

Corporate governance statement Orphazyme A/S, CVR no. 32 26 63 55

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1 Orphazyme A/S – Statement on Corporate Governance

This corporate governance statement for Orphazyme A/S, company registration no. 32 26 63 55 ("Orphazyme" or the "Company") has been prepared in connection with the initial public offering of the shares in the Company (the "IPO") and will be effective as from the date of the first day of trading on Nasdaq Copenhagen A/S ("Nasdaq Copenhagen") (the "Admission" or "IPO").

The statement includes a description of the Company's management structure and a review of how the Company considers the Danish Recommendations on Corporate Governance issued by the Committee on Corporate Governance in May 2013 (as amended in November 2014).

2 Board of Directors

Orphazyme has a two-tier management structure consisting of the Board of Directors and the Executive Management. The Board of Directors supervises the work of the Executive Management and is responsible for the overall management and strategic direction whereas the Executive Management handles the day-to-day management of the Company. No person serves as a member of both corporate bodies.

With effect from the Admission, the Board of Directors will consist of eight members elected by the general meeting and the Board of Directors will have elected a Chairman among its members. Under the current Recommendations on Corporate Governance, all of the board members, including the Chairman, are currently considered independent. The members of the Board of Directors comprise a group of professionally skilled business people also representing diversity (in a broader sense) and international experience.

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.

The Board of Directors normally holds at least five regular meetings annually, including a strategy review, plus ad hoc meetings as required.

3 Board Committees

With effect from Admission, the Board of Directors will have established an Audit Committee, a Remuneration Committee and a Nomination Committee, each of which has a charter outlining the purpose and responsibilities of such committee.

3.1 Audit Committee

The Audit Committee consists of three members of the Board of Directors. The overall purpose of the Audit Committee is to review accounting and audit matters and assesses the internal control and risk management system of the Company. The Audit Committee's duties also include supervision of the Company's external auditors and review of the audit process.

3.2 Remuneration Committee

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The Remuneration Committee consists of three members of the Board of Directors. The overall purpose of the Remuneration Committee is to ensure that the Company maintains a remuneration policy for the members of the Board of Directors and the Executive Management, including incentive guidelines, and to evaluate and make recommendations for the remuneration of the members of the Board of Directors and the Executive Management.

3.3 Nomination Committee

The Nomination Committee consists of three members of the Board of Directors. The overall purpose of the Nomination Committee is to 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.

4 Executive Management

The Board of Directors has appointed an Executive Management currently consisting of two members, who are responsible for the day-to-day management and compliance with the procedures, instructions, guidelines and recommendations issued by the Board of Directors. The Executive Management's responsibilities include inter alia organisation of the Company as well as allocation of resources, determination and implementation of strategies and policies and ensuring timely reporting to the Board of Directors. The Executive Management also presents and recommends proposals on the overall strategy and objectives to the Board of Directors.

5 Recommendations for good corporate governance

Orphazyme is subject to the Recommendations on Corporate Governance from May 2013 (as amended in November 2014), which are available on the Committee on Corporate Governance's website www.corporategovernance.dk. As a company listed on Nasdaq Copenhagen, Orphazyme will be required to report on its compliance with these recommendations according to the "comply or explain" principle. Orphazyme's position on each recommendation is described in the following.

Orphazyme intends to comply with the Recommendations on Corporate Governance in all material respects, except that Orphazyme has opted to deviate in the following area:

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 Recommendations on Corporate Governance 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.
- Orphazyme's current long-term incentive programme is 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 board members should have a maturity of one year from the date of allocation.
- Under the Orphazyme's current long-term incentive programme, 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.

Adopted by the Board of Directors of Orphazyme A/S on 2 November 2017.

Page 5 Corporate Governance Overview

Recommendation	Complies	Partially complies	Deviates	Comments where Orphazyme deviates
1. Communication and interaction by the company with its investors and other stakeholders				
1.1. Dialogue between company, shareholders and other stakeholders				
1.1.1. The Committee recommends that the board of directors ensure ongoing dialogue between the company and its shareholders in order for the shareholders to gain relevant insight into the company's potential and policies, and in order for the board of directors to be aware of the shareholders' views, interests and opinions on the company.	V			
1.1.2. The Committee recommends that the board of directors adopt policies on the company's relationship with its stakeholders, including shareholders and other investors, and that the board ensures that the interests of the shareholders are respected in accordance with company policies.	V			
1.1.3. The Committee recommends that the company publish quarterly reports.			÷	For the time being, the Company has decided only to publish half-yearly financial reports, as the Company is stil in a developmental phase unti 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.

Recommendation	Complies	Partially complies	Deviates	Comments where Orphazyme deviates
1.2. General meeting				
1.2.1. The Committee recommends that, when organising the company's general meeting, the board of directors plans the meeting to support active ownership.	V			
1.2.2. The Committee recommends that proxies granted for the general meeting allow shareholders to consider each individual item on the agenda.	V			
1.3. Takeover bids				
1.3.1. The Committee recommends that the company set up contingency procedures in the event of takeover bids from the time that the board of directors has reason to believe that a takeover bid will be made. According to such contingency procedures, the board of directors should not without the acceptance of the general meeting, attempt to counter the takeover bid by making decisions which in reality prevent the shareholders from deciding on the takeover bid themselves.	√			
2. Tasks and responsibilities of the board of directors				
2.1. Overall tasks and responsibilities				
2.1.1. The Committee recommends that at least once a year the board of directors take a position on the matters related to the board's performance of its responsibilities.	V			
2.1.2. The Committee recommends that at least once a year the board of directors take a position on the overall strategy of the company with a view to ensuring value creation in the company.	V			
2.1.3. The Committee recommends that the board of directors ensure that the company has a capital and share structure ensuring that the strategy and long-term value creation of the company are in the best interest of the shareholders and the company, and that the board of directors presents this in the management commentary on the company's annual report and/or on the company's website.	√			
2.1.4. The Committee recommends that the board of directors annually review and approve guidelines for the executive board; this includes establishing requirements	V			

Recommendation	Complies	Partially complies	Deviates	Comments where Orphazyme deviates
for the executive board on timely, accurate and adequate reporting to the board of directors.				
2.1.5. The Committee recommends that at least once a year the board of directors discuss the composition of the executive board, as well as developments, risks and succession plans.	√			
2.1.6. The Committee recommends that once a year the board of directors discuss the company's activities to ensure relevant diversity at management levels, including setting specific goals and accounting for its objectives and progress made in achieving the objectives in the management commentary on the company's annual report and/or on the website of the company.	V			
2.2. Corporate social responsibility				
2.2.1. The Committee recommends that the board of directors adopt policies on corporate social responsibility.	√			
2.3. Chairman and vice-chairman of the board of directors				
2.3.1. The Committee recommends appointing a vice-chairman of the board of directors who will assume the responsibilities of the chairman in the event of the chairman's absence, and who will also act as effective sparring partner for the chairman.	V			
2.3.2. The Committee recommends ensuring that, if the board of directors, in exceptional cases, asks the chairman of the board of directors to perform special operating activities for the company, including briefly participating in the day-to-day management, a board resolution to that effect be passed to ensure that the board of directors maintains its independent, overall management and control function. Resolutions on the chairman's participation in day-to-day management and the expected duration hereof should be published in a company announcement.	√			
3. Composition and organisation of the board of directors				
3.1. Composition				

Recommendation	Complies	Partially complies	Deviates	Comments where Orphazyme deviates
 3.1.1. The Committee recommends that the board of directors annually accounts for: the skills it must have to best perform its tasks, the composition of the board of directors, and the special skills of each member. 	V			
3.1.2. The Committee recommends that the selection and nomination of candidates for the board of directors be carried out through a thoroughly transparent process approved by the overall board of directors. When assessing its composition and nominating new candidates, the board of directors must take into consideration the need for integration of new talent and diversity in relation to age, international experience and gender.	√			
 3.1.3. The Committee recommends that a description of the nominated candidates' qualifications, including information about the candidates' other executive functions, e.g. memberships in executive boards, boards of directors, and supervisory boards, including board committees in foreign enterprises, be accompanied by the notice convening the general meeting when election of members to the board of directors is on the agenda. demanding organisational tasks, and information about whether candidates to the board of directors are considered independent. 	√			
3.1.4. The Committee recommends that the company's articles of association stipulate a retirement age for members of the board of directors.			÷	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

Recommendation	Complies	Partially complies	Deviates	Comments where Orphazyme deviates
				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 Recommendations on Corporate Governance expected to enter into force in January 2018.
3.1.5. The Committee recommends that members of the board of directors elected by the general meeting be up for election every year at the annual general meeting.	V			
3.2 Independence of the board of directors				
3.2.1. The Committee recommends that at least half of the members of the board of directors elected by the general meeting be independent persons, in order for the board of directors to be able to act independently of special interests. To be considered independent, this person may not:				
 be or within the past five years have been member of the executive board, or senior staff member in the company, a subsidiary undertaking or an associate, 	√			
 within the past five years, have received larger emoluments from the company/group, a subsidiary undertaking or an associate in another capacity than as member of the board of directors, 				

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 represent the interests of a controlling shareholder, 				
 within the past year, have had significant business relations (e.g. personal or indirectly as partner or employee, shareholder, customer, supplier or member of the executive management in companies with corresponding connection) with the company, a subsidiary undertaking or an associate, 				
 be or within the past three years have been employed or partner at the external auditor, 				
 have been chief executive in a company holding cross-memberships with the company, 				
 have been member of the board of directors for more than 12 years, or 				
 have been close relatives with persons who are not considered independent. 				
3.3. Members of the board of directors and the number of other executive functions				
3.3.1. The Committee recommends that each member of the board of directors assesses the expected time commitment for each function in order that the member does not take on more functions than he/she can manage satisfactorily for the company.	V			
3.3.2. The Committee recommends that the management commentary, in addition to the provisions laid down by legislation, includes the following information about the members of the board of directors:				
 the position of the relevant person, 	,			
 the age and gender of the relevant person, 	V			
 whether the member is considered independent, 				
 the date of appointment to the board of directors of the member, 				
 expiry of the current election period, 				

Recommendation	Complies	Partially complies	Deviates	Comments where Orphazyme deviates
 other executive functions, e.g. memberships in executive boards, boards of directors, and supervisory boards, including board committees in foreign enterprises, 				
 demanding organisational tasks, and 				
 the number of shares, options, warrants and similar in the company as well as changes in the portfolio of the member of the securities mentioned which have occurred during the financial year. 				
3.4. Board committees				
3.4.1. The Committee recommends that the company publish the following on the company's website:				
 the terms of reference of the board committees, 				
 the most important activities of the committees during the year, and the number of meetings held by each committee, and 	√			
 the names of the members of each committee, including the chairmen of the committees, as well as information on which members are independent members and which members have special qualifications. 				
3.4.2. The Committee recommends that a majority of the members of a board committee be independent.	√			
3.4.3. The Committee recommends that the board of directors set up a formal audit committee composed such that				
 the chairman of the board of directors is not chairman of the audit committee, and between them, the members should possess such expertise and experience as to provide an updated insight into and experience in the financial, accounting and audit agreets of companies whose shows are admitted to trading on a regulated. 	√			
audit aspects of companies whose shares are admitted to trading on a regulated market.				

Recommendation	Complies	Partially complies	Deviates	Comments where Orphazyme deviates
 3.4.4. The Committee recommends that, prior to the approval of the annual report and other financial reports, the audit committee monitors and reports to the board of directors about: significant accounting policies, significant accounting estimates, related party transactions, and uncertainties and risks, including in relation to the outlook for the current year. 	√			
 3.4.5. The Committee recommends that the audit committee: annually assesses the need for an internal audit, and in such case, makes recommendations on selecting, appointing and removing the head of the internal audit function and on the budget of the internal audit function, and monitor the executive board's follow-up on the conclusions and recommendations of the internal audit function. 	√			
 3.4.6. The Committee recommends that the board of directors establish a nomination committee chaired by the chairman of the board of directors with at least the following preparatory tasks: describe the qualifications required by the board of directors and the executive board, and for a specific membership, state the time expected to be spent on having to carry out the membership, as well as assess the competences, knowledge and experience of the two governing bodies combined, annually assess the structure, size, composition and results of the board of directors and the executive board, as well as recommend any changes to the board of directors, annually assess the competences, knowledge and experience of the individual members of management, and report to the board of directors in this respect, 	√			

Recommendation	Complies	Partially complies	Deviates	Comments where Orphazyme deviates
 consider proposals from relevant persons, including shareholders and members of the board of directors and the executive board for candidates for the board of directors and the executive board, and propose an action plan to the board of directors on the future composition of 				
the board of directors, including proposals for specific changes.				
3.4.7. The Committee recommends that the board of directors establish a remuneration committee with at least the following preparatory tasks:				
 to recommend the remuneration policy (including the general guidelines for incentive-based remuneration) to the board of directors and the executive board for approval by the board of directors prior to approval by the general meeting, 				
 make proposals to the board of directors on remuneration for members of the board of directors and the executive board, as well as ensure that the remuneration is in compliance with the company's remuneration policy and the assessment of the performance of the persons concerned. The committee should have information about the total amount of remuneration that members of the board of directors and the executive board receive from other companies in the group, and 	V			
 recommend a remuneration policy applicable for the company in general. 				
3.4.8. The Committee recommends that the remuneration committee do not consult with the same external advisers as the executive board of the company.	√			
3.5. Evaluation of the performance of the board of directors and the executive board				
3.5.1. The Committee recommends that the board of directors establish an evaluation procedure where contributions and results of the board of directors and the individual members, as well as collaboration with the executive board are annually evaluated. Significant changes deriving from the evaluation should be included in the management commentary or on the company's website.	V			

Recommendation	Complies	Partially complies	Deviates	Comments where Orphazyme deviates
3.5.2. The Committee recommends that in connection with preparation of the general meeting, the board of directors consider whether the number of members is appropriate in relation to the requirements of the company. This should help ensure a constructive debate and an effective decision-making process in which all members are given the opportunity to participate actively.	V			
3.5.3. The Committee recommends that at least once every year the board of directors evaluate the work and performance of the executive board in accordance with predefined clear criteria.	V			
3.5.4. The Committee recommends that the executive board and the board of directors establish a procedure according to which their cooperation is evaluated annually through a formalised dialogue between the chairman of the board of directors and the chief executive officer and that the outcome of the evaluation be presented to the board of directors.	V			
4. Remuneration of management				
4.1. Form and content of the remuneration policy				
 4.1.1. The Committee recommends that the board of directors prepare a clear and transparent remuneration policy for the board of directors and the executive board, including a detailed description of the components of the remuneration for members of the board of directors and the executive board, the reasons for choosing the individual components of the remuneration, and 	√			
 a description of the criteria on which the balance between the individual components of the remuneration is based. 				
The remuneration policy should be approved by the general meeting and published on the company's website.				
4.1.2. The Committee recommends that, if the remuneration policy includes variable components,	V			

Recommendation	Complies	Partially complies	Deviates	Comments where Orphazyme deviates
 limits be set on the variable components of the total remuneration package, a reasonable and balanced linkage be ensured between remuneration for governing body members, expected risks and the value creation for shareholders in the short and long terms, there be clarity about performance criteria and measurability for award of variable components, there be criteria ensuring that qualifying periods for variable components in remuneration agreements are longer than one calendar year, and an agreement is made which, in exceptional cases, entitles the company to reclaim in full or in part variable components of remuneration that were paid on the basis of data, which proved to be misstated. 				
4.1.3. The Committee recommends that remuneration of members of the board of directors does not include share options.			÷	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.

Recommendation	Complies	Partially complies	Deviates	Comments where Orphazyme deviates
4.1.4. The Committee recommends that if share-based remuneration is provided, such programmes be established as roll-over programmes, i.e. the options are granted periodically and should have a maturity of at least three years from the date of allocation.			÷	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 board members should have a maturity of one year from the date of allocation. Under the Orphazyme's current long-term incentive programme, 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.

Recommendation	Complies	Partially complies	Deviates	Comments where Orphazyme deviates
4.1.5. The Committee recommends that agreements on termination payments should not amount to more than two years' annual remuneration.	V			
4.2. Disclosure of the remuneration policy				
4.2.1. The Committee recommends that the company's remuneration policy and compliance with this policy be explained and justified annually in the chairman's statement at the company's general meeting.	V			
4.2.2. The Committee recommends that the proposed remuneration for the board of directors for the current financial year be approved by the shareholders at the general meeting.	V			
4.2.3. The Committee recommends that the total remuneration granted to each member of the board of directors and the executive board by the company and other companies in the group, including information on the most important contents of retention and retirement/resignation schemes, be disclosed in the annual report and that the linkage with the remuneration policy be explained.	V			
5. Financial reporting, risk management and audits				
5.1. Identification of risks and transparency about other relevant information				
5.1.1. The Committee recommends that the board of directors in the management commentary review and account for the most important strategic and business-related risks, risks in connection with the financial reporting as well as for the company's risk management.	V			
5.2. Whistle-blower scheme				
5.2.1. The Committee recommends that the board of directors decide whether to establish a whistle-blower scheme for expedient and confidential notification of possible or suspected wrongdoing.	V			Given the current size of the Company and the nature of its present operations, the Board of Directors has decided not to establish a formal whistle-

Recommendation	Complies	Partially complies	Deviates	Comments where Orphazyme deviates
				blower scheme, but generally encourage all employees of the Company approach the Executive Management or Board of Directors with any matters that they wish to discuss.
5.3. Contact to auditor				
5.3.1. The Committee recommends that the board of directors ensure regular dialogue and exchange of information between the auditor and the board of directors, including that the board of directors and the audit committee at least once a year meet with the auditor without the executive board present. This also applies to the internal auditor, if any.	V			
5.3.2. The Committee recommends that the audit agreement and auditors' fee be agreed between the board of directors and the auditor on the basis of a recommendation from the audit committee.	V			