Principia Achieves $10 Million in Additional Milestones Related to Development of PRN2246/SAR442168

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-- Additional milestones achieved under the Sanofi collaboration which is focused on treating diseases of the central nervous system --

SOUTH SAN FRANCISCO, Calif., October 16, 2018 – Principia Biopharma Inc. (Nasdaq: PRNB), a clinical-stage biopharmaceutical company dedicated to bringing transformative oral therapies to patients with significant unmet medical needs in immunology and oncology, today announced the achievement of $10 million in additional milestones related to successful development activities conducted by Principia as part of the clinical development of PRN2246. PRN2246, also known as SAR442168, is a Bruton's tyrosine kinase (BTK) inhibitor that crosses the human blood-brain barrier and modulates immune cell function in the brain for the potential treatment of central nervous system (CNS) diseases. PRN2246 is being developed under a collaboration agreement with Sanofi, a company committed to discovering and developing new treatment options for people living with multiple sclerosis (MS).

"Principia is pleased that, with the achievement of these additional milestones, we have now earned, in the aggregate, $25 million in milestones in 2018 related to our successful development efforts for PRN2246," said Martin Babler, Chief Executive Officer of Principia. "We intend to present the results of our Phase 1 clinical trial at a future scientific conference and we look forward to Sanofi commencing Phase 2 development in MS following Principia's completion of our remaining Phase 1 activities."

As previously disclosed, in the Phase 1 clinical trial, PRN2246 was considered generally safe and well-tolerated following both single and multiple dose administration to healthy human volunteers for 10 days. There were no serious adverse events or deaths reported in the trial, and all drug-related adverse events were classified as mild. Importantly, in a dedicated arm of the trial, pharmacologically-relevant cerebral spinal fluid (CSF) exposure to PRN2246 was confirmed, highlighting the potential for PRN2246 to impact B-cell driven inflammation in both the periphery and the CNS.

About PRN2246
PRN2246 is being developed to potentially treat MS and other CNS diseases, in part by penetrating the blood-brain barrier and modulating B cells and other immune cells in the CNS. During neuro-inflammation, the number of B cells in the brain increases, which is thought to play a central role in the pathology of MS and other CNS diseases. In late 2017, Principia formed a collaboration with Sanofi under which Principia granted Sanofi an exclusive, worldwide license to develop and commercialize PRN2246. Principia is responsible for completion of Phase 1 activities.

About Principia Biopharma
Principia is a clinical-stage biopharmaceutical company dedicated to bringing transformative oral therapies to patients with significant unmet medical needs in immunology and oncology. Principia's proprietary Tailored Covalency® platform enables Principia to design and develop reversible and irreversible covalent, small molecule inhibitors with potencies and selectivities that have the potential to rival those of injectable biologics, yet maintain the convenience of a pill. PRN1008, a reversible covalent BTK inhibitor, is being evaluated in a Phase 2 clinical trial in patients with pemphigus, an orphan autoimmune disease, and in a Phase 2 clinical trial in patients with immune thrombocytopenic purpura, a rare hematological disease. PRN2246, a covalent BTK inhibitor which crosses the blood-brain barrier, has completed a Phase 1 clinical trial in healthy volunteers, and has been partnered with Sanofi for development in multiple sclerosis and, potentially, for other diseases of the CNS. PRN1371, a covalent inhibitor of Fibroblast Growth Factor Receptor (FGFR), is being evaluated in a Phase 1 trial in patients with solid tumors. For more information, please visit www.principiabio.com

Forward-Looking Statements
This press release contains forward-looking statements. All statements other than statements of historical facts contained herein are forward-looking statements reflecting 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 status and timing of presenting Phase 1 results for PRN2246, and the commencement of Phase 2 development and completion of Principia's remaining Phase 1 activities. 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 any future results, performance, or achievements expressed or implied by the forward-looking statements. Such risks and uncertainties include, among others, the risks and uncertainties of the clinical development process and of clinical trial recruitment; statements about the efficacy, safety and tolerability of our product candidates; early research or clinical results may be materially different from future clinical results; Principia's reliance on third-party organizations, such as contract research organizations, contract manufacturing organizations, and partners such as Sanofi Genzyme; risk of third party claims alleging infringement of patents and proprietary rights or seeking to invalidate Principia's patents or proprietary rights; and the risk that Principia's proprietary rights may be insufficient to protect its technologies and product candidates. For a further 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 Principia's business in general, see Principia's risk factors set forth in Principia's Registration Statement on Form S-1 that is on file with the Securities and Exchange Commission (SEC) and the prospectus dated September 13, 2018 relating to its initial public offering of common stock. 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.

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