognized	(8,451)	(4,173)	(3,156)
Total income tax benefit for the period	**5,500**	**2,750**	**5,875**

Deferred tax in the statement of financial position

(TDKK)	31 December 2016	31 December 2015	30 June 2015
Tax deductible losses	29,716	21,425	17,848
Other temporary differences	(113)	(41)	(80)
	29,603	21,384	17,768
Deferred tax asset not recognized	(29,603)	(21,384)	(17,768)
Carrying amount included on statement of financial position	0	0	0

The Company had net tax loss carry-forwards in Denmark for income tax purposes of MDKK 135, MDKK 97 and MDKK 81 million as of 31 December 2016 and 2015 and 30 June 2015.

Income tax benefit for the year includes a tax credit for research and development at the applicable tax rate under the Danish Corporate Income Tax Act.

The tax loss carry forwards have no expiry date. The Company's ability to use tax loss carry forwards in any one year is limited to 100% of the first MDKK 7.5 of taxable income plus 60% of taxable income above MDKK 7.5.

The Company recognizes deferred tax assets, including the tax base of tax loss carry forwards, if management assesses that these tax assets can be offset against positive taxable income within a foreseeable future. Significant management judgment is required to determine the amount of deferred tax assets that can be recognized, based upon the likely timing and the level of future taxable profits together with future tax planning strategies. This judgment is made periodically after considering current facts and circumstances, budgets and business plans as well as the risks and uncertainty associated with the Company's ability successfully commercialize and defend its intellectual property.

Significant judgment
The development of therapeutic products within the biopharmaceutical industry is subject to significant risks and uncertainties and there is no assurance a therapeutic product will be successfully developed. As the result of this uncertainty and since the Company has reported significant losses since inception, has no commercial products or revenues and does not expect to generate revenues or profits for the foreseeable future, management has concluded that deferred tax assets should not be recognized as of 31 December 2016 or at any other prior date. The tax assets are currently not deemed to meet the criteria for recognition as management is not able to provide convincing positive evidence that taxable profits will be available in the future to utilize the benefit from the tax assets.

As of 31 December 2016 there are no tax audits in process nor has management been notified of any pending tax audit. As of 31 December 2016, the tax years that remain open for audit by the Danish tax authorities include 2012 through 2015.

Note 12 – Loss per share

The following reflects the net loss attributable to both preferred and ordinary shareholders and share data used in the basic and diluted loss per share computations for the twelve months ended 31 December 2016 and 2015 and the six months ended 31 December 2015:

(TDKK)	Twelve months ended 31 December 2016	Six months ended 31 December 2015	Twelve months ended 30 June 2015
Loss for the period	(57,935)	(26,732)	(32,592)
Preference dividend attributable to B shares	(18,367)	(8,746)	(15,902)
Preference dividend attributable to C shares	(10,982)	(4,702)	(3,574)
Loss attributable to all share classes	(87,284)	(40,180)	(52,068)
Weighted average A shares outstanding	125,000	125,000	125,000
Weighted average B shares outstanding	2,050,208	2,050,208	2,050,208
Weighted average C shares outstanding	1,171,410	1,048,145	391,059
Total weighted average shares outstanding	**3,346,618**	**3,223,353**	**2,566,267**
Loss attributable to all share classes	(87,284)	(40,180)	(52,068)
Total weighted average shares outstanding	3,346,618	3,223,353	2,566,267
Loss per share, A shares	**(26.08)**	**(12.47)**	**(20.29)**
Preference dividend attributable to B shares	18,367	8,746	15,902
Pro rata share of loss	(53,471)	(25,557)	(41,597)
Loss attributable to B shares	(35,104)	(16,811)	(25,695)
Weighted average B shares outstanding	2,050,208	2,050,208	2,050,208
Loss per share, B shares	**(17.12)**	**(8.20)**	**(12.53)**
Preference dividend attributable to C shares	10,982	4,702	3,574
Pro rata share of loss	(30,551)	(13,066)	(7,934)
Loss attributable to C shares	(19,569)	(8,364)	(4,360)
Weighted average C shares outstanding	1,171,410	1,048,145	391,059
Loss per share, C shares	**(16.71)**	**(7.98)**	**(11.15)**

Basic loss per share amounts are calculated by dividing the net loss for the period attributable to each share class by the weighted average number of shares outstanding during each the period. Due to the fact that the Company has incurred losses for each period presented, the potential A class shares issuable related to outstanding warrants have been excluded from the calculation of diluted loss per share as the effect of such shares is anti dilutive. There are not outstanding warrants related to the B or C class shares, therefore, basic and diluted loss per share are the same for each period presented.

Subsequent to 31 December 2016, the Company has finished two capital increases increasing the number of shares with a total of 534,007, which will have an impact on the calculation of earnings per share from 2017 and forward.

Note 13 – Capital management

For the purpose of the Company's capital management, capital includes issued capital, share premium and all other equity reserves attributable to the equity holders of the Company. The primary objective of the Company's capital management is to maximize shareholder value. The board of directors' policy is to maintain a strong capital base so as to maintain investor, creditor and market confidence, and a continuous advancement of the Company's intellectual property, product pipeline and business. Cash, cash equivalents and financial assets are monitored on a regular basis by management and the board of directors in assessing current and long term capital needs. As of 31 December 2016 the Company held cash and cash equivalents totaling MDKK 14.3 that together with the subsequent capital increases will be sufficient to provide adequate funding to allow the Company to meet its planned operating activities, including increased levels of research and development activities, in the normal course of business for the next twelve months. The Company currently has no significant planned capital expenditures.

Subsequent to 31 December 2016, the Company has finished two capital increases for a total of MDKK 152, where MDKK 65 has been paid in cash and thereby increase the cash and cash equivalents and MDKK 87 is recognized as a receivable. MDKK 52 is due when the Board of Directors requests the funds. The remaining MDKK 35 is due upon on reaching a specific milestone where MDKK 9 is immediately due and the remaining MDKK 26 is due when the Board of Directors requests the funds.

The Company's activities expose it to a number of financial risks whereby future events, which can be outside the control of the Company, could have a material effect on the Company's financial position and results of operations. The known risks include foreign currency, interest and credit risk and there could be other risks currently unknown to management. The Company historically has not hedged its financial risks.

Foreign Currency

The Company maintains operations in Denmark and uses the DKK as its functional currency. The Company conducts cross border transactions where the functional currency is not always used. Accordingly, future changes in the exchange rates of the DKK, the EUR, the USD and/or the GBP will expose the Company to currency gains or losses that will impact the reported amounts of assets, liabilities, income and expenses and the impact could be material. For the year ended 31 December 2016 and 2015 and the six months ended 31 December 2015 the impact on the Company's statement of loss for possible changes in the EUR, USD and GBP exchange rates against the Company's functional currency of DKK would be as follows:

Currency, (TDKK)	Possible change	Twelve months ended 31 December 2016	Six months ended 31 December 2015	Twelve months ended 30 June 2015
EUR	+/2%	+177 / (177)	+611 / (611)	+704 / (704)
USD	+/10%	(21) / +21	+26 / (26)	+591 / (591)
GBP	+/10%	(327) / +327	(153) / +153	+486 / (486)

Interest Rate Risk

The Company has no interest bearing debt.

Credit Risk

The Company's credit risk is associated with cash held in banks. The Company does not trade financial assets for speculative purposes and invests with the objective of preserving capital.

The Company's cash and cash equivalents are held primarily at two banks in Denmark with Moody's long term credit ratings exceeding of A1.

Note 14 – Contractual obligations and contingencies

Contractual obligations
The Company has the following non-cancelable contractual obligations related to its lease and other rent liabilities:

Contractual obligations, (TDKK)	Twelve months ended 31 December 2016	Six months ended 31 December 2015	Twelve months ended 31 June 2015
0 – 1 years	418	284	278
1 – 5 years	-	-	-
More than 5 years	-	-	-
Total	**418**	**284**	**278**

Total expense under non-cancelable contractual obligations was TDKK 848, TDKK 397 and TDKK 647 for the twelve months 31 December 2016, the six months ended 31 December 2015 and the twelve months ended 30 June 2015, respectively.

The Company furthermore has contracts with CRO's (Clinical Research Organizations), where the CRO will carry out clinical trials for the Company. There is a contract in place between the parties but the amounts are not fixed as it depends on reaching milestones, number of enrolled patients etc.

Contingencies
Contingencies are assets and liabilities that arise from past events but whose existence will only be confirmed by the occurrence or non occurrence of future events that in some situations are beyond the Company's control. As of 31 December 2016 and 2015 and 30 June 2015 there are no contingent assets or liabilities.

Note 15 – Related party disclosures

The Company is not ultimately controlled by any of the investors. See Note 5 for additional related party transactions, related to the remuneration paid to key management. In addition to Novo A/S, Kurma Biofund II, Idinvest, Sunstone Life Science Ventures Fund II K/S and Cooperative Aescap Venture I U.A. all owns more than 5%.

There have been no transactions between related parties in the 12 months ended 31 December 2016 and the six months ended 31 December 2015 besides capital increases as described in note 10. In the twelve months ended 30 June 2015 in addition to capital increases there was also a convertible debt, that was converted to shares in the Company. The interest expense on the convertible debt was TDKK 1,245 up to the date of conversion. Reference is made to note 17.

Terms and conditions of transactions with related parties
Amounts due to related parties are uncollateralized and interest free. There have been no guarantees provided or received for any related party receivables or payables. For the twelve months ended 31 December 2016, the six months ending 31 December 2015 and the twelve months ended 30 June 2015 the Company has not recorded any impairment of receivables relating to amounts owed by related parties. There are no related party receivables at any of the balance sheet dates.

Transactions with key management
The Company has not granted any loans, guarantees, or other commitments to or on behalf of any of the members of the board of directors or key management personnel.

Other than the remuneration described in Note 5, no other significant transactions have taken place with key management personnel during the period presented herein.

Compensation paid to members of the board of directors
Compensation paid to members of the board of directors are classified as administrative expense within the statement of loss. The following table lists compensation paid to members of the board of directors:

(TDKK)	Twelve months ended 31 December 2016	Six months ended 31 December 2015	Twelve months ended 30 June 2015
Director fees	524	795	522
Warrants	0	0	0
Other fees	0	0	0
Total compensation paid to the board of directors	**524**	**795**	**522**

Certain members of the board of directors have received warrants in the Company (reference is made to note 6). Besides warrants and the director fees presented below, they have not received any other remuneration for their services.

Transactions with shareholders and affiliates

There have been no transactions with shareholders or affiliates of shareholders during the twelve months ending 31 December 2016, the six months ended 31 December 2015 or the twelve months ending 30 June 2015, except for the capital increase disclosed in Note 10. In the twelve months ended 30 June 2015 the Company finished a capital increase with Cooperative Aescap Venture I U.A., Novo A/S and Sunstone Life Science Ventures Fund II K/S for a total of MDKK 74.

Note 16 – Subsequent events

Management has evaluated its financial statements for potential subsequent events occurring after the balance sheet date of 31 December 2016 but prior to the date that these financial statements were issued. During Q1 2017, the Company's 2015 financing round was extended, with 534,007 C class preferred shares issued to existing shareholders for total net proceeds received of MDKK 48 including issuance costs of TDKK 79.

On March 8, 2017 the Company completed a MDKK 104 financing round by issuing new shares to LSP V Coöperatieve U.A. and ALS Investment Fund. MDKK 69 has been invested and MDKK 35 is dependent on certain future milestones.

Other than the event disclosed above, there were no other events that were required to be reported or disclosed that are not already included within these financial statements.

Note 17 – First time adoption of IFRS

These financial statements are the first financial statements presented in accordance with IFRS as adopted by the European Union. The date of transition is 1 July 2014. As the Company is presenting financial statements in accordance with IFRS as adopted by the European Union for the first time, estimates for each balance sheet date has been revisited and updated with the most recent information.

Under IFRS, fair value of warrants issued to employees as remuneration for their services is recognized as an expense over the vesting period. Under previous GAAP, no expense has been recognized for these warrants.

Convertible bonds issued and converted into equity in 2014/2015 comprises a conversion option with a conversion price which was not fixed. Consequently, the conversion option is a derivative financial liability which shall be accounted for separately from the host debt contract and measured at fair value through profit or loss. Under previous GAAP, the convertible bond was classified in full as a financial liability.

The change in accounting policy has resulted in an increase in financial expenses of TDKK 638 in 2014/2015. It has not affected equity on any of the balance sheet dates presented because the bond was issued and converted within the same financial year.

Note 18 – Fees paid to auditors appointed at the annual general meeting

(TDKK)	Twelve months ended 31 December 2016	Six months ended 31 December 2015	Twelve months ended 30 June 2015
Fee for statutory audit	200	40	77
Assurance engagements	0	0	0
Tax consultancy	0	10	52
Other assistance	40	0	287
	240	**50**	**416**

Statement by the Executive Management and the Board of Directors on the 2015 Comparative Financial Statements for the period 1 January – 31 December 2015

The Board of Directors and the Executive Management have today considered and approved the 2015 Comparative Financial Statements.

The 2015 Comparative Financial Statements have been prepared in accordance with the basis of preparation as set out in note 1.

In our opinion, the 2015 Comparative Financial Statements have in all material respects been prepared in accordance with the basis of preparation and gives a fair presentation of the Company's assets, liabilities and financial position at 31 December 2015 and of the results of the Company's operations and cash flows for the period 1 January - 31 December 2015.

Copenhagen, 6 November 2017

Board of Directors

Georges Gemayel
Chairman

Bo Jesper Hansen
Deputy Chairman

Martin Bonde
Board Member

Martin Rahbek Kornum
Board Member

Martijn Kleijwegt
Board Member

Nanna Lüneborg
Board Member

Patrick J.H. Krol
Board Member

Rémi Droller
Board Member

Sten Verland
Board Member

Executive Management

Anders Mørkeberg Hinsby
CEO

Anders Vadsholt
CFO

Independent auditor's review report on the 2015 unaudited Comparative Financial Statements

To the shareholders and potential shareholders
We have reviewed the financial statements for the period 1 January - 31 December 2015 (the "2015 Comparative Financial Statements"), which comprise statement of profit or loss and other comprehensive income, statement of financial position, statement of changes in shareholders' equity, statement of cash flows and notes.

The 2015 Comparative Financial Statements have been prepared by management in accordance with the basis of preparation set out in note 1 of the 2015 Comparative Financial Statements.

Management's responsibilities for the 2015 Comparative Financial Statements
Management is responsible for the preparation of the 2015 Comparative Financial Statements and for such internal control as Management determines is necessary to enable the preparation of the 2015 Comparative Financial Statements that are free from material misstatement, whether due to fraud or error.

Auditor's responsibilities
Our responsibility is to express a conclusion on the 2015 Comparative Financial Statements. We conducted our review in accordance with the International Standard on Review of Interim Financial Information Performed by the Independent Auditor of the Entity and additional requirements applicable in Denmark.

This requires us to conclude whether anything has come to our attention that causes us to believe that the 2015 Comparative Financial Statements, taken as a whole, are not prepared, in all material respects, in accordance with the basis of preparation set out in note 1 of the 2015 Comparative Financial Statements. This standard also requires us to comply with ethical requirements.

A review of the 2015 Comparative Financial Statements in accordance with the International Standard on Review of Interim Financial Information Performed by the Independent Auditor of the Entity is a limited assurance engagement. The auditor performs procedures primarily consisting of making enquiries of Management and others within the company, as appropriate, applying analytical procedures and evaluate the evidence obtained.

The procedures performed in a review are substantially less that those performed in an audit conducted in accordance with the International Standards on Auditing. Accordingly, we do not express an audit opinion on the 2015 Comparative Financial Statements.

Conclusion
Based on our review, nothing has come to our attention that causes us to believe that the financial statements for the period 1 January - 31 December 2015 are not prepared, in all material respects, in accordance with the basis of preparation set out in note 1 of the 2015 Comparative Financial Statements.

Copenhagen, 6 November 2017

ERNST & YOUNG
Godkendt Revisionspartnerselskab
CVR no. 30 70 02 28

Christian Schwenn Johansen
State Authorised Public Accountant

Lars Hansen
State Authorised Public Accountant

Statement of Profit or Loss and Other Comprehensive Income

(TDKK)	Note	Twelve months ended 31 December 2015
Research and development expenses	2	(45,865)
Administrative expenses	2	(7,220)
Operating loss		**(53,085)**
Net financials		(317)
Net loss before tax		**(53,402)**
Income tax benefit	4	5,688
Net loss for the period		**(47,714)**
Other comprehensive income/(loss)		-
Total comprehensive loss		**(47,714)**
Loss per share, basic and diluted	5	
Class C preferred shares		(14.56)
Class B preferred shares		(15.05)
Class A ordinary shares		(23.19)

Statement of Financial Position

(TDKK)	Note	31 December 2015
Assets		
Non-current assets		
Property, plant and equipment		1,487
Corporation tax receivable	4	2,750
Deposits		211
Total non-current assets		**4,448**
Current assets		
Corporation tax receivable	4	5,875
Other receivables		644
Prepayments		5,970
Cash and cash equivalents		68,015
Total current assets		**80,504**
Total assets		**84,952**
Equity and liabilities		
Equity		
Share capital		3,346
Share premium		224,999
Accumulated deficit		(154,202)
Total equity	**3**	**74,143**
Current liabilities		
Bank debt		-
Trade payables		2,447
Other payables		8,362
Total current liabilities		**10,809**
Total equity and liabilities		**84,952**

Statement of Changes in Shareholders' Equity

(TDKK)	Note	Share Capital	Share Premiuim	Accumulated deficit	Total
Balance as of 1 January 2015		**2,175**	**124,250**	**(106,488)**	**19,937**
Net loss for the period		-	-	(47,714)	(47,714)
Other comprehensive loss for the period		-	-	-	-
Transactions with owners					
Capital increase		955	85,015	-	85,970
Conversion of debt		216	15,949	-	16,165
Expenses, capital increase		-	(215)	-	(215)
Share-based payment expense		-	-	-	-
Balance as of 31 December 2015	**3**	**3,346**	**224,999**	**(154,202)**	**74,143**

Statement of Cash Flows

(TDKK)	Twelve months ended 31 December 2015
Operating activities	
Net loss before tax	(53,402)
Adjustments to reconcile loss before tax to cash flows from operating activities	
Share-based payment expense	0
Depreciation and write-down	554
Gain/loss on sale and disposal of assets	-
Change in other receivables	(277)
Change in prepayments	(4,969)
Change in trade payables	858
Change in other payables	4,330
Cash flows from taxes	6,250
Interest paid	241
Net cash flow used in operating activities	**(46,414)**
Investing activities	
Investment in property, plant and equipment	(495)
Net cash flow used in investing activities	**(495)**
Financing activities	
Capital contributions from shareholders	85,970
Cash from convertible loan	-
Bank loans	-
Expenses related to capital contributions	(215)
Net cash provided by financing activities	**85,755**
Net change in cash and cash equivalents	**38,846**
Net foreign exchange differences	-
Cash and cash equivalents at the beginning of the period	29,169
Cash and cash equivalents at the end of the period	**68,015**

Notes to the comparative financial statements

Note 1 – Accounting policies

Basis of Presentation and accounting policies
The Company has previously undertaken statutory financial reporting based on a financial year covering the period from 1 July to 30 June; however, with effect from the calendar year 2016, the Company's financial year was converted to the period 1 January to 31 December.

The 2015 Comparative Financial Statements have been prepared in order to present the Company's historical financial information on a comparative basis with the statutory financial statements for the calendar year 2016.

The 2015 Comparative Financial Statements were prepared and approved by the Executive Management and the Board of Directors on 6 November 2017.

The 2015 Comparative Financial Statements comprise statement of profit or loss and other comprehensive income, statement of financial position, statement of changes in shareholders' equity, statement of cash flows and certain selected notes and as such these have not been prepared in accordance with International Financial Reporting Standards ("**IFRS**") as a number of note disclosures are not included.

However, the 2015 Comparative Financial Statements have in respect of recognition and measurement been prepared in accordance with the accounting policies applied in the financial statements at 31 December 2016 and for the period 1 January - 31 December 2016 with comparative figures for the financial year 1 July 2015 - 31 December 2015 and 1 July 2014 - 30 June 2015 as set out on pages F-21 to F-24, which was prepared in accordance with IFRS.

Management Judgments and Estimates
The preparation of the 2015 Comparative Financial Statements requires management to make judgments, estimates and assumptions that affect the reported amounts of income, expense, asset and liabilities as well as accompanying disclosures. The most significant judgments, estimates and assumptions include, among other things, accrual and prepaid costs for clinical trial development costs and deferred tax assets. For additional descriptions of significant judgments and estimates, refer to note 2 in audited financial statements at and for the financial year 1 January 2016 - 31 December 2016 with comparative figures for the financial year 1 July 2015 - 31 December 2015 and 1 July 2014 - 30 June 2015 discussed on pages F-24 to F-25.

Note 2 – Warrants

The Company has issued warrants to employees, consultants providing similar services and key management. The warrants can be settled by subscribing for A-shares of the Company at an exercise price of DKK 44 per share. Due to the liquidation preference to B and C shares as discussed in note 10, the exercise price for the warrants were significantly above the fair value of one A-share at the respective issuance dates.

Management has applied a Black Scholes option valuation model to determine fair value of the warrants. Fair value of the warrants granted in the period 1 January 2015 - 31 December 2015 amounts to TDKK 6.

The most significant assumption applied is the underlying share price. Fair value of one A-share has been determined on the basis of the share of fair value of Orphazyme attributable to A-shares. Fair value of Orphazyme has been determined as the implied fair value, which can be derived from the subscription price in the most recent capital increase round prior to granting the warrants. Fair value per A-share has been determined to be in the range DKK 1.87 – DKK 3.72.

The table below summarizes the activity related to the warrants for the year ended 31 December 2015:

(TDKK)	Key management	Employees	Board of Directors	Consultants	Total Warrants	Warrants exercisable
Outstanding at 31 December 2014	153,278	80,238	97,075	-	330,591	204,207
Granted	58,601	23,989	27,047	9,700	119,337	
Forfeited	-	(28,051)	-	-	(28,051)	
Outstanding at 31 December 2015	**211,879**	**76,176**	**124,122**	**9,700**	**421,877**	**266,621**

The weighted average remaining contractual life of the warrants outstanding as of 31 December 2015 was 5.6 years.

Note 3 – Equity

On 6 January 2015, the Company issued 1,704,554 indemnification warrants to the investors subscribing for C class shares at this date. The warrants entitle the holders to subscribe for additional C class shares at DKK 1 per share if certain liabilities of the Company prove higher than warranted by the Company in the Investment agreement. The warrants expiry on 6 January 2018 and it is not expected to be realised. The warrants are considered as an adjustment mechanism to the subscription price and not as separate derivative liabilities.

On 5 February 2015, the share capital was increased by 827,165 C class shares through issuance of shares for TDKK 74,445 in cash and issuance of 215,658 C class shares through conversion of convertible debt instruments with a principal amount of TDKK 15,527. The convertible bonds were issued on June 27, 2014 for proceeds of TDKK 14,920 that were received during July 2014. The conversion ratio was the lower of DKK 76.44 per share and the subscription price less 20% in a subsequent capital increase in excess of MEUR 10. The coupon rate was 8% p.a. Due to the variability in the conversion price, the conversion option was classified as a derivative financial instrument. Upon exercise, fair value of the conversion option TDKK 638 (for further details please refer to note 17 in audited financial statements at and for the financial year 1 January 2016 - 31 December 2016 with comparative figures for the financial year 1 July 2015 - 31 December 2015 and 1 July 2014 - 30 June 2015 discussed on page F-36) was transferred to equity and presented as part of the proceeds from the capital increase as they fulfill the requirements according to IFRS. In connection with the capital increase, the Company incurred expenses totaling TDKK 185.

As part of the subscription of C class shares in the twelve months ended 31 December 2015, the subscribers received 1,704,554 indemnification warrants to subscribe for additional C class shares at DKK 1 per share. The warrants may only be exercised in the event of the occurrence of one or more Warranty Claims as defined in the Investment Agreement dated December 30, 2014. Exercise of the warrants is further subject to the submission to the Company by the Owner of written notice of one or more Warranty Claims on or before 6 January 2016, or, in the event of a Warranty Claim which relates to tax issues, before 6 January 2018. The number of warrants exercisable is determined by the amount of the Claim. The warrants are accounted for as equity instruments.

On 15 December 2015, the share capital was increased by 127,724 C class shares though issuance of shares for TDKK 11,525 in cash. In connection with the capital increase, the Company incurred expenses totaling TDKK 30.

Note 4 – Income tax and deferred tax

Income tax benefit for the year includes an estimated tax credit for research and development at the applicable tax rate under the Danish Corporate Income Tax Act.

The tax loss carry forwards have no expiry date. The Company's ability to use tax loss carry forwards in any one year is limited to 100% of the first MDKK 7.5 of taxable income plus 60% of taxable income above MDKK 7.5.

The Company recognizes deferred tax assets, including the tax base of tax loss carry forwards, if management assesses that these tax assets can be offset against positive taxable income within a foreseeable future. Significant management judgment is required to determine the amount of deferred tax assets that can be recognized, based upon the likely timing and the level of future taxable profits together with future tax planning strategies. This judgment is made periodically after considering current facts and circumstances, budgets and business plans as well as the risks and uncertainty associated with the Company's ability successfully commercialize and defend its intellectual property.

Note 5 – Loss per share

The following reflects the net loss attributable to both preferred and ordinary shareholders and share data used in the basic and diluted loss per share computations for the twelve months ended 31 December 2015:

(TDKK)	Twelve months ended 31 December 2015
Loss for the period	(47,714)
Preference dividend attributable to B shares	(16,697)
Preference dividend attributable to C shares	(8,274)
Loss attributable to all share classes	(72,685)
Weighted average A shares outstanding	125,000
Weighted average B shares outstanding	2,050,208
Weighted average C shares outstanding	958,701
Total weighted average shares outstanding	3,133,909
Loss attributable to all share classes	(72,685)
Total weighted average shares outstanding	3,133,909
Loss per share, A shares	**(23.19)**
Preference dividend attributable to B shares	16,697
Pro rata share of loss	(47,550)
Loss attributable to B shares	(30,853)
Weighted average B shares outstanding	2,050,205
Loss per share, B shares	**(15.05)**
Preference dividend attributable to C shares	8,274
Pro rata share of loss	(22,235)
Loss attributable to C shares	**(13,961)**
Weighted average C shares outstanding	958,701
Loss per share, C shares	**(14.56)**

Basic loss per share amounts are calculated by dividing the net loss for the period attributable to each share class by the weighted average number of shares outstanding during each the period. Due to the fact that the Company has incurred losses for each period presented, the potential A class shares issuable related to outstanding warrants have been excluded from the calculation of diluted loss per share as the effect of such shares is anti dilutive. There are not outstanding warrants related to the B or C class shares, therefore, basic and diluted loss per share are the same for each period presented.

Note 6 – Related party disclosures

The Company is not ultimately controlled by any of the investors. See note 15 in audited financial statements at and for the financial year 1 January 2016 - 31 December 2016 with comparative figures for the financial year 1 July 2015 - 31 December 2015 and 1 July 2014 - 30 June 2015 discussed on pages F-35 to F-36 for additional information on related party transactions. In addition to Novo A/S, Kurma Biofund II, Idinvest, Sunstone Life Science Ventures Fund II K/S and Cooperative Aescap Venture I U.A. all owns more than 5%.

There have been no transactions between related parties in the twelve months ended 31 December 2015 besides capital increases as described in note 3. In the twelve months ended 31 December 2015 in addition to capital increases there was also a convertible debt, that was converted to shares in the Company. The interest expense on the convertible debt was TDKK 1,245 up to the date of conversion.

Terms and conditions of transactions with related parties

Amounts due to related parties are uncollateralized and interest free. There have been no guarantees provided or received for any related party receivables or payables. For the twelve months ended 31 December 2015 the Company has not recorded any impairment of receivables relating to amounts owed by related parties. There are no related party receivables at any of the balance sheet dates.

Transactions with key management

The Company has not granted any loans, guarantees, or other commitments to or on behalf of any of the members of the board of directors or key management personnel.

Other than the remuneration, no other significant transactions have taken place with key management personnel during the period presented herein.

Compensation paid to members of the board of directors

Compensation paid to members of the board of directors are classified as administrative expense within the statement of loss. The following table lists compensation paid to members of the board of directors:

(TDKK)	Twelve months ended 31 December 2015
Director fees	765
Warrants	0
Other fees	0
Total compensation paid to the board of directors	**765**

Certain members of the board of directors have received warrants in the Company (reference is made to note 2). Besides warrants and the director fees presented below, they have not received any other remuneration for their services.

Transactions with shareholders and affiliates

There have been no transactions with shareholders or affiliates of shareholders during the twelve months ending 31 December 2015, except for the capital increase disclosed in Note 3. In the twelve months ended 31 December 2015 the Company finished a capital increase with Cooperative Aescap Venture I U.A., Novo A/S and Sunstone Life Science Ventures Fund II K/S for a total of MDKK 74.

Note 7 – Subsequent events

Subsequent events are discussed in the audited financial statements at and for the financial year 1 January 2016 - 31 December 2016 with comparative figures for the financial year 1 July 2015 - 31 December 2015 and 1 July 2014 - 30 June 2015 on pages F-17 to F-36 and in the condensed interim financial statements at and for the six months ended 30 June 2017 and 2016 on pages F-5 to F-13.

Annex A – Excerpt from Articles of Association

Articles of Association

Orphazyme A/S
Registration (CVR) no. 32266355

1 Name and objects

1.1. The Company's name is Orphazyme A/S.

1.2. The Company's objects are research, development, production, marketing, sales and/or licensing of medicinal products for treatment of various disorders, including lysosomal storage diseases (LSD), neuromuscular disorders and other related diseases, as well as to carry out associated activities. Furthermore, the Company may, within its line of business, participate in partnerships or co-operate with other businesses.

2 Share capital and shares

2.1. The Company's nominal share capital is DKK 11,590,092, divided into shares of DKK 1 each or multiples thereof.

2.2. The share capital has been fully paid up.

2.3. The shares shall be issued in the name of the holder and shall be recorded in the name of the holder in the Company's register of shareholders.

2.4. The register of shareholders is kept by VP SECURITIES A/S, CVR no. 21 59 93 36.

2.5. The shares are negotiable instruments. No restrictions shall apply to the transferability of the shares.

2.6. No shares shall carry special rights.

2.7. No shareholder shall be under an obligation to have his/her shares redeemed in whole or in part by the Company or by any third party.

2.8. The shares are registered with and issued in dematerialised form through VP SECURITIES A/S, CVR no. 21 59 93 36. Dividend is paid out through VP SECURITIES A/S and is deposited at the registered dividend accounts at VP SECURITIES A/S. Rights concerning the shares shall be notified to VP Securities A/S in accordance with applicable rules.

3 Increase of share capital

3.1. In the period until 31 December 2017, the Board of Directors is authorised to increase the Company's share capital in one or more issues of new shares without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 9,400,000. The capital increase shall take place at market price as determined through a book-building process and shall be effected by cash payment.

3.2. In the period until 31 December 2017, the Board of Directors is authorised to increase the Company's share capital in one or more issues of new shares without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 1,500,000. The capital increase shall take place at the offer price as determined through a book-building process in connection with the Company's initial public offering and admission to trading and official listing on Nasdaq Copenhagen A/S and shall be effected by cash payment.

3.3. In the period until 31 December 2017, the Board of Directors is authorised to increase the Company's share capital in one or more issues without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 1,700,000 in connection with directed issues of bonus shares to one or more shareholders. The capital increase shall take place at par value (i.e. below market price).

3.4. In the period until 2 November 2022, the Board of Directors is authorised to increase the Company's share capital in one or more issues of new shares without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 5,000,000. The capital increase shall take place at market price and may be effected by cash payment, conversion of debt or by contribution of other assets than cash.

3.5. In the period until 2 November 2022, the Board of Directors is authorised to increase the Company's share capital in one or more issues without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 1,300,000 in connection with the issue of new shares to members of the Board of Directors, executives and/or employees of the Company. The new shares shall be issued against cash payment at a subscription price to be determined by the Board of Directors, which may be below the market price.

3.6. In the period until 2 November 2022, the Board of Directors is authorised to increase the Company's share capital in one or more issues without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 15,750,000 in connection with directed issues of bonus shares , and/or directed issues of new shares effected by cash payment, to Kansas Life Sciences Development Inc. and UCL Business PLC (or entities designated by them), respectively. The capital increase shall take place at par value (i.e. below market price). The value of such new shares to be issued can in any case not exceed a maximum of USD 2.5 million with a fixed exchange rate of DKK 6.30 per 1 USD based on the average closing price of the Company's shares on Nasdaq Copenhagen A/S for the 30 days immediately prior to the date of issuance.

3.7. New shares issued pursuant to Articles 3.1, 3.2, 3.3, 3.4, 3.5 and 3.6 shall be fully paid up, issued in the name of the holder and shall be recorded in the name of the holder in the Company's register of shareholders, shall be negotiable instruments and shall in every respect carry the same rights as the existing shares. The Board of Directors is authorised to lay down the terms and conditions for capital increases pursuant to the above authorisations and to make any such amendments to the Company's Articles of Association as may be required as a result of the Board of Directors' exercise of said authorisations.

4. Warrants

4.1. Until 26 January 2022, the Board of Directors shall be authorised to decide on one or more issues of warrants – to the Company's employees, external consultants and/or members of the Company's Board of Directors – with rights of subscription of 1 share per warrant in the Company in the total nominal amount of up to DKK 383,000 with addition of DKK 245,000, DKK 376,250 and DKK 389,026 (in total nominally DKK 1,393,276) without pre-emptive subscription right for the Company's shareholders.

Upon a full issue of the Board of Directors under this authorisation, the exercise of minimum 462,701 warrants shall be conditional upon that the warrant recipient has not exercised certain warrants previously granted, and shall be on terms entailing that such previously granted warrants cannot be exercised.

The Board of Directors is overall – including already issued warrants – at the time of any future issuance of warrants under the authorisation at the most authorised to issue war-rants with rights of subscription of up to 14 % of the entire issued share capital of the Company at such time, however, with addition of shares which may be subscribed by exercise of warrants upon full exercise of this capped authorisation, i.e. the cap of the authorisation is calculated as the Company's entire issued share capital with addition of this capped authorisation, corresponding to 830,592 warrants as per 29 June 2017, and subject to potential adjustment of the number of warrants pursuant to the warrant terms. Warrants, which as per 29 June 2017 have been issued with an exercise price of DKK 44 per share, shall not be included under this cap.

The terms applicable to the issued warrants shall include,

that shares subscribed on the basis of warrants shall not belong to a specific share class, but be ordinary shares,

that in the event of exercise of the issued warrants, the shareholders of the Company shall not have any pre-emptive subscription rights, cf. Section 162 of the Companies Act,

that any new shares subscribed on the basis of the issued warrants shall be negotiable instruments and freely transferable, cf. article 2.5 of the Articles of Association, and

that any new shares subscribed on the basis of the issued warrants shall be registered in the name of the holder and be entered in the Company's register of shareholders.

The terms of the issued warrants, including the subscription/exercise price, shall be subject to the decision of the Board of Directors. It is noted that warrants may be issued with an exercise price down to par, i.e. DKK 1 per share of DKK 1.

In the event that warrants lapse without having been used for the subscription of shares, the Board of Directors may reissue these warrants without further reducing this authorisation.

The Board of Directors has issued a total of 1,293,293 warrants under the authorisation however only with a right to subscribe for up to nominally DKK 916,773 shares.

The Board of Directors is authorised to delete this Clause 4.1 (including Clauses 4.1.1-4.1.4 and the related appendices to the Articles of Association) upon completion of an admission of the Company's shares to trading and official listing on Nasdaq Copenhagen A/S ("IPO") and registration of the capital increases related to the IPO and the exercise of the warrants issued pursuant to Clause 4.1.

4.1.1. On 3 September 2010, on 25 November 2010, on 15 June 2011, on 29 August 2011, on 1 May 2012, on 28 February 2013, on 2 September 2014, on 8 April 2015, and on 9 September 2015, the Board of Directors has decided to issue 32,000 warrants, 3,000 warrants, 26,080 warrants, 6,020 warrants, 166,416 warrants, 63,865 warrants, 45,983 warrants, 11,900 warrants, and 107,437 warrants, respectively, in accordance with the authorisation in article 4.1. The warrants give right to subscribe for shares in the Company at a price of 4,400, corresponding to DKK 44 per share of nominally DKK 1.

Each warrant gives the right to subscribe for 1 share of nominally DKK 1. The maximum total nominal amount of the capital increase resulting from exercise of the warrants is DKK 462,701, subject to potential adjustment of the number of warrants pursuant to the warrant terms.

The recipient has the right to exercise granted warrants for subscription of shares in the Company until 21 August 2021. If notice of exercise has not been submitted on 21 August 2021, all issued warrants shall lapse.

The time limits (exercise periods) for subscription of shares are stated in appendix 4.1.1, 4.1.1.a, 4.1.1.b, 4.1.1.c, 4.1.1.d and 4.1.1.e, respectively.

When exercising warrants, the subscription amount shall be paid to the Company not later than the time of subscription, unless otherwise set out in the warrant terms.

The recipient's legal position in the event of changes in the Company's capital structure, issue of new warrants or convertible bonds, liquidation of the Company, merger or de-merger is as stated in appendix 4.1.1, 4.1.1.a, 4.1.1.b, 4.1.1.c, 4.1.1.d and 4.1.1.e, respectively, of the Articles of Association.

Any new shares subscribed on the basis of warrants shall be subject to the same rights as the existing shares of the Company, as provided in these Articles of Association. Such new shares shall carry financial and administrative rights from the time of subscription.

The further terms of the issued warrants are set out in appendix 4.1.1, 4.1.1.a, 4.1.1.b, 4.1.1.c, 4.1.1.d and 4.1.1.e, respectively.

In connection with the issue of warrants, the Board of Directors has adopted the cash capital increase of up to nominally DKK 462,701 which may result from conversion of warrants. The capital increase is not subject to pre-emptive rights of subscription for the other shareholders of the Company.

4.1.2. On 31 March 2017, the Board of Directors has decided to issue 551,573 warrants in accordance with the authorisation in article 4.1. The warrants give the right to subscribe for shares in the Company at a price of 100, corresponding to DKK 1 per share of nominally DKK 1.

Exercise of warrants issued under this article 4.1.2 is conditional upon that the recipient has not exercised warrants previously issued to such recipient and entails that the recipient cannot exercise a corresponding number of warrants previously issued to such recipient.

Thus, exercise of warrants issued under this article 4.1.2 is conditional upon that the recipient's exercise of warrants issued to the recipient under article 4.1.1 entails that the recipient's right to exercise warrants under this article 4.1.2 will be reduced by a 1:1 ratio, and that the recipient's exercise of warrants issued to the recipient under this article 4.1.2 entails that the recipient's right to exercise warrants issued to the recipient under article 4.1.1 will be equally reduced by a 1:1 ratio.

Each warrant gives the right to subscribe for 1 share of nominally DKK 1. The maximum total nominal amount of the capital increase resulting from exercise of the warrants under article 4.1.1 and this article 4.1.2 is DKK 551,573, subject to potential adjustment of the number of warrants pursuant to the warrant terms.

The recipient has the right to exercise granted warrants for subscription of shares in the Company until 21 August 2021. If notice of exercise has not been submitted on 21 August 2021, all issued warrants shall lapse.

The time limits (exercise periods) for subscription of shares are stated in appendix 4.1.2.a and 4.1.2.b, respectively.

When exercising warrants, the subscription amount shall be paid to the Company not later than the time of subscription, unless otherwise set out in the warrant terms.

The recipient's legal position in the event of changes in the Company's capital structure, issue of new warrants or convertible bonds, liquidation of the Company, merger or de-merger is as stated in appendix 4.1.2.a and 4.1.2.b, respectively, of the Articles of Association.

Any new shares subscribed on the basis of warrants shall be subject to the same rights as the existing shares of the Company, as provided in these Articles of Association. Such new shares shall carry financial and administrative rights from the time of subscription.

The further terms of the issued warrants are set out in appendix 4.1.2.a and 4.1.2.b, respectively.

In connection with the issue of warrants, the Board of Directors has adopted the cash capital increase of up to nominally DKK 551,573 which may result from conversion of warrants. The capital increase is not subject to pre-emptive rights of subscription for the other shareholders of the Company.

4.1.3. On 20 september 2017, the Board of Directors has decided to issue 148,478 warrants in accordance with the authorisation in article 4.1. The warrants give the right to subscribe for shares in the Company at a price of 100, corresponding to DKK 1 per share of nominally DKK 1.

Each warrant gives the right to subscribe for 1 share of nominally DKK 1. The maximum total nominal amount of the capital increase resulting from exercise of the warrants under this article 4.1.3 is DKK 148,478 subject to potential adjustment of the number of warrants pursuant to the warrant terms.

The recipient has the right to exercise granted warrants for subscription of shares in the Company until 21 August 2021. If notice of exercise has not been submitted on 21 August 2021, all issued warrants shall lapse.

The time limits (exercise periods) for subscription of shares are stated in appendix 4.1.3.a and 4.1.3.b, respectively.

When exercising warrants, the subscription amount shall be paid to the Company not later than the time of subscription, unless otherwise set out in the warrant terms.

The recipient's legal position in the event of changes in the Company's capital structure, issue of new warrants or convertible bonds, liquidation of the Company, merger or de-merger is as stated in appendix 4.1.3.a and 4.1.3.b, respectively, of the Articles of Association.

Any new shares subscribed on the basis of warrants shall be subject to the same rights as the existing shares of the Company, as provided in these Articles of Association. Such new shares shall carry financial and administrative rights from the time of subscription.

The further terms of the issued warrants are set out in appendix 4.1.3.a and 4.1.3.b, respectively.

In connection with the issue of warrants, the Board of Directors has adopted the cash capital increase of up to nominally DKK 148,478 which may result from conversion of warrants. The capital increase is not subject to pre-emptive rights of subscription for the other shareholders of the Company.

4.1.4. On 20 september 2017, the Board of Directors has decided to issue 130,541 warrants in accordance with the authorisation in article 4.1. The warrants give the right to subscribe for shares in the Company at a price of 100, corresponding to DKK 1 per share of nominally DKK 1.

Each warrant gives the right to subscribe for 1 share of nominally DKK 1. The maximum total nominal amount of the capital increase resulting from exercise of the warrants under this article 4.1.4 is DKK 130,541 subject to potential adjustment of the number of warrants pursuant to the warrant terms.

The recipient has the right to exercise granted warrants for subscription of shares in the Company until 31 December 2018 and subject to the occurrence of certain exercise events as set out in the warrant terms. If notice of exercise has not been submitted on 31 December 2018, all issued warrants shall lapse.

The time limits (exercise periods) for subscription of shares are stated in appendix 4.1.4.a and 4.1.4.b, respectively.

When exercising warrants, the subscription amount shall be paid to the Company not later than the time of subscription, unless otherwise set out in the warrant terms.

The recipient's legal position in the event of changes in the Company's capital structure, issue of new warrants or convertible bonds, liquidation of the Company, merger or de-merger is as stated in appendix 4.1.4.a and 4.1.4.b, respectively, of the Articles of Association.

Any new shares subscribed on the basis of warrants shall be subject to the same rights as the existing shares of the Company, as provided in these Articles of Association. Such new shares shall carry financial and administrative rights from the time of subscription.

The further terms of the issued warrants are set out in appendix 4.1.4.a and 4.1.4.b, respectively.

In connection with the issue of warrants, the Board of Directors has adopted the cash capital increase of up to nominally DKK 130,541 which may result from conversion of warrants. The capital increase is not subject to pre-emptive rights of subscription for the other shareholders of the Company.

4.2. At an extraordinary general meeting on 6 January 2015 and at an extraordinary general meeting on 26 January 2017, the Company has issued 1,704,554 and 1,158,302 warrants (Indemnification Warrants), respectively, each entitling the holder to subscribe 1 share in the Company. All warrants are issued free of charge.

The holder shall be entitled to exercise the warrants allotted to the issue of shares in the Company until 26 January 2018, or, in the event that on that date a dispute is pending before an arbitral tribunal in regard to the exercise of warrants, until 30 days from the rendering of a final decision of such dispute. The warrants shall lapse if notice of exercise has not been given within the periods stated above.

Each warrant shall entitle the holder to issue 1 share of DKK 1, nominal value at a subscription price to be determined in accordance with the warrant agreements, cf. appendices 4.2.a and 4.2.b. The maximum capital increase, which can be effected by exercising the above warrants, amounts to a total of DKK 2,862,586 nominal value, subject to adjustment pursuant to the warrant terms.

In the event that the holder's terms are impaired because of changes in the Company's capital situation, the exercise price and possibly also the number of warrants will be adjusted in order to compensate the holder, as specified in the warrant terms.

The holder's legal position in the event of changes in the Company's capital structure, issue of new warrants or convertible bonds, liquidation of the Company, merger or de-merger is as stated in the warrant agreements, of which a template is adopted as appendices 4.2.a and 4.2.b to the Articles of Association.

The new shares shall carry the same rights as existing shares, cf. the provisions of the Articles of Association.

The terms and conditions governing the issued warrants are set out in further detail in the warrant agreements, cf. appendices 4.2.a and 4.2.b.

The shareholders general meetings have together with the decisions to issue warrants adopted the ancillary capital increases of up to nominally DKK 2,862,586, to be paid in cash without pre-emptive subscription rights for the existing shareholders, subject to adjustment pursuant to the warrant terms.

The Board of Directors is authorised to delete this Clause 4.2 (including the related appendices to the Articles of Association) upon completion of an IPO.

5. General meeting, venue and notice

5.1. The general meetings of the Company shall be held in the Capital Region of Denmark.

5.2. The annual general meeting of the Company shall be held each year in due time for the audited and approved annual report to be received by the relevant authorities before the applicable statutory time limit. The Company shall no later than eight weeks before the contemplated date of the annual general meeting publish the date of the general meeting and the deadline for submitting requests for specific proposals to be included on the agenda.

5.3. Extraordinary general meetings shall be held when determined by the Board of Directors or requested by the Company's auditor. Furthermore, an extraordinary general meeting shall be held when requested by shareholders possessing no less than five per cent of the share capital. Such request shall be submitted in writing to the Board of Directors and be accompanied by a specific proposal for the business to be transacted. The Board of Directors convenes an extraordinary general meeting no later than two weeks after such request has been made.

5.4. General meetings shall be convened by the Board of Directors with at least three weeks' and not more than five weeks' notice. The notice shall be published on the Company's website. Furthermore, a notice of the general meeting shall be sent to all shareholders recorded in the Company's register of shareholders who have so requested.

5.5. For a period of three weeks prior to the general meeting, including the date of the general meeting, the following information shall be available on the Company's website:

a. The notice convening the general meeting

b. The aggregate number of shares and voting rights as at the date of the notice

c. The documents to be presented at the general meeting

d. The agenda and the complete proposals as well as, for annual general meetings, the audited annual report

e. The forms to be used for voting by proxy or by postal vote

5.6. General meetings shall be held in English. The Board of Directors may decide to offer simultaneous interpretation into Danish. Documents prepared in connection with or following a general meeting shall be in English and to the extent required by law or if decided by the Board of Directors, in Danish.

5.7. The general meeting shall be presided over by a chairman elected by the Board of Directors who shall ensure that the general meeting is conducted in a proper and efficient manner.

6. Agenda for the annual general meeting

6.1. The agenda for the annual general meeting shall include the following:

a. The Board of Directors election of the chairman of the meeting

b. The Board of Directors' report on the Company's activities in the past financial year

c. Presentation and adoption of the annual report

d. Distribution of profit or covering of loss according to the adopted annual report

e. Resolution to grant discharge of liability to the Board of Directors and the Executive Management

f. Approval of remuneration of the Board of Directors for the current financial year

g. Election of members to the Board of Directors

h. Election of auditor

i. Authorisation to acquire treasury shares

j. Any proposals from the Board of Directors or shareholders

k. Any other business

6.2. Every shareholder shall be entitled to have a specific subject considered at the annual general meeting. Such proposals must be submitted in writing to the Board of Directors not later than six weeks prior to the annual general meeting.

7. Shareholders' attendance and voting rights at the general meeting

7.1. The right of a shareholder to attend and vote at a general meeting is determined by the shares held by the shareholder at the record date. The record date is one week prior to the general meeting. The shares held by each shareholder at the record date are calculated based on the registration of the number of shares held by that shareholder in the Company's register of shareholders as well as any notification of ownership received by the Company for the purpose of registration in the Company's register of shareholders, but which have not yet been registered.

7.2. A shareholder who is entitled to attend the general meeting pursuant to Article 7.1 and who wants to attend the general meeting shall request to receive an admission card not later than three days prior to the date of the general meeting.

7.3. A shareholder may attend in person or by proxy, and the shareholder or the proxy may attend together with an adviser.

7.4. The right to vote may be exercised by a written and dated instrument of proxy in accordance with applicable laws.

7.5. A shareholder who is entitled to participate in the general meeting pursuant to Article 7.1 may vote by postal vote in accordance with the provisions of the Danish Companies Act. Such postal votes shall be received by the Company not later than the business day before the general meeting. Postal votes cannot be withdrawn.

7.6. Each share of the nominal value of DKK 1 shall carry 1 vote.

8. Resolutions at general meetings

8.1. Resolutions by the general meeting shall be passed by a simple majority of votes cast unless otherwise prescribed by law or by these Articles of Association.

8.2. Adoption of changes to these Articles of Association, dissolution of the Company, merger or demerger requires that the decision is adopted with at least 2/3 of the votes cast as well as the share capital represented at the general meeting, unless applicable laws prescribe stricter or less strict adoption requirements or applicable laws confer specific authority to the Board of Directors or other bodies.

9. Board of Directors

9.1. The Board of Directors consists of not less than six and not more than nine members elected by the general meeting.

9.2. The members of the Board of Directors elected by the general meeting are elected for a term of one year. Re-election of board members may take place.

9.3. The Board of Directors elects a Chairman and, if so decided by the Board of Directors, a Deputy Chairman among its members. If the Chairman of the Board of Directors resigns during a term of election, the Deputy Chairman (if elected) shall take up the position as Chairman until a new Chairman is elected among the members of the Board of Directors.

9.4. Any employee representatives on the Board of Directors and their alternates, if any, are elected in accordance with applicable law thereon in force from time to time.

9.5. Resolutions of the Board of Directors are passed by simple majority. In the event of equal votes, the Chairman or, in his/her absence, the Deputy Chairman shall have a casting vote.

9.6. The Board of Directors forms a quorum when more than half of its members are represented, including the Chairman or the Deputy Chairman.

10. Executive Management

10.1. The Board of Directors appoints an Executive Management consisting of one to three members to be in charge of the day-to-day management of the Company.

11. Rules of signature

11.1. The Company shall be bound (i) by the joint signatures of the Chairman and a member of the Executive Management, (ii) by the joint signatures of the Chairman and two members of the Board of Directors or (iii) by the joint signatures of two members of the Executive Management.

12. Overall guidelines on incentive pay

12.1. The Company has adopted overall guidelines on incentive pay to the Board of Directors and the Executive Management. The overall guidelines on incentive pay, which have been approved by the general meeting, are available on the Company's website.

13. Electronic communication

13.1. All communication from the Company to the individual shareholders, including notices convening general meetings, may take place electronically by posting on the Company's website or by email. General notices shall be published on the Company's website and in such other manner as may be prescribed by applicable laws. The Company may as an alternative choose to send notices, etc., by ordinary post.

13.2. Communication from a shareholder to the Company may take place by email or by ordinary post.

13.3. Each shareholder is responsible for ensuring that the Company has the correct email address at all times. The Company is not obliged to verify such contact information or to send notices in any other way.

13.4. The Company's website, www.orphazyme.com, contains information about system requirements and electronic communication procedures.

13.5. Company announcements shall be prepared in English and, if decided by the Board of Directors, in Danish.

14. Annual report

14.1. The Company's annual accounts shall be audited by a state-authorised public accountant elected by the general meeting for a one-year term. Re-election may take place to the extent permitted under applicable law.

14.2. Annual reports shall be prepared in English and, if decided by the Board of Directors, in Danish.

15 Financial year

15.1. The Company's financial year is the calendar year.

As adopted at the Company's extraordinary general meeting held on 2 November 2017.

As chairman of the general meeting:

Rikke Schiøtt Petersen

Appendices:

Appendix 4.1.1	Terms governing Ordinary Warrants
Appendix 4.1.1.a	Compiled template (employees and board members)
Appendix 4.1.1.b	Compiled template showing time of grants of warrants (employees and board members)
Appendix 4.1.1.c	Template Warrant Agreement (advisory board)
Appendix 4.1.1.d	Template Warrant Agreement (employees)
Appendix 4.1.1.e	Template Warrant Agreement (board members)
Appendix 4.1.2.a	Template Warrant Agreement (employees)
Appendix 4.1.2.b	Template Warrant Agreement (board members)
Appendix 4.1.3.a	Template Warrant Agreement (employees)
Appendix 4.1.3.b	Template Warrant Agreement (board members)
Appendix 4.1.4.a	Template Warrant Agreement (employees)
Appendix 4.1.4.b	Template Warrant Agreement (board members)
Appendix 4.2.a	Terms governing Indemnification Warrants
Appendix 4.2.b	Terms governing Indemnification Warrants

ORPHAZYME

Annex B – Application Form

Application form (only one form per custody account)	**Offering of up to 10,781,250 Offer Shares (including the Overallotment Shares) of DKK 1 nominal value each**

Application for subscription of Offer Shares in Orphazyme A/S, registration (CVR) no. 32266355

Selling agents:	Carnegie Investment Bank, filial af Carnegie Investment Bank AB (publ), Sverige	Danske Bank A/S
Joint Global Coordinators and Joint Bookrunners:	Carnegie Investment Bank, filial af Carnegie Investment Bank AB (publ), Sverige and Danske Bank A/S	
Co-Lead Manager	Oddo BHF SCA	
Offer Period:	6 November 2017 to 16 November 2017 at 12:00 p.m. (noon) (CET) unless the Offering is closed earlier in whole or in part. The Offer Period for order applications up to and including DKK 3 million may be closed before the remainder of the Offering. The Offering will not be closed before 15 November 2017 at 00:01 a.m. (CET).	
Offer Price Range:	DKK 64 to DKK 80 per Offer Share	
ISIN:	Permanent ISIN for the Shares: DK0060910917 Temporary ISIN for the Temporary Purchase Certificates: DK0060911055	

The Offering Circular dated 6 November 2017 includes, *inter alia*, the Articles of Association of Orphazyme A/S, the financial statements for the financial years 1 July 2014 to 30 June 2015; 1 July 2015 to 31 December 2015; and 1 January 2016 to 31 December 2016, respectively, as included in the statutory annual report for 2016; and the interim financial statements for the six months ended 30 June 2017 and 2016 for Orphazyme A/S as well as the terms and conditions for the subscription of Offer Shares.

Both binding order applications and expressions of interest can be submitted with specification of a maximum price. If the Offer Price is determined at a higher level than the stated maximum price, no Offer Shares will be allocated to the subscriber.

Applications should be made for a number of Temporary Purchase Certificates representing Offer Shares or for an aggregate amount rounded to the nearest Danish kroner amount. The minimum subscription/purchase amount is one Offer Share.

For binding orders up to and including DKK 3 million, the application form is submitted to the subscriber's own account-holding institution duly filled in and signed.

The application form shall be submitted within an appropriate amount of time for the account-holding institution to process and forward the application form so that the application form reaches Danske Bank A/S no later than 16 November 2017 at 12:00 p.m. (noon) (CET) or such earlier time as the Offering may be closed in whole or in part.

Expressions of interest to purchase Offer Shares for more than DKK 3 million can be submitted to one of the Managers, e.g., by using this application form.

On the terms and conditions stated in the Offering Circular dated 6 November 2017, including in "*Risk Factors*" and "*Selling Restrictions*", I/we hereby submit an order application for the subscription of Offer Shares in Orphazyme A/S and simultaneously declare to have received a copy of the Offering Circular; and that I/we have solely based my/our investment decision on the contents of the Offering Circular. The Offer Price will be fixed upon closing of the Offering through a book-building process. See "*The Offering—Offer Price*". Only one application form per custody account with VP SECURITIES A/S (VP) will be accepted.

[Please complete order form on the back]

ORPHAZYME

Application submitted as a binding application (for orders up to and including DKK 3 million)

I/we accept that the Managers may demand information about my/our name(s), address(es) and application and are entitled to pass on such information to the Main Shareholders, Orphazyme A/S and the Managers. I/we undertake to pay the equivalent of the Offer Shares allocated at the Offer Price fixed.

Field (1) or (2) should be completed

(1) For Danish kroner (DKK)	(2) Number of Offer Shares	(3) Maximum price per Offer Share, if any

Expression of interest submitted pursuant to the book-building process (for orders above DKK 3 million)

I/we accept that the application form and information about my/our name(s) and address(es) are entitled to be passed on to the Main Shareholders, Orphazyme A/S and the Managers. I/we accept that I/we during the Offer Period can amend or revoke this expression of interest but that this expression of interest will automatically be converted into a binding subscription order upon expiry of the Offer Period.

Field (1) or (2) should be completed

(1) For Danish kroner (DKK)	(2) Number of Offer Shares	(3) Maximum price per Offer Share if any

If the aggregate applications to subscribe and expressions of interest exceeds the total number of Offer Shares, a reduction will be completed as further described in the Offering Circular. See "*Plan of Distribution*". Neither submission of application orders nor submission of expressions of interest entitles one to any Offer Shares. Settlement of the Offering will be effected by way of registration of Temporary Purchase Certificates representing the allocated number of Offer Shares on your custody account with VP SECURITIES A/S (VP) against payment in DKK, which is expected to take place on or before 21 November 2017. All dealings in the Temporary Purchase Certificates and/or the Offer Shares prior to settlement of the Offering will be for the account of, and at the sole risk of, the parties involved.

Information and signature

Name:	VP custody account no.:	
Address:	Settlement account no.:	
Postal code and city:	Custodian bank:	
Telephone:		
Date:		
This application form was submitted to (to be completed by account-holding institution):		
	Reg. no.:	*Participant ID no. (CD-ident.):*
	Date:	*Tel.:*

Signature *Company stamp and signature*

Please complete the form overleaf when opening a new VP custody account.

Opening of new VP custody account **(This box should be filled in when opening a new VP custody account and any related settlement account)**
Civil registration (CPR) no./company registration (CVR) no.:
Name:
Address:
Postal code and city:
Tel.:
Position:
Existing account no. for settlement, if any:

Ordreblanket (Kun én blanket pr. depot)	Udbud af op til 10.781.250 Aktier (inklusive Overallokeringsaktierne) à nom. DKK 1

Ordre om tegning af Udbudte Aktier i Orphazyme A/S, CVR-nr. 32266355

Salgssteder:	Carnegie Investment Bank, filial af Carnegie Investment Bank AB (publ), Sverige	Danske Bank A/S
Joint Global Coordinators og Joint Bookrunners:	Carnegie Investment Bank, filial af Carnegie Investment Bank AB (publ), Sverige og Danske Bank A/S	
Co-Lead Manager	Oddo BHF SCA	
Udbudsperiode:	6. november 2017 til 16. november 2017 kl. 12.00 dansk tid, medmindre Udbuddet helt eller delvist lukkes tidligere. Udbudsperioden for ordre op til og med DKK 3 mio. kan lukkes før resten af Udbuddet. Udbuddet vil tidligst blive lukket 15. november 2017 kl. 00:01 dansk tid.	
Udbudskursinterval:	DKK 64 til DKK 80 pr. Udbudt Aktie	
ISIN kode:	Permanent ISIN for Aktierne: DK0060910917 Midlertidig ISIN kode for de Midlertidige Købsbeviser: DK0060911055	

Dette Prospekt dateret 6. november 2017 indeholder blandt andet vedtægter for Orphazyme A/S, årsregnskabet for henholdsvis 1. juli 2014 til 30. juni 2015, 1. juli 2015 til 31. december 2015 og 1. januar 2016 til 31. december 2016, som indeholdt i årsrapporten for 2016, samt delårsregnskabet for seks-månedersperioden indtil 30. juni 2017 og 2016 for Orphazyme A/S, og vilkårene for tegning af Udbudte Aktier.

Både bindende ordrer og interessetilkendegivelser kan afgives med angivelse af en eventuel maksimumkurs. Fastsættes Udbudskursen højere end den anførte maksimumkurs, vil ordregiver ikke blive tildelt nogen Udbudte Aktier.

Ordrer skal afgives for et antal Udbudte Aktier eller for et samlet beløb afrundet til nærmeste kronebeløb. Der skal som minimum tegnes/købes 1 stk. Udbudt Aktie.

For bindende ordrer til og med DKK 3 mio. indleveres ordreblanketten til ordregivers eget kontoførende institut i udfyldt og underskrevet stand.

Ordreblanketten skal indleveres i så god tid, at det kontoførende institut har mulighed for at behandle og videresende ordren, således at den er Danske Bank A/S i hænde senest den 16. november 2017 kl. 12:00 dansk tid eller et sådant tidligere tidspunkt, hvor Udbuddet måtte blive lukket helt eller delvist.

Interessetilkendegivelser på mere end DKK 3 mio. skal afgives til en af Emissionsbankerne evt. ved brug af denne ordreblanket.

På vilkår som anført i Prospektet dateret den 6. november 2017, herunder afsnittene "*Risikofaktorer*" og "*Salgsbegrænsninger*", afgiver jeg/vi hermed tilbud om tegning af Udbudte Aktier i Orphazyme A/S og bekræfter samtidig at have fået udleveret et eksemplar af Prospektet, og at jeg/vi alene har baseret min/vores investeringsbeslutning på indholdet af Prospektet. Udbudskursen fastsættes efter lukning af Udbuddet via bookbuilding-metoden, jf. afsnittet "*Udbudsbetingelser*". Der kan kun afgives én ordreblanket pr. depot hos VP SECURITIES A/S (VP).

[Venligst udfyld ordreblanket på bagsiden]

Ordre afgivet som bindende ordre (for ordrebeløb til og med DKK 3 mio.)

Jeg/vi accepterer, at Emissionsbankerne kan kræve oplysninger om mit/vort navn, adresse og ordre, og er berettiget til at videregive denne information til de Betydelige Aktionærer, Selskabet og Emissionsbankerne. Jeg/vi forpligter mig/os hermed til at betale modværdien af tildelte Udbudte Aktier til den fastsatte udbudskurs.

Felt 1) eller 2) skal udfyldes

(1) For kroner (DKK):	(2) Antal Udbudte Aktier (stk.)	(3) Evt. maksimumkurs pr. Udbudt Aktie:

Interessetilkendegivelse afgivet efter bookbuilding-metoden (for ordrebeløb større end DKK 3 mio.)

Jeg/vi accepterer, at ordreblanketten samt navn og adresse videregives til de Betydelige Aktionærer, Orphazyme A/S og Emissionsbankerne. Jeg/vi accepterer, at jeg/vi i Udbudsperioden løbende kan ændre eller tilbagekalde interessetilkendegivelsen, men at denne bliver til en bindende ordre ved lukning af Udbuddet.

Felt 1) eller 2) skal udfyldes

(1) For kroner (DKK):	(2) Antal Udbudte Aktier (stk.)	(3) Evt. maksimumkurs pr. Udbudt Aktie:

Overstiger de samlede ordrer og interessetilkendegivelser det samlede antal Udbudte Aktier, vil der ske reduktion som anført i det Engelsksprogede Prospekt, jf. afsnittet "*Udbudsbetingelserne—Tildeling og reduktion*". Afgivelse af ordrer eller interessetilkendegivelser medfører ingen sikkerhed for hel eller delvis tildeling af Udbudte Aktier. Afvikling af Udbuddet sker ved registrering af Midlertidige Købsbeviser repræsenterende det antal tildelte Udbudte Aktier på Deres depot i VP SECURITIES A/S (VP) mod kontant betaling i DKK, hvilket forventes at finde sted senest 21. november 2017. Al handel med de Midlertidige Købsbeviser og/eller de Udbudte Aktier forud for afvikling sker for de involverede parters egen regning og risiko.

Oplysninger og underskrift

Navn:	VP-depotnr.:	
Addresse:	Kontonr. Til afregning.:	
Postnr. og by:	Kontoførende institut:	
Telefon		
Dato:		
	Ordren er indleveret hos (udfyldes af kontoførende institut):	
	Reg.nr.:	*CD-ident:*
	Dato:	*Telefon:*

Underskrift *Firmastempel og underskrift*

Udfyld nedenfor ved oprettelse af et nyt VP-depot.

Oprettelse af nyt VP-depot **(Denne rubrik udfyldes i forbindelse med oprettelse af nyt VP-depot og evt. tilhørende afregningskonto)**
CPR/CVR-nr.:
Navn:
Adresse:
Postnr. og by:
Telefon:
Stilling:
Evt. eksisterende kontonr. til afregning:

THE COMPANY

Orphazyme A/S
Ole Maaløes Vej 3
DK-2200 Copenhagen N
Denmark

MANAGERS

Joint Global Coordinators and Joint Bookrunners

Carnegie Investment Bank, filial af Carnegie Investment Bank AB (publ), Sverige
Overgaden neden Vandet 9B
DK-1414 Copenhagen K
Denmark

Danske Bank A/S
Holmens Kanal 2-12
DK-1092
Copenhagen K
Denmark

Co-Lead Manager
Oddo BHF SCA
12, boulevard de la Madeleine
75440 Paris Cedex 09
France

LEGAL ADVISOR

To the Company:
Gorrissen Federspiel Advokatpartnerselskab
Axel Towers
Axeltorv 2
DK-1609 Copenhagen V
Denmark

To the Managers:
Plesner Advokatpartnerselskab
Amerika Plads 37
DK-2100 Copenhagen Ø
Denmark

AUDITORS OF THE COMPANY

Ernst & Young Godkendt Revisionspartnerselskab
Osvald Helmuths Vej 4
DK-2000 Frederiksberg
Denmark