PRESS RELEASE

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Emapalumab's efficacy confirmed by sensitivity analysis presented at ESID2020

- Treatment goal in primary haemophagocytic lymphohistiocytosis (HLH), a rare and life-threatening disorder, is to stabilise the disease and bring patients to haematopoietic stem cell transplantation (HSCT), the only curative therapy
- Unmet medical need remains high, with current standard-of-care therapy associated with adverse events and no decrease in mortality rates in the past 20 years
- Pivotal study of emapalumab in primary HLH patients who had no satisfactory response to prior treatments, was the first HLH study to use clinically objective response criteria
- Data from sensitivity analysis utilising various definitions of treatment response support the primary analysis and the use of clinically objective response criteria
- All analyses support the primary endpoint efficacy of a 63 per cent overall response rate (ORR) in patients with insufficient response to standard of care in the pivotal study.

Sobi™ today presented results from the sensitivity analysis from the pivotal phase 2/3 study (NCT01818492) of emapalumab in patients with primary HLH at the 19th meeting of the European Society of Immunodeficiencies (ESID). Analyses of the efficacy of emapalumab in primary HLH utilising various definitions of treatment response all support the study primary endpoint of a 63 per cent overall response rate (ORR) in patients with insufficient response to standard of care, as published in NEJM in May 2020[1].

"This analysis confirms the methodology and use of clinically objective response criteria in the pivotal study assessing treatment efficacy of emapalumab in primary HLH, where there are currently no validated endpoints for efficacy assessment" said Ravi Rao, Head of R&D and Chief Medical Officer at Sobi. "We remain committed to making emapalumab accessible to patients affected by this severe condition around the world."

A high unmet medical need exists among these patients despite today's standard-of-care therapy, glucocorticoids and etoposide, with or without cyclosporine. Conventional therapies are associated with adverse events as cytotoxicity and opportunistic infections, and no decrease in mortality has been shown in the past 20 years using the current standard-of-care therapies.

No standard approach to measuring response rate to treatment has been established due to the rarity of the disease. The pivotal study used clinically objective response criteria to define the

primary endpoint of overall response rate to emapalumab in primary HLH. The sensitivity analysis supports the use of the clinically objective ORR as a primary endpoint in studies of primary HLH.

Emapalumab is the first therapy approved by the US Food & Drug Administration (FDA) for primary HLH. Emapalumab is under review by the European Medicines Agency (EMA).

About emapalumab
Emapalumab is a monoclonal antibody that binds to and neutralises interferon gamma (IFNγ). In the US, emapalumab is indicated for the treatment of adult and paediatric (new-born and older) patients with primary hemophagocytic lymphohistiocytosis (HLH) with refractory, recurrent or progressive disease or intolerance with conventional HLH therapy. Primary HLH is a 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 phase 2/3 studies (NCT01818492 and NCT02069899). Emapalumab is indicated for administration through intravenous infusion over one hour twice per week until haematopoietic stem cell transplantation (HSCT). For more information please see www.gamifant.com including the full US Prescribing Information. Emapalumab is under review for primary HLH by the European Medicines Agency (EMA). In September 2020, emapalumab received Orphan Drug Designation (ODD) by the FDA for prevention of graft failure following haematopoietic stem cell transplantation.

About Sobi™
Sobi is a specialised international biopharmaceutical company transforming the lives of people with rare diseases. Sobi is providing sustainable access to innovative therapies in the areas of haematology, immunology and specialty indications. Today, Sobi employs approximately 1,400 people across Europe, North America, the Middle East, Russia and North Africa. In 2019, Sobi's revenues amounted to SEK 14.2 billion. Sobi's share (STO:SOBI) is listed on Nasdaq Stockholm. You can find more information about Sobi at sobi.com.

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[1]Locatelli F, Jordan MB, Allen C, et al. Emapalumab in children with primary hemophagocytic lymphohistiocytosis. N Engl J Med 2020;382:1811-22.