



Principia Announces Expansion of its BTK Franchise with PRN473 Topical

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Company expands focus on immune-mediated diseases and suspends FGFR program

Company outlines anticipated key corporate milestones

SOUTH SAN FRANCISCO, Calif., Jan. 09, 2020 (GLOBE NEWSWIRE) -- Principia Biopharma Inc. (Nasdaq: PRNB), a late-stage biopharmaceutical company focused on developing treatments for immune-mediated diseases, today announced an expansion of its Bruton tyrosine kinase (BTK) franchise with PRN473 Topical. The company also announced suspension of PRN1371, an FGFR inhibitor for bladder cancer to focus the portfolio on immune-mediated diseases.

"PRN473 Topical broadens Principia's portfolio of purpose-designed BTK inhibitors, with the goal of expanding the options for patients and the knowledge of BTK's role across immune-mediated diseases," said Martin Babler, Principia's president and chief executive officer. "Principia remains at the forefront of BTK immune therapy with three separate molecules and approaches (systemic, topical, and CNS), each with distinctive rationales and potential future commercial opportunities."

By commencing clinical development of PRN473 Topical, a reversible covalent BTK inhibitor, Principia believes topical administration will be an important addition to the expected utility of systemic BTK inhibition (rilzabrutinib) and CNS-penetrant BTK therapy (SAR442168/PRN2246). Each of the three programs were discovered by Principia and have separate intellectual property estates.

About PRN473 Topical

PRN473 Topical, an investigational drug, is entering into a Phase 1, randomized, double blind, placebo-controlled, single and multiple dose trial to evaluate its safety, tolerability and pharmacokinetics. The trial will be conducted in Australia and is expected to be completed in 2020.

BTK is involved in innate and adaptive immune responses and is a critical signaling molecule in immune-mediated diseases. BTK inhibitors have demonstrated an ability to block inflammatory immune cells, eliminate autoantibody destructive signaling, and prevent new autoantibody production without depleting B cells. Given the abundance of immune cells in skin and their role in immune-mediated diseases, PRN473 Topical is being developed for localized skin applications. PRN473 Topical is the company's third BTK inhibition molecule to enter the clinic, joining rilzabrutinib, an oral, reversible covalent BTK inhibitor designed to treat systemic diseases (pemphigus, immune thrombocytopenia, and IgG4-RD) and SAR442168/PRN2246, the company's partnered central nervous system (CNS) penetrating BTK inhibitor that targets immune and inflammatory diseases of the CNS (multiple sclerosis).

Anticipated key corporate milestones

Principia also announced anticipated key milestones for the company's clinical development programs:

- **Rilzabrutinib**
 - 1H20 – Presentation of pemphigus Phase 2 Part B trial
 - 1H20 – Initiation of Phase 2 trial for IgG4-RD
 - 2H20 – Presentation of ITP Phase 2 trial
 - 2H21 – PEGASUS Phase 3 trial results (accelerated from 1H22)
- **PRN473 Topical**
 - 1H20 – Initiation of Phase 1 trial
 - 2020 – Phase 1 trial results
- **SAR442168 / PRN2246**
 - 1Q20 – Sanofi anticipates Phase 2b trial data in multiple sclerosis
 - 2020 – Principia Phase 3 co-funding decision

About Principia Biopharma

Principia is a late-stage biopharmaceutical company dedicated to bringing transformative therapies to patients with significant unmet medical needs in immune-mediated diseases. Principia's proprietary Tailored Covalency[®] platform differentiates the company's investigational therapies from traditional small molecules and provides the potential to deliver the potency, selectivity and safety of injectable drugs while maintaining the convenience of a pill or cream. This highly reproducible approach enables the company to pursue multiple programs efficiently. Rilzabrutinib, a reversible covalent BTK inhibitor, is being evaluated in a Phase 3 clinical trial in patients with pemphigus -- an orphan autoimmune disease -- a Phase 2 clinical trial in patients with immune thrombocytopenia (ITP), and the company plans to initiate a Phase 2 clinical trial in patients with IgG4-related diseases. PRN2246/SAR442168, a covalent BTK inhibitor which crosses the blood-brain barrier, is being evaluated in a Phase 2 clinical trial in patients with multiple sclerosis and is partnered with Sanofi. PRN473 Topical, a reversible covalent BTK inhibitor plans to initiate a Phase 1 clinical trial for immune-mediated diseases. For more information, please visit www.principiabiopharma.com.

Forward-Looking Statements

This press release contains forward-looking statements. These forward-looking statements reflect the current beliefs and expectations of management made pursuant to the safe harbor provisions of the Private Securities Litigation Reform Act of 1995, including, but not limited to, Principia's

expectations regarding the Principia pipeline of product candidates, the design, progress, initiation, and timing of, and results from, its clinical trials and information regarding the timing, scope and success of clinical results and potential commercial opportunities. Such forward-looking statements involve known and unknown risks, uncertainties, and other important factors that may cause Principia's actual results, performance, or achievements to be materially different from those expressed or implied by the forward-looking statements. For a description of the risks and uncertainties that could cause actual results to differ from those expressed in these forward-looking statements, as well as risks relating to the Principia's business in general, see the risk factors set forth in Principia's reports filed with the Securities and Exchange Commission. Any forward-looking statements contained in this press release speak only as of the date hereof, and Principia specifically disclaims any obligation to update any forward-looking statement, whether as a result of new information, future events or otherwise.

Investor Contact

Christopher Chai, CFO

ir@principiabiotech.com

Media Contact

Paul Laland, VP of Corporate Communications

paul.laland@principiabiotech.com

415.519.6610



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