AB2 Bio signs U.S. option and licensing agreement with Nippon Shinyaku for Tadekinig alfa for an ultra-rare autoimmune disease

- Agreement grants Nippon Shinyaku an option to acquire exclusive U.S. rights to commercialize AB2 Bio's Tadekinig alfa to treat Primary Monogenic IL-18 driven Hyperinflammatory Syndrome in patients with NLRC4 mutation and XIAP deficiency
- AB2 Bio receives early payments of up to USD 36 million (including an initial payment of USD 6 million upon signature) and would be eligible for development milestone payments of up to USD 150 million and and commercial milestone and royalty payments of up to USD 500 million
- AB2 Bio to continue to prepare Biologics License Application (BLA) and seek marketing authorization for the U.S.
- AB2 Bio retains rights to commercialize Tadekinig alfa for all other indications in the U.S. and all indications in other markets

Lausanne (Switzerland), January 27, 2025 — AB2 Bio Ltd., a biotechnology company developing innovative therapies for the treatment of severe systemic hyperinflammatory diseases and conditions driven by IL-18, announced today it 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 U.S.

Under the terms of the agreement, Nippon Shinyaku received an option to acquire exclusive U.S. rights to commercialize 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 U.S. Biologics License Application (BLA) approval for Tadekinig alfa in the indication.

AB2 Bio receives early payments of up to USD 36 million (including an initial payment of USD 6 million upon closing) and would be eligible for development milestone payments of up to USD 150 million and and commercial milestone and royalty payments of up to USD 500 million. Nippon Shinyaku would have the exclusive right to commercialize Tadekinig alfa for its lead indication, Primary Monogenic IL-18 driven Hyperinflammatory Syndrome in patients with NLRC4 mutation and XIAP deficiency, in the U.S. (including Guam, Puerto Rico and U.S. Virgin Islands). AB2 Bio retains rights to Tadekinig alfa for all other indications in the U.S. (including Guam, Puerto Rico and U.S. Virgin Islands) and all indications in the rest of the world.

"This partnership will accelerate bringing Tadekinig alfa to young patients suffering from this rare and devastating disease and potentially provide a significant improvement in treatment," said Dr. Djordje Filipovic, CEO of AB2 Bio. "Nippon Shinyaku has an impressive and established track record of commercializing rare disease therapies in the U.S., and their expertise will be invaluable for maximizing the potential of Tadekinig alfa."

"AB2 Bio is a global pioneer in developing therapies for IL-18 driven diseases and conditions," said Dr. Toru Nakai, President & Representative Director of Nippon Shinyaku. "Monogenic IL-18 driven hyperinflammation is a rare, serious, and potentially life-threatening condition without any FDA-approved treatment options. I am delighted to enter into an agreement with AB2 Bio to bring this extremely promising therapy to the U.S. patients that need it."

"As the Chair of AB2 Bio, I am very pleased to welcome our new partner Nippon Shinyaku and start this exciting chapter in the history of the Company," stated Dr. Andrea Pfeifer, Chairwoman of AB2 Bio. "Tadekinig alfa is a potential treatment option for patients suffering from IL-18 driven hyperinflammation with no current standard of care and we look forward to completing all activities required for U.S. BLA submission."

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.

In addition to, completing the Phase 3 program in the lead indication, Tadekinig alfa has established clinical proof-of-concept in three life-threatening orphan diseases and obtained Orphan Drug Designations in both Europe and the U.S. as well as U.S. Breakthrough Therapy and Pediatric Rare Disease Designations, making it potentially eligible for a Priority Review Voucher.

About Primary monogenic IL-18 driven Hyperinflammatory Syndrome in patients with NLRC4 and XIAP deficiency.

Primary monogenic IL-18 driven Hyperinflammatory Syndrome is a potentially life-threatening disease with no approved treatments. If left untreated, the disease may rapidly progress to multiple-organ failure and death. The Phase 3 program enrolled patients with verified NLRC4 or XIAP mutations who continued to suffer from severe, life-threatening hyperinflammation despite symptomatic treatment. Mutations in the NLRC4 or XIAP genes are associated with extremely high systemic levels of the pro-inflammatory cytokine IL-18, the therapeutic target of Tadekinig alfa. The excessive release of IL-18 drives the pathology of this clearly defined subgroup of primary hemophagocytic lymphohistiocytosis (HLH). The resulting hyperactivation of immune cells leads to a multiorgan pathology and non-reversible organ damage in fatal cases. This ultra-rare disease occurs most often in infants and young children.

About Nippon Shinyaku

Based on its business philosophy, “Helping people lead healthier, happier lives,” Nippon Shinyaku aims to be an organization trusted by the community through creating unique medicines that will bring hope to patients and families suffering from illness. Please visit the website (https://www.nippon-shinyaku.co.jp/english/) for products or detailed information.

About AB2 Bio Ltd

AB2 Bio is a Phase 3 clinical-stage biotech company developing innovative therapies for the treatment of severe systemic hyperinflammatory syndromes driven by IL-18. The company is advancing Tadekinig alfa in a wide range of IL-18 mediated hyperinflammatory diseases and conditions, including rare orphan diseases with high unmet medical needs. AB2 Bio was founded in 2010 and is headquartered in the Innovation Park at the Ecole Polytechnique Fédérale de Lausanne (EPFL), Switzerland. More information can be found on www.ab2bio.com.

For further information, please contact:

AB2 Bio
Zoltán Czigler
CFO, AB2 Bio
Phone +41 79 419 9111
zoltan.czigler@ab2bio.com

Media & Investors
Chris Maggos
Cohesion Bureau
Phone +41 79 367 6254
chris.maggos@cohesionbureau.com