

Code of business conduct and ethics

Introduction

Purpose and Scope

The Board of Directors of Orphazyme A/S (together with its subsidiaries as existing from time-to-time, “**Orphazyme**”) have established this Code of Business Conduct and Ethics (the “**Code**”) to aid Orphazyme’s directors, officers and employees in making ethical and legal decisions when conducting Orphazyme’s business and performing their day-to-day duties.

Orphazyme’s Board of Directors, or any committee of the Board delegated by the Board, shall be responsible for administering this Code. The Board of Directors has delegated day-to-day responsibility for administering and interpreting the Code to a Compliance Officer. Orphazyme’s Chief Financial Officer has been appointed as our Compliance Officer under this Code.

Orphazyme expects its directors, officers and employees to exercise reasonable judgment when conducting its business. Orphazyme encourages its directors, officers and employees to refer to this Code frequently to ensure that they are acting within both the letter and the spirit of this Code, and to promptly report any suspected violations hereof to the Compliance Officer. Orphazyme also understands that this Code will not contain the answer to every situation you may encounter or every concern you may have about conducting Orphazyme’s business ethically and legally. In these situations, or if you otherwise have questions or concerns about this Code, we encourage each officer and employee to speak with his or her supervisor (if applicable) or, if you are uncomfortable doing that, with the Compliance Officer under this Code.

Contents of this Code

This Code has two sections which follow this Introduction. The first section, “*Standards of Conduct,*” contains the actual guidelines that Orphazyme’s directors, officers and employees are expected to adhere to in the conduct of Orphazyme’s business. The second section, “*Compliance Procedures,*” contains specific information about how this Code functions, including who administers the Code, who can provide guidance under the Code and how violations may be reported, investigated and punished. This section also contains a discussion about waivers of and amendments to this Code.

A Note About Other Obligations

Orphazyme’s directors, officers and employees generally have other legal and contractual obligations to Orphazyme. This Code is not intended to reduce or limit the other obligations that you may have to Orphazyme. Instead, the standards in this Code should be viewed as the

minimum standards that Orphazyme expects from its directors, officers and employees in the conduct of its business.

1. Standards of conduct

Conflicts of Interest

Orphazyme recognizes and respects the right of its directors, officers and employees to engage in outside activities, unless otherwise specifically agreed, which they may deem proper and desirable, provided that these activities do not impair or interfere with the performance of their duties to Orphazyme or their ability to act in Orphazyme's best interests. In most, if not all, cases this will mean that our directors, officers and employees must avoid situations that present a potential or actual conflict between their personal interests and Orphazyme's interests.

A "conflict of interest" occurs when a director's, officer's or employee's personal interest, including financial interest interferes with Orphazyme's interests. Conflicts of interest may arise in many situations. For example, conflicts of interest can arise when a director, officer or employee takes an action or has an outside interest, responsibility or obligation that may make it difficult for him or her to perform the responsibilities of his or her position objectively and/or effectively in Orphazyme's best interests. Conflicts of interest may also occur when a director, officer or employee or his or her immediate family member receives a personal benefit (whether improper or not) as a result of the director's, officer's or employee's position with Orphazyme. Each individual's situation is different and in evaluating his or her own situation, a director, officer or employee will have to consider many factors.

Further, no director or officer may participate in the transaction of business involving any agreement between Orphazyme and the director or officer or any legal proceedings against the director or officer or any agreement between Orphazyme and a third party or any legal proceedings against a third party if the director or officer has a material interest in such business that may conflict with the interests of Orphazyme.

Any material transaction or relationship that reasonably could be expected to give rise to a conflict of interest should be reported promptly to the Compliance Officer. The Compliance Officer may notify the Board of Directors or a committee thereof as he or she deems appropriate.

Compliance with Laws, Rules and Regulations

Orphazyme seeks to conduct its business in compliance with applicable laws, rules and regulations. No director, officer or employee shall engage in any unlawful activity in conducting Orphazyme's business or in performing his or her day-to-day company duties, nor shall any director, officer or employee instruct others to do so.

Anti-Bribery and Anti-Corruption

Orphazyme has a zero tolerance policy against bribery and corruption and condemns it in all forms. Orphazyme recognises that corruption and other unethical practices are undermining to its business and therefore takes active measures to ensure that bribery and corruption does not occur in its business activities.

Orphazyme seeks to conduct its business in compliance with the U.S. Foreign Corrupt Practices Act of 1977 (the “**FCPA**”), Sections 122 and 299(2) of the Danish Criminal Code consolidated by Act no. 976 of 17 September 2019 (in Danish: *Straffeloven*) and other applicable anti-bribery and anti-corruption laws, rules and regulations, including the laws of such countries in which Orphazyme A/S has subsidiaries. To ensure compliance with such anti-corruption and anti-bribery laws, Orphazyme has adopted an Anti-Corruption Policy containing guidelines, standards and procedures applicable to directors, officers, and employees as well as business partners. Please direct any questions or concerns related hereto to Orphazyme’s Compliance Officer.

Protection and Proper Use of Orphazyme’s Assets

Loss, theft and misuse of Orphazyme’s assets has a direct impact on Orphazyme’s business and its profitability. Employees, officers and directors are expected to protect Orphazyme’s assets that are entrusted to them and to protect Orphazyme’s assets in general. Employees, officers and directors are also expected to take steps to ensure that Orphazyme’s assets are only used for legitimate business purposes.

Corporate Opportunities

Employees, officers and directors owe a duty to Orphazyme to act loyally towards Orphazyme and to advance its legitimate business interests when the opportunity to do so arises. Each employee, officer and director is prohibited from:

- diverting to himself or herself or to others any opportunities that are discovered through the use of Orphazyme’s property or information as a result of his or her position with Orphazyme unless such opportunity has first been presented to, and rejected by, Orphazyme;
- using Orphazyme’s property or information or his or her position for improper personal gain; or
- competing with Orphazyme.

Confidentiality

Confidential information generated and gathered in Orphazyme's business plays a vital role in Orphazyme's business, prospects and ability to compete. "Confidential information" includes all non-public information that might be of use to competitors or harmful to Orphazyme, its partners or collaborators or its customers if disclosed. Directors, officers and employees may not disclose or distribute Orphazyme's confidential information, except when disclosure is authorized by Orphazyme or required by applicable law, rule or regulation or pursuant to an applicable legal proceeding. Directors, officers and employees shall use confidential information solely for legitimate business purposes. Directors, officers and employees must return or provide evidence of destruction of all of Orphazyme's confidential and/or proprietary information in their possession to Orphazyme when they cease to be employed by or to otherwise serve Orphazyme.

Fair Dealing

Competing vigorously, yet lawfully, with competitors and establishing advantageous, but fair, business relationships with business partners is a part of the foundation for long-term success. However, unlawful and unethical conduct, which may lead to short-term gain, may damage a company's reputation and long-term business prospects. Accordingly, it is Orphazyme's policy that directors, officers and employees must endeavor to deal ethically and lawfully with Orphazyme's suppliers and other business partners, competitors and employees in all business dealings on Orphazyme's behalf. No director, officer or employee should take unfair advantage of another person in business dealings on Orphazyme's behalf through the abuse of privileged or confidential information or through improper manipulation, concealment or misrepresentation of material facts.

Accuracy of Records

The integrity, reliability and accuracy in all material respects of Orphazyme's books, records and financial statements is fundamental to Orphazyme's continued and future commercial success. No director, officer or employee may cause us to enter into a transaction with the intent to document or record it in a deceptive or unlawful manner. In addition, no director, officer or employee may create any false or artificial documentation or book entry for any transaction entered into by Orphazyme. Similarly, officers and employees who have responsibility for accounting and financial reporting matters have a responsibility to accurately record all funds, assets and transactions on our books and records.

Quality of Public Disclosures

Orphazyme is committed to providing its shareholders with information about its financial condition and results of operations as required by the securities laws of Denmark, the United States and all other relevant jurisdictions. It is Orphazyme's policy that the reports and documents it files with or submits to the U.S. Securities and Exchange Commission (the "SEC"),

Nasdaq Copenhagen and (if relevant) the Danish Financial Supervisory Authority, and the earnings releases and similar public communications made by Orphazyme, include fair, timely, understandable, full and accurate disclosure. Officers and employees who are responsible for these filings and disclosures, including Orphazyme’s principal executive, financial and accounting officers, must use reasonable judgment and perform their responsibilities honestly, ethically and objectively in order to ensure that this disclosure policy is fulfilled. Orphazyme’s senior management is primarily responsible for monitoring Orphazyme’s public disclosure.

Code of Ethics for the CEO and Senior Financial Officers

The CEO and all senior financial officers, including the CFO, the Director of Finance and persons performing similar functions– (the “**Senior Financial Officers**” and together with the CEO, the “**Named Officers**”) shall:

- engage in and promote honest and ethical conduct, including the ethical handling of actual or apparent conflicts of interest between personal and professional relationships;
- avoid conflicts of interest and disclose to the chairman of the Audit Committee any material transaction or relationship that reasonably could be expected to give rise to such a conflict;
- take all reasonable measures to protect the confidentiality of non-public information about Orphazyme and its customers obtained or created in connection with their activities and prevent the unauthorized disclosure of such information unless required by applicable law or regulation or legal or regulatory process;
- take all reasonable measures to achieve responsible use of and control over Orphazyme’s assets and resources;
- promote full, fair, accurate, timely, and understandable disclosure in material respects in reports and documents that Orphazyme files with, or submits to, the SEC, Nasdaq Copenhagen, the Danish Financial Supervisory Authority and other regulators and in other public communications made by Orphazyme in accordance with the following guidelines:
 - all accounting records, and the reports produced from such records, must be in accordance with all applicable laws;
 - all accounting records must fairly and accurately reflect the transactions or occurrences to which they relate;
 - all accounting records must fairly and accurately reflect in reasonable detail in accordance with generally accepted accounting principles Orphazyme’s assets, liabilities, revenues and expenses;
 - all accounting records must not contain any materially false or intentionally misleading entries;
 - no transactions should be intentionally misclassified as to accounts, departments or accounting periods;
 - all transactions must be supported by accurate documentation in reasonable detail and in all material respects to be recorded in the proper account and in the proper accounting period;

- no information should be concealed from the auditors; and compliance with Orphazyme’s system of internal controls is required;
- comply with all governmental laws, rules and regulations applicable to Orphazyme’s business, including taking necessary steps to avoid and, where possible, prevent any violations of the securities laws; and
- promptly report to the Chairman of the Audit Committee (or, if the Chairman is unavailable, to all other members of the Audit Committee) any fraud, whether or not material, involving management or other employees of Orphazyme who have a significant role in Orphazyme’s disclosures or internal controls over financial reporting; and promptly report any possible violation of this Section to the Chairman of the Audit Committee.

2. Compliance procedures

Communication of Code

All directors, officers and employees will be supplied with a copy of the Code upon beginning service at Orphazyme and will, on a periodic basis, be asked to review and sign an acknowledgment that he or she has: (i) received a copy of the Code; (ii) read and understood the Code; (iii) not violated the Code; and (iv) agreed to comply with the Code. Updates of the Code will be provided from time-to-time. A copy of the Code is also available to all directors, officers and employees by requesting one from the Compliance Officer.

Monitoring Compliance and Disciplinary Action

Orphazyme’s management, under the supervision of its Board of Directors or a committee thereof or, in the case of accounting, internal accounting controls, auditing or securities law matters, the Audit Committee, shall take reasonable steps from time to time to (i) monitor compliance with the Code, (ii) when appropriate, impose and enforce appropriate disciplinary measures for violations of the Code, and (iii) when appropriate, report violators to the appropriate authorities.

Any failure to comply with this Code may result in Orphazyme taking disciplinary actions against those involved in such non-compliance.

Orphazyme’s management shall periodically report to the Board of Directors or a committee thereof on these compliance efforts including, without limitation, periodic reporting of alleged violations of the Code and the actions taken with respect to any such violation.

Reporting Concerns/Receiving Advice - Communication Channels

Be Proactive

Every employee is expected to act proactively by asking questions, seeking guidance and reporting suspected violations of the Code and other policies and procedures of Orphazyme, as well as any violation or suspected violation of applicable law, rule or regulation arising in the conduct of Orphazyme's business or occurring on its property. **If any employee believes that actions have taken place, may be taking place, or may be about to take place that violate or would violate the Code or any law, rule or regulation applicable to Orphazyme, he or she must bring the matter to the attention of Orphazyme.**

Seeking Guidance

The best starting point for an officer or employee seeking advice on ethics-related issues or reporting potential violations of the Code will usually be his or her supervisor. However, if the conduct in question involves his or her supervisor, if the employee has reported the conduct in question to his or her supervisor and does not believe that he or she has dealt with it properly, or if the officer or employee does not feel that he or she can discuss the matter with his or her supervisor, the employee may raise the matter with the Compliance Officer.

Communication Alternatives

Any officer or employee may communicate with the Compliance Officer by any of the following methods:

- in writing (which may be done anonymously as set forth below under "Reporting; Anonymity; Retaliation"), addressed to the Compliance Officer by mail to Orphazyme A/S, Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark; or
- via the independent external whistleblower portal (Link to whistleblower portal: <https://orphazyme.whistleblownetwork.net>).

Reporting by an officer or employee through the above methods may be done anonymously, and the reporting should preferably be made in the company languages (English and Danish).

Reporting Accounting, Securities Law and Similar Concerns

Any concerns or questions regarding potential violations of the Code, any other company policy or procedure or applicable law, rules or regulations involving accounting, internal accounting controls, auditing or securities law (including the Foreign Corrupt Practices Act) matters should be directed to (i) the Audit Committee or a designee of the Audit Committee. Officers and employees may communicate with the Audit Committee or its designee: in writing to: Audit Committee, Orphazyme A/S, Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark, or (ii)

the independent external whistleblower portal (<https://orphazyme.whistleblownetwork.net>), in accordance with Orphazyme's Whistleblower Policy.

Reporting by an officer or employee through the above methods may be done anonymously, and the reporting should preferably be made in the company languages (English and Danish).

Cooperation

Within the parameters of applicable law, employees are expected to cooperate with Orphazyme in any investigation of a potential violation of the Code, any other company policy or procedure, or any applicable law, rule or regulation.

Misuse of Reporting Channels

Employees must not use these reporting channels in bad faith or in a false or unreasonable manner.

Reporting; Anonymity; Retaliation

When reporting suspected violations of the Code, Orphazyme prefers that officers and employees identify themselves in order to facilitate its ability to take appropriate steps to address the report, including conducting any appropriate investigation. However, we also recognize that some people may feel more comfortable reporting a suspected violation anonymously.

If an officer or employee wishes to remain anonymous, he or she may do so, and Orphazyme will use reasonable efforts to protect the confidentiality of the reporting person subject to applicable law, rule or regulation or to any applicable legal proceedings. In the event the report is made anonymously, however, Orphazyme may not have sufficient information to look into or otherwise investigate or evaluate the allegations. Accordingly, persons who make reports anonymously should provide as much detail as possible to permit Orphazyme to evaluate the matter(s) set forth in the anonymous report and, if appropriate, commence and conduct an appropriate investigation.

No Retaliation

Orphazyme expressly forbids any retaliation against any officer or employee who, acting in good faith on the basis of a reasonable belief, reports suspected misconduct. Specifically, Orphazyme will not discharge, demote, suspend, threaten, harass or in any other manner discriminate against, such an officer or employee in the terms and conditions of his or her employment. Any person who participates in any such retaliation is subject to disciplinary action, including termination.

Waivers and Amendments

No waiver of any provisions of the Code for the benefit of a director or an executive officer (which includes, without limitation, for purposes of this Code, Orphazyme's principal executive, financial and accounting officers) shall be effective unless (i) approved by the Board of Directors or, if permitted, a committee thereof, and (ii) if applicable, such waiver is promptly disclosed to Orphazyme's shareholders in accordance with applicable U.S. securities laws and/or the rules and regulations of the exchange or system on which Orphazyme's shares are traded or quoted, as the case may be.

Any waivers of the Code for other employees may be made by the Compliance Officer, the Board of Directors or, if permitted, a committee thereof.

All amendments to the Code must be approved by the Board of Directors or a committee thereof and, if applicable, must be promptly disclosed to Orphazyme's shareholders in accordance with applicable U.S. securities laws and/or the rules and regulations of the exchange or system on which Orphazyme's shares are traded or quoted, as the case may be.

Approved on August 20, 2020