AB2 Bio Completes Enrolment in Pivotal Phase 3 Trial of Tadekinig alfa to Treat Primary Monogenic IL-18 Driven HLH

- **Topline pivotal Phase 3 results expected during second half of 2023**
- **Potential new treatment option in ultra-rare, life-threatening, primarily pediatric disease with no approved therapies**

Lausanne (Switzerland), March 7, 2023 -- AB2 Bio Ltd., a biotechnology company developing innovative therapies for the treatment of severe systemic autoinflammatory diseases and conditions driven by IL-18, announced today the completion of enrolment of its ongoing pivotal Phase 3 study of Tadekinig alfa. The study is designed to show the efficacy and safety of Tadekinig alfa (r-hIL-18BP) for the treatment of primary monogenic IL-18 driven HLH, an ultra-rare and life-threatening condition with no approved therapies, mainly affecting children. AB2 Bio anticipates topline results during second half of 2023.

"This is a truly exciting time as we pioneer development of the first targeted treatment option for IL18 driven diseases and conditions," said Dr. Djordje Filipovic, CEO of AB2 Bio. "We are pleased to have completed the targeted enrollment in this pivotal study which represents a significant milestone for AB2 Bio and the patient community. This randomized, placebo-controlled study will provide the dataset required by regulatory authorities to evaluate the efficacy and safety of Tadekinig alfa in patients suffering from primary monogenic IL-18 driven HLH, which will be the basis to support marketing authorization submissions."

"We are grateful to all the patients and their caregivers for participating in this study and the dedication of the investigators and their collaborators," added Dr. Eduardo Schiffrin, Medical Director of AB2 Bio. "Tadekinig alfa is an important potential new treatment option for patients suffering from IL-18 driven autoinflammatory diseases with no current standard of care. We look forward to communicating top-line data from this study later this year."

About Primary monogenic IL-18 driven HLH in patients with NLRC4 mutation and XIAP deficiency

Primary monogenic IL-18 driven HLH (Hemophagocytic lymphohistiocytosis) is a potentially lifethreatening disease characterized by severe systemic inflammation that, if left untreated, may rapidly evolve into multiple-organ failure and death. 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 HLH, characterized by severe systemic inflammation caused by the detrimental hyperactivation of immune cells leading to a multiorgan pathology and non-reversible organ damage in fatal cases. This ultra-rare disease is most often occurring in infants and young children.

About Tadekinig alfa double-blind placebo controlled pivotal Phase 3 trial

The pivotal Phase 3 study conducted in the United States of America, Canada, and Europe, is a multicenter, double-blind, placebo-controlled, randomized withdrawal trial evaluating the efficacy and safety of Tadekinig alfa (r-hIL-18BP) in 15 primary monogenic IL-18 driven HLH patients with verified NLRC4 or XIAP mutations, suffering from severe, life-threatening hyperinflammation despite

symptomatic treatment with current care. The study has an overall duration of approximately 34 weeks. During the first 18-week single-arm, open-label (SAOL) phase, Tadekinig alfa is being administered in addition to treatments previously used in an attempt to control inflammation and related sequelae with limited success. Patients who complete the SAOL phase are enrolled in a randomized withdrawal (1:1) phase, during which they receive Tadekinig alfa or placebo for up to 16 weeks. The design, size, and endpoints of the study were validated with the US Food and Drug Administration (FDA) and the European Medicines Agency (EMA). Further details of the study can be reviewed on www.clinicaltrials.gov website using Identifier NCT03113760.

About Tadekinig alfa

Tadekinig alfa is a novel, recombinant human interleukin-18 binding protein (IL-18 BP) inhibiting IL18, a major proinflammatory cytokine. In healthy people, a large excess of naturally occurring endogenous IL-18 Binding Protein is keeping levels of systemic free IL-18 undetectable. Dysregulation of this balance results in high systemic free IL-18 levels which leads to pathological hyperinflammation. Tadekinig alfa, captures excess free IL-18 mimicking the normal physiological situation, and therefore represents a well differentiated approach for the treatment of IL-18-opathies (diseases and conditions characterized by high systemic IL-18 levels).

Tadekinig alfa has established clinical Proof-of-Concept in three life-threatening orphan diseases and obtained EMA's Orphan Drug Designation and U.S. FDA's Orphan Drug Designation, Breakthrough Therapy and Pediatric Rare Disease Designations, making it eligible for a Priority Review Voucher.

Tadekinig alfa is presented as a liquid in a vial and is being administered subcutaneously at patient's homes.

About AB2 Bio Ltd

AB2 Bio is a Phase 3 clinical-stage 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 preclinical 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 www.ab2bio.com.

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