

ULTRAGENYX PHARMACEUTICAL INC.

CORPORATE GOVERNANCE GUIDELINES

Approved: March 30, 2017

The Board of Directors (the “Board”) of Ultragenyx Pharmaceutical Inc. (the “Company”) has adopted the Corporate Governance Guidelines (the “Guidelines”) below to assist the Board in serving the best interests of the Company and its stockholders. The Guidelines are intended to be a framework for conducting the Board’s business, and are not a set of legally binding obligations. These Guidelines may be modified by the Board from time to time upon recommendation of the Company’s Nominating and Corporate Governance Committee.

Selection and Composition of Board

Size of the Board

The Company’s Certificate of Incorporation provides that the number of directors of the Company be fixed by the Board from time to time. The Company believes a board should be small enough to permit thorough discussion of issues but large enough to provide a mix of perspectives and properly staff all Board committees. The Nominating and Corporate Governance Committee will periodically review and recommend to the Board the appropriate size and mix of the Board in light of the Company’s need for particular expertise, skills, perspectives, and competencies.

Selection of New Directors

The Board has delegated to the Nominating and Corporate Governance Committee the task of identifying, reviewing, and recommending a slate of director nominees to be proposed by the Board to the stockholders, and recommending any director nominees to be elected by the Board to fill interim vacancies.

Board Membership Criteria

The Nominating and Corporate Governance Committee is responsible for reviewing with the entire Board from time to time the appropriate skills and characteristics required of directors in the context of the current make-up of the Board. It is the policy of the Board that directors should possess strong personal and professional ethics, integrity, and values; be experienced and genuinely interested in the Company; and be committed to representing the long-term interests of the stockholders. The Board also believes that its membership should reflect a diversity of talents, skills, backgrounds, including with respect to age, gender, national origin, sexual orientation and identification, race, ethnicity and culture, and expertise necessary to provide sound and prudent oversight with respect to the operations and interests of the business.

Director Independence

The Board believes that, as a matter of policy, a majority of the members of the Board should be independent as defined by the rules of The NASDAQ Stock Market (the “NASDAQ Listing Rules”). In addition, the members of the Audit Committee may not receive, directly or indirectly, any fees from the Company or any Company subsidiary other than those described below under “Board Compensation Policy” and may not be “affiliated persons” (as defined in Rule 10A-3 under the Securities Exchange Act of 1934, as amended) of the Company. At least annually, the Board will evaluate significant relationships between the Company and each director, and significant relationships between a member of senior management and a director, in light of relevant facts and circumstances, for the purposes of determining whether a material relationship exists that might signal a potential conflict of interest.

Non-Independent Directors

The Board recognizes that individuals who are not independent may make significant contributions as directors and is willing to entertain their nomination for election to the Board.

Lead Director

When the Chair of the Board is not an independent director, a Lead Director will be elected annually by the Board. The Lead Director shall be an independent director.

Separation of Board Chair and CEO Positions; Rotation of Chair

The Company currently separates the positions of CEO and Chair of the Board. The Board expects that the Chair of the Board shall rotate every five years, unless the Nominating and Corporate Governance Committee recommends otherwise.

Term of Director Service

The Nominating and Corporate Governance Committee reviews each director’s continuation on the Board at least once every three years to enable each director to have the opportunity to confirm his or her desire to continue as a director, and allow the Company to replace directors as needed.

Directors Who Change Their Present Job Responsibility

A director who retires or otherwise changes from the principal occupation or principal background association held when the director was originally invited to join the Board will offer to tender his or her resignation from the Board by submitting such offer in writing to the Chair of the Nominating and Corporate Governance Committee. Upon receipt of such notice, the Nominating and Corporate Governance Committee, together with the CEO, shall either confirm with the director that they do not believe that the change in the director’s status would inhibit the director’s ability to continue to serve the best interests of the Company or its stockholders, or accept the director’s offer to tender his or her resignation. In addition, when the Ultragenyx CEO no longer holds the CEO position, he or she must offer to tender his or her resignation from

the Board. Whether that individual continues to serve on the Board is a matter for discussion at that time between the Board and the new CEO.

Joining New Boards or Other Associations

The Company expects that each of its directors will be able to dedicate the time and resources sufficient to ensure the diligent performance of his or her duties on the Company's behalf, including attending board and applicable committee meetings. The Board does not believe, however, that explicit limits on the number of other boards of directors on which the directors may serve, or on other activities the directors may pursue, are appropriate. The Board does recognize that excessive time commitments can interfere with an individual's ability to perform his or her duties effectively; therefore, in connection with its assessment of director candidates for nomination, as well as authorizing a member of the Board to become a member of the board of directors of another public company (as discussed in further detail below), the Board will assess whether the performance of any director has been or is likely to be adversely impacted by excessive time commitments, including service on other boards of directors.

The Company also expects that each director will avoid circumstances that create an actual or perceived conflict of interest. Accordingly, a director shall notify the Chair of the Nominating and Corporate Governance Committee if he or she wishes to accept an invitation to:

- i. become a member of the board of directors of a public company; or
- ii. join a governmental commission, a private company board of directors, a company advisory board or similar body, or the governing board of a non-profit entity if the director reasonably believes, or the Board or CEO reasonably assumes, that the activities of such organization or company could be competitive with the Company, or otherwise impact the Company in a material manner.

Upon receipt of such notice, the Chair of the Nominating and Corporate Governance Committee, together with the CEO and Chair of the Board, shall confirm with the director (without the necessity of holding a formal meeting) that they do or do not believe joining such organization would represent a conflict of interest or otherwise inhibit the director's ability to serve the best interests of the Company and its stockholders. In the case of a disagreement, the Nominating and Corporate Governance Committee shall meet to make a final determination.

Board Compensation and Performance

Board Compensation Policy

It is the policy of the Board that a significant portion of director compensation will be in the form of stock or stock-based instruments in order to align interests of directors with those of stockholders. The Compensation Committee has the responsibility of recommending to the Board the compensation and benefits for non-employee directors. The Compensation Committee shall report from time to time to the entire Board on the status of director compensation. An executive officer of the Company serving as a member of the Board does not receive additional compensation for his or her service as a director.

Any proposed changes in director compensation come at the recommendation of the Compensation Committee, but with discussion and concurrence by the full Board, and where appropriate, approval of the stockholders.

Evaluation of Board Performance

The Nominating and Corporate Governance Committee recommends criteria for assessment of the performance of the Board as a whole, for each Board committee, and for individual directors. Based on these criteria, the Board, each committee, and each member of the Board shall conduct an annual self-assessment of performance in accordance with the process specified by the Nominating and Corporate Governance Committee.

Attendance at Annual Meeting of Stockholders

Each director who is up for election at an annual meeting of stockholders or who has a term that continues after such annual meeting is encouraged to attend the annual meeting of stockholders.

Interactions with Institutional Investors, Press, Customers, etc.

The Board believes that management speaks for the Company. The Chair of the Board may, from time to time, meet or otherwise communicate with various constituencies that are involved with the Company, but it is expected that the Chair of the Board would do this only with the knowledge of management and, in most instances, at the request of management.

Stockholders and other interested parties may communicate with directors by writing to them in care of the Secretary of the Company, who will receive the correspondence and respond on behalf of the Board following consultation with the named Board members.

Meetings of the Board

Participation in Board Meetings

The Company expects directors to rigorously prepare for, attend, and participate in all board and applicable committee meetings. Each director is expected to ensure that other commitments do not materially interfere with service as a director.

Meetings of the Independent Directors

It is the policy of the Board to have a separate meeting session for the independent directors generally during every regularly scheduled meeting of the full Board to review matters concerning the relationship of the Board with management and such other matters as it deems appropriate. Any independent director may request a meeting of the independent directors at any time. The Chair of the Board, or the Lead Director, if the Chair of the Board is not an independent director, presides at all meetings of independent directors at which he or she is present.

Scheduling and Selection of Agenda Items for Board Meetings

The Chair of the Board, in consultation with the CEO, determines the frequency and length of meetings of the Board. It is the expectation of the Board that regular, in-person meetings at appropriate intervals are desirable for the performance of their responsibilities, but meetings may also be conducted via teleconference. In addition to regularly scheduled meetings, additional unscheduled meetings may be called upon appropriate notice at any time to address any special needs.

The Chair of the Board, in consultation with the CEO, establishes the agenda for each meeting. Each director is free to suggest the inclusion of items on an agenda, to raise at any meeting subjects that are not on the agenda for that meeting, or to request the presence of or a report by any member of management. During at least one meeting each year, the long-term strategic plan for the Company and the principal issues that it expects to face in the future, as well as the Company's risk management and compliance program, are presented to, and discussed by, the Board.

Board Material and Presentations

Materials that are important to an understanding of the business and matters to be considered at a meeting are distributed in advance to directors. As a general rule, materials on specific subjects are sent to directors sufficiently in advance so directors will be prepared to discuss questions that they may have about the material.

The Board encourages the CEO to schedule members of management to present at meetings who can provide additional insight into the specific matters being discussed.

Access to Management and Advisors

Each director is encouraged to keep himself or herself informed of the affairs of the Company between board meetings through direct contact with members of senior management and outside advisors, and each director has access to any such member of senior management and outside advisor. It is expected that the CEO shall be informed of such contact, and each director will use judgment to assure that such access is not distracting to the business operation of the Company.

Committees of the Board

Number of Committees

Our Board establishes committees from time to time to facilitate and assist in the execution of its responsibilities. These committees generally address issues that, because of their complexity and technical nature, level of detail or time requirements or because of proper corporate governance principles are suitable for committee oversight.

We currently have four standing committees: (1) the Compensation Committee, (2) the Audit Committee, (3) the Nominating and Corporate Governance Committee and (4) the Research and Development Committee. The Company has made these Guidelines and the charter for each of its Compensation, Audit, Nominating and Corporate Governance and Research and Development

Committees publicly available on its web site for review by its stockholders. There will, from time to time, be occasions on which the Board may want to form a new committee or disband a current committee, depending upon the circumstances. Subject to the phase-in provisions of the NASDAQ Listing Rules, only independent directors may serve on the Compensation Committee, Audit Committee, and Nominating and Corporate Governance Committee.

Assignment and Term of Service of Committee Members

The Board is responsible for the appointment of committee members and committee chairs, taking into account the desires of individual members and the recommendations of the Nominating and Corporate Governance Committee. The Nominating and Corporate Governance Committee shall make such recommendations based on the competencies of each member, and the need to ensure that an appropriate succession plan is in place for each committee chair. In making such appointments, the Board considers it appropriate to rotate committee membership at least as often as every five years, unless the Nominating and Corporate Governance Committee recommends otherwise. The Board also feels that it is appropriate and valuable to rotate committee chairs every three to five years, unless the Nominating and Corporate Governance Committee recommends that the current committee chair should continue to serve as chair for an additional period.

Frequency and Length of Committee Meetings and Committee Agenda

The committee chair determines the frequency and length of committee meetings and develops the agenda for committee meetings. The meeting minutes of the committees will be shared with the full Board. Any director who is not a member of a particular committee may attend any committee meetings with the concurrence of the committee chair. In addition, at least one member of management acts as a liaison for each committee.

Leadership Development

Formal Evaluation of Chief Executive Officer

The independent directors evaluate the CEO at least annually, and the evaluation is led by and communicated to the CEO by the Chair of the Board, or by the Lead Director if the Chair of the Board is not an independent director. The evaluation is based on clearly articulated criteria established by the Board, including performance of the business, accomplishment of long-term strategic objectives, and development of senior management. The evaluation is to be used by the Compensation Committee when considering the compensation of the CEO.

Succession Planning and Management Development

The Board has the sole responsibility for the evaluation, hiring, or termination of the CEO, as well as the development of policies and principles for selection of a new CEO, including succession in the event of an emergency. The current CEO reviews senior management succession planning and management development with the Board and the Compensation Committee on an annual basis.