

## **AB2 Bio signs licensing agreement worth \$500 M with Nippon Shinyaku for ultra-rare autoimmune disease treatment**

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AB2 Bio, a biotechnology company developing innovative therapies for the treatment of severe systemic hyperinflammatory diseases and conditions driven by IL-18, has entered into an option and licensing agreement with Nippon Shinyaku Co. Ltd., a leading Japanese pharmaceutical company with extensive experience in marketing rare disease therapeutics in the US.

Under the terms of the agreement, Nippon Shinyaku received an option to acquire exclusive US rights to commercialise Tadekinig alfa to treat Primary Monogenic IL-18 driven Hyperinflammatory Syndrome, a rare and potentially life-threatening pediatric disease that, if left untreated, may rapidly lead to multiple-organ failure and death.

AB2 Bio will continue to prepare for filing for US Biologics License Application (BLA) approval for Tadekinig alfa in the indication.

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Nippon Shinyaku would have the exclusive right to commercialise Tadekinig alfa for its lead indication, Primary Monogenic IL-18 driven Hyperinflammatory Syndrome in patients with NLRC4 mutation and XIAP deficiency, in the US (including Guam, Puerto Rico and U.S. Virgin Islands). AB2 Bio retains rights to Tadekinig alfa for all other indications in the US (including Guam, Puerto Rico and U.S. Virgin Islands) and all indications in the rest of the world.

Tadekinig alfa is a novel, recombinant human interleukin-18 Binding Protein (IL-18 BP) that binds and inhibits IL-18, a major proinflammatory cytokine. In healthy people, a large excess of naturally occurring endogenous IL-18 Binding Protein keeps levels of systemic free IL-18 undetectable. Dysregulation of this balance in certain autoinflammatory diseases results in high

systemic levels of free IL-18 leading to dangerous pathological hyperinflammation. Tadekinig alfa treatment restores the IL-18 BP/IL-18 balance by capturing excess free IL-18, thereby reducing inflammation. This is a novel and promising approach for the treatment of several autoimmune diseases and conditions characterized by high systemic IL-18 levels.