PRESS RELEASE

AB2 Bio Ltd receives Orphan Drug Designation from the European Medicines Agency for the treatment of Haemophagocytic Lymphohistiocytosis (HLH)

- **Important regulatory milestone recognizing the potential therapeutic benefits of IL-18BP in treating HLH, a broad family of severe autoinflammatory diseases with high unmet medical need**
- **EMA ODD status supports accelerated Phase III development of Tadekinig alfa (IL-18BP)**

Lausanne (Switzerland), November 29, 2016. AB2 Bio Ltd, a clinical-stage Swiss biotech company, specialized in developing innovative therapies for the treatment of severe systemic autoinflammatory diseases, today announced that the European Medicines Agency (EMA) has granted Orphan Drug Designation (ODD) to Tadekinig alfa (IL-18BP) for the treatment of Haemophagocytic Lymphohistiocytosis (HLH).

Dr. Andrew Sleight, CEO of AB2 Bio Ltd, commented: "We are pleased that the EMA has provided ODD to Tadekinig alfa for the treatment of HLH. It validates the potential of our therapy in addressing high unmet medical need in serious autoinflammatory diseases for which there are no approved therapies."

Dr. Eduardo Schiffrin, Medical Director of AB2 Bio Ltd, added: "ODD is an important regulatory tool that can help in facilitating the development of new therapies with significant benefit for patients suffering from this serious and rare condition in Europe."

Orphan Drug Designation for Tadekinig alfa (IL-18BP)

Following on from the extraordinary case report presented at the Annual Meeting of the American College of Rheumatology in November 2015, in which AB2 Bio treated a critically ill baby girl suffering from a genetic mutation of the NLRC4 gene with IL-18BP under a compassionate use basis, AB2 Bio is currently initiating a pivotal Phase III clinical trial in patients affected by the same condition. NLRC4 mutations are part of a broad family of HLH diseases ranging from Macrophage Activation Syndrome, to patients carrying mutations of the X-linked inhibitor of apoptosis protein (XIAP) which could also benefit from Tadekinig alfa.

About Orphan Drug Designation

To receive Orphan Drug Designation from the EMA, a medicinal product must be intended for the treatment of a life-threatening or a chronically debilitating rare disease affecting not more than 5 in 10,000 individuals in the European Union (EU), and have the potential to be of significant benefit to those affected by that condition. Orphan Drug Designation provides incentives designed to facilitate development, including protocol assistance, reduced fees for regulatory activities and up to ten years of market exclusivity in the EU upon marketing approval for the designated indication.

About Haemophagocytic Lymphohistiocytosis (HLH)

HLH and related disorders are potentially life-threatening conditions of severe systemic autoinflammation. People with HLH usually develop symptoms within the first months or years of life which may include fever, pancytopenia, coagulopathy, and hemophagocytosis. These disorders can be either inherited (genetic) or secondary to other conditions such as cancer. HLH is caused by an

overactivation of macrophages. A key aspect of Tadekinig alfa's therapeutic profile is that it specifically targets the core of HLH by controlling macrophages and preventing them from becoming overactivated.

About Interleukin-18 Binding Protein (IL-18BP), a safe and transformative potential treatment in severe autoinflammatory diseases

While the time-limited inflammatory response is a natural mechanism intended to limit harm to the body, dysregulated and persistent inflammatory processes are the basis of several chronic inflammatory and autoimmune diseases. IL-18BP is a human protein with a high affinity for IL-18, a major inflammatory cytokine. In healthy people, there is a large excess of naturally occurring IL-18BP keeping levels of free IL-18 low. However, in patients with certain inflammatory diseases, the IL-18/IL-18BP balance is disrupted, resulting in high levels of free and active IL-18, which in turn leads to pathological inflammation. Administration of AB2 Bio's exogenous recombinant human IL-18BP restores the IL-18/IL-18BP balance, removing free IL-18 and thereby reducing inflammation. AB2 Bio has developed the first and unique proprietary assay detecting free IL-18 allowing the identification of clinical entities that are driven by free IL-18. As patients with high levels of free IL-18 can be identified, the clinical impact of treatment with IL-18BP will be maximized. The patients unlikely to respond to the treatment will not be unnecessarily exposed to ineffective medicines. Extensive Phase I, Ib and II clinical trial results have demonstrated that IL-18BP is very well tolerated and has an excellent safety profile.

About NLRC4 mutations and pivotal Phase III clinical trial

Recently, single point mutations in the NLRC4 gene have been identified. These genetic and gain of function mutations give rise to severe, life threatening systemic inflammation as they are associated with extremely high levels of IL-18, the therapeutic target of AB2 Bio.

In summer 2015, AB2 Bio successfully treated, on a compassionate use basis, a critically ill baby girl carrying an NLRC4 mutation and with major systemic inflammation with its experimental drug, IL-18BP. In September 2015, it was reported that the patient has entered into full remission. On November 9, 2015, this extraordinary case report was presented at the Annual Meeting of the American College of Rheumatology and was very well received. Building on this proof of concept, AB2 Bio is currently initiating a pivotal Phase III clinical trial with its experimental drug IL-18BP in patients with the same condition.

About AB2 Bio Ltd

AB2 Bio Ltd, located on the Innovation Park at the École polytechnique fédérale de Lausanne (EPFL), is specialised in the development of treatments against autoinflammatory diseases. The Swiss clinical-stage biotech company is developing drugs that will not only treat the symptoms but particularly target the underlying causes of inflammation-based diseases.

AB2 Bio is initiating a pivotal Phase III clinical trial in patients carrying mutations of the NLRC4 gene or the XIAP gene. Also, AB2 Bio has just completed a Phase II clinical trial in Adult onset Still's disease and is currently analysing the clinical data. Please find further information on www.ab2bio.com.

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