PRESS RELEASE

Stockholm, Sweden, 10 May 2019

Individualised extended half-life factor replacement therapy controls haemostasis in orthopaedic surgery

Today, Sobi™ and Sanofi will present data demonstrating that Elocta® (efmoroctocog alfa) and Alprolix® (eftrenonacog alfa) can provide perioperative haemostatic control across a wide spectrum of major and minor surgeries in subjects of all ages with severe haemophilia A and B respectively. The presentation will take place at the World Federation of Hemophilia's 16th International Musculoskeletal Congress (MSK) 10-12 May 2019, in Madrid, Spain.

One of the major complications for people living with haemophilia is recurrent bleeding into joints and subsequent deteriorating joint health. Joint surgery and rehabilitation of damaged joints can improve the quality of life for these patients. However, people with haemophilia are at risk during surgical procedures due to the increased risk of bleeding during the procedure and the reduced clotting of the blood which characterises haemophilia. It is possible to minimise blood loss during surgery by treating patients with an individualised and optimal factor replacement therapy to control the haemostasis. The risk of bleeding in a haemophilia patient is inversely proportional to the factor level activity in the blood[1] and treatment guidelines recommend monitoring factor levels post-surgery.

"Orthopaedic surgery is common in haemophilia since a large majority of haemophilia patients experience joint damage due to suboptimal levels of protection, leading to reoccurring bleeding in the joints," says Armin Reininger, Head of Scientific and Medical Affairs at Sobi. "Therefore, it is of great importance to offer an individualised prophylactic treatment to reduce the need for and occurrence of surgery as well as an optimal and measurable factor treatment during surgery. Furthermore, to maintain joint health, it is important to reduce pain and increase mobility of patients so that they can fully participate in everyday life"

The data presented at MSK support the use of individualised treatment with rFVIIIFc (efmoroctocog alfa) and rFIXFc (eftrenonacog alfa) to provide perioperative haemostatic control across a wide spectrum of major and minor surgeries in subjects of all ages with severe haemophilia A and B respectively. Most major procedures required one injection to maintain haemostasis during surgery, and all haemostatic responses were assessed as excellent or good by the investigator/surgeon. Both treatments were well tolerated.

Results from the studies:

The data present final pooled post-hoc analysis of A-LONG, Kids A-LONG and ASPIRE subjects who underwent surgery and were treated with an investigator-determined rFVIIIFc regimen as well as the final pooled post-

[1] Fijnvandraat K, Cnossen MH, Leebeek FW, Peters M. Diagnosis and management of haemophilia. BMJ. 2012;344:e2707.

hoc analysis of B-LONG, Kids B-LONG and B-YOND subjects treated with investigator-determined rFIXFc regimens during major and minor surgery.

Forty-six major and 90 minor surgeries were performed on respectively 32 and 70 subjects with haemophilia A. rFVIIIFc was administered on the day of surgery for 44 major (including 33 orthopaedic surgeries) and 84 minor procedures. Most major (87 per cent) and minor (89 per cent) surgeries required ≤1 injection of rFVIIIFc (including loading dose) to maintain haemostasis during surgery. All haemostatic responses (where available) were assessed as excellent (major surgery: 93 per cent; minor surgery: 85 per cent) or good. No adverse events were deemed related to rFVIIIFc treatment.

Thirty-five major and 62 minor surgeries were performed on respectively 22 and 37 subjects with haemophilia B. rFIXFc was administered on the day of surgery for 35 major (including 24 orthopaedic procedures) and 55 minor procedures. Most major (82.9 per cent) and minor (87.1 per cent) surgeries required ≤1 injection of rFIXFc (including the loading dose) to maintain perioperative haemostasis. Haemostatic responses were assessed as excellent/good for all major surgeries and 95 per cent of minor surgeries. No adverse events were deemed related to rFIXFc treatment.

The safety and efficacy of prophylactic rFVIIIFc (efmoroctocog alfa) regimens in patients with severe haemophilia A has been demonstrated by the A-LONG, Kids A-LONG, and the ASPIRE extension phase 3 studies. The safety and efficacy of rFIXFc (eftrenonacog alfa) prophylaxis has been demonstrated in patients with severe haemophilia B in the B-LONG and Kids B-LONG phase 3 trials and the B-YOND extension studies.

Abstracts:

- Poster P-03: Recombinant Factor IX Fc Fusion Protein (rFIXFc) for Perioperative Haemostatic Management in Severe Haemophilia B
- Poster P-14: Recombinant Factor VIII Fc Fusion Protein (rFVIIIFc) Efficacy for Perioperative Haemostatic Management in Severe Haemophilia A

About Elocta®
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). 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 Sanofi

in the United States, Japan and Canada. It is also approved in Australia, New Zealand, Brazil and other countries, where Sanofi 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, 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). 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. It is also approved in the United States, Canada, Japan, Australia, New Zealand, Brazil and other countries where Sanofi 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 196,700 people are currently diagnosed with haemophilia 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 Sanofi collaboration
Sobi and Sanofi 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). Sanofi 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 Sanofi have optimised the technology and are the first companies to utilise it in the treatment of haemophilia. In 2014, Sobi elected to add the rFVIIIFc-XTEN-vWF fusion molecule for potential treatment of haemophilia A, to the collaboration agreement but has not yet opted-in.

About Sobi™
At Sobi, we are transforming the lives of people affected by rare diseases. As a specialised international biopharmaceutical company, we provide sustainable access to innovative therapies in the areas of haematology, immunology and specialty care. We bring something rare to rare diseases – a belief in the strength of focus, the power of agility and the potential of the people we are dedicated to serving. The hard work and dedication of our approximately 1050 employees around the globe has been instrumental in our success across Europe, North America, the Middle East, Russia and North Africa, leading to total revenues of SEK 9.1 billion in 2018. Sobi's share (STO:SOBI) is listed on Nasdaq Stockholm. You can find more information about Sobi at www.sobi.com.

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[i] World Federation of Hemophilia. Annual Global Survey 2017, published in October 2018. Available at: http://www1.wfh.org/publications/files/pdf-1714.pdf. Accessed on 9 May 2019.
[ii] World Federation of Hemophilia. About Bleeding Disorders – Frequently Asked Questions. Available at: http://www.wfh.org/en/page.aspx?pid=637. Accessed on 9 May 2019.
[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 9 May 2019.