Sobi to present new data at the ASH 2022 Annual Meeting

Sobi® will present new data at the 64th Annual Meeting of the American Society of Hematology (ASH) taking place from 10 to 13 December 2022. Sobi’s commitment to developing innovative treatments for people living with rare diseases is highlighted in studies spanning several rare disorders, including haemophilia, paroxysmal nocturnal haemoglobinuria (PNH), cold agglutinin disease (CAD), immune thrombocytopenia (ITP) and primary haemophagocytic lymphohistiocytosis (pHLH).

Results from the long-term extension study of Aspaveli®/Empaveli™ (pegcetacoplan) in adults with PNH will be presented, demonstrating robust and sustained improvements for approximately two years in key markers of disease across a broad population of PNH patients.

In haemophilia A, new data on efanesoctocog alfa will be presented. Efanesoctocog alfa, an investigational new class of factor therapy for people with haemophilia A, has the potential to elevate standards of care by providing high sustained factor VIII activity into the near-normal range for the majority of the week with once-weekly prophylaxis.

New data also will be presented from the REAL-HLH study, which is the first study describing the clinical characteristics, treatment patterns and treatment outcomes of a larger cohort of patients who received Gamifant® (emapalumab) in a real-world clinical setting.

New information will also be presented supporting the role of Doptelet® (avatrombopag) in the treatment of ITP and describing the design of a study with Aspaveli/Empaveli for the treatment of CAD.

“The breadth of data being presented at ASH this year showcases our ongoing commitment to providing innovative treatments for those affected by rare, ultra-rare and life-threatening conditions,” said Anders Ullman, Head of Research & Development and Chief Medical Officer at Sobi. “We look forward to collaborating and connecting in person at this year’s meeting.”

ADC Therapeutics to present new data on loncastuximab tesirine
Sobi collaboration partner ADC Therapeutics will present eight abstracts at ASH. Multiple presentations will highlight the clinical utility of loncastuximab tesirine in diffuse large B-cell lymphoma. For more information please visit: ir.adctherapeutics.com/press-releases.
### Key Sobi data to be presented at ASH 2022

<table>
<thead>
<tr>
<th><strong>Haemophilia</strong></th>
<th><strong>Efficacy of Efanesoctocog Alfa on Physical Functioning: Results from the XTEND-1 Phase 3 Clinical Trial in Previously Treated Patients with Hemophilia A</strong></th>
<th>Poster presentation #2468 Sunday, 11 Dec, 6:00-8:00 PM Joint with Sanofi</th>
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<tr>
<td>efanesoctocog alfa</td>
<td><strong>Efficacy of Efanesoctocog Alfa on Pain in Patients with Hemophilia A: Results from the XTEND-1 Phase 3 Clinical Trial in Previously Treated Patients with Hemophilia A</strong></td>
<td>Poster presentation #2474 Sunday, 11 Dec, 6:00-8:00 PM Joint with Sanofi</td>
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<td></td>
<td><strong>A Population Pharmacokinetic (PopPK) Model to Characterize Efanesoctocog Alfa (BIVV001) Factor VIII (FVIII) Activity Levels in Patients with Severe Hemophilia A</strong></td>
<td>Poster presentation #3788 Monday, 12 Dec, 6:00-8:00 PM Joint with Sanofi</td>
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<th><strong>Paroxysmal nocturnal haemoglobinuria</strong></th>
<th><strong>Aspaveli/Empaveli (pegcetacoplan)</strong></th>
<th><strong>Long-Term Safety and Efficacy of Pegcetacoplan Treatment in Adults with Paroxysmal Nocturnal Hemoglobinuria</strong></th>
<th>Poster presentation #1248 Saturday, 10 Dec, 5:30-7:30 PM Joint with Apellis</th>
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<td></td>
<td><strong>Intensive Pegcetacoplan Dosing in the Management of Acute Hemolysis As Part of the 307 Open-Label Extension Study</strong></td>
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<td>Poster presentation #1255 Saturday, 10 Dec, 5:30-7:30 PM Joint with Apellis</td>
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| **Cold agglutinin disease** | **Aspaveli/Empaveli (pegcetacoplan)** | **Trial in Progress: Randomized, Double-Blind, Placebo-Controlled Multicenter Phase 3 Study to Evaluate the Efficacy and Safety of Pegcetacoplan in Patients with Cold Agglutinin Disease (CASCADE)** | Poster presentation #2333 Sunday, 11 Dec, 6:00-8:00 PM Joint with Apellis |

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<th><strong>Immune thrombocytopenia</strong></th>
<th><strong>Doptelet (avatrombopag)</strong></th>
<th><strong>Comparative Length of Therapy and Persistence Among Thrombopoietin Receptor Agonists (TPO-RA) in Immune Thrombocytopenia (ITP): United States Healthcare Claims and Specialty Pharmacy Prescription Analyses</strong></th>
<th>Poster presentation #3769 Monday, 12 Dec, 6:00-8:00 PM</th>
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<td></td>
<td><strong>Avatrombopag Treatment Response in Patients with Immune Thrombocytopenia: A Real-World Evidence Study</strong></td>
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<td>Poster presentation #4892 Monday, 12 Dec, 6:00-8:00 PM</td>
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Haemophagocytic lymphohistiocytosis

| Gamifant (emapalumab) | Real-World Demographics, Clinical Characteristics, and Treatment Patterns of Patients Treated with Emapalumab for Primary Hemophagocytic Lymphohistiocytosis in the United States: The REAL-HLH Study | Poster presentation #1086 Saturday, 10 Dec, 5:30-7:30 PM |

All abstracts can be accessed via the official ASH website.

**About efanesoctocog alfa (BIVV001)**
Efanesoctocog alfa is a novel and investigational recombinant factor VIII therapy with the potential to deliver near-normal factor activity levels for most of the week, extending bleed protection in a once-weekly dose for people with haemophilia A. Efanesoctocog alfa builds on the innovative Fc fusion technology by adding a region of von Willebrand factor and XTEN® polypeptides to potentially extend its time in circulation. It is the only therapy that has been shown to break through the von Willebrand factor ceiling, which is believed to impose a half-life limitation on current factor VIII therapies. Efanesoctocog alfa is currently under clinical investigation and its safety and efficacy have not been reviewed by any regulatory authority. Efanesoctocog alfa was granted orphan drug designation by the US Food & Drug Administration (FDA) in August 2017 and the European Commission in June 2019. The FDA has accepted for priority review the Biologics License Application for efanesoctocog alfa for the treatment of haemophilia A. The target action date for the FDA decision is 28 February 2023. Regulatory submission in the EU will follow availability of data from the ongoing XTEND-Kids paediatric study, expected in 2023.

**About the Sobi and Sanofi collaboration**
Sobi and Sanofi collaborate on the development and commercialisation of Alprolix® and Elocta®/Eloctate®. The companies also collaborate on the development and commercialisation of efanesoctocog alfa, an investigational factor VIII therapy with the potential to provide high sustained factor activity levels with once-weekly dosing for people with haemophilia A. Sobi has final development and commercialisation rights in the Sobi territory (essentially Europe, North Africa, Russia and most Middle Eastern markets). Sanofi has final development and commercialization rights in North America and all other regions in the world excluding the Sobi territory.

**About Aspaveli®/Empaveli™**
Aspaveli/Empaveli (pegcetacoplan) is a targeted C3 therapy designed to regulate excessive activation of the complement cascade, part of the body’s immune system, which can lead to the onset and progression of many serious diseases. It is approved as Aspaveli for the treatment of adults with paroxysmal nocturnal haemoglobinuria (PNH) who are anaemic after treatment with a C5 inhibitor for at least three months in the European Union and the United Kingdom and as Empaveli for treatment of adult patients with PNH in the United States and Saudi Arabia. Empaveli is also approved in Australia for treatment of adult patients with PNH who have an inadequate response to, or are intolerant of, a C5 inhibitor.

**About the Sobi and Apellis Collaboration**
Sobi and Apellis collaborate to develop and commercialize systemic Aspaveli/Empaveli. Sobi has exclusive ex-US commercialisation rights for systemic Aspaveli/Empaveli. Apellis has exclusive US commercialisation rights for systemic Empaveli. The companies have global co-development rights for systemic Aspaveli/Empaveli.

**About Doptelet®**
Doptelet (avatrombopag) is an orally administered thrombopoietin receptor agonist (TPO-RA) that mimics the biologic effects of TPO in stimulating the development and maturation of megakaryocytes, resulting in increased platelet count. It is approved by the European Medicines Agency and the US Food & Drug Administration for the treatment of severe thrombocytopenia in adult patients with chronic liver disease who are scheduled to undergo an invasive procedure and for the treatment of thrombocytopenia in adult patients with primary chronic immune thrombocytopenia (ITP) who have had an insufficient response to a previous treatment. Chronic ITP is a rare autoimmune bleeding disorder characterised by low number of platelets. The incidence of primary ITP in adults is 3.3/100 000 adults per year with a prevalence of 9.5 per 100 000 adults.\(^1\)
About Gamifant®
Gamifant (emapalumab) is an anti-interferon gamma (IFNγ) monoclonal antibody that binds to and neutralises IFNγ. In the USA, Gamifant is indicated for the treatment of adult and paediatric (newborn and older) patients with primary haemophagocytic 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). Gamifant is indicated for administration through intravenous infusion over one hour twice per week until haematopoietic stem cell transplantation (HSCT).

About loncastuximab tesirine
Loncastuximab tesirine (US brand name ZYNLONTA®) is a CD19-directed antibody-drug conjugate (ADC). Once bound to a CD19-expressing cell, ZYNLONTA is internalised by the cell, where enzymes release a pyrrolobenzodiazepine (PBD) payload. The potent payload binds to DNA minor groove with little distortion, remaining less visible to DNA repair mechanisms. This ultimately results in cell cycle arrest and tumour cell death.

The US Food and Drug Administration (FDA) has approved ZYNLONTA (loncastuximab tesirine-lpyl) for the treatment of adult patients with relapsed or refractory (r/r) large B-cell lymphoma after two or more lines of systemic therapy, including DLBCL not otherwise specified, DLBCL arising from low-grade lymphoma and also high-grade B-cell lymphoma. The LOTIS-2 study included a broad spectrum of heavily pre-treated patients (median three prior lines of therapy) with difficult-to-treat disease, including patients who did not respond to first-line therapy, patients refractory to all prior lines of therapy, patients with double/triple hit genetics and patients who had stem cell transplant and CAR-T therapy prior to their treatment with ZYNLONTA. This indication is approved by the FDA under accelerated approval based on overall response rate and continued approval for this indication may be contingent upon verification and description of clinical benefit in a confirmatory study.

ZYNLONTA is also being evaluated as a therapeutic option in combination studies in other B-cell malignancies and earlier lines of therapy.

Sobi®
Sobi is a specialised international biopharmaceutical company transforming the lives of people with rare diseases. Providing sustainable access to innovative medicines in the areas of haematology, immunology and specialty care, Sobi has approximately 1,600 employees across Europe, North America, the Middle East and Asia. In 2021, revenue amounted to SEK 15.5 billion. Sobi’s share (STO:SOBI) is listed on Nasdaq Stockholm. More about Sobi at sobi.com, LinkedIn and YouTube.

Contacts
For details on how to contact the Sobi Investor Relations Team, please click here. For Sobi Media contacts, click here.