

AB2 Bio receives \$30 million from Nippon Shinyaku for exclusive U.S. commercialization rights to Tadekinig Alfa

- **Nippon Shinyaku exercises its exclusive U.S. commercialization option for Tadekinig alfa after AB2 Bio's positive interactions with U.S. FDA**
- **License covers lead indication in the U.S., Primary Monogenic IL-18-Driven Hyperinflammatory Syndrome in patients with NLRC4 and XIAP mutations**
- **AB2 Bio retains world-wide rights to Tadekinig alfa for all other indications and all ex-U.S. rights for the lead indication**
- **AB2 Bio is eligible to receive up to USD 600 million in milestones and royalties**
- **AB2 Bio continues to lead Biologics License Application (BLA) preparation and U.S. regulatory activities**

Lausanne, Switzerland, 30 July 2026 — AB2 Bio Ltd., a clinical-stage biotechnology company developing innovative therapies targeting IL-18-driven inflammatory diseases, today announced that Nippon Shinyaku Co., Ltd. has exercised its exclusive U.S. commercialization option under the companies' 2025 option and license agreement for Tadekinig alfa, AB2 Bio's lead investigational therapy. The agreement covers treatment of Primary Monogenic IL-18-Driven Hyperinflammatory Syndrome in patients with NLRC4 and XIAP mutations, an ultra-rare, potentially life-threatening pediatric disease with no FDA-approved treatment options.

"This progress represents an important milestone for AB2 Bio and, more importantly, for patients and families affected by this devastating disease," said Dr. Djordje Filipovic, Chief Executive Officer of AB2 Bio. "Nippon Shinyaku's decision reflects the strength of our clinical program and our shared confidence in Tadekinig alfa. Together, we are well positioned to accelerate the delivery of this potential therapy to patients in the United States."

Under the agreement, AB2 Bio will receive a USD 30 million option exercise payment and is eligible to receive up to USD 100 million in development milestone payments and up to USD 500 million in commercial milestone and royalty payments. AB2 Bio received a USD 6 million option fee last year. Nippon Shinyaku now holds exclusive commercialization rights in the United States for the licensed indication, while AB2 Bio retains exclusive rights to all other indications in the United States and to all indications outside the United States. AB2 Bio will continue to lead Biologics License Application (BLA) preparation and other U.S. regulatory activities. Closing of the transaction may require clearance under the Hart-Scott-Rodino Antitrust Improvements Act of 1976, as amended, depending upon the circumstances at the time of exercise.

"AB2 Bio has established a leading position in the development of therapies targeting IL-18-driven diseases," said Dr. Toru Nakai, President and Representative Director of Nippon Shinyaku. "There are currently no FDA-approved therapies for this rare and potentially life-threatening disease. We are pleased to expand our partnership with AB2 Bio and look forward to bringing Tadekinig alfa to patients in the United States."

"This agreement validates Tadekinig alfa's clinical development program and underscores the strength of AB2 Bio's long-term strategy," said Dr. Andrea Pfeifer, Chair of AB2 Bio. "By combining our leadership in IL-18 biology with Nippon Shinyaku's proven rare disease expertise, we are accelerating the path to patients while preserving significant long-term value through our retained global rights and the continued development of Tadekinig alfa across additional IL-18-driven indications."

Tadekinig alfa is a recombinant human interleukin-18 binding protein (IL-18BP) designed to neutralize excess free IL-18, a key driver of hyperinflammation. By restoring the natural balance between IL-18 and its endogenous inhibitor, Tadekinig alfa is designed to reduce excessive inflammation and represents a novel therapeutic approach for IL-18-driven diseases.

AB2 Bio has successfully completed its Phase 3 clinical program in its lead indication and has demonstrated clinical proof of concept in three additional life-threatening orphan diseases. Tadekinig alfa has received Orphan Drug Designation in the United States and Europe, as well as Breakthrough Therapy and Rare Pediatric Disease Designations from the U.S. Food and Drug Administration (FDA), making it potentially eligible for a Priority Review Voucher upon approval.

Addressing a Critical Unmet Need

Primary Monogenic IL-18-Driven Hyperinflammatory Syndrome is an ultra-rare, potentially life-threatening disease for which there are currently no approved therapies. Patients with NLRC4 or XIAP mutations develop markedly elevated levels of free IL-18, driving severe systemic hyperinflammation that can rapidly progress to multiorgan dysfunction, irreversible organ damage and, in the most severe cases, death. The disease primarily affects infants and young children.

The Phase 3 clinical program enrolled patients with genetically confirmed NLRC4 or XIAP mutations who continued to experience severe, life-threatening hyperinflammation despite standard supportive care. By targeting excess free IL-18, Tadekinig alfa is designed to address the underlying disease mechanism rather than its symptoms, offering the potential for a first targeted treatment option for these patients.

About Nippon Shinyaku

Guided by its business philosophy, “Helping people lead healthier, happier lives,” Nippon Shinyaku aims to be an organization trusted by the community by creating unique medicines that bring hope to patients and families suffering from illness. Please visit the website (<https://www.nippon-shinyaku.co.jp/english/>) for information on products or for more 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 at www.ab2bio.com.

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