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Late-breaking abstract at ASH supports evidence on transformative first-ever treatment in primary HLH

Swedish Orphan Biovitrum AB (publ) (Sobi™) and Novimmune SA are pleased to announce that the abstract "*Safety and efficacy of emapalumab in paediatric patients with primary hemophagocytic lymphohistiocytosis*" was accepted as a late-breaking abstract at the 60th Annual Meeting of the American Society of Hematology in San Diego, California, 1-4 December 2018, for oral presentation. The presentation of the data will be given on Tuesday, 4 December, by the first author Professor Franco Locatelli, Head of the Department of Onco-Haematology, Bambino Gesù Children's Hospital IRCCS, Rome, Italy. Late-breaking abstracts highlight novel and substantive studies of high impact.

Emapalumab is an interferon gamma (IFN?) blocking antibody which has just been approved under the trade name Gamifant® by the FDA in the US for the treatment of paediatric (newborn and older) and adult patients with primary haemophagocytic lymphohistiocytosis (HLH) with refractory, recurrent or progressive disease or intolerance to conventional HLH therapy.

"We are thrilled that the clinical data on emapalumab in primary HLH has been selected for presentation at this important medical congress. We appreciate this recognition from ASH as it confirms the relevance of the underlying science to further the understanding of HLH, and the spotlight this shines on a very rare life-threathening disease. This would not have been possible without the support of the study investigators and participanting patients over a number of years," say Cristina de Min, Chief Medical Officer at Novimmune, and Milan Zdravkovic, Chief Medical Officer and Head of Research & Development at Sobi.

"The data obtained in the pivotal phase 2/3 study will give the opportunity to appreciate the innovative approach offered by emapalumab in controlling the HLH manifestations. Indeed, this new medicine specifically neutralises the cytokine playing a key role in HLH pathophysiology, namely interferon-gamma, thus representing the first targeted therapy for this rare disease. Paediatricians and haematologists now have a new therapeutic option for blocking the hyperinflammation typical of HLH. Moreover, thanks to the use of emapalumab, patients can receive the allograft, the only available curative option for these patients, in better condition and without the need for high-dose steroids," says Professor Locatelli.

Oral presentation:

- Safety and Efficacy of Emapalumab in Pediatric Patients with Primary Hemophagocytic Lymphohistiocytosis: Tuesday 4 December, during the Late-Breaking Abstracts oral session, 7:30-9:15. Abstract #120810. Hall AB (San Diego Convention Center).

Details of the abstract can be accessed at the ASH website:
https://ash.confex.com/ash/2018/webprogram/Paper120810.html

Satellite symposium:
Preceding the 60th ASH Annual Meeting and Exposition there will be a Friday Satellite Symposium (FSS):

- Hemophagocytic Lymphohistiocytosis – New Treatment Horizons: Friday 30 November, 18:00-22:00. Hyatt, Coronado Ballroom A-C.

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 (IFNy) 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 underdiagnosis. 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 (IFNy). In the US, Gamifant is indicated for paediatric (newborn 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.

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 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.

For more information please contact

Sobi

Media relations
Charlotte af Klercker, Head of Communications (acting)
+46 707 297 327
charlotte.afklercker@sobi.com

Investor relations
Jörgen Winroth, Vice President, Head of Investor Relations
+1 347 224 0819, +1 212 579 0506
jorgen.winroth@sobi.com

Novimmune

Chief Business Officer
Adrian Mills
+41 791 956 162
amills@novimmune.com

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Milan Zdravkovic, Chief Medical Officer and Head of Research & Development at Sobi We are thrilled that the clinical data on emapalumab in primary HLH has been selected for presentation at this important

medical congress. We appreciate this recognition from ASH as it confirms the relevance of the underlying science to further the understanding of HLH, and the spotlight this shines on a very rare life-threathening disease. This would not have been possible without the support of the study investigators and participanting patients over a number of years