PRESS RELEASE

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Sobi strengthens haemophilia position with potential once-weekly haemophilia A treatment

Swedish Orphan Biovitrum AB (publ) (Sobi™) (STO:SOBI) has entered into an expanded agreement with Sanofi to exercise early opt-in for the development and commercialisation of BIVV001, an investigational extended half-life factor VIII therapy with the potential to provide extended protection from bleeds with once-weekly dosing for people with haemophilia A. Sobi will make a payment to Sanofi of USD 50 million and will become a development partner in this programme. Upon potential approval in the EU, Sobi will pay a royalty on revenues in Sobi's territories and receive royalties on Sanofi revenues in its territories. Royalty rates are described below.

- Early opt-in for the BIV001 development programme, creating a development partnership with Sanofi
- BIVV001 is an investigational once-weekly haemophilia A replacement therapy with the potential to deliver a new standard of care in personalised treatment
- BIVV001 phase 3 pivotal trial anticipated to start later this year
- As part of the extended Sanofi agreement, a new supply agreement now in place until 2027

"The accelerated opt-in demonstrates our commitment to haemophilia and our confidence in BIVV001," says Guido Oelkers, Sobi President and CEO. "Our position as development partners brings us closer to the haemophilia community. "We strive to provide access to treatments that bring factor levels closer to normal. We are delighted at the prospect of bringing this potential new treatment to patients in our territory, in our efforts to enable patients to live their lives beyond their haemophilia."

Professor John Pasi, Haemophilia Centre Director at Barts Health NHS Trust and Professor of Haemostasis and Thrombosis at Queen Mary University of London, says the phase 1/2 results for BIVV001 have been encouraging. "The therapy has the potential to further develop haemophilia A treatment and enable personalised therapy to achieve potentially near-normal factor levels for a part of the week, with physiological benefits that this may bring to patients in the short and longer term," he says.

In 2014, Sobi elected to add the BIVV001 development programme to the collaboration agreement . BIVV001 (rFVIIIFc-VWF-XTEN) is the first investigational factor VIII therapy to break through the von Willebrand factor ceiling in haemophilia A and is designed to provide extended protection from bleeds with once-weekly prophylactic dosing. Data from the BIVV001 phase 1/2a study was presented at the 27th congress of the International Society on Thrombosis and Haemostasis (ISTH), in July 2019, and underscored the potential for once-weekly dosing with sustained high factor levels in

haemophilia A. BIVV001 was granted orphan drug designation by the US Food and Drug Administration in August 2017 and the European Commission in June 2019.

In the collaboration agreement with Sanofi, Sobi holds the commercial rights for joint haemophilia programmes for Europe, North Africa, certain countries in the Middle East, and Russia (Sobi's territory). In connection with the expansion of the collaboration agreement, a new supply contract with Sanofi until 2027 regarding Elocta and Alprolix has been agreed, with the potential for expansion to include BIVV001.

Payment and royalty structure

Upon EU regulatory approval of BIVV001, Sobi will be liable to pay the balance of the development costs incurred for BIVV001. The total payment obligation is currently estimated to reach approximately USD 280-290 million (including the USD 50 million payment).

The royalty structure under the agreement states that Sobi will pay Sanofi 9 per cent of direct sales in the Sobi territory, and Sanofi will pay Sobi 8 per cent of direct sales in North America and 13 per cent of direct sales in other markets.

About BIVV001

BIVV001 (rFVIIIFc-VWF-XTEN) is a novel and investigational recombinant factor VIII therapy that is designed to extend protection from bleeds with once-weekly prophylactic dosing for people with hemophilia A. BIVV001 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. BIVV001 was granted orphan drug designation by the US Food and Drug Administration in August 2017 and the European Commission in June 2019.

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 1,300 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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