PRESS RELEASE

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At ASH, extended half-life therapies Elocta® and Alprolix® demonstrate proven efficacy and well-characterised safety over four years

ASPIRE and B-YOND extension studies show no inhibitor development and consistently low annualised bleeding rates in study participants over four years with Elocta and Alprolix, respectively.

Swedish Orphan Biovitrum AB (publ) (Sobi™) and Bioverativ Inc., a Sanofi-company, today announced the final results of ASPIRE and B-YOND, the most comprehensive long-term studies of extended half-life factor therapies in haemophilia. The data from both studies confirm the established safety and sustained efficacy of Elocta® (efmoroctocog alfa), marketed as ELOCTATE® in the United States and other countries, and Alprolix® (eftrenonacog alfa) over four years of treatment in previously treated adult, adolescent, and paediatric patients with severe haemophilia A and B, respectively. These results were presented at the 60th Annual Meeting of the American Society of Hematology (ASH).

Factor replacement therapy is the cornerstone of haemophilia care and results from ASPIRE and B-YOND demonstrated that long-term prophylactic treatment with efmoroctocog alfa and eftrenonacog alfa consistently improved annualised bleed rates, including joint bleeds, across all patient populations studied and at extended dosing intervals. No inhibitors were observed in subjects enrolled in either of the two extension studies and the overall safety profile was consistent with the pivotal phase 3 studies. Inhibitor development has been observed with efmoroctocog alfa and eftrenonacog alfa post market.

“These data add to a significant body of evidence showing that efmoroctocog alfa and eftrenonacog alfa provide protection from all types of haemophilia-related bleeds with individualised and flexible dosing regimens across all study populations,” said Tim Harris, Ph.D. DSc., Executive Vice President, Research and Development at Bioverativ. “We remain focused on and committed to providing complete protection for people with haemophilia.”

Debilitating joint disease, which is caused by repeated bleeds into joints over time, is one of the most common complications for people with haemophilia. In ASPIRE and B-YOND, subjects on prophylactic treatment experienced low joint and spontaneous joint annualised bleed rates (ABRs) across all dosing regimens. These results support that prophylactic dosing with efmoroctocog alfa and eftrenonacog alfa can effectively manage and control all types of joint bleeds.

“Joint protection remains a significant challenge in the long-term treatment of haemophilia keeping individuals from living a life without constraints of their disease and these results confirm that our therapies can play a role in the effective prevention of joint bleeds,” said Milan Zdravkovic, Head of Research &

Development and Chief Medical Officer at Sobi. “In addition to providing the clinical evidence to support the long-term use of our therapies, we continue to explore the impact of Fc fusion on joint health.”

An interim, post-hoc analysis of ASPIRE published in *Haemophilia* found that a prophylactic regimen of efmoroctocog alfa can lead to continuous improvement in joint health, regardless of prior treatment regimen, severity of joint damage, or target joints. This retrospective study evaluated joint health in adult and adolescent participants (n=47) using a modified version of the Hemophilia Joint Health Score (mHJHS), a first-line assessment tool that grades joints by specific domains including swelling, muscle atrophy, alignment, range of motion, joint pain, strength and global gait. Additional studies will be needed to confirm these findings and the mHJHS will require further validation.

Efmoroctocog alfa and eftrenonacog alfa are leading extended half-life therapies in the United States and Europe. They have been proven to treat all types of bleeds and can be used in all treatment scenarios, including acute, surgical and emergency situations. The safety and efficacy of both therapies have been studied over hundreds of exposure days in adult, adolescent, and paediatric patients with haemophilia since 2010.

About ASPIRE

ASPIRE is an open-label, non-randomised, multi-year extension study for people who completed the pivotal, phase 3 A-LONG or Kids A-LONG studies. The study enrolled 211 males, including 150 (98 per cent) of those who completed A-LONG and 61 (91 per cent) of those who completed Kids A-LONG. The primary endpoint is the development of inhibitors. Secondary endpoints include the annualised number of bleeding episodes per subject, Elocta exposure days and a participant’s assessment of response to treatment of a bleeding episode. Key findings include:

- Overall median ABRs for those on prophylactic treatment remained low throughout ASPIRE, particularly in the individualised dosing arm.
- Zero spontaneous joint bleeds were reported in subjects in all age groups in the individualised dosing arm. Median joint ABRs of <0.66 were also reported in the cohort.
- In the study, adult and adolescent subjects (n=72) treated prophylactically with efmoroctocog alfa experienced a mean improvement in modified hemophilia joint health (mHJHS) score of -2.5 (negative shows improvement) compared to baseline score in A-LONG.
- Over 92 per cent of subjects either lengthened or experienced no change in dosing intervals during the length of the study.
- Low ABRs as well as improved joint health scores reported in ASPIRE demonstrate clinical benefit of efmoroctocog alfa that goes beyond just bleed prevention.

About B-YOND

B-YOND is an open-label, non-randomised, multi-year extension study for people who completed the pivotal, phase 3 B-LONG or Kids B-LONG studies. B-YOND enrolled 116 previously-treated males, including 93 participants (81%) who completed B-LONG, and 27 (100%) of those who completed Kids B-LONG. The primary outcome measure is development of inhibitors. Secondary endpoints

include the annualised number of bleeding episodes per subject (including spontaneous joint bleeding rates), Alprolix exposure days per participant, Alprolix consumption (total IU/kg per subject per year), and the participant's assessment of response to treatment of a bleeding episode. Key findings include:

- For subjects on prophylactic treatment, ABRs remained low throughout the study across all age groups, especially related to joint and spontaneous joint bleeds.
- In adult and adolescent subjects following a prophylactic regimen with eftrenonacog alfa, joint and spontaneous joint median ABRs were <1.58 and <0.38, respectively.
- In study participants<12 years on prophylactic treatment joint and spontaneous joint median ABRs were <0.85 and zero, respectively.
- Data showed 85 per cent of adult and 93 per cent of paediatric subjects either lengthened or experienced no change in dosing intervals during the extension study.
- Eftrenonacog alfa provides flexible dosing while maintaining consistently low bleeding rates with extended interval dosing up to 14 days.
- The B-YOND study reflects real world use of eftrenonacog alfa with flexible dosing and adjustments based on individualised preference and clinical needs.

About Elocta®/ELOCTATE®
Elocta® (efmoroctocog alfa) is a recombinant clotting factor therapy developed for haemophilia A using Fc fusion technology to prolong circulation in the body. It is engineered by fusing factor VIII to the Fc portion of immunoglobulin G subclass 1, or IgG1 (a protein commonly found in the body), enabling Elocta to use a naturally occurring pathway to extend the time the therapy remains in the body (half-life). While Fc fusion technology has been used for more than 15 years, Sobi and Bioverativ have optimised the technology and are the first companies to utilise it in the treatment of haemophilia Elocta is manufactured using a human cell line in an environment free of animal and human additives.

Elocta is approved and marketed by Sobi for the treatment of haemophilia A in the EU, Iceland, Kuwait, Liechtenstein, Norway, Saudi Arabia and Switzerland. It is approved and marketed as ELOCTATE® [Antihemophilic Factor (Recombinant), Fc Fusion Protein] by Bioverativ in the United States, Japan and Canada. It is also approved in Australia, New Zealand, Brazil and other countries, where Bioverativ has the marketing rights.

As with any factor replacement therapy, allergic-type hypersensitivity reactions and development of inhibitors may occur in the treatment of haemophilia A. Inhibitor development has been observed with Elocta/ELOCTATE, including in previously untreated patients. For more information, please see the full U.S. prescribing information for ELOCTATE. Note that the indication for previously untreated patients is not included in the EU Product Information for Elocta.

About Alprolix®
Alprolix® (eftrenonacog alfa), is a recombinant clotting factor therapy developed for haemophilia B using Fc fusion technology to prolong circulation in the body. It is engineered by fusing factor IX to the Fc portion of immunoglobulin G subclass 1, or IgG1 (a protein commonly found in the body), enabling Alprolix to use a naturally occurring pathway to extend the time the therapy remains in the body (half-life). While Fc fusion technology has been used for more than 15 years, Sobi and Bioverativ have optimised the technology and are the first companies to utilise it in the treatment of haemophilia. Alprolix is manufactured using a human cell line in an environment free of animal and human additives.

Alprolix is approved and marketed by Sobi for the treatment of haemophilia B in the EU, Iceland, Kuwait, Liechtenstein, Norway, Saudi Arabia and Switzerland, as well as in the United States, Canada, Japan, Australia, New Zealand, Brazil and other countries where Bioverativ has the marketing rights.

Allergic-type hypersensitivity reactions and development of inhibitors have been observed with Alprolix in the treatment of haemophilia B, including in previously-untreated patients. For more information, please see the full U.S. prescribing information for

Alprolix. Note that the indication for previously-untreated patients is not included in the EU Product Information.

About haemophilia A and B
Haemophilia is a rare, genetic disorder in which the ability of a person's blood to clot is impaired. Haemophilia A occurs in about one in 5,000 male births annually, and more rarely in females. Haemophilia B occurs in about one in 25,000 male births annually, and more rarely in females. The World Federation of Hemophilia estimates that approximately 180,000 people are currently diagnosed with haemophilia A and B worldwide.[i]

People with haemophilia A or B experience bleeding episodes that can cause pain, irreversible joint damage and life-threatening haemorrhages. Prophylactic infusions of factor VIII or IX can temporarily replace the clotting factors that are needed to control bleeding and prevent new bleeding episodes.[ii] The World Federation of Hemophilia recommends prophylaxis as the optimal therapy as it can prevent bleedings and joint destruction.[iii]

About the Sobi and Bioverativ collaboration
Sobi and Bioverativ, a Sanofi company, collaborate on the development and commercialisation of Alprolix and Elocta/ELOCTATE. Sobi has final development and commercialisation rights in the Sobi territory (essentially Europe, North Africa, Russia and most Middle Eastern markets). Bioverativ has final development and commercialisation rights in North America and all other regions in the world excluding the Sobi territory and has manufacturing responsibility for Elocta/ELOCTATE and Alprolix. While Fc fusion technology has been used for more than 15 years, Sobi and Bioverativ have optimised the technology and are the first companies to utilise it in the treatment of haemophilia. Sobi has elected to add the rFVIIIFc-VWF-XTEN fusion molecule and the rFIXFc-XTEN for the potential treatment of haemophilia A and B respectively to its collaboration agreement with Bioverativ.

About Bioverativ
Bioverativ, a Sanofi company, is dedicated to transforming the lives of people with hemophilia and other rare blood disorders through world-class research, development, and commercialization of innovative therapies. Bioverativ is committed to actively working with the blood disorders community, and its hemophilia therapies when launched represented the first major advancements in hemophilia treatment in more than two decades. For more information, visit www.bioverativ.com or follow @bioverativ on Twitter.

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 make a significant difference 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.

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[i] World Federation of Hemophilia. Annual Global Survey 2015, published in October 2016. Available at: http://www1.wfh.org/publication/files/pdf-1669.pdf. Accessed on May 23, 2017.
[ii] World Federation of Hemophilia. About Bleeding Disorders – Frequently Asked Questions. Available at: http://www.wfh.org/en/page.aspx?pid=637. Accessed on May 23, 2017.
[iii] World Federation of Hemophilia. Guideline for the management of hemophilia, 2nd edition. Available at: http://www1.wfh.org/publication/files/pdf-1472.pdf. Accessed on May 23, 2017.