

Please bring this card with you to the Meeting and present it at Shareholder registration/accreditation.

Additional Holders:

ADDITIONAL HOLDER 1
ADDITIONAL HOLDER 2
ADDITIONAL HOLDER 3
ADDITIONAL HOLDER 4

The Chairman of Nightstar Therapeutics plc invites you to attend the Court Meeting of the Company to be held at the offices of Skadden, Arps, Slate, Meagher & Flom (UK) LLP, at 40 Bank Street, London E14 5DS on 8 May 2019 at 10.40 am.

MR SAM SAMPLE
DESIGNATION (IF ANY)
MR JOINT HOLDER 1
ADD1
ADD2
ADD3
ADD4

000001

SG349

Shareholder Reference Number

C1234567890



Please detach this portion before posting this proxy form.

Form of Proxy - Court Meeting to be held on 8 May 2019



Cast your Proxy online...It's fast, easy and secure!

www.eproxyappointment.com

You will be asked to enter the Control Number, Shareholder Reference Number (SRN) and PIN shown opposite and agree to certain terms and conditions.

Control Number: 915717

SRN: C1234567890

PIN: 1234



To view the Court Meeting Notice & Explanatory Notes online visit: www.nightstartx.com

To be effective, all proxy appointments must be lodged with the Company's Registrars at:
Computershare Investor Services PLC, The Pavilions, Bridgwater Road, Bristol BS99 6ZY by 10.40 am on 3 May 2019.

Explanatory Notes:

- Every holder has the right to appoint some other person(s) of his/her/its choice, who need not be a shareholder, as his/her/its proxy to exercise all or any of his/her/its rights to attend, speak and vote at the meeting. If you wish to appoint a person other than the Chairman, please insert the full name of your chosen proxy holder in the space provided (see reverse). If a shareholder wishes a proxy to speak on his/her/its behalf at the meeting, the shareholder will need to appoint his/her/its own choice of proxy (not the Chairman) and give instructions directly to that proxy. If the proxy is being appointed in relation to less than your full voting entitlement, please enter in the box next to the proxy holder's name (see reverse) the number of shares in relation to which he/she/it is authorised to act as your proxy.
- A shareholder may appoint more than one proxy provided each proxy is appointed to exercise rights attached to different shares. A shareholder may not appoint more than one proxy to exercise rights attached to any one share. To appoint more than one proxy, (an) additional proxy form(s) may be obtained by contacting the Registrar's helpline on 0370 703 6033 or you may photocopy this form. Please indicate in the box next to the proxy holder's name (see reverse) the number of shares in relation to which they are authorised to act as your proxy. If no number of shares is entered, the proxy will be authorised to act on your behalf in relation to your entire shareholding in the Company. Please also indicate by marking the box provided if the proxy instruction is one of multiple instructions being given. All forms must be signed and should be returned together in the same envelope.
- Entitlement to attend and vote at the meeting and the number of votes which may be cast thereat will be determined by reference to the register of members of the Company at close of business on the day which is two days before the day of the meeting. For the purpose of calculating this two day period, no account shall be taken of part of a day that is not a working day. Changes to entries on the register of members after that time shall be disregarded in determining the rights of any person to attend and vote at the meeting.
- To appoint one or more proxies or to give instruction to a proxy (whether previously appointed or otherwise) via the CREST system, CREST messages must be received by the issuer's agent (ID number 3RA50) not later than 48 hours before the time appointed for holding the meeting. For the

purpose of calculating this 48 hour period, no account shall be taken of any part of a day that is not a working day. The time of receipt will be taken to be the time (as determined by the timestamp generated by the CREST system) from which the issuer's agent is able to retrieve the message. The Company may treat as invalid a proxy appointment sent by CREST in the circumstances set out in Regulation 35(5)(a) of the Uncertificated Securities Regulations 2001.

- The above is how your address appears on the register of members. If this information is incorrect please ring the Registrar's helpline on 0370 703 6033 to request a change of address form or go to www.investorcentre.co.uk to use the online Investor Centre service.
- Any alterations made to this form should be initialled.
- The completion and return of this form will not preclude a shareholder from attending the meeting, or any adjournment thereof, and voting in person.
- If more than one joint holder is present at the meeting, whether in person or by proxy, the person whose name appears first in the register of members of the Company on the record date shall alone be entitled to vote.
- If two or more valid forms of proxy are delivered in respect of the same share, the one which was delivered last (regardless of its date of execution) will be valid.
- Terms defined in the Scheme Document issued on 9 April 2019 shall have the same meanings when used in this form of proxy, unless the context otherwise requires.
- If this form of proxy is not returned by the relevant time, it may be handed to the Chairman before the start of the Court Meeting and will still be valid.
- If you have any questions relating to the Form of Proxy, please call the Registrar's helpline on 0370 703 6033 between 8.30 am and 5.30 pm. Monday to Friday (except public holidays).

This proxy is solicited on behalf of the board of directors of Nightstar Therapeutics plc

Kindly Note: This form is issued only to the addressee(s) and is specific to the unique designated account printed hereon. This personalised form is not transferable between different: (i) account holders; or (ii) uniquely designated accounts. The Company and Computershare Investor Services PLC accept no liability for any instruction that does not comply with these conditions.

All Named Holders

MR A SAMPLE
<Designation>
ADDITIONAL HOLDER 1
ADDITIONAL HOLDER 2
ADDITIONAL HOLDER 3
ADDITIONAL HOLDER 4



Nightstar Therapeutics plc - Court Meeting

Form of Proxy

Before completing the Form of Proxy below, please read carefully the Notice of Court Meeting and section entitled "Actions to be taken" in the Scheme Document which has been sent to shareholders.

Please complete this box only if you wish to appoint a third party proxy other than the Chairman.

Please leave this box blank if you want to select the Chairman as your proxy. Do not insert your own name(s).



C1234567890



I/We hereby appoint the Chairman of the Meeting OR the person indicated in the box above as my/our proxy to attend, speak and vote in respect of my/our full voting entitlement* on my/our behalf at the Court Meeting of Nightstar Therapeutics plc to be held **at the offices of Skadden, Arps, Slate, Meagher & Flom (UK) LLP, at 40 Bank Street, London E14 5DS on 8 May 2019 at 10.40 am**, and at any adjourned meeting.

* For the appointment of more than one proxy, please refer to Explanatory Note 2 (see front).

Please mark here to indicate that this proxy appointment is one of multiple appointments being made.

Please sign and place a cross in the appropriate box below, Place a cross in **ONE** box only

IMPORTANT: if you wish to vote for the Scheme, select the box marked "FOR". If you wish to vote against the Scheme, select the box "AGAINST".

Signature

For Against

Date

In the case of a corporation, this proxy must be given under its common seal or be signed on its behalf by an attorney or officer duly authorised, stating their capacity (e.g. director, secretary).

