

KINIKSA PHARMACEUTICALS, LTD.

CORPORATE GOVERNANCE GUIDELINES

The Board of Directors (the “**Board**”) of Kiniksa Pharmaceuticals, Ltd. (the “**Company**”) has adopted the following Corporate Governance Guidelines (the “**Guidelines**”) to assist the Board in the exercise of its responsibilities and to serve the interests of the Company and its shareholders. These Guidelines should be interpreted in the context of all applicable laws and the Company’s Bye-laws and other corporate governance documents. These Guidelines acknowledge the leadership exercised by the Board’s standing committees and their chairs and are intended to serve as a flexible framework within which the Board may conduct its business and not as a set of legally binding obligations. The Guidelines are subject to modification from time to time by the Board as the Board may deem appropriate and in the best interests of the Company and its shareholders or as required by applicable laws and regulations.

I. THE BOARD

A. Size of the Board

The Company’s Bye-laws provides that the number of directors will be no less than five and no more than such maximum number determined from time to time by the Board. The Nominating and Corporate Governance Committee will periodically review the size of the Board, and may make recommendations to the Board regarding the size that is most effective in relation to future operations.

B. Independence of the Board

Except as otherwise permitted by the applicable rules of The Nasdaq Stock Market LLC (the “**Nasdaq Rules**”), the Board will be comprised of a majority of directors who qualify as independent directors (the “**Independent Directors**”) as required under Nasdaq Rules.

C. Separate Sessions of Independent Directors

The Independent Directors will meet in executive session without non-Independent Directors or management present on a regularly scheduled basis, but no less than twice per year. Each executive session of the Independent Directors will be presided over by the Chairman of the Board, if the Chairman of the Board qualifies as independent, or by the lead director, if the Chairman of the Board does not qualify as independent.

D. Lead Director

If the Chairman of the Board is a member of management or does not otherwise qualify as independent, the Independent Directors may elect a lead director. The lead director’s responsibilities include, but are not limited to: presiding over all meetings of the Board at which

the Chairman of the Board is not present, including any executive sessions of the Independent Directors; approving Board meeting schedules and agendas; and acting as the liaison between the Independent Directors and the Chief Executive Officer and Chairman of the Board. At such times as the Chairman of the Board is an Independent Director, the Chairman of the Board will serve as lead director. The Board may modify its leadership structure in the future as it deems appropriate.

E. Director Qualification Standards

In evaluating the suitability of individual candidates (both new candidates and current Board members), the Nominating and Corporate Governance Committee, in recommending candidates for election, and the Board, in approving (and, in the case of vacancies, appointing) such candidates, may take into account many factors, including: personal and professional integrity, ethics and values; experience in corporate management, such as serving as an officer or former officer of a publicly held company; strong finance experience; relevant social policy concerns; experience relevant to the Company's industry; experience as a board member or executive officer of another publicly held company; relevant academic expertise or other proficiency in an area of the Company's operations; diversity of expertise and experience in substantive matters pertaining to the Company's business relative to other board members; diversity of background and perspective, including, but not limited to, with respect to age, gender, race, place of residence and specialized experience; practical and mature business judgment, including, but not limited to, the ability to make independent analytical inquiries; and any other relevant qualifications, attributes or skills. The Board evaluates each individual in the context of the Board as a whole, with the objective of assembling a group that can best perpetuate the success of the business and represent stockholder interests through the exercise of sound judgment using its diversity of experience in these various areas. In determining whether to recommend a director for re-election, the Nominating and Corporate Governance Committee may also consider the director's past attendance at meetings and participation in and contributions to the activities of the Board.

F. Selection of New Directors

The Board is divided into three (3) classes. As a result, approximately one-third (1/3) of the Board will stand for election by the shareholders of the Company each year at the Company's annual meeting for three (3) year terms. Each year, at the annual meeting, the Board will recommend a slate of directors for election by the shareholders. In accordance with the Bye-laws of the Company, the Board will also be responsible for filling vacancies or newly-created directorships on the Board that may occur between annual meetings of shareholders. The Nominating and Corporate Governance Committee is primarily responsible for identifying, screening and recommending candidates to the entire Board for Board membership.

G. Director Orientation and Continuing Education

Management will provide an orientation process for new directors, including background material on the Company and its business. As appropriate, management will provide

opportunities for additional educational sessions for directors on matters relevant to the Company and its business.

H. No Specific Limitation on Other Board Service

The Board does not believe that its members should be prohibited from serving on boards of other organizations and has not adopted any guidelines limiting such activities. However, the Nominating and Corporate Governance Committee may take into account the nature of and time involved in a director's service on other boards and/or committees in evaluating the suitability of individual director candidates and current directors. Prior to accepting any new position on the board of directors of any organization, whether for-profit or not-for-profit, current directors should notify the Chairman of the Board and the Company's General Counsel.

Service on other boards and/or committees should be consistent with the Company's conflict of interest policies set forth below.

I. Directors Who Resign or Materially Change Their Current Positions With Their Own Company or Become Aware of Circumstances that May Adversely Reflect upon the Director or the Company

When a director, including any director who is currently an officer or employee of the Company, resigns or materially changes his or her position with his or her employer or becomes aware of circumstances that may adversely reflect upon the director or the Company, such director should notify the Nominating and Corporate Governance Committee of such circumstances. The Nominating and Corporate Governance Committee will consider the circumstances, and may in certain cases recommend that the Board request that the director submit his or her resignation from the Board if, for example, continuing service on the Board by the individual is not consistent with the criteria deemed necessary for continuing service on the Board.

J. Term Limits

As each director is periodically subject to election by shareholders, the Board does not believe it is in the best interests of the Company to establish term limits at this time. Additionally, such term limits may cause the Company to lose the contribution of directors who have been able to develop, over a period of time, increasing insight into the Company's business and therefore can provide an increasingly significant contribution to the Board.

K. Director Responsibilities

The business and affairs of the Company will be managed by or under the direction of the Board, including through one or more of its committees as set forth in the Company's Bye-laws and committee charters. Each director is expected to spend the time and effort necessary to properly discharge his or her responsibilities. These include:

- overseeing the conduct of the Company’s business, to evaluate whether the business is being managed properly and in conformity with applicable laws and regulations;
- reviewing and, where appropriate, approving the Company’s major financial objectives, plans and actions;
- reviewing and, where appropriate, approving major changes in, and determinations of other major issues respecting, the appropriate auditing and accounting principles and practices to be used in the preparation of the Company’s financial statements;
- reviewing and, where appropriate, approving major changes in, and determinations under, these Guidelines, the Company’s Code of Business Conduct and Ethics and other Board-approved policies of the Company;
- reviewing and, where appropriate, approving actions to be undertaken by the Company that would result in a material change in the financial structure or control of the Company, the acquisition or disposition of any businesses or asset(s) material to the Company or the entry of the Company into any major new line of business;
- reviewing the performance of the Chief Executive Officer and other executive officers, considering any input from the Compensation Committee;
- planning for succession with respect to the position of Chief Executive Officer and monitoring management’s succession planning for other key executives;
- setting a “tone at the top” that emphasizes compliance with the highest standards of ethical conduct;
- becoming and remaining well-informed about the Company’s business and operations and general business, industry and economic trends affecting the Company; and
- Ensuring that the business of the Company is conducted so as to further the best interests of its shareholders.

L. Compensation

The Company’s executive officers shall not receive additional compensation for their service as directors.

Except as otherwise permitted by the applicable Nasdaq Rules, members of the Audit Committee and Compensation Committee may not directly or indirectly receive any compensation from the Company other than their directors’ compensation (which compensation includes any compensation for service on committees of the Board and the receipt of equity incentive awards).

M. Share Ownership

The Company encourages directors to own shares of the Company. However, the number of shares of the Company owned by any director is a personal decision and, at this time, the Board has chosen not to adopt a policy requiring ownership by directors of a minimum number of shares.

N. Conflicts of Interest

Directors are expected to avoid any action, position or interest that conflicts with the interests of the Company or gives the appearance of a conflict. If an actual or potential conflict of interest develops, the director should immediately report all facts regarding the matter to the Board. Any significant conflict must be resolved or the director should resign.

O. Interaction with Institutional Investors, the Press and Customers

The Board believes that management speaks for the Company. Each director should refer all inquiries from institutional investors, the press or customers regarding the Company's operations to management. Individual Board members may, from time to time at the request of management, meet or otherwise communicate with various constituencies that are involved with the Company. If comments from the Board are appropriate, they should, in most circumstances, come from the Chairman of the Board.

P. Board Access to Senior Management

The Board will have complete access to Company management in order to ensure that directors can ask any questions and receive all information necessary to perform their duties. Directors should exercise judgment to ensure that their contact with management does not distract managers from their jobs or disturb the business operations of the Company. Any meetings or contacts that a director wishes to initiate may be arranged through the Chief Executive Officer, the Chairman of the Board or lead director, or if none is available or none is appropriate, directly by the director. To the extent appropriate, such contact, if in writing, should be copied to the Chief Executive Officer.

Q. Board Access to Independent Advisors

The Board committees may hire independent advisors as set forth in their applicable charters. The Board as a whole shall have access to such advisors and such other independent advisors that the Company retains or that the Board considers necessary to discharge its responsibilities.

R. Board and Committee Self-Evaluation

The Nominating and Corporate Governance Committee will periodically oversee an assessment of the Board and its committees.

II. BOARD MEETINGS

A. Frequency of Meetings

The Board will meet at least four (4) times annually. In addition, special meetings may be called from time to time as determined by the needs of the Company. It is the responsibility of the directors to attend meetings.

B. Director Attendance

A director is expected to spend the time and effort necessary to properly discharge his or her responsibilities. Accordingly, a director is expected to regularly prepare for and attend meetings of the Board and all committees on which the director sits (including separate meetings of the Independent Directors), with the understanding that, on occasion, a director may be unable to attend a meeting. A director who is unable to attend a meeting is expected to notify the Chairman of the Board or the Chairman of the appropriate committee in advance of such meeting, and, whenever possible, participate in such meeting via teleconference.

C. Attendance of Non-Directors

The Board encourages the Chairman of the Board or of any committee to invite Company management and outside advisors or consultants from time to time to participate in Board and/or committee meetings to (i) provide insight into items being discussed by the Board which involve the manager, advisor or consultant, (ii) make presentations to the Board on matters which involve the manager, advisor or consultant, and (iii) bring managers with high potential into contact with the Board. Attendance of non-directors at Board meetings is at the discretion of the Board.

D. Advance Receipt of Meeting Materials

Information regarding the topics to be considered at a meeting is essential to the Board's understanding of the business and the preparation of the directors for a productive meeting. To the extent feasible, the meeting agenda and any written materials relating to each Board meeting will be distributed to the directors sufficiently in advance of each meeting to allow for meaningful review of such agenda and materials by the directors. Directors are expected to have reviewed and be prepared to discuss all materials distributed in advance of any meeting.

III. COMMITTEE MATTERS

A. Number, Name, Responsibilities and Independence of Committees

The Board currently has four (4) committees: (i) the Audit Committee, (ii) the Compensation Committee, (iii) the Nominating and Corporate Governance Committee, and (iv) the Science and Research Committee. The Audit Committee, Compensation Committee, Nominating and Corporate Governance Committee and the Science and Research Committee are each composed entirely of Independent Directors, subject to any exceptions provided by the

Nasdaq Rules and applicable laws and regulations, as applicable. From time to time and depending upon the circumstances, the Board may form a new committee or disband a current committee. Each committee will perform its duties as assigned by the Board in compliance with the Company's Bye-laws and the committee's charter. It is the responsibility of the directors to attend the meetings of the committees on which they serve.

B. Appointment and Rotation of Committee Members

Committee members and committee chairs will be recommended by the Nominating and Corporate Governance Committee and appointed by the Board according to criteria set forth in the applicable committee charter and such other criteria that the Board determines to be appropriate in light of the responsibilities of each committee. Committee membership and the position of committee chair will not be rotated on a mandatory basis unless the Board determines that rotation is in the best interest of the Company.

C. Succession Planning

The Board (and/or a committee delegated by the Board) will be responsible for overseeing the process for and working with the Chief Executive Officer (together with any other Board committee or Board members to the extent desirable) on a periodic basis to develop and evaluate the Company's succession plans for the Chief Executive Officer and other executive officers, including an emergency succession plan for the Chief Executive Officer, and to review and discuss such succession plans with the Board.

IV. RISK MANAGEMENT

As provided in the Audit Committee Charter, the Audit Committee is responsible for discussing the Company's policies with respect to risk assessment and risk management, including guidelines and policies to govern the process by which the Company's exposure to risk is handled. In accordance with those policies, the Board and the Board committees shall have an active role in overseeing management of the Company's risks. The Board shall regularly review information regarding the Company's credit, liquidity and operations, as well as the risks associated with each. The Company's Compensation Committee shall be responsible for overseeing the management of risks relating to the Company's executive compensation plans and arrangements as well as risks related to human capital management. The Company's Audit Committee shall oversee management of financial and cybersecurity risks. The Nominating and Corporate Governance Committee shall manage risks associated with the independence of the Board and potential conflicts of interest. The Science and Research Committee assesses and monitors risks associated with our research and development initiatives, programs and related investments. While each committee shall be responsible for evaluating certain risks and overseeing the management of such risks, the entire Board is regularly informed through committee reports about such risks.

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