

NEWS RELEASE



January 27, 2025

Nippon Shinyaku and AB2 BIO enter into an Option Agreement for Commercialization of Tadekinig alfa for the treatment of NLRC4 mutation and XIAP deficiency in the U.S.

KYOTO, Japan, January 27, 2025 – Nippon Shinyaku Co., Ltd. (Headquarters: Kyoto, Japan, President: Toru Nakai) announced that Nippon Shinyaku and AB2 BIO Ltd. (Headquarters: Lausanne, Switzerland, CEO: Djordje Filipovic) have entered into an option agreement for the rights of commercialization of Tadekinig alfa for the treatment of NLRC4 mutation and XIAP deficiency in the U.S. by Nippon Shinyaku. AB2 BIO Ltd. is developing Tadekinig alfa for the treatment of NLRC4 mutation and XIAP deficiency in the U.S. and Europe.

NLRC4 mutation and XIAP deficiency are types of rare and serious hereditary autoinflammatory diseases, and elevated levels of interleukin 18 (IL-18) cause a variety of inflammatory symptoms. Most patients show symptoms in infancy and the inflammation persists throughout their lives. The number of patients with both NLRC4 mutation and XIAP deficiency is small, and the long-term prognosis is not clear, but these are serious diseases that make it difficult for many patients to reach adulthood. There are no drugs on the market for the treatment of either of these diseases, and new effective treatments are needed.

Tadekinig alfa is a recombinant human IL-18 binding protein, and it suppresses IL-18 related immune and inflammatory responses by specifically binding to excessively produced IL-18. Tadekinig alfa has received Orphan Drug Designation, Breakthrough Therapy Designation and Rare Pediatric Disease Designation in the U.S., and Orphan Drug Designation in Europe. In addition, AB2 BIO Ltd. has completed Phase III trials in North America and Europe, and is currently preparing for submission of a Biologics License Application (BLA) in the U.S.

After Nippon Shinyaku exercises its option rights and AB2 BIO Ltd. obtains the BLA approval in the U.S., NS Pharma, Inc. (New Jersey, USA, President: Yukiteru Sugiyama), a wholly owned subsidiary of Nippon Shinyaku, will market Tadekinig alfa.

Nippon Shinyaku is working to develop new treatments for intractable and rare diseases and aims to be a company that is trusted by society through the creation of unique drugs.

About Nippon Shinyaku

Based on Nippon Shinyaku's business philosophy, "Helping people lead healthier, happier lives," we aim 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 our website (<https://www.nippon-shinyaku.co.jp/english/>) for products or detailed information.

About AB2 BIO Ltd.

AB2 Bio is a biotech company developing innovative therapies for the treatment of severe systemic autoinflammatory diseases and conditions 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, at clinical and pre-clinical phase.

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 <http://www.ab2bio.com>.

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