PRESS RELEASE

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FDA approves Gamifant® (emapalumab), the first and only treatment for primary haemophagocytic lymphohistiocytosis (HLH)

Swedish Orphan Biovitrum AB (publ) (Sobi™) (STO: SOBI), an international biopharmaceutical company dedicated to rare diseases, and Novimmune SA, a Swiss biotech company, today announce that the US Food and Drug Administration (FDA) has approved Gamifant® (emapalumab-lzsg), an interferon gamma (IFNγ) blocking antibody for the treatment of paediatric (new born and older) and adult patients with primary haemophagocytic lymphohistiocytosis (HLH) with refractory, recurrent or progressive disease or intolerance to conventional HLH therapy. Primary HLH is an ultra-rare syndrome of hyperinflammation with high morbidity and mortality and for which there was previously no approved drug. Gamifant represents a major advance in the treatment of these patients through a targeted mode of action.

"The FDA approval of Gamifant marks an important milestone in Sobi's vision to bring transformative treatments to patients with a high medical need," says Sobi President and CEO Guido Oelkers. "Gamifant will add significant strength to Sobi's Immunology franchise, along with Kineret and the recent acquisition of the US rights to Synagis®. We would like to acknowledge the excellent work done by Novimmune to develop and bring this important treatment to approval and Sobi will now focus on ensuring a successful commercialisation of Gamifant for the benefit of HLH patients in the US."

The FDA approval is based on data from the pivotal phase 2/3 study which enrolled patients with primary HLH. The study primary endpoint in patients with either refractory, recurrent, or progressive disease during conventional HLH therapy or who were intolerant of conventional HLH therapy was achieved, with a clinically meaningful and statistically significant proportion of patients demonstrating an overall response at the end of treatment. In addition, 70 per cent of patients proceeded to haematopoietic stem-cell transplantation (HSCT). The most commonly reported adverse reactions (≥ 20 per cent) were infections, hypertension, infusion-related reactions and fever. Results from the pivotal study will be presented at forthcoming international meetings.

Milan Zdravkovic, Chief Medical Officer and Head of Research & Development at Sobi, says: "Primary HLH is a very rare disease with significant morbidity and mortality. The approval of Gamifant as the first treatment for primary HLH is a significant step in our journey of helping these patients."

"We are extremely glad to make a new medicine available for patients suffering from primary HLH," says Cristina de Min, Chief Medical Officer at Novimmune. "Gamifant is the first drug specifically targeted to neutralise IFNγ. We would like to extend our heartfelt thanks to the patients, families and the healthcare providers who participated in the emapalumab clinical study and whose efforts helped make today's approval possible. We would also like to thank the FDA for their continuous support during emapalumab

development. Based on the clinical validation of this new target, additional clinical studies are ongoing or being planned with emapalumab in diseases for which IFNγ is considered pathogenic."

"HLH is a disorder of immune regulation in which many cytokines are deranged, but interferon gamma appears to play a critical role. While we have long understood the pivotal role of this cytokine in HLH, until emapalumab's approval we did not have a medicine that could specifically hit this target," says Michael Jordan, MD, a physician-scientist in the division of Bone Marrow Transplantation and Immune Deficiency at Cincinnati Children's Hospital Medical Center HLH Center of Excellence, and Primary Investigator in the emapalumab clinical trial. "Emapalumab represents an entirely new approach to treating primary HLH and helping these very sick patients reach haematopoietic stem cell transplant."

In the US, Gamifant was reviewed under Priority Review and received Orphan Drug Designation, Breakthrough Therapy Designation and Rare Pediatric Disease Designation from the FDA. Novimmune is eligible to receive a Priority Review Voucher (PRV) with the approval. In Europe, emapalumab was accepted for review by the European Medicines Agency (EMA) in August 2018 and has been granted orphan designation and PRIority MEdicine (PRIME) status by the EMA.

Gamifant was developed and submitted for approval to the FDA by Novimmune. Sobi acquired the global rights to Gamifant from Novimmune through an exclusive licensing agreement announced in July 2018.

About primary haemophagocytic lymphohistiocytosis (HLH)
Primary haemophagocytic lymphohistiocytosis (HLH) is an ultra-rare, rapidly progressive, often-fatal syndrome of hyperinflammation in which massive hyperproduction of interferon gamma (IFNγ) is thought to drive immune system hyperactivation, ultimately leading to organ failures. It is estimated that fewer than 100 cases of primary HLH are diagnosed each year in the US, but this is believed to represent under diagnosis. Diagnosis is challenging due to the variability in signs and symptoms, which may include fevers, swelling of the liver and spleen, severe low red and white blood cell counts, bleeding disorders, infections, neurological symptoms, organ dysfunction and organ failure. Primary HLH can rapidly become fatal if left untreated, with median survival of less than two months. The immediate goal of treatment is to quickly control the hyperinflammation and to prepare for haematopoietic stem-cell transplant. The current conventional treatment prior to transplant includes steroids and chemotherapy and are not specifically approved to treat primary HLH.

About Gamifant® (emapalumab)
Emapalumab is a monoclonal antibody (mAb) that binds to and neutralises interferon gamma (IFNγ). In the US, Gamifant is indicated for paediatric (new born and older) and adult primary haemophagocytic lymphohistiocytosis (HLH) patients with refractory, recurrent or progressive disease, or intolerance to standard-of-care HLH therapy. Gamifant is the first and only medicine approved in the US for primary HLH, an ultra-rare syndrome of hyperinflammation that usually occurs within the first year of life and can rapidly become fatal unless diagnosed and treated. The FDA approval is based on data from the pivotal phase 2/3 study (NCT01818492). Gamifant is indicated to be administered through intravenous (IV) infusion over one hour twice per week until haematopoietic stem cell transplant (HSCT). Visit www.gamifant.com for more information, including full US Prescribing Information.

About Sobi™
Sobi™ is an international speciality healthcare company dedicated to rare diseases. Our vision is to be recognised as a global leader in providing access to innovative treatments that transform lives for individuals with rare diseases. The product portfolio is primarily focused on treatments in Haemophilia and Specialty Care. Partnering in the development and commercialisation of products in specialty care is a key element of our strategy. Sobi has pioneered in biotechnology with world-class capabilities in protein biochemistry and biologics manufacturing. In 2017, Sobi had total revenues of SEK 6.5 billion and approximately 850 employees. The share (STO:SOBI) is listed on Nasdaq Stockholm. More information is available at www.sobi.com.

About Novimmune
Novimmune SA is a privately held, Swiss biopharmaceutical company focused on discovering and developing antibody-based drugs targeted for the treatment of inflammatory diseases, immune-related disorders and cancer. Founded in 1998 by the renowned immunologist Professor Bernard Mach, Novimmune has more than 150 employees and operates in two sites in Geneva and Basel (Switzerland). Since its foundation, Novimmune has built a significant R&D pipeline of drug candidates, of which emapalumab is the most advanced. The development program of Gamifant was supported by a FP7 grant from the European Commission (FIGHT HLH). Novimmune has also developed a bispecific antibody generation platform designed to streamline the identification, production and characterization of fully-human bispecific antibodies. More information is available at www.novimmune.com.

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