



## SpringWorks Therapeutics Announces Clinical Collaboration with Janssen to Evaluate Nirogacestat in Combination with Teclistamab in Patients with Relapsed or Refractory Multiple Myeloma

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### Third collaboration to evaluate nirogacestat in combination with BCMA therapies across modalities

STAMFORD, Conn., Sept. 14, 2020 (GLOBE NEWSWIRE) -- SpringWorks Therapeutics, Inc. (Nasdaq: SWTX), a clinical-stage biopharmaceutical company focused on developing life-changing medicines for patients with severe rare diseases and cancer, today announced that the company has entered into a clinical collaboration and supply agreement with Janssen Biotech, Inc. (Janssen) to evaluate SpringWorks Therapeutics' investigational gamma secretase inhibitor (GSI), nirogacestat, in combination with Janssen's bispecific antibody targeting B-cell maturation antigen (BCMA) and CD3, teclistamab, in patients with relapsed or refractory multiple myeloma.

Gamma secretase inhibition prevents the cleavage and shedding of BCMA from the surface of myeloma cells. In preclinical models, nirogacestat has been shown to increase the cell surface density of BCMA and reduce levels of soluble BCMA, which may enhance the activity of BCMA-targeted therapies, including CD3 bispecific antibodies.

"We are delighted to enter into this collaboration with Janssen to study nirogacestat in combination with teclistamab," said Saqib Islam, Chief Executive Officer of SpringWorks Therapeutics. "We now have three collaborations with industry-leading BCMA developers to evaluate nirogacestat in combinations across modalities, including with an antibody-drug conjugate, an allogeneic CAR-T cell therapy and now a bispecific antibody. This collaboration is an important step in continuing to advance our goal of developing nirogacestat as a best-in-class BCMA potentiator."

Under the terms of the agreement, Janssen will sponsor and conduct the Phase 1 study to evaluate the safety, tolerability and preliminary efficacy of the combination, and will assume all costs associated with the study, other than expenses related to the manufacturing of nirogacestat. SpringWorks Therapeutics will also form a joint oversight committee with Janssen. Pending discussions with regulators, the study is anticipated to commence by early 2021.

In addition to its ongoing clinical collaborations with BCMA-directed therapies, SpringWorks is also currently conducting a global Phase 3, double-blind, randomized, placebo-controlled clinical trial (the DeFi Trial) to evaluate nirogacestat in adults with progressing desmoid tumors.

### About Nirogacestat

Nirogacestat is an investigational, oral, selective, small molecule, gamma secretase inhibitor in Phase 3 clinical development for desmoid tumors, which are rare and often debilitating and disfiguring soft-tissue tumors. Gamma secretase cleaves multiple transmembrane protein complexes, including Notch, which is believed to play a role in activating pathways that contribute to desmoid tumor growth.

In addition, gamma secretase has been shown to directly cleave membrane-bound BCMA, resulting in the release of the BCMA extracellular domain, or ECD, from the cell surface. By inhibiting gamma secretase, membrane-bound BCMA can be preserved, increasing target density while reducing levels of soluble BCMA ECD, which may serve as decoy receptors for BCMA-directed therapies. Nirogacestat's ability to enhance the activity of BCMA-directed therapies has been observed in preclinical models of multiple myeloma. SpringWorks is pursuing a combination therapy approach to evaluate nirogacestat as a BCMA potentiator across modalities by collaborating with industry leaders. In addition to today's announcement, SpringWorks has entered into two other clinical collaborations to evaluate nirogacestat in combination with GlaxoSmithKline's BCMA antibody-drug conjugate BLENREP (belantamab mafodotin-blmf) and with Allogene's allogeneic BCMA CAR-T cell therapy ALLO-715.

Nirogacestat has received Orphan Drug Designation from the U.S. Food and Drug Administration (FDA) for the treatment of desmoid tumors (June 2018) and from the European Commission for the treatment of soft tissue sarcoma (September 2019). The FDA also granted Fast Track and Breakthrough Therapy Designations for the treatment of adult patients with progressive, unresectable, recurrent or refractory desmoid tumors or deep fibromatosis (November 2018 and August 2019).

### About SpringWorks Therapeutics

SpringWorks is a clinical-stage biopharmaceutical company applying a precision medicine approach to acquiring, developing and commercializing life-changing medicines for underserved patient populations suffering from devastating rare diseases and cancer. SpringWorks has a differentiated portfolio of small molecule targeted oncology product candidates and is advancing two potentially registrational clinical trials in rare tumor types, as well as several other programs addressing highly prevalent, genetically defined cancers. SpringWorks' strategic approach and operational excellence in clinical development have enabled it to rapidly advance its two lead product candidates into late-stage clinical trials while simultaneously entering into multiple shared-value partnerships with industry leaders to expand its portfolio. For more information, please visit [www.springworkstx.com](http://www.springworkstx.com), and follow @SpringWorksTx on [Twitter](https://twitter.com/SpringWorksTx) and [LinkedIn](https://www.linkedin.com/company/springworks-therapeutics).

### SpringWorks Forward-Looking Statements

This press release contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995, as amended, including, without limitation, statements regarding SpringWorks' clinical trials and its strategy, business plans and focus. The words "may," "will," "could," "would," "should," "expect," "plan," "anticipate," "intend," "believe," "estimate," "predict," "project," "potential," "continue," "target" and similar expressions are intended to identify forward-looking statements, although not all forward-looking statements contain these identifying words. Any forward-looking statements in this press release are based on management's current expectations and beliefs and are subject to a number of risks, uncertainties and important factors that may cause actual events or results to differ materially from those expressed or implied by any forward-looking statements contained in this press release, including, without limitation, those related to SpringWorks' financial results, the timing for completion of SpringWorks' clinical trials of its product candidates, whether and when, if at all, SpringWorks' product candidates will receive approval from the U.S.

Food and Drug Administration, or FDA, or other foreign regulatory authorities, uncertainties and assumptions regarding the impact of the COVID-19 pandemic on SpringWorks' business, operations, clinical trials, supply chain, strategy, goals and anticipated timelines, competition from other biopharmaceutical companies, and other risks identified in the section entitled "Risk Factors" in Item 1A of Part II of SpringWorks' Quarterly Report on Form 10-Q for the quarter ended June 30, 2020, as well as discussions of potential risks, uncertainties and other important factors in SpringWorks' subsequent filings with the Securities and Exchange Commission. SpringWorks cautions you not to place undue reliance on any forward-looking statements, which speak only as of the date they are made. SpringWorks disclaims any obligation to publicly update or revise any such statements to reflect any change in expectations or in events, conditions or circumstances on which any such statements may be based, or that may affect the likelihood that actual results will differ from those set forth in the forward-looking statements. Any forward-looking statements contained in this press release represent SpringWorks' views only as of the date hereof and should not be relied upon as representing its views as of any subsequent date.

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