UNITED STATES SECURITIES
AND EXCHANGE COMMISSION
Washington, D.C. 20549

FORM 6-K

REPORT OF FOREIGN PRIVATE ISSUER
PURSUANT TO RULE 13a-16 OR 15d-16
UNDER THE SECURITIES EXCHANGE ACT OF 1934

For the Month of August 2021

Commission File Number: 001-39545

Orphazyme A/S

(Translation of registrant's name into English)

Ole Maaløes Vej 3, DK-2200
Copenhagen N
Denmark
(Address of principal executive office)

Indicate by check mark whether the registrant files or will file annual reports under cover of Form 20-F or Form 40-F:
☒ Form 20-F ☐ Form 40-F

Indicate by check mark if the registrant is submitting the Form 6-K in paper as permitted by Regulation S-T Rule 101(b)(1): ☐

Indicate by check mark if the registrant is submitting the Form 6-K in paper as permitted by Regulation S-T Rule 101(b)(7): ☐

INCORPORATION BY REFERENCE

This Report on Form 6-K (the "Report") and Exhibit 99.1 to this Report is hereby expressly incorporated by reference into the registrant's registration statements on Form S-8 (File nos. 333-249407 and 333-255661) and to be a part thereof from the date on which this Report is filed, to the extent not superseded by documents or reports subsequently furnished.

Half-Year Financial Report for the Six Months Ended June 30, 2021

The Company's half-year financial report, including its unaudited condensed consolidated financial statements as of and for the six months ended June 30, 2021, is attached to this Report on Form 6-K as Exhibit 99.1 hereto and is incorporated by reference herein.

EXHIBIT LIST

Exhibit	Description
99.1	Interim Report First Half 2021

SIGNATURES

Pursuant to the requirements of the Securities Exchange Act of 1934, the registrant has duly caused this report to be signed on its behalf by the undersigned, thereunto duly authorized.

Orphazyme A/S

Date: August 31, 2021

By: /s/ Anders Vadsholt

Name Anders Vadsholt

Title: Chief Financial Officer

Company announcement
No. 21/2021

Orphazyme A/S
Ole Maaløes Vej 3
DK-2200 Copenhagen N
CVR No.: 32266355

Interim Report First Half 2021

Copenhagen, Denmark, August 31, 2021 – Orphazyme A/S (ORPHA.CO; ORPH) ("the Company"), a late-stage biopharmaceutical company, today announces its Interim Report First Half 2021 for the period January 1 – June 30, 2021.

"We remain steadfast in our belief that arimoclomol holds significant potential for people living with NPC and we remain committed to pursuing a path to regulatory approval in Europe and the United States," said Christophe Bourdon, Chief Executive Officer of Orphazyme. *"While maintaining our patients on the Early Access Programs across key countries, we have executed on our restructuring plan, enabling significant cost savings to the company and are pursuing approval for arimoclomol. In the fourth quarter of 2021, we expect a CHMP opinion from EMA on arimoclomol and expect to provide further detail on our path forward in the US following the conclusion of a Type A meeting with the FDA. We are assessing different possibilities for obtaining additional funding to sustain operations in 2022 and beyond".*

Pipeline Updates First Half 2021

- Announced top-line results from both the Phase 2/3 trial of arimoclomol in Inclusion Body Myositis and the Phase 3 trial of arimoclomol in Amyotrophic Lateral Sclerosis; neither trial met its primary endpoint and we have ceased further development of arimoclomol in these indications
- Received a Complete Response Letter from FDA following its review of the new drug application for arimoclomol in NPC; we are assessing the path forward with FDA
- Marketing authorization application for arimoclomol for NPC remains underway with the European Medicines Agency and a CHMP opinion is expected during Q4 2021
- Our Early Access Programs (EAP) for arimoclomol in the United States, Germany and France continued through the first half of 2021, with approx. 100 patients participating in the EAP as of June 30, 2021
- Presented 12-month and 24-month results from the open-label extension of the Phase 2/3 trial of arimoclomol in NPC at the 17th Annual WORLDSymposium Scientific meeting and the Parseghian Scientific Conference for NPC Research respectively. The results demonstrate that arimoclomol provided a sustained benefit to study participants through 36 months and are consistent with the previously reported safety profile

Financial and Business Highlights First Half 2021

- Appointed Christophe Bourdon as Chief Executive Officer, effective April 1, 2021
- Announced company restructuring that will result in a significant headcount reduction (~70 remaining FTEs expected by year-end), substantial cost savings, and a focus on activities to support potential approval of arimoclomol in Europe and in the US
- For the first six months of 2021, Orphazyme reported a net loss of DKK 463.8.0 million or DKK 13.27 per share (basic and diluted) compared to a net loss of DKK 251.4 million or DKK 9.77 per share (basic and diluted) for the same period in 2020
- Beginning in 2021 and for the six-month period ended June 30, 2021, Orphazyme recognized net revenue of DKK 13.2 million from the sale of arimoclomol for treatment of NPC under the ATU (remunerated early access program) in France

- Research and development expenses for the period totaled DKK 264.7 million compared to DKK 167.0 million for the same period in 2020 mainly due to the increased activity in our development functions for most of the period before receipt of the Complete Response Letter from the FDA; and recognition of restructuring provisions for the close-out of the clinical trials for IBM and ALS and related impairment charges recognized following the negative trial results.
- General and administrative expenses for the period totaled DKK 214.2 million compared to DKK 78.6 million for the same period in 2020 due to the build-up of the commercial organization in preparation for commercial launch during most of the period, including expenses related to supporting functions, before receipt of the Complete Response Letter from the FDA. In addition, this amount includes a restructuring provision for the commercial and administrative employees who were made redundant
- As of June 30, 2021, Orphazyme held cash totaling DKK 334.2 million compared to DKK 610.4 million as of June 30, 2020 and DKK 726.9 million as of December 31, 2020

Subsequent Events

- Announced publication of 12-month data from the double-blind portion of the Phase 2/3 trial in NPC in the Journal of Inherited Metabolic Disease (JIMD). Arimoclomol was well-tolerated with a statistically significant and clinically meaningful effect on disease progression (mean treatment effect in favor of arimoclomol of -1.40 points on 5-domain NPCCSS (95% CI: −2.76, −0.03; p = 0.046))

Outlook

The company maintains its revised outlook for 2021, as published on June 18, 2021. Operating expenses are anticipated to be in the range of DKK 700 -720 million; net operating loss is anticipated to be in the range of DKK 670–700 million; and our cash position at year-end 2021 is anticipated to be approximately DKK 50 million. We anticipate reaching net revenues of between DKK 30 and DKK 40 million by year-end December 31, 2021.

Conference Call

Orphazyme will host an investor call during which Management will present the Interim Report First Half 2021. The presentation will be followed by a Q&A session.

The call will be held on **Tuesday, August 31, 2021 at 2.00 PM CEST/8.00 AM EDT.**

Dial-in details:

- Denmark: +45 3272 0417
- United States: +1 6467 413 167
- Standard International: +44 (0) 2071 928338
- United Kingdom: +44 (0) 8444819752
- France: +33 (0) 170700781
- Netherlands: +31 (0) 207956614
- Sweden: +46 (0) 856618467

Event Title: Orphazyme Interim Report First Half 2021

Confirmation code: 4182747

The presentation will also be available via webcast: ***https://edge.media-server.com/mmc/p/vmsh6rph***. After the call, the presentation will be available via the webcast link above.

Condensed Consolidated Key Figures

DKK (000)	As of and for the six-months ended Jun 30, 2021	As of and for the six-months ended Jun 30, 2020	As of and for the year ended Dec 31, 2020
Statement of profit or loss and other comprehensive income			
Net revenue	13,152	-	-
Research and development expenses	(264,683)	(166,980)	(361,284)
General and administrative expenses	(214,167)	(78,575)	(247,250)
Operating loss	**(465,698)**	**(245,555)**	**(608,534)**
Net financial items	(400)	(7,841)	(26,627)
Loss before tax	**(466,098)**	**(253,396)**	**(635,161)**
Income tax benefit	2,318	1,981	1,915
Net loss for the period	**(463,780)**	**(251,415)**	**(633,246)**
Total comprehensive loss	**(463,704)**	**(251,550)**	**(632,641)**
Loss per share, basic and diluted (DKK)	(13.27)	(9.77)	(22.32)
Statement of financial position			
Intangible assets	2,194	10,773	12,454
Right-of-use assets & Property, plant and equipment	13,928	19,789	19,546
Other non-current assets	8,214	7,608	6,829
Cash	334,184	610,448	726,929
Other current assets	69,511	27,742	56,735
Total assets	428,031	676,360	822,493
Share capital	34,952	27,045	34,698
Total equity	166,761	506,135	620,525
Non-current liabilities	37,610	48,419	35,341
Current liabilities	223,660	121,806	166,627
Cash flow statement			
Net cash used in operating activities	(381,724)	(204,169)	(539,076)
Net cash used in investing activities	(806)	(1,760)	(5,101)
Net cash provided by (used in) financing activities	(15,527)	692,944	1,159,422
Other			
Share price (DKK)	46.84	89.30	67.10
Total outstanding shares	34,952,241	27,044,929	34,697,703
Market capitalization (DKK million)[1]	1,637.2	2,415.1	2,328.2
Equity ratio[2]	39.0%	74.8%	75.4%
Equity per share (DKK)[3]	4.77	18.71	17.88
Purchase of property, plant and equipment (DKK 000)	-	1,170	2,365
Average number of employees	164	106	117
Number of full-time employees (FTEs), end of period	180	114	141

1 Market capitalization is calculated as the share price multiplied with the total outstanding shares as of the balance sheet date.
2 Equity ratio is calculated as the equity divided by total assets as of the balance sheet date.
3 Equity per share is calculated as the total equity divided by the total outstanding shares as of the balance sheet date.

Outlook

In millions of DKK	2021 guidance	2020 actual
Operating expenses	700 – 720	609
Operating loss	670 – 700	609
Cash position at year-end	~50	727

Orphazyme is maintaining its revised outlook for 2021 as published on June 18, 2021.

We anticipate reaching net revenues of between DKK 30 and DKK 40 million by year-end December 31, 2021. The net revenue pertains to the ATU in France.

On June 18, 2021, Orphazyme announced that it received a Complete Response Letter (CRL) from the U.S. Food and Drug Administration (FDA) following the FDA's review of the new drug application for arimoclomol, a heat shock protein amplifier intended for the treatment of Niemann-Pick disease type C (NPC). As a result of arimoclomol for NPC not being approved by the FDA, Orphazyme cannot start the commercialization of its drug product in the U.S., which would have contributed to revenue and to reduced operating loss. Furthermore, Orphazyme was not able to obtain a priority review voucher in 2021.

Receiving the CRL from the FDA and the resulting consequences for our cash position introduces a going concern risk and consequently, Management has made an assessment of Orphazyme's ability to continue as a going concern for the twelve-month period following the balance sheet date of June 30, 2021. In connection with this assessment, several uncertainties have been identified and, as a result, mitigating actions have been initiated.

The following significant uncertainties were identified as of June 30, 2021:

- ***Path forward with the FDA:*** Following the receipt of the CRL from the FDA, Orphazyme will request a Type-A meeting with the FDA to assess a path forward for arimoclomol for NPC in the U.S. There is no guarantee that Orphazyme will ultimately obtain an approval for arimoclomol for NPC in the U.S.

- ***Outcome of CHMP assessment and subsequent regulatory approval by EMA:*** Orphazyme expects to receive an opinion from the European Committee for Medicinal Products for Human Use (CHMP) in Q4 2021. The CHMP is conducting an initial assessment of our application for EU-wide marketing authorizations for arimoclomol for NPC. Whereas a positive response from CHMP does not automatically result in an approval from the European Medicines Agency (EMA), a positive response increases the likelihood of receiving approval from EMA. Orphazyme expects to receive feedback on potential approval from EMA in the first quarter of 2022. There is no guarantee that Orphazyme will obtain positive feedback from CHMP or approval from EMA.

- ***Cash usage***: With the current cash position and the current estimated outlook, Orphazyme expects to have approximately DKK 50 million in cash at the end of December 2021. Following our restructuring and close-out of clinical trials, we anticipate to significantly reduce our monthly cash usage to sustain our reduced organization and activities. Any significant deviation in our cash usage can materially impact our anticipated cash position at year-end 2021. There is no guarantee that the estimated cash usage will materialize as expected.

- ***Additional funding:*** The current expectation for future cash requirements indicates that Orphazyme will need to raise additional funding. The Group is therefore currently assessing different possibilities for obtaining additional funding. However, if further funding is not obtained, Orphazyme will not have sufficient cash to fund its activities and meet its obligations.

In response to these uncertainties, the following mitigating actions have been taken:

- ***Restructuring program:*** Following receipt of the CRL from the FDA, a restructuring program reducing the global workforce by approximately two thirds was initiated and finally concluded by July 31, 2021. The company terminated certain employees in the Group's subsidiaries and in headquarters, who were not deemed to be essential. Following our restructuring, we have strategically maintained medical and commercial personnel to strengthen our footprint in areas with the most significant market potential for NPC. We have maintained a core team in the U.S. to effectively act on any actions agreed with the FDA; and we have maintained a core team in Europe and the UK to continue to support our early access programs and establish our commercial presence in those respective territories.

- ***Termination of contracts and agreements:*** Management has initiated termination of contracts and agreements not deemed essential in the continued pursuit of regulatory approval in Europe and assessing a path forward in the U.S. This includes initiating the close-out of all clinical trial activities for IBM, ALS and Gaucher.

- ***Additional funding:*** Management continuously assesses options to ensure sufficient funding is in place to support its corporate priorities. We are currently evaluating a number of strategic options including (i) debt and/or equity financing; (ii) new potential commercial relationships, and (iii) monetization of non-core assets.

With the uncertainties listed above, there can be no assurance that Orphazyme will achieve or sustain profitability or positive cash flows from operations. If the Group is unable to attract additional funding, future operations will need to be significantly scaled back or discontinued. These conditions raise substantial doubt about the Group's ability to continue as a going concern. These interim condensed consolidated financial statements do not include any adjustments that might result from the outcome of this uncertainty. Based on the mitigating actions described above, Management is of the view that a going concern assumption is appropriate as of June 30, 2021.

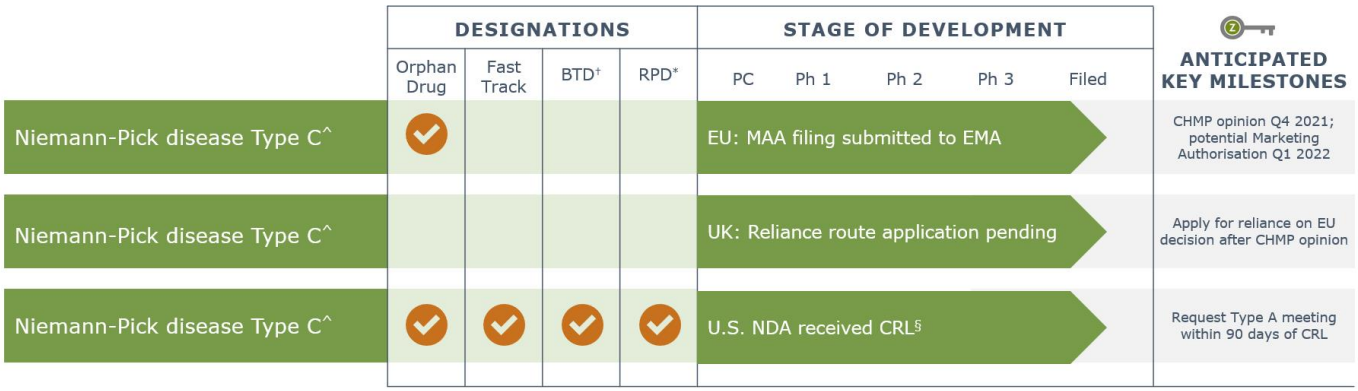
COVID-19

At this time, there is no material impact directly related to COVID-19 on the Group's consolidated financial statements, including the judgements and estimates applied. Specifically, Orphazyme is not currently conducting any clinical trials and is in the process of closing out the existing trial extensions following the disappointing results of the Phase 2/3 clinical trial related to IBM and the Phase 3 clinical trial related to ALS. However, as Orphazyme is assessing a potential path forward with the FDA following receiving a complete response letter from the FDA on arimoclomol for the treatment of NPC and is actively pursuing the European regulatory approval, other parts of the business and operations may be adversely impacted by the effects of COVID-19, for example: our third-party manufacturers and other third parties; the productivity of our staff; ability to attract, integrate, manage and retain qualified personnel or key employees; our global supply chains and relationships with vendors and other parties; significant disruption of global financial markets; and reduced ability to secure additional funding. We will continue to monitor the COVID-19 pandemic and its potential impact on our business and financials.

Priorities H2 2021

Priority	Targeted milestone	Estimated timing
NPC registration EU and UK	•Marketing authorization in Europe and UK	•EMA review underway; anticipate CHMP opinion in Q4 2021 and potential Marketing Authorisation in Q1 2022
NPC registration U.S.	•Assess path forward with the FDA	•Ongoing

Product Pipeline



^Early Access Program in US, FR, and DE ongoing; † Breakthrough Therapy Designation; * Rare Pediatric Disease Designation; § Complete Response Letter

Niemann-Pick disease Type C (NPC)
Orphazyme remains committed to developing new treatment options for patients with NPC. In 2020, the Company submitted applications to the U.S. and EU regulators seeking approval for its investigational treatment, arimoclomol, for NPC. Arimoclomol remains under review with the European Medicines Agency and a CHMP opinion is expected in Q4 2021, with potential marketing authorization approval in Q1 2022. In June 2021, Orphazyme received a Complete Response Letter (CRL) from the U.S. Food and Drug Administration (FDA) following its review of the new drug application for arimoclomol for NPC. The FDA issued the CRL based on needing additional qualitative and quantitative evidence to further substantiate the validity and interpretation of NPCCSS and, in particular, the swallow domain. Further, the FDA noted in the CRL that additional data are needed to bolster confirmatory evidence beyond the single phase 2/3 clinical trial to support the benefit-risk assessment of the NDA. Orphazyme is assessing the path forward for arimoclomol for NPC in the U.S. with the FDA and anticipates requesting a Type A meeting with the FDA within 90 days of receiving the CRL.

A primary endpoint of the Phase 2/3 clinical trial in NPC was progression in disease severity as measured by the 5-domain NPC Clinical Severity Scale (NPCCSS). This is a disease-specific measure of disease progression consisting of the five clinically most relevant domains to patients with NPC, caregivers, and physicians. A change of 1 point or greater on the NPCCSS scale has been assessed to be a clinically meaningful change based on a survey with NPC clinicians, individuals with NPC and caregivers.

In February 2021, Orphazyme presented 12-month interim results from the Phase 2/3 open-label extension (OLE) trial for arimoclomol in NPC at the WORLDSymposium Scientific meeting. Additionally, in June 2021, Orphazyme presented 24-month results from the OLE trial at the Parseghian Scientific Conference for Niemann-Pick disease type C Research. Both 12- and 24 months OLE-results demonstrated that arimoclomol provided a sustained benefit reducing disease progression in study participants on continuous arimoclomol treatment. The mean change in the 5-domain NPCCSS from baseline to 36-months was 3.5 points in the arimoclomol group and an estimated 5.2 points in the natural history group (based on a statistical model combining placebo data from the NPC-002 double-blind study and prospective data from the observational NPC-001 study; mean treatment difference of 1.7 points). The effect of arimoclomol on disease progression was consistent across pre-specified subgroups, including among participants more than four years of age and participants receiving miglustat as part of routine clinical care. During the 12 months double-blind part of the trial the group of patients who received placebo had a mean change in 5-domain NPCCSS score of +2.0. After initiation of arimoclomol (OLE phase), the mean change in 5-domain NPCCSS score throughout 24 months was +0.9 points. This also supports a slowing of disease progression after initiation arimoclomol.

Arimoclomol demonstrated a consistent safety profile throughout the 36-month treatment period. Adverse events observed during the OLE phase were similar to those observed in the double-blind phase.

After the period, data from the 12-month double-blind portion of the Phase 2/3 trial NPC-002 was published in the peer-reviewed journal, Journal of Inherited Metabolic Disease (JIMD). In this placebo controlled part of the study, fifty patients aged 2–18 years were randomized 2:1 to arimoclomol:placebo, stratified by miglustat use. Routine clinical care was maintained. Arimoclomol was administered orally three times daily, (weight-adjusted 1200mg daily dose). The results demonstrated that at 12-months, a significant treatment effect in favor of arimoclomol of −1.40 points (95% CI: −2.76, −0.03; p = 0.046) was observed corresponding to a 65% relative reduction in annual disease progression (previously calculated as -1.34 (95% CI -2.71, 0.02; p=0.0537; updated to reflect a clinical data correction). In the prespecified subgroup of patients receiving miglustat as routine care, arimoclomol resulted in stabilization of disease severity with a treatment difference of −2.06 in favor of arimoclomol (p = 0.006). In the pre-specified subgroup of patients ≥4 years of age the mean treatment difference was −1.80 in favor of arimoclomol (p=0.016), corresponding to an 82% relative reduction in annual disease progression.

Arimoclomol is currently available to NPC patients in the United States, Germany and France through our early access program, or EAP, with approximately 100 patients on treatment as of June 30, 2021. We also plan to establish additional early access or compassionate use programs in other locations in H2 2021.

Gaucher disease
Orphazyme reported positive top-line results from a randomized, double-blind, dose-ranging phase 2 clinical trial investigating arimoclomol in patients with Gaucher disease Type 1 and 3 naïve to therapy in June 2020. Arimoclomol was observed to be well-tolerated and demonstrated a relative reduction in serum chitotriosidase activity from baseline to six months, the primary endpoint, across all dosages compared to placebo, although statistical significance was not achieved. A statistically significant and clinically meaningful dose-dependent reduction in liver size was observed, in addition to a clinically meaningful dose-dependent reduction in spleen size. At this time, the Company does not intend to conduct any further trials in Gaucher disease.

Amyotrophic Lateral Sclerosis (ALS)
In May 2021, Orphazyme announced that the Phase 3 ORARIALS-01 pivotal trial of arimoclomol in ALS did not meet its primary and secondary endpoints. No important safety signals were reported in the trial. The phase 3 ALS trial was an 18-month, placebo-controlled trial including 245 patients randomized (2:1 ratio) to receive either arimoclomol (248 mg three times daily) or placebo for up to 76 weeks. The primary endpoint was to determine the efficacy of chronic treatment with arimoclomol compared to placebo in participants with ALS as assessed by the combined assessment of function and survival (CAFS). This endpoint was selected to illustrate the overall treatment effect based on survival and the change in the ALS Functional Rating Scale-Revised (ALSFRS-R) score. Secondary endpoints included survival, change in ALSFRS-R, and slow vital capacity (SVC). Topline data were presented at the virtual European Network to Cure ALS (ENCALS) meeting in May 2021, and complete data from the study is expected to be submitted for publication in a peer-reviewed journal in due course. As a result of the findings from the Phase 3 trial in ALS, Orphazyme has ceased further development of arimoclomol in this indication.

Inclusion Body Myositis (IBM)
In March 2021, Orphazyme announced that the phase 2/3 trial evaluating arimoclomol for the treatment of inclusion body myositis (IBM), a progressively debilitating muscle-wasting disease, did not meet its primary and secondary endpoints. The primary goal was to evaluate the treatment effect on disease progression as measured by the inclusion body myositis functional rating scale (IBMFRS). The randomized, placebo-controlled trial was conducted among 150 IBM patients at 12 sites in North America and Europe, in partnership with University College of London and the University of Kansas. Participants were randomized (1:1 ratio) to receive either arimoclomol citrate (400 mg three times daily) or placebo for up to 20 months. No important safety concerns were detected in the trial. Complete findings from the study will be shared in a future scientific forum. As a result of the data from the Phase 2/3 trial in IBM, Orphazyme has ceased further development of arimoclomol in this indication.

New Molecular Entities (NMEs)
Orphazyme is focusing its development activities on the clinical and regulatory activities needed to support approval of arimoclomol in NPC and has therefore limited its research on next generation Heat-Shock Protein (HSP) amplifiers and lysosome biology-targeting compounds, while exploring potential avenues to realize value from these assets going forward.

Financial Review

Income statement
The net result for the first six months of 2021 was a loss of DKK 463.8 million compared to a loss of DKK 251.4 million for the same period in 2020. The increased net loss was due to higher costs recognized during the period as we accelerated preparation for commercial launch in the U.S. during most of the period before receipt of the Complete Response Letter (CRL) from the FDA for arimoclomol for the treatment of NPC in mid-June; as well as costs recognized for accelerated close out of the IBM and ALS activities following unfavorable trial results in March 2021 and May 2021, respectively. The unfavorable trial results led to the recognition of impairment losses of licenses relating to IBM and ALS recognized within intangible assets. Further, following the CRL from the FDA, Orphazyme initiated a restructuring program including a reduction of the total global workforce of approximately two thirds and terminating contracts not deemed necessary in the continued pursuit of regulatory approval in Europe and the U.S.

Net revenue
Net revenue amounted to DKK 13.2 million and represents revenue from sales in France as part of the remunerated early access program (“nATU”). During the current six-month period, Orphazyme has recognized revenue for the first time and therefore there are no comparatives for 2020. The revenue is recognized net of any estimated repayment to be made once the drug product is approved by the French authorities. Cost of goods sold relating to products sold under the French nATU is recognized in research and development expenses, as the drug products sold have not yet received approval for commercial sale, consequently any related inventory is impaired and written off.

Research and development expenses
Research and development (R&D) expenses totaled DKK 264.7 million for the first six months of 2021 compared to DKK 167.0 million for the same period in 2020. The increase of DKK 97.7 million is mainly attributed to manufacturing costs in the amount of DKK 52 million; the recognition of expenses for development activities related to IBM and ALS, including a provision for accelerated trial close-out costs and recognition of impairment losses on licenses and right-of-use assets after receiving unfavorable trial results; In addition, following the CRL from the FDA, we have initiated an employee restructuring program to make approximately two-thirds of our global workforce redundant, which led us to recognize DKK 1.4 million for our R&D workforce.

General and administrative expenses
General and administrative (G&A) expenses totaled DKK 214.2 million for the first six month of 2021 compared to DKK 78.6 million for the same period in 2020. The increase of DKK 135.6 million was primary due to the build-up of the commercial organization, including commercial launch preparation activities as well as expenses related to support functions during most of the period before receipt of the CRL from the FDA in mid-June. General and administrative expenses are also impacted by an impairment charge for software and the employee restructuring program, which resulted in the recognition of DKK 11.4 million attributable to the redundancy of our G&A work force.

Net financial items
Net financial items totaled an expense of DKK 0.4 million for the first six months of 2021 compared to an expense of DKK 7.8 million for the same period in 2020. Financial income of DKK 6.5 million related mainly to currency gains. Financial expenses of DKK 6.9 million related mainly to interest expense of DKK 4.1 million attributed to the loan agreement with Kreos and DKK 0.6 million resulting from the fair value adjustment on the facilitation fee accounted for as an embedded call option. The remaining amount relates to interest expense on cash and bank balances and lease liabilities.

Income tax benefit
Income tax benefit totaled DKK 2.3 million for the first six months of 2021 compared to DKK 2.0 million for the same period in 2020. Income tax benefit in both periods relates to the tax credit for research and development expenses at the applicable tax rate under the Danish Corporate Income Tax Act. The amount of the tax benefit has been reduced by income tax expenses for the current year in the subsidiaries in the U.S. and in Switzerland.

Statement of financial position

Cash
As of June 30, 2021, Orphazyme held cash of DKK 334.2 million compared to DKK 726.9 million as of December 31, 2020. The decrease is the result of operating expenses recognized during the period.

Equity
As of June 30, 2021, total equity amounted to DKK 166.8 million compared to DKK 620.5 million as of December 31, 2020. The decrease is due to the net loss for the period.

Cash flows

Cash flow from operating activities
Net cash flow from operating activities amounted to an outflow of DKK 381.7 million in the six-month period ended June 30, 2021 compared to DKK 204.2 million in the six-month period ended June 30, 2020. Net cash flow from operating activities is impacted by the net loss for the period off-set by a positive impact from the change in provisions, trade payables, accruals, and other liabilities due to the recognition of costs relating to the restructuring program and onerous contracts.

Cash flow from investing activities
Net cash flow from investing activities amounted to an outflow of DKK 0.8 million in the six-month period ended June 30, 2021 compared to DKK 1.8 million in the six-month period ended June 30, 2020. In 2020, cash flow from investing activities was mainly impacted by the acquisition of the ERP system.

Cash flow from financing activities
Net cash flow from financing activities amounted to an outflow of DKK 15.5 million in the six-month period ended June 30, 2021 compared to an inflow of DKK 692.9 million in the six-months period ended June 30, 2020. In 2021, cash flow from financing activities was impacted by payments to Kreos on the loan agreement; whereas in 2020, cash flow from financing activities was significantly impacted by the proceeds from the financing round completed in February 2020.

Consolidated Statements of Profit or Loss and Other Comprehensive Income

	Six months ended Jun 30, 2021 DKK (000)	Six months ended Jun 30, 2020 DKK (000)
Net revenue (Note 4)	13,152	-
Research and development expenses (Note 5)	(264,683)	(166,980)
General and administrative expenses (Note 6)	(214,167)	(78,575)
Operating loss	**(465,698)**	**(245,555)**
Financial income	6,521	126
Financial expenses	(6,921)	(7,967)
Loss before tax	**(466,098)**	**(253,396)**
Income tax benefit	2,318	1,981
Net loss for the period	**(463,780)**	**(251,415)**
Exchange differences from translation of foreign operations	76	(135)
Total comprehensive loss	**(463,704)**	**(251,550)**
Loss per share, basic and diluted (Note 12)	(13.27)	(9.77)

The accompanying notes are an integral part of these interim financial statements.

Consolidated Statements of Financial Position

	Jun 30, 2021 DKK (000)	Dec 31, 2020 DKK (000)
ASSETS		
Non-current assets		
Intangible assets (Note 8)	2,194	12,454
Right-of-use assets (Note 8)	10,012	14,859
Property, plant, and equipment	3,916	4,687
Corporation tax receivable	5,500	2,750
Deferred tax assets	-	2,065
Prepayments and deposits	2,714	2,014
Total non-current assets	**24,336**	**38,829**
Current assets		
Corporation tax receivable	7,600	5,500
Trade receivables (Note 4)	26,304	-
Prepayments and other receivables	35,607	51,235
Cash	334,184	726,929
Total current assets	**403,695**	**783,664**
TOTAL ASSETS	**428,031**	**822,493**
EQUITY & LIABILITIES		
Equity		
Share capital (Note 11)	34,952	34,698
Share premium (Note 11)	2,082,486	2,082,254
Other reserves	3,277	6,494
Accumulated deficit	(1,953,954)	(1,502,921)
Total equity	**166,761**	**620,525**
Non-current liabilities		
Borrowings	12,861	23,830
Lease liabilities	9,743	9,877
Other non-current liabilities (Note 4)	15,006	1,634
Total non-current liabilities	**37,610**	**35,341**
Current liabilities		
Provisions (Note 9)	49,842	-
Borrowings	33,933	33,349
Lease liabilities	2,053	3,657
Trade payable and accruals	98,984	72,135
Corporation tax payable	115	4,159
Other liabilities	38,733	53,327
Total current liabilities	**223,660**	**166,627**
TOTAL EQUITY AND LIABILITIES	**428,031**	**822,493**

The accompanying notes are an integral part of these interim financial statements.

Consolidated Statements of Changes in Shareholders' Equity

			Other reserves			
	Share capital DKK (000)	Share premium DKK (000)	Foreign currency translation reserve DKK (000)	Share-based compensation – acquisition of intangible assets DKK (000)	Accumulated deficit DKK (000)	Total DKK (000)
Balance as of December 31, 2019	**19,984**	**924,021**	**109**	**7,873**	**(899,018)**	**52,969**
Net loss for the period	-	-	-	-	(251,415)	(251,415)
Other comprehensive loss for the period	-	-	(135)	-	-	(135)
Total other comprehensive loss	-	-	*(135)*	-	*(251,415)*	*(251,550)*
Transactions with owners						
Capital increase, Bonus Shares	21	-	-	(2,094)	2,073	-
Capital increase, exercise of RSUs	7	394	-	-	-	401
Capital increase, private placement	7,033	738,458	-	-	-	745,491
Transaction costs	-	(51,243)	-	-	-	(51,243)
Share-based payment costs	-	-	-	-	10,067	10,067
Total transactions with owners	*7,061*	*687,609*	-	*(2,094)*	*12,140*	*704,716*
Balance as of June 30, 2020	**27,045**	**1,611,630**	**(26)**	**5,779**	**(1,138,293)**	**506,135**

			Other reserves			
	Share capital DKK (000)	Share premium DKK (000)	Foreign currency translation reserve DKK (000)	Share-based compensation – acquisition of intangible assets DKK (000)	Accumulated deficit DKK (000)	Total DKK (000)
Balance as of December 31, 2020	**34,698**	**2,082,254**	**714**	**5,780**	**(1,502,921)**	**620,525**
Net loss for the period	-	-	-	-	(463,780)	(463,780)
Other comprehensive loss for the period	-	-	76	-	-	76
Total other comprehensive loss	-	-	*76*	-	*(463,780)*	*(463,704)*
Transactions with owners						
Capital increase, issuance of Matching Shares, net of costs (Note 7)	170	-	-	-	-	170
Capital increase, Bonus Shares (Note 7)	22	-	-	(1,645)	1,623	-
Cash settlement of Bonus Shares (Note 7)				(1,648)		(1,648)
Capital increase, issuance of sign-on bonus shares to former CEO (Note 7)	58	-	-	-	-	58
Capital increase, exercise of RSUs (Note 7)	4	232	-	-	-	236
Share-based payment costs (Note 7)	-	-	-	-	11,123	11,123
Total transactions with owners	*254*	*232*	-	*(3,293)*	*12,746*	*9,939*
Balance as of June 30, 2021	**34,952**	**2,082,486**	**790**	**2,487**	**(1,953,955)**	**166,761**

The accompanying notes are an integral part of these interim financial statements.

Consolidated Statements of Cash Flow

	Six months ended Jun 30, 2021 DKK (000)	Six months ended Jun 30, 2020 DKK (000)
Operating activities		
Operating loss	(465,698)	(245,555)
Adjustments to reconcile loss before tax to cash flows from operating activities:		
Equity-settled share-based compensation expense (Note 7)	11,123	10,067
Depreciation, amortization and impairment losses	16,812	2,250
Change in trade receivables, prepayments, deposits and other receivables	(11,377)	(9,061)
Change in provisions, trade payables, accruals and other liabilities	75,468	37,400
Corporation taxes received / (paid)	(4,181)	5,500
Interest received / (paid), net	(3,871)	(4,770)
Net cash used in operating activities	**(381,724)**	**(204,169)**
Investing activities		
Purchase of intangible assets	(806)	(590)
Purchase of property, plant, and equipment	-	(1,170)
Net cash used in investing activities	**(806)**	**(1,760)**
Financing activities		
Proceeds from issuance of shares (Note 7)	464	745,892
Cash settlement of Bonus Shares (Note 7)	(1,648)	-
Transaction costs related to issuance of shares	-	(51,243)
Repayment of borrowings	(12,540)	-
Repayment of lease liabilities	(1,803)	(1,705)
Net cash provided by (used in) financing activities	**(15,527)**	**692,944**
Net change in cash and cash equivalents	**(398,057)**	**487,015**
Cash balance at beginning of period	726,929	123,588
Effect of changes in exchange rates	5,312	(155)
Cash balance at end of period	**334,184**	**610,448**

The accompanying notes are an integral part of these interim financial statements.

Notes to the Financial Statement

NOTE 1 – CORPORATE INFORMATION

Orphazyme A/S (the "Company") is a late-stage biopharmaceutical company. The Company is a limited liability company publicly traded on Nasdaq Copenhagen with headquarters in Copenhagen, Denmark. The company's American Depositary Shares are listed on the Nasdaq Global Select Market in the U.S. The group consists of the headquarters in Copenhagen, Orphazyme A/S; a wholly-owned subsidiary in the U.S., Orphazyme U.S., Inc.; and a wholly-owned subsidiary in Switzerland, Orphazyme Schweiz GmbH (together "Orphazyme" or "the Group").

Following receipt of a Complete Response Letter from the FDA for arimoclomol for NPC, a restructuring program reducing our global workforce by approximately two thirds was initiated and finally concluded by July 31, 2021. Employees in the Group's subsidiaries and in headquarters were affected. Following our restructuring, we have strategically maintained medical and commercial personnel to strengthen our footprint in areas with the most significant market potential for NPC. We have maintained a core team in the U.S. to effectively act on any actions agreed with the FDA; and we have maintained a core team in Europe and the UK to continue to support our early access programs and establish our commercial presence in those respective countries.

NOTE 2 – BASIS OF PREPARATION AND UPDATES TO THE GROUP'S ACCOUNTING POLICIES

Basis of preparation
The interim condensed consolidated financial statements for the six months ended June 30, 2021 have been prepared in accordance with IAS 34 *Interim Financial Reporting* as issued by the International Accounting Standards Board (IASB) and as adopted by the EU, and additional Danish disclosure requirements for interim reports of companies listed on the Nasdaq Copenhagen.

The interim condensed consolidated financial statements do not include all the information and disclosures required in annual financial statements and should be read in conjunction with Orphazyme A/S' latest consolidated annual financial statements as of December 31, 2020. These interim condensed consolidated financial statements have been prepared in accordance with the going concern assumption (Note 3).

COVID-19

At this time, there is no material impact directly related to COVID-19 on the Group's consolidated financial statements, including the judgements and estimates applied. Specifically, Orphazyme is not currently conducting any clinical trials and in the process of closing out the existing trial extensions following the disappointing results of the Phase 2/3 clinical trial related to IBM and the Phase 3 clinical trial related to ALS. However, as Orphazyme is assessing the potential path forward with the FDA following receiving a complete response letter from the FDA on arimoclomol for the treatment of NPC and is actively pursuing the European regulatory approval, other parts of the business and operations may be adversely impacted by the effects of COVID-19, for example: our third-party manufacturers and other third parties; the productivity of our staff; ability to attract, integrate, manage and retain qualified personnel or key employees; our global supply chains and relationships with vendors and other parties; significant disruption of global financial markets; and reduced ability to secure additional funding. We will continue to monitor the COVID-19 pandemic and its potential impact on our business and financials.

Updates to the Group's accounting policies
The accounting policies used in the preparation of the interim condensed consolidated financial statements are consistent with those used in the preparation of Orphazyme's annual consolidated financial statements for the year ended December 31, 2020, with any additions included in the respective notes below. No new or amended standards became applicable for the current reporting period; and consequently, there is no resulting impact on the interim condensed consolidated financial statements of the Group.

Significant accounting estimates and judgements
In addition to the significant accounting estimates and judgements disclosed in note 1.4 of the consolidated financial statements for the year ended December 31, 2020, Management has prepared these interim condensed consolidated financial statements under the going concern assumption (Note 3); and the company has estimated the revenue recognized using the 'expected value' method (see Note 4).

NOTE 3 - GOING CONCERN

On June 18, 2021, Orphazyme announced that it received a Complete Response Letter (CRL) from the U.S. Food and Drug Administration (FDA) following the FDA's review of the new drug application for arimoclomol, a heat shock protein amplifier intended for the treatment of Niemann-Pick disease type C (NPC). As a result of arimoclomol for NPC not being approved by the FDA, Orphazyme cannot start the commercialization of its drug product in the U.S., which would have contributed to revenue and to reduced operating loss. Furthermore, Orphazyme was not able to obtain a priority review voucher in 2021.

Receiving the CRL from the FDA and the resulting consequences for our cash position introduces a going concern risk and consequently, Management has made an assessment of Orphazyme's ability to continue as a going concern for the twelve-month period following the balance sheet date of June 30, 2021. In connection with this assessment, several uncertainties have been identified and, as a result, mitigating actions have been initiated.

The following significant uncertainties were identified as of June 30, 2021:

- ***Path forward with the FDA:*** Following the receipt of the CRL from the FDA, Orphazyme will request a Type-A meeting with the FDA to assess a path forward for arimoclomol for NPC in the U.S. There is no guarantee that Orphazyme will ultimately obtain an approval for arimoclomol for NPC in the U.S.

- ***Outcome of CHMP assessment and subsequent regulatory approval by EMA:*** Orphazyme expects to receive an opinion from the European Committee for Medicinal Products for Human Use (CHMP) in Q4 2021. The CHMP is conducting an initial assessment of our application for EU-wide marketing authorizations for arimoclomol for NPC. Whereas a positive response from CHMP does not automatically result in an approval from the European Medicines Agency (EMA), a positive response increases the likelihood of receiving approval from EMA. Orphazyme expects to receive feedback on potential approval from EMA in the first quarter of 2022. There is no guarantee that Orphazyme will obtain positive feedback from CHMP or approval from EMA.

- ***Cash usage***: With the current cash position and the current estimated outlook, Orphazyme expects to have approximately DKK 50 million in cash at the end of December 2021. Following our restructuring and close-out of clinical trials, we anticipate to significantly reduce our monthly cash usage to sustain our reduced organization and activities. Any significant deviation in our cash usage can materially impact our anticipated cash position at year-end 2021. There is no guarantee that the estimated cash usage will materialize as expected.

- ***Additional funding:*** The current expectation for future cash requirements indicates that Orphazyme will need to raise additional funding. The Group is therefore currently assessing different possibilities for obtaining additional funding. However, if further funding is not obtained, Orphazyme will not have sufficient cash to fund its activities and meet its obligations.

In response to these uncertainties, the following mitigating actions have been taken:

- ***Restructuring program:*** Following receipt of the CRL from the FDA, a restructuring program reducing the global workforce by approximately two thirds was initiated and finally concluded by July 31, 2021. The company terminated certain employees in the Group's subsidiaries and in headquarters, who were not deemed to be essential. Following our restructuring, we have strategically maintained medical and commercial personnel to strengthen our footprint in areas with the most significant market potential for NPC. We have maintained a core team in the U.S. to effectively act on any actions agreed with the FDA; and we have maintained a core team in Europe and the UK to continue to support our early access programs and establish our commercial presence in those respective territories.

- ***Termination of contracts and agreements:*** Management has initiated termination of contracts and agreements not deemed essential in the continued pursuit of regulatory approval in Europe and assessing a path forward in the U.S. This includes initiating the close-out of all clinical trial activities for IBM, ALS and Gaucher.

- ***Additional funding:*** Management continuously assesses options to ensure sufficient funding is in place to support its corporate priorities. We are currently evaluating a number of strategic options including (i) debt and/or equity financing; (ii) new potential commercial relationships, and (iii) monetization of non-core assets.

With the uncertainties listed above, there can be no assurance that Orphazyme will achieve or sustain profitability or positive cash flows from operations. If the Group is unable to attract additional funding, future operations will need to be significantly scaled back or discontinued. These conditions raise substantial doubt about the Group's ability to continue as a going concern. These interim condensed consolidated financial statements do not include any adjustments that might result from the outcome of this uncertainty. Based on the mitigating actions described above, Management is of the view that a going concern assumption is appropriate as of June 30, 2021.

NOTE 4 – NET REVENUE

Orphazyme recognizes revenue in accordance with IFRS 15 *Revenue from Contracts with Customers* and, as a result, follows the five-step model when recognizing revenue: 1) identifying a contract; 2) identifying the performance obligations; 3) determining the transaction price; 4) allocating the price to the performance obligations; and 5) recognizing revenue when the performance obligations have been fulfilled.

Net revenue comprises revenue from the sale of arimoclomol for the treatment of NPC under the remunerated early access compassionate use program ("nATU") in France. An early access compassionate use program is a program giving specific patients access to a drug, which is not yet on the market. Only drugs targeting serious or rare indications and for which there is currently no appropriate treatment are considered for early access compassionate use programs. Further, to be considered for the early access compassionate use program, the drug must have proven efficacy and safety and must either be undergoing price negotiations or seeking marketing approval. Orphazyme is currently pursuing regulatory approval from the European Medicines Agency (EMA).

Revenue is recognized when the drug products are sold to the customer, i.e., at the time when control over the drug product is transferred to the third-party customer.

Under the French nATU, the manufacturer can set its own price for the drug products until a price agreement with the authorities is in place. Any excess in the price charged by the manufacturer compared to the price agreed with the health authorities once the drug product is approved in France must be repaid. Orphazyme is therefore recognizing net revenue in accordance with IFRS 15 *Revenue from Contracts with Customers* using the 'expected value'-method, which is based on probability-weighted consideration of different agreed-upon sales prices. The estimated net revenue is recognized less VAT and other sales related taxes. A long-term liability is recognized for the amount estimated to be repaid.

For the six-month period ended June 30, 2021, Orphazyme recognized net revenue of DKK 13.2 million (DKK 0 for the six-month period ended June 30, 2020), which is net of certain estimated repayment amounts that may become due following potential approval of our drug product from the French authorities. The estimated amount to be repaid is accrued as a long-term liability under the line item Other non-current liabilities.

Trade receivables in the amount of DKK 26.3 million are recognized in the balance sheet at the total invoiced amount less any expected credit losses. Due to the nature of the revenue transactions, expected credit losses are very limited.

Cost of goods sold relating to products sold under this French nATU is recognized as research and development expenses. As the drug products sold have not yet received approval for commercial sale, any related inventory is impaired and written off (Note 5).

NOTE 5 – RESEARCH AND DEVELOPMENT EXPENSES

Research and development expenses of DKK 264.7 million (June 30, 2020: DKK 167.0 million) include employee costs for research and development personnel (i.e. salaries, bonuses, employer contributions to pension schemes, share-based compensation); external costs for further development of our pipeline, including ongoing clinical trials and clinical pharmacology registration trials; and pre-launch inventory costs. Further, research and development expenses include costs of goods sold relating to products sold under the French early access compassionate use program (Note 4). For the six-months ended June 30, 2021, an impairment charge was recognized for intangible assets and right-of-use assets (Note 8).

Included in total research and development expenses is an amount of DKK 37.7 million attributable to restructuring activities, i.e. clinical trial close-out costs (DKK 36.3 million) and employee redundancies (DKK 1.4 million).

NOTE 6 – GENERAL AND ADMINISTRATIVE EXPENSES

General and administrative expenses of DKK 214.2 million (June 30, 2020: DKK 78.6 million) include pre-launch costs of DKK 123.8 million (June 30, 2020: DKK 44.1 million) associated with establishing a commercial organization and the escalation of launch preparation activities, including hiring a commercial team in our subsidiaries in the U.S. and Switzerland and medical affairs activities to further engage with the scientific community through communication and education programs. Furthermore, included in general and administrative expenses is DKK 61.8 million (June 30, 2020: DKK 34.5 million) relating to salaries for administrative employees and Executive Management, remuneration to the Board of Directors, share-based compensation costs, audit fees, legal costs and investor relations costs; as well as an impairment charge related to software used in pre-launch and administrative activities (Note 8). Included in these amounts is DKK 12.1 million attributable to restructuring activities, i.e. employee redundancies (DKK 11.4 million) and termination of onerous contracts (DKK 0.7 million).

NOTE 7 – SHARE-BASED COMPENSATION COSTS

Please refer to Note 2.5 of the Group's consolidated financial statements included in the 2020 Annual Report for a description of the share-based compensation programs and the accounting policies and estimates applied. The activities in the respective programs are outlined below. The respective share-based compensation programs allow the vesting of awards through the employment release date. As such, there are no forfeitures to be disclosed or expense to be reversed due to the termination of employees as a result of our restructuring.

a) Long-term incentive programs (equity-settled)

2021 Long-term incentive program ("2021 LTIP"):

Similar to the 2017 LTIP, the 2019 LTIP and the 2020 LTIP described in Note 2.5 of the consolidated financial statements for the year ended December 31, 2020, the 2021 LTIP offers all permanent employees of Orphazyme a certain number of Restricted Share Units (RSUs) contingent on continued employment and Performance Share Units (PSUs) depending on the development of the Company's share price. Although the grant date of these awards was in April 2021, management has determined that the service commencement date of the participants was the earlier of January 1, 2021, or the employee's employment date, and consequently the compensation expense is recognized from the service commencement date.

The fair value of the 2021 LTIP awards was estimated separately for the RSUs and PSUs using a Black-Scholes model and a Monte-Carlo simulation, respectively, at the grant date of April 21, 2021. The following inputs were used in the valuation models:

	RSUs	**PSUs**
Dividend yield (%)	-	-
Expected volatility (%)	55.6%	55.6%
Risk-free interest rate (%)	(0.53%)	(0.53%)
Expected life (years)	2.69	2.69
Share price (DKK)	59.05	59.05
Fair value of awards at measurement date (DKK 000)	19,653	6,080

The risk-free interest rate has been estimated based on Danish government bonds with similar maturities. Expected volatility has been determined based on Orphazyme's own historic volatility. Based on the fair value of the 2021 LTIP, an expense of DKK 6.0 million was recognized for the six-month period ending June 30, 2021.

In addition, for the six-month period ending June 30, 2021, an aggregate expense of DKK 5.1 million was recognized for the 2020 LTIP, 2019 LTIP and 2017 LTIP for the six-month period ending June 30, 2021 compared to DKK 10.1 million recognized for the six-month period ended June 30, 2020.

In January 2021, the Matching Shares from the 2020 LTIP fully vested and were issued to the participants in exchange for the nominal value of DKK 1 per share. This resulted in cash received of DKK 170,131 and a capital increase of the same number of shares.

b) Phantom share-based incentive programs (cash-settled)

As described in Note 2.5 of the consolidated financial statements for the year ended December 31, 2020, the Phantom programs are cash-settled and their fair value is re-assessed at each reporting date. Management used a Monte-Carlo simulation model with the following inputs to estimate the fair value as of June 30, 2021:

Valuation Date:	**June 30, 2021**	**June 30, 2021**	**June 30, 2021**	**June 30, 2**
Program	2020-1	2019-2	2019-1	20
Dividend yield (%)	-	-	-	
Expected volatility (%)	79.1%	91.6%	91.6%	11(
Risk-free interest rate (%)	(0.46%)	(0.52%)	(0.52%)	(0.5
Expected life (years)	3.58	2.58	2.58	
Share price (DKK)	46.84	46.84	46.84	4
Fair value at measurement date (DKK 000)	283	28	196	

Valuation Date:	**Dec 31, 2020**	**Dec 31, 2020**	**Dec 31, 2020**
Program	2020	2019	2018
Dividend yield (%)	-	-	-
Expected volatility (%)	47.1%	47.3%	54.3%
Risk-free interest rate (%)	(0.59%)	(0.61%)	(0.61%)
Expected life (years)	4.08	3.08	2.08
Share price (DKK)	67.10	67.10	67.10
Fair value at measurement date (DKK 000)	406	293	160

The risk-free interest rate has been estimated based on Danish government bonds with similar maturities. Expected volatility has been determined based on Orphazyme's own historic volatility. Based on the fair value of the awards on June 30, 2021, a credit of DKK 10 thousand was recognized for the six-month period ended June 30, 2021 compared to an expense of DKK 0.2 million recognized for the six-month period ended June 30, 2020.

c) Restricted share units (cash-settled)

In March 2020, the 2019 RSUs granted to the Board of Directors in 2019 fully vested. As of June 30, 2021, all but one board member exercised the fully vested RSUs.

Also in March 2020, the 2020 RSU program was announced, granting the Board of Directors an aggregate of 15,177 RSUs under similar terms and conditions as the 2019 RSUs described in Note 2.5 of the consolidated financial statements for the year ended December 31, 2020. In September 2020, a new RSU incentive program was announced (2020-2 RSU program), which comprised 22,993 RSUs in total, including an on-boarding grant to a new board member in accordance with the Company's remuneration policy. The 2020-2 RSU program runs in parallel with the 2020 RSU program and board members can only exercise RSUs under one of the programs. As of June 30, 2021, these RSUs were fully vested but unexercised.

In May 2021, an aggregate of 30,450 RSUs were granted to the Board of Directors under the 2021 RSU program under similar terms and conditions as the 2019 RSUs described in Note 2.5 of the consolidated financial statements for the year ended December 31, 2020. The 2021 RSUs fully vest in March 2022.

Management used a Black-Scholes model with the following inputs to estimate the fair value of the cash-settled 2021 RSUs and 2020 RSUs as of June 30, 2021:

	2021 RSUs	2020 RSUs
Dividend yield (%)	-	-
Expected volatility (%)	117.6%	249.9%
Risk-free interest rate (%)	(0.54%)	(0.51%)
Expected life (years)	1.25	0.25
Share price (DKK)	46.84	46.84
Fair value at measurement date (DKK 000)	1,396	1,054

The risk-free interest rate has been estimated based on Danish government bonds with similar maturities. Expected volatility has been determined based on Orphazyme's own historic volatility. Based on the fair value of the 2021 RSUs on June 30, 2021, an expense of DKK 0.3 million was recognized for the six-month period ended June 30, 2021. In addition, an amount of DKK 0.3 million was recognized for the 2020 RSUs for the six-month period ended June 30, 2021 compared to DKK 0.1 million recognized for the six-month period ended June 30, 2020.

d) ***Bonus shares***

As part of the license agreement with KLSDC and UCL described in Note 3.1 of the consolidated financial statements for the year ended December 31, 2020, consideration to KLSDC and UCL is payable in shares of the Company ("Bonus Shares") each January and is based on incurred costs reported by KLSDC and UCL for the previous year. Subsequently, it was agreed, that as an alternative to the issuance of shares, a cash equivalent of the value of the Bonus Shares may be paid instead. As at December 31, 2020 the aggregate costs incurred by KLSDC and UCL amounted to USD 0.5 million (DKK 3.2 million). A total of 22,553 Bonus Shares ("2021 Bonus Shares") were issued to KLSDC based on the average 30-day closing price of Orphazyme's shares and an amount of USD 0.3 million (DKK 1.6 million) was paid to UCL. At the time of the share issuance and the cash transfer, the equity reserve was decreased by DKK 3.3 million, which represents the market value of the shares issued and the cash paid.

NOTE 8 - IMPAIRMENT OF ASSETS

As disclosed in Note 3.1 of the consolidated financial statements for the year ended December 31, 2020, Orphazyme entered into a license agreement with KU Center for Technology Commercialization Inc., University of Kansas, Kansas Life Sciences Development Company, Inc. (KLSDC) and UCL Business PLC (UCL) in 2017, granting Orphazyme the right to develop and commercialize products under all data generated in the course of the Phase 2/3 clinical trial on arimoclomol for the treatment of IBM worldwide. In March 2021, it was announced, that the phase 2/3 trial evaluating arimoclomol for the treatment of inclusion body myositis (IBM), a progressively debilitating muscle-wasting disease, did not meet its primary and secondary endpoints. As a result, Orphazyme has recognized an impairment loss of DKK 7.5 million corresponding to the remaining carrying amount of the license agreement. The impairment loss was recognized under research and development expenses.

Further, as also disclosed in Note 3.1. of the consolidated financial statements for the year ended December 31, 2020, Orphazyme entered into an exclusive license agreement with the University of Miami in 2019, granting Orphazyme a global royalty-bearing, exclusive license to all data, know-how, inventions and technology generated by the University of Miami and certain other institutions in a Phase 2 clinical trial of arimoclomol in ALS with the A4V SOD1 mutation to research, develop, make, use or sell certain

pharmaceutical products or processes containing arimoclomol. In May 2021, it was announced, the ORARIALS-01 pivotal trial of arimoclomol in amyotrophic lateral sclerosis (ALS) did not meet its primary and secondary endpoints to show benefit in people living with ALS. As a result, Orphazyme has recognized an impairment loss of DKK 0.5 million corresponding to the remaining carrying amount of the license agreement. The impairment loss was recognized under research and development activities.

In addition, Orphazyme has impaired leased laboratory equipment classified as a right-of-use asset in the amount of DKK 3.2 million. This amount is recognized under research and development activities.

Lastly, Orphazyme has impaired software and other assets recognized within intangible assets at a total amount of DKK 2.6 million. The impairment loss was recognized under general and administrative expenses, as the assets were used in pre-launch or administrative activities.

NOTE 9 – PROVISIONS FOR RESTRUCTURING COSTS

Provisions for restructuring are recognized when the plan for the restructuring has been prepared, and the employees affected have a valid expectation that the restructuring program will be fulfilled. Provisions for onerous contracts are recognized to the extent that obligations under the contract exceed the economic benefit from the contract.

In June 2021, Orphazyme announced a restructuring program, including a reduction of the global workforce by approximately two thirds and terminating contracts and agreements not deemed necessary in the continued pursuit of regulatory approval in Europe and in the U.S. A total amount of DKK 49.8 million has been recognized as restructuring costs, of which DKK 12.9 million relates to the reduction of the global workforce and DKK 36.9 million relates to termination of contracts and agreements. As a result of the phase 2/3 trial evaluating arimoclomol for the treatment of inclusion body myositis (IBM) and the ORARIALS-01 pivotal trial of arimoclomol in amyotrophic lateral sclerosis (ALS) not meeting the primary and secondary endpoints, several contracts with CROs and other vendors have been terminated.

The restructuring costs were expensed under research and development expenses or general and administrative expenses, depending on their nature (see Notes 5 and 6).

NOTE 10 – FINANCIAL LIABILITIES

As disclosed in Note 3.6 of the consolidated financial statements for the year ended December 31, 2020, the structured debt facility with Kreos (“Loan Agreement”) entered into in August 2019 includes a Facilitation Fee that is due and payable by Orphazyme at the sole discretion of the lender. The Facilitation Fee is an amount equal to the greater of (i) 10% of the aggregate amount of the amount borrowed and (ii) the percentage increase in the Company’s share price between the 30-day volume-weighted average share price on the date of the Loan Agreement and the closing share price on the day immediately preceding the date of the payment request notification by the lender applied to the aggregate amount of amounts borrowed. The variability arising from the change in Orphazyme’s share price is not closely related to the host debt instrument characterized mainly by interest rate and credit risk. Therefore, the embedded equity-linked amount is separated from the host debt instrument and accounted for as an embedded written call option at fair value through profit and loss. The call option is measured at fair value at level 2 in the fair value hierarchy using a Black-Scholes option valuation model. In measuring the fair value, various observable and unobservable inputs are required. Observable input mainly relates to the market price of Orphazyme’s shares, and risk-free interest rate. Unobservable inputs mainly relate to the expected volatility of Orphazyme’s share price and the term of the option.

The table below shows the inputs used in the valuation of the call option and the estimated fair value on June 30, 2021 and December 31, 2020:

	June 30, 2021	Dec 31, 2020
Dividend yield (%)	-	-
Expected volatility (%)	110.6%	54%
Risk-free interest rate (%)	(0.54%)	(0.61%)
Expected life (years)	1.75	2.2
Share price (DKK)	46.84	67.1
Fair value of call option (DKK 000)	1,422	838

The change in fair value of the call option is recognized as a finance expense in the statement of profit or loss. Based on the fair value of the call option on June 30, 2021, an expense of DKK 0.6 million (June 30, 2020: DKK 0.7 million) was recognized for the period.

NOTE 11 – EQUITY

The following table summarizes the Company's share activity:

	Ordinary Shares
December 31, 2019	**19,984,799**
Issuance of Bonus Shares as part of license agreement	20,650
Issuance of shares in related to directed issue and private placement	7,032,937
Issuance of shares due to exercise of restricted share-units	6,543
June 30, 2020	**27,044,929**
Issuance of Matching Shares	31,250
Issuance of shares related to U.S. listing	7,616,146
Issuance of shares due to exercise of restricted share-units	5,378
December 31, 2020	**34,697,703**
Issuance of Bonus Shares as part of license agreement	22,553
Issuance of Matching Shares	170,131
Issuance of sign-on bonus shares to former CEO, Kim Stratton	58,000
Issuance of shares due to exercise of restricted share-units	3,854
June 30, 2021	**34,952,241**

As disclosed in Note 4.8 of the consolidated financial statements for the year ended December 31, 2020, in February 2021, the Company issued 170,131 Matching Shares to participants in the 2020 LTIP. In February 2021, 58,000 ordinary shares were issued to the former CEO, Kim Stratton, and also in February 2021, 22,553 Bonus Shares were issued as part of consideration payable to KLSDC relating to the license agreement (see Note 7d above).

As discussed in Note 7 above, in March 2021, certain members of the Board of Directors exercised restricted share units and the Company issued 3,854 shares in exchange for DKK 0.2 million.

Following the above activity, the total nominal share capital of the Company as of June 30, 2021, was DKK 34,952,241, representing 34,952,241 ordinary shares each with a nominal value of DKK 1.

In January 2020 the Company issued 20,650 shares as part of consideration payable to KLSDC and UCL relating to the license agreement.

On February 7, 2020, Orphazyme completed an offering of 7,032,937 shares in a directed issue and private placement and raised gross proceeds of approximately DKK 745 million and net proceeds of approximately DKK 694 million. The net proceeds of the directed issue and private placement is expected to support the U.S. and European filings for approval of arimoclomol for the treatment of Niemann-Pick disease Type C (NPC), as well as preparations for commercial launch.

The transaction consisted of a directed issue and private placement of up to 3,961,264 new shares of a nominal value of DKK 1 each (the "New Shares") and a private placement of up to 3,071,673 existing shares of a nominal value of DKK 1 each (the "Existing Shares" and together with the New Shares, the "Offer Shares") at an offer price of DKK 106 per Offer Share, as determined by the Board of Directors of the Company through a book-building process (the "Offering"). The New Shares were issued without pre-emption rights for existing shareholders.

The offering of Existing Shares was facilitated by a share loan to the Company from related parties Novo Holdings A/S and Orpha Pooling B.V. (the "Lending Shareholders") pursuant to a stock lending and subscription agreement with an obligation for the Company to redeliver new shares of an equivalent number as the Existing Shares borrowed by the Company from each of the Lending Shareholders (the "Replacement Shares"), which were issued without pre-emption rights for existing shareholders. The Lending Shareholders did not participate in the Offering and were only facilitating the loan of the Lending Shares for purposes of the Company's offering of Existing Shares in the Offering.

In April 2020 certain members of the Board of Directors exercised their 2019 RSUs and the Company issued 6,543 shares in exchange for DKK 0.4 million.

Following the above activity, the total nominal share capital of the Company as of June 30, 2020 was DKK 27,044,929, representing 27,044,929 ordinary shares each with a nominal value of DKK 1.

NOTE 12 – 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 June 30, 2021 and 2020:

	Six months ended June 30, 2021 DKK (000)	Six months ended June 30, 2020 DKK (000)
Loss for the period	(463,780)	(251,415)
Weighted-average shares outstanding	34,903,711	25,447,748*
Loss per share, basic and diluted	**(13.27)**	**(9.77)***

*Recalculated retrospectively as a result of Bonus Shares issued subsequently to June 30, 2020.

Basic loss per share is calculated by dividing the net loss attributable to ordinary shareholders for the period by the weighted-average number of ordinary shares outstanding during each period. Diluted loss per share is calculated by dividing the net loss by the weighted-average number of ordinary shares outstanding during the period increased by the dilutive effect of the assumed issuance of any outstanding share-based awards. Due to the fact that Orphazyme has incurred losses for each period presented, the potential shares issuable related to outstanding equity awards 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.

As disclosed in Note 4.3 of the consolidated financial statements for the year ended December 31, 2020, in February 2021, Bonus Shares were issued to KLSDC under the terms of the license agreement entered into in October 2017. Basic and diluted loss per share for the comparative period presented has been adjusted retrospectively to include these Bonus Shares in the number of weighted average shares outstanding for the six-months ended June 30, 2020.

NOTE 13 - COMMITMENTS AND CONTINGENCIES

There have been the following material changes in commitments and contingencies compared to year-end 2020.

Contractual obligations towards Contract Research Organizations (CROs)
As a result of the phase 2/3 trial evaluating arimoclomol for the treatment of inclusion body myositis (IBM) and the ORARIALS-01 pivotal trial of arimoclomol in amyotrophic lateral sclerosis (ALS) not meeting the primary and secondary endpoints, several contracts with CROs and other vendors have been terminated. A provision in the amount of DKK 36.3 million has been recognized to cover the onerous part of these contracts.

Contingency related to early access compassionate use program in France
Net revenue comprises revenue from the sale of arimoclomol for the treatment of NPC under the early access compassionate use program (“nATU”) in France. Under the French nATU, the manufacturer can set its own price for the drug products until a price agreement with the authorities is in place. Any excess in the price charged by the manufacturer compared to the price agreed with the health authorities once the drug product is approved in France must be repaid. Orphazyme is recognizing net revenue in accordance with *IFRS 15 Revenue from Contracts with Customers* using the ‘expected value’ method. The ‘expected value’ method is based on probability-weighted consideration of different agreed-upon sales prices. A long-term liability is recognized for the amount estimated to be repaid. However, Orphazyme has a contingent liability if the final price agreed with the health authorities is less than the price estimated by Orphazyme, which has been recognized as revenue.

Putative class action lawsuit
On July 9, 2021, a putative class action lawsuit was filed against the company and certain of its current and former directors and officers in the United States District Court for the Northern District of Illinois. This lawsuit alleges that certain representations about arimoclomol in the Company's U.S. IPO offering documents and in subsequent public statements were false and misleading, in violations of U.S. securities. Orphazyme does not believe these claims have merit and intends to vigorously defend itself against the lawsuit.

NOTE 14 – RELATED PARTIES

Orphazyme's related parties are Orphazyme A/S' subsidiaries, Orphazyme U.S. Inc. and Orphazyme Schweiz GmbH, Board of Directors, Executive Management, and close members of the family of these persons. Effective April 1, 2021, Christophe Bourdon was appointed Chief Executive Officer and a member of the Executive Management.

During the six-month period ended June 30, 2021, remuneration to Executive Management and the Board of Directors was made and share-based compensation awards were granted to Executive Management and the Board of Directors in line with the Company's remuneration policy. There were no material related party transactions during the first half of 2021.

NOTE 15 – SUBSEQUENT EVENTS

The most recent French Social Security law of December 14, 2020 (Article 78- La Loi de financement de la sécurité sociale, FSSL) for the healthcare plan of 2021, implemented changes to the existing ATU (remunerated early access program) system. The ATU program has now been grouped into two subgroups: An early access authorization program (Accès précoce aux médicaments, EAP) which includes the cohort ATU and a compassionate access program (Accès compassionnel aux médicaments, CAP) which includes nominative ATU, among others. The reforms will come into force no later than July 1, 2021 and ATUs with an expiry date after this date will not be eligible for renewal and will have to reapply.

Statement by the Board of Directors and the Executive Management

The Board of Directors and the Executive Management have today reviewed and approved the interim financial report of Orphazyme A/S for the period January 1 - June 30, 2021. The interim financial report has not been reviewed or audited by the Company's independent auditors.

The interim financial report for the period January 1 - June 30, 2021 has been prepared in accordance with IAS 34 *Interim Financial Reporting*. The accounting policies used in the interim financial report are consistent with those accounting policies used in Orphazyme's 2020 Annual Report with the additions described and additional Danish disclosure requirements for interim reports of listed companies.

In our opinion, the interim condensed consolidated financial statements give a true and fair view of Orphazyme's assets, liabilities, and financial position at June 30, 2021 and of the results of its operations and cash flows for the period January 1 - June 30, 2021. Furthermore, in our opinion, Management's Review gives a true and fair account of the development and performance of the Group's activities.

Copenhagen, August 31, 2021.

Board of Directors

Georges Gemayel
Chairman of the Board

Bo Jesper Hansen
Deputy Chairman of the Board

Carrolee Barlow

Catherine Moukheibir

Martin Bonde

Stephanie Smith Okey

Executive Management

Christophe Bourdon
Chief Executive Officer

Anders Vadsholt
Chief Financial Officer

Disclaimer

This company announcement may contain certain forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995, including in respect of the timing of the company's clinical trials and the results thereof, anticipated regulatory developments and approvals for the company's product candidates, the company's anticipated operating performance, financial position and ability to operate as a going concern. Although the Company believes its expectations are based on reasonable assumptions, all statements other than statements of historical fact included in this company announcement about future events are subject to (i) change without notice and (ii) factors beyond the Company's control, including adverse developments in the company's clinical program, actions by regulatory agencies, effects of the global COVID-19 pandemic, technical and scientific developments in the indications that the company's product candidates are designed to treat, regulatory developments and the impact of the Company's restructuring. These statements may include, without limitation, any statements preceded by, followed by, or including words such as "target," "believe," "expect," "aim," "intend," "may," "anticipate," "estimate," "plan," "project," "will," "can have," "likely," "should," "would," "could", and other words and terms of similar meaning or the negative thereof. Forward-looking statements are subject to inherent risks and uncertainties beyond the Company's control that could cause the Company's actual results, performance, or achievements to be materially different from the expected results, performance, or achievements expressed or implied by such forward-looking statements. In light of these risks and uncertainties, and other risks and uncertainties that are described in the Risk Factors section and other sections of the Company's Annual Report on Form 20-F, filed with the Securities and Exchange Commission (SEC) on March 2, 2021, and other filings that the Company makes with the SEC from time to time (which are available at http://www.sec.gov), the events and circumstances discussed in such forward-looking statements may not occur, and the Company's actual results could differ materially and adversely from those anticipated or implied thereby. Although the Company's forward-looking statements reflect the good faith judgment of its management, these statements are based only on facts and factors currently known by the Company. As a result, you are cautioned not to rely on these forward-looking statements. Except as required by law, the Company assumes no obligation to update these forward-looking statements publicly, or to update the reasons actual results could differ materially from those anticipated in the forward-looking statements, even if new information becomes available in the future.