PRESS RELEASE

Stockholm, Sweden, and Geneva, Switzerland, 20 July 2018

Sobi™ strengthens inflammation franchise by acquiring global rights for emapalumab from Novimmune

- Strategic partnership to develop and commercialise emapalumab, a highly attractive late stage orphan drug candidate that addresses a high unmet medical need in primary Haemophagocytic lymphohistiocytosis (HLH)
- Emapalumab provides an attractive near-term commercial opportunity for Sobi with sales potential from 2019 onwards, and with an estimated annual SEK 2.5-3.0 B peak sales potential
- Emapalumab application for US regulatory approval was filed with the FDA in March 2018 with a regulatory decision expected towards the end of 2018. Breakthrough Designation has been granted by the FDA. In Europe, emapalumab has been granted eligibility for the PRIME (PRIority MEdicine) scheme by the EMA. A European filing with EMA is planned later in 2018
- Emapalumab has potential therapeutic value in other serious medical conditions, with studies in secondary HLH and haematopoietic stem cell transplant (HSCT) ongoing or being planned
- Upfront payment of CHF 50 M (SEK 450 M) in cash, with a total of CHF 400 M (SEK 3,600 M) in additional payments over an eight year period. Upon payment of all additional amounts, the intellectual property related to emapalumab, including patent rights, data and know-how, will be transferred to Sobi. The additional payments may be accelerated by either party any time after 1 July 2019.
- A non-binding letter of intent for a possible subsequent acquisition of all emapalumab assets has been signed

Swedish Orphan Biovitrum AB (publ) (Sobi™) (STO: SOBI), an international biopharmaceutical company dedicated to rare diseases, and Novimmune SA have entered into an exclusive licence agreement for the perpetual global rights to emapalumab, a late-stage orphan drug candidate for the treatment of primary Haemophagocytic lymphohistiocytosis (HLH), developed by Novimmune SA, a privately-held, pre-commercial drug discovery and development company focused on rare inflammatory diseases, immune disorders and immuno-oncology.

Further, Sobi has signed a non-binding letter of intent with the majority shareholders of Novimmune to explore a broader collaboration regarding a possible subsequent acquisition of all assets relating to emapalumab, which would also include the transfer of relevant employees, and options for exclusive

development and commercialisation rights to NI-1801 and Novimmune's interest in NI-1701[1], two preclinical products in the field of immuno-oncology.

"We believe this transaction is an excellent fit for Sobi and consistent with our strategy of expanding our commercial Specialty Care portfolio, strengthening our geographic footprint, particularly in the US, and building our R&D pipeline. The addition of emapalumab to Sobi's portfolio is a natural fit that builds upon our expertise with Kineret® (anakinra) and focuses our business in inflammation, immunology and immuno-oncology. Our strategic goal is to build our Specialty Care business area as a complement to Haemophilia, allowing Sobi to advance our position to become global leaders in rare diseases," comments Sobi President and CEO Guido Oelkers.

Novimmune's Chairman and CEO Eduardo Enrico Holdener adds, "We are delighted to announce this collaboration with Sobi. Novimmune has brought emapalumab all the way from discovery through development to the brink of regulatory approval. Now is the right time to bring in a partner with substantial commercial capabilities to take the next step of making the product available for patients in need. In Sobi we see a strong partner with a proven ability to further develop, commercialise and provide access to innovative treatments for rare disease patients across European and US markets. Pending approval by the FDA, this means that patients who suffer from HLH will be able to benefit from emapalumab, the only targeted medicine specifically developed for the treatment of HLH."

About emapalumab

Emapalumab is an anti-interferon-gamma (IFN-ɣ) monoclonal antibody (mAb), currently under FDA review for the treatment of primary Haemophagocytic lymphohistiocytosis (HLH), a life-threatening syndrome of immune activation with high unmet medical need for which there is currently no approved drug therapy. In the US, emapalumab has received Orphan Drug Designation, Breakthrough Designation and Rare Pediatric Disease Designation from the FDA. Novimmune will be eligible to receive a Priority Review Voucher (PRV) upon approval. The Biologic License Application (BLA) for primary HLH was filed with the FDA in March 2018 and the file was accepted in May 2018. A regulatory decision in the US is expected towards the end of 2018. In Europe, emapalumab has been granted orphan designation and PRIME (PRIority MEdicine) status by the EMA. The EMA filing is expected to be submitted later in 2018. Studies in secondary HLH are ongoing, and emapalumab carries potential therapeutic value in other serious medical conditions, with studies in secondary HLH and acute graft failure in haematopoietic stem cell transplant (HSCT) ongoing or being planned.

[1] Novimmune entered into a joint venture and licence option agreement with TG Therapeutics Inc. to develop NI-1701 on 18 June 2018.

About HLH

Haemophagocytic lymphohistiocytosis, HLH, is a rare and life-threatening syndrome of extreme immune activation. The primary form of the disease (pHLH, inherited) mainly occurs in infants and young children and the secondary form of the disease (sHLH, acquired) is acquired from or associated with infection, autoimmune diseases or malignancy. The estimated patient population in the US, EU and Japan amounts to around 5,000 people across both primary and secondary HLH. There is currently no approved drug therapy for HLH, with a current treatment protocol consisting of immuno-chemotherapy (in particular dexamethasone and etoposide) being used to suppress the immune reaction.

Terms of the transaction

Sobi has entered into an exclusive licensing agreement with Novimmune under which an exclusive global licence for emapalumab is granted to Sobi. An upfront payment of CHF 50 M (SEK 450 M) in cash will be paid by Sobi for the global licence, with a total of CHF 400 M (SEK 3,600 M) in additional payments over an eight year period. The additional unpaid payments may, however, be accelerated by either party at any time after 1 July 2019. Upon payment of the final fixed amount (over time or following the exercise of the acceleration option), all intellectual property that is owned or controlled by Novimmune that is related to emapalumab will be transferred to Sobi. Sobi will have full responsibility for all future development and commercialisation costs from September 2018, with final control in the event of a dispute. Closing of the licensing deal is expected in Q3 2018, subject to customary approvals of the relevant competition authorities.

Financial implications

The upfront payment of CHF 50 M (SEK 450 M) will be paid in cash. The licence agreement for emapalumab is expected to contribute to Sobi's top-line sales growth from 2019 onwards and is expected to be accretive to earnings from 2021 onwards. The emapalumab-related development and pre-launch costs for Sobi during 2018 are expected to be approximately SEK 250-350 M.

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. 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.

About Sobi™
Sobi™ is an international biopharmaceutical 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.

Forward looking statements
This press release contains forward-looking statements, including statements regarding the potential benefits that may be derived from the licencing agreement regarding emapalumab, plans and expected timing with respect to the potential approval of emapalumab in the US, as well as potential future sales of emapalumab. These forward-looking statements are based on management expectations and assumptions as of the date of this press release, and actual results may differ materially from those in these forward-looking statements as a result of various factors. These factors include risks that the filings with the FDA are not approved and that adequate pricing and reimbursement for emapalumab in the US and Europe is not available. Forward-looking statements speak only as of the date of this press release and Sobi does not undertake any obligation to update or revise these statements, except as may be required by law or regulation.

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:
Chairman and CEO
Ed Holdener
+41 79 309 8726
eholdener@novimmune.com

Chief Business Officer
Adrian Mills
+41 79 195 6162
amills@novimmune.com

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