sobi
Pioneer in Rare Diseases

Geoffrey McDonough

Stockholm | 4 May 2017

A strong platform for new and transformative treatments

Forward looking statements

In order to utilize the 'Safe Harbor' provisions of the United States Private Securities Litigation Reform Act of 1995, Swedish Orphan Biovitrum is providing the following cautionary statement. This presentation contains forward-looking statements with respect to the financial condition, results of operations and businesses of Swedish Orphan Biovitrum. By their nature, forward-looking statements and forecasts involve risk and uncertainty because they relate to events and depend on circumstances that will occur in the future. There are a number of factors that could cause actual results and developments to differ materially from that expressed or implied by these forward-looking statements. These factors include, among other things, the loss or expiration of patents, marketing exclusivity or trade marks; exchange rate fluctuations; the risk that R&D will not yield new products that achieve commercial success; the impact of competition, price controls and price reductions; taxation risks; the risk of substantial product liability claims; the impact of any failure by third parties to supply materials or services; the risk of delay to new product launches; the difficulties of obtaining and maintaining governmental approvals for products; the risk of failure to observe ongoing regulatory oversight; the risk that new products do not perform as we expect; and the risk of environmental liabilities.

Dedicated to rare diseases

- Our key therapeutic areas are **Inflammation, Genetic diseases** and **Haemophilia**
- We deliver products to specialist physicians and their patients through **our integrated team approach** to sales and marketing, medical affairs and patient access
- We leverage our **world-class capabilities in protein biochemistry and biologics development and supply** to develop next generation biological products

Our vision

We are inspired to pioneer a world in which rare disease patients are diagnosed at birth, receive effective and sustainable therapy, and go on to live full and healthy lives

The Sobi way
– lessons from two decades of sustainable transformation

93%

2 YEAR SURVIVAL
WITH DX < 2 YEARS OLD

29%

2 YEAR SURVIVAL
WITH DX < 2 YEARS OLD

1000

PEOPLE TYROSENEMIA
ARE GROWING UP
AROUND THE WORLD

OUR DREAM IS THAT THEY ONE DAY BECOME

GRAND-PARENTS

Birth

Childhood

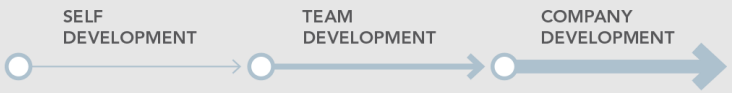
Adolescence

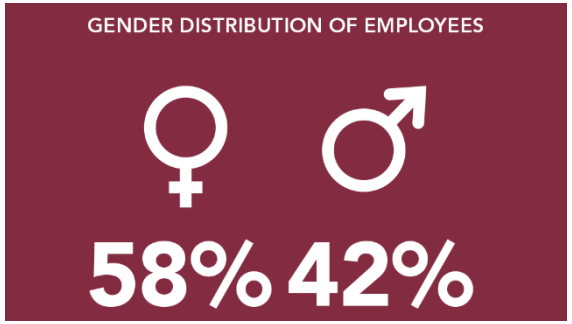
Mid-life

Mature adulthood

Our employees – our most important asset

- Our culture is our strategy → acting on our values to build value for stakeholders
- Actions based on employee feedback
- Aligning compliance with our corporate principles
- A culture that promotes the open discussion of business ethics
- Labour rights
- Diversity and development





Building a sustainable future

2011

REVENUE
SEK 1,911 M

Sobi established in the Middle East and the US

2012

REVENUE
SEK 1,923 M

- Operational focus on key therapeutic areas.
- Pivotal phase 3 studies completed and paediatric trials initiated for Elocta and Alprolix.
- Research agreement signed with Affibody AB.
- ReFacto agreeement extended and Nordic commercial rights sold to Pfizer.

Expanded partner portfolio with Xiapex® and Pharmaswiss portfolio

2013

REVENUE
SEK 2,177 M

- New momentum in operating business.
- Aquired full rights for Kineret and received approval for CAPS indication in the EU.

Eloctate® and Alprolix launched in the US by our partner Bioverativ

2014

REVENUE
SEK 2,607 M

- Positive contributions from all parts of the business.
- Development pipeline setbacks as Kiobrina phase 3 study did not meet primary end-points.
- Orfadin direct sales in the US.

Elocta approved in the EU

2015

REVENUE
SEK 3,228 M

- Haemophilia launch organisation in place.
- Implemented new distribution model for Kineret in the US.

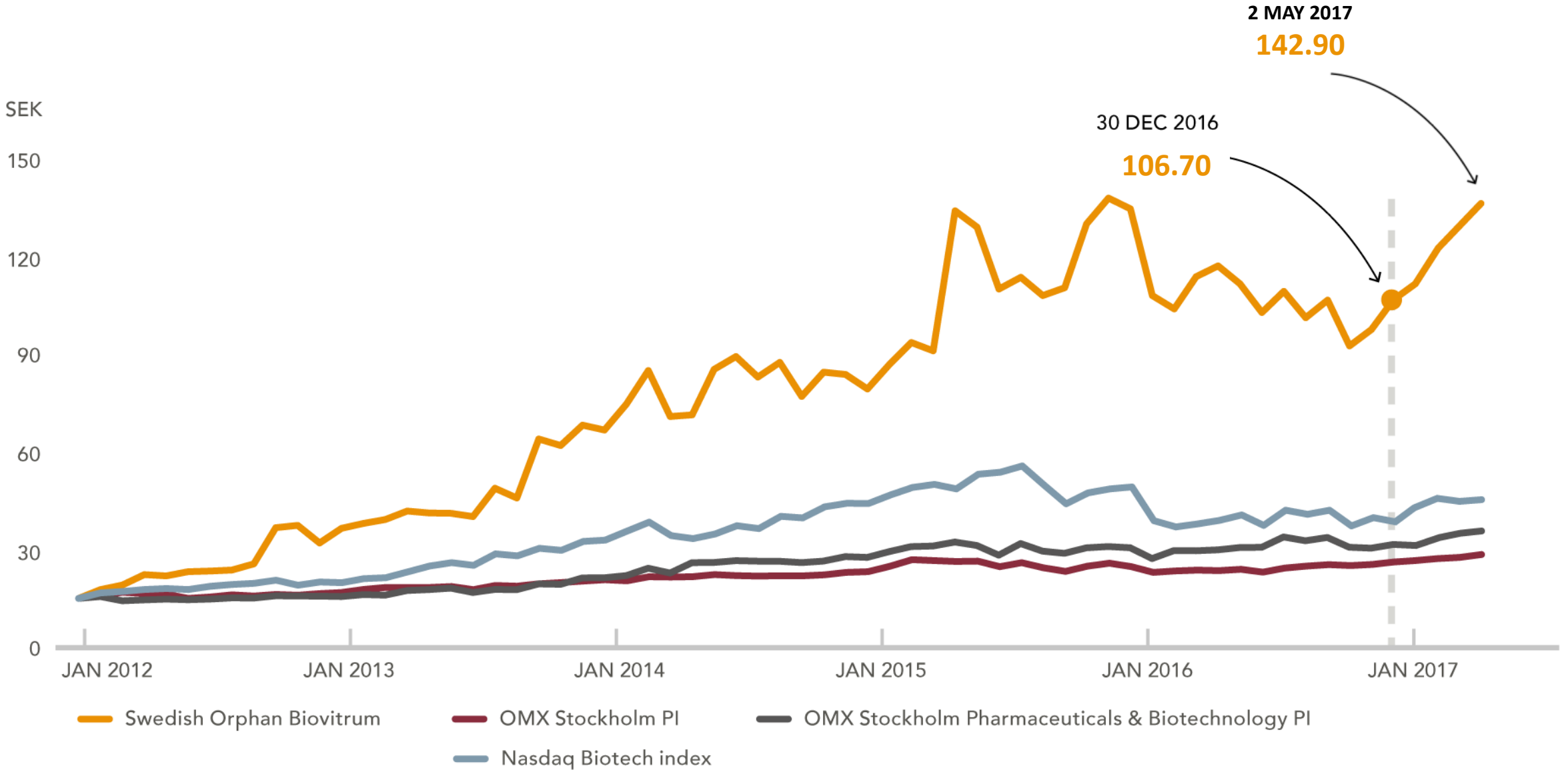
2016

REVENUE
SEK 5,204 M

Launch of Elocta and Alprolix

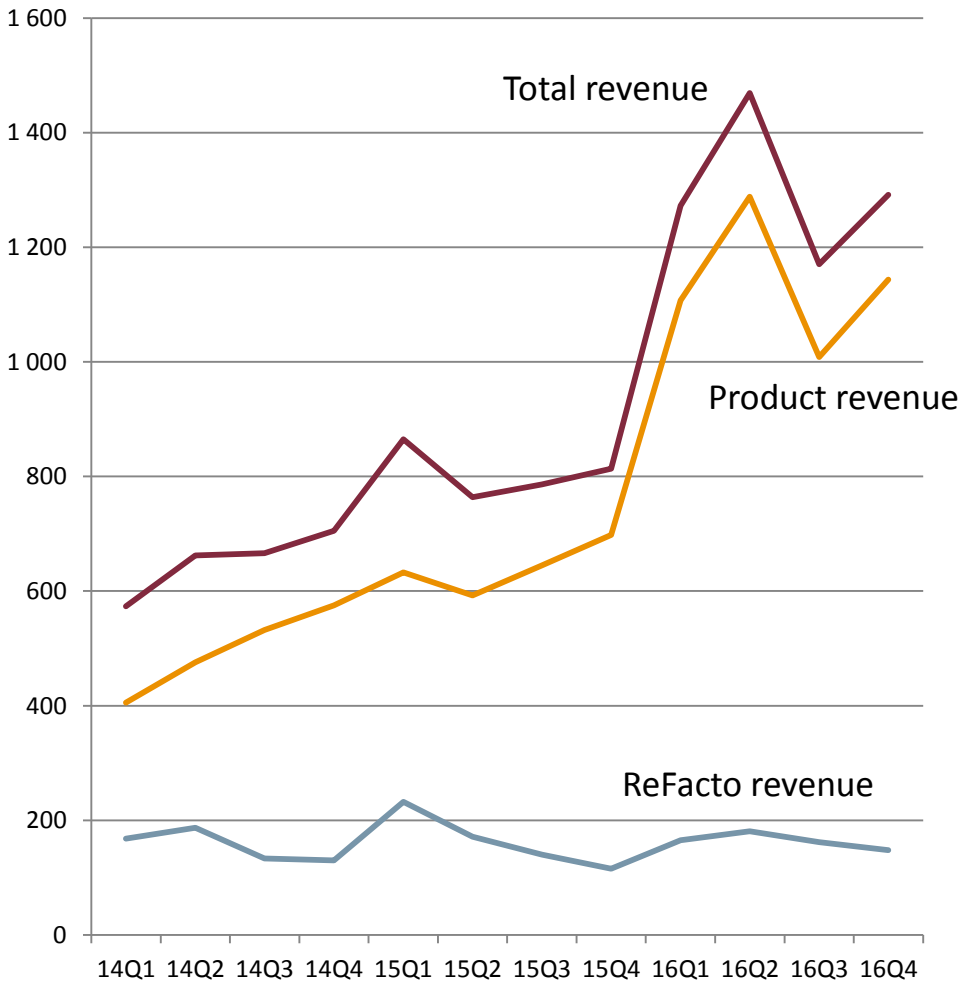
- Alprolix approved in the EU.
- Launch of two next-generation haemophilia products – Elocta and Alprolix in Europe and the Middle East.
- Pipeline strengthened with two candidate drugs in early stage development and new planned clinical programmes.
- ReFacto agreeement extended to 2023.

Share development 2012 – 2 May 2017



Financial highlights FY 2016

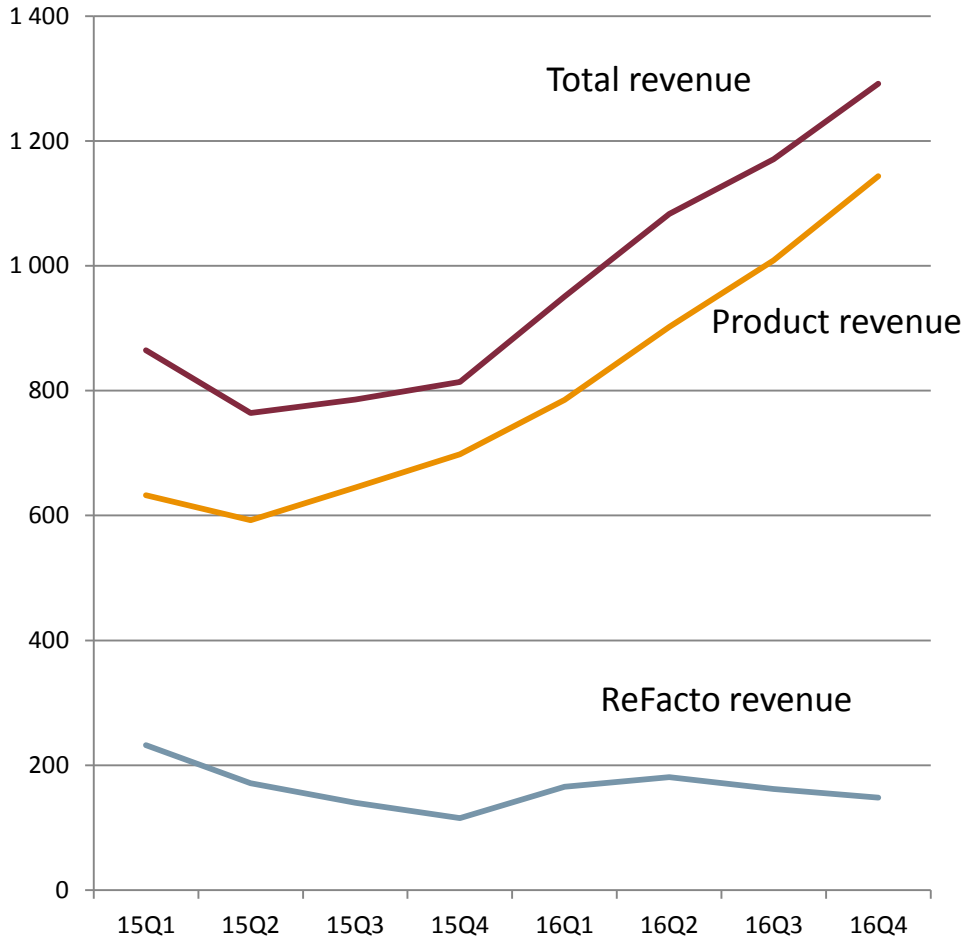
- Total revenue: SEK 5,204 M (3,228)
  - 61% growth
- Product revenue: SEK 4,548 M (2,568)
  - 77% growth
- ReFacto revenue: SEK 656 M (660)
- Gross margin: 70% (62%)
- EBITA: SEK 1,543 M (433)
- Cash flow operations: SEK 343 M (507)



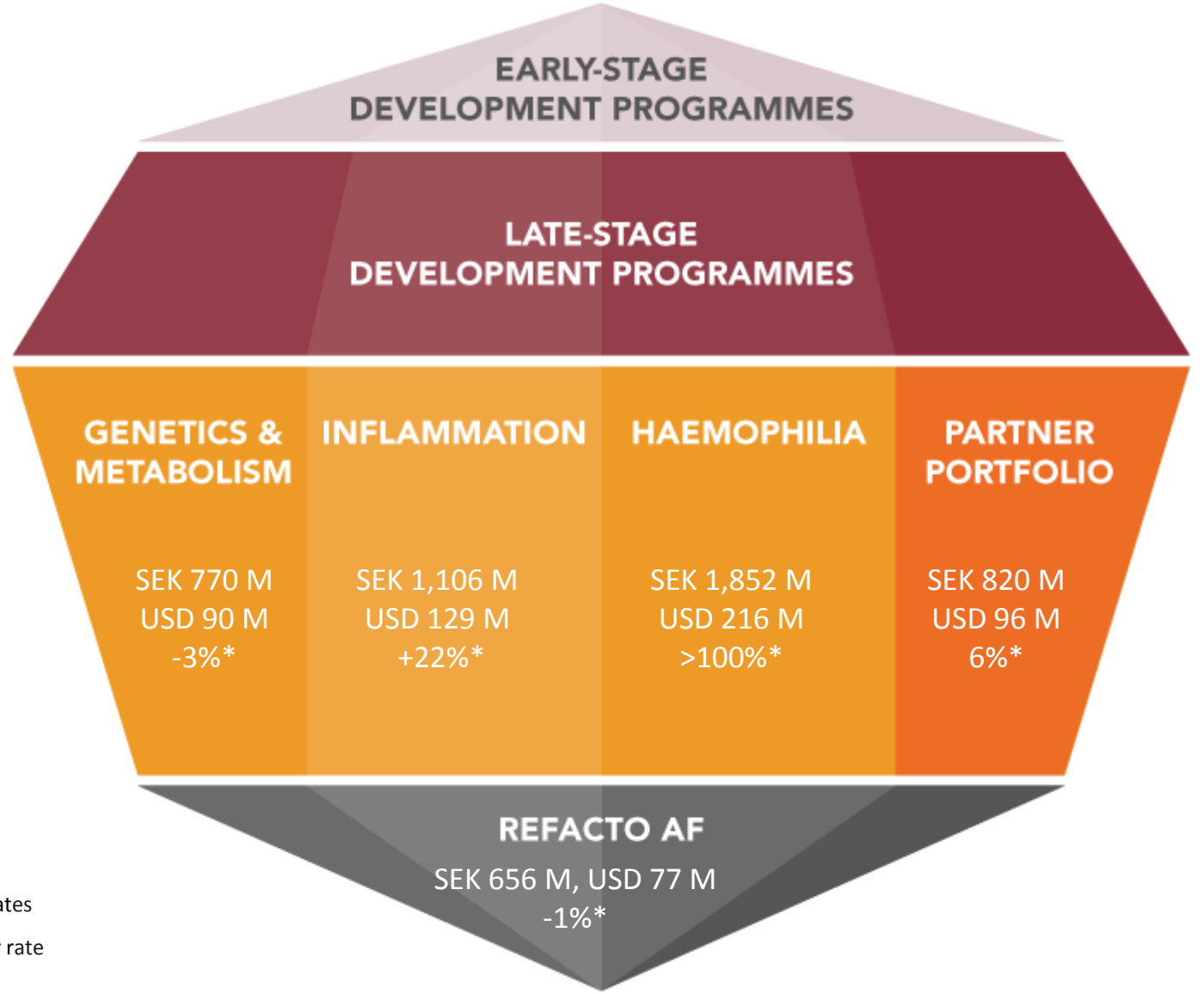
Financial highlights FY 2016

[excluding one-time credits of SEK 708 M]

- Total revenue: SEK 4,496 M (3,228)
  - 39% growth
- Product revenue: SEK 3,840 M (2,568)
  - 50% growth (49% at CER)
- ReFacto revenue: SEK 656 M (660)
- Gross margin: 66% (62%)
- EBITA: SEK 835 M (433)
- Cash flow operations: SEK 343 M (507)



Revenues by business line FY 2016

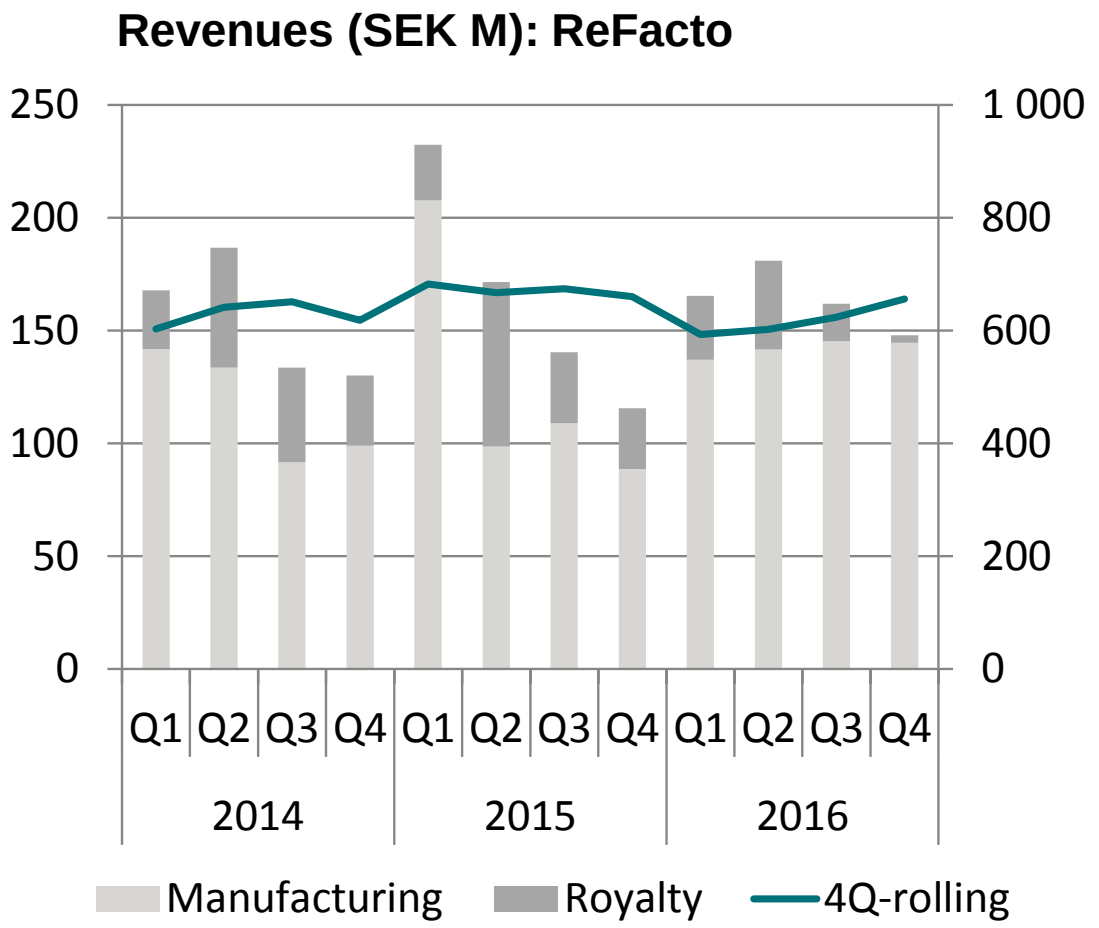


*Growth at Constant Exchange Rates

USD 1 = SEK 8,5613 (average year rate

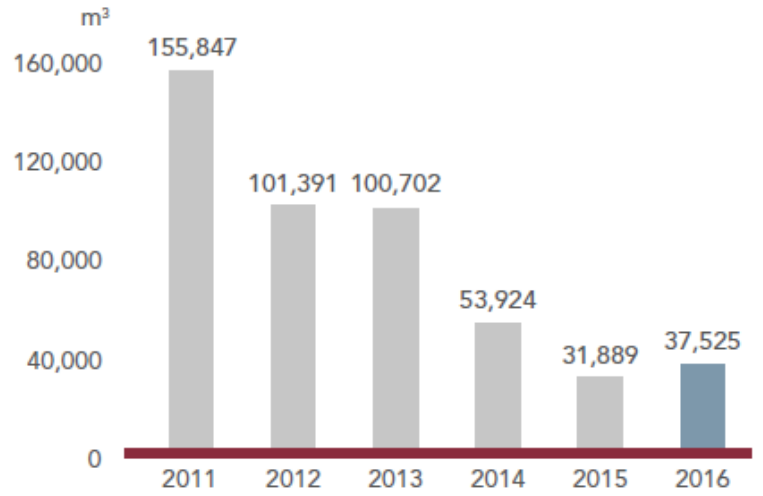
ReFacto manufacturing

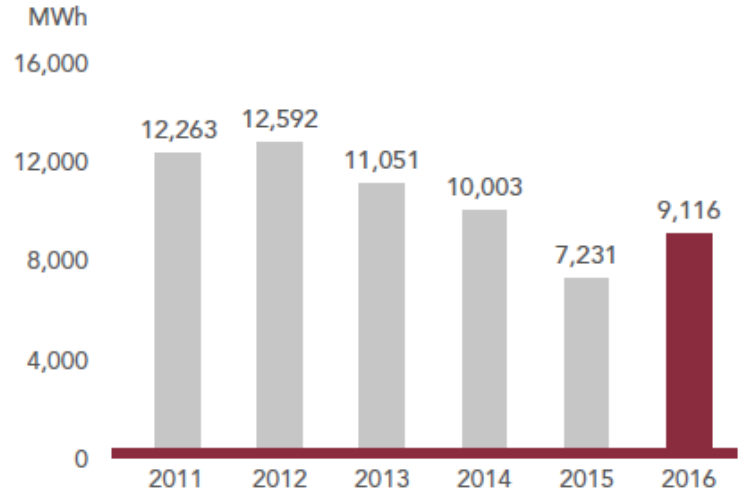
- FY revenue was SEK 656 M (660)
- New manufacturing agreement with Pfizer



Decreased relative energy and water consumption

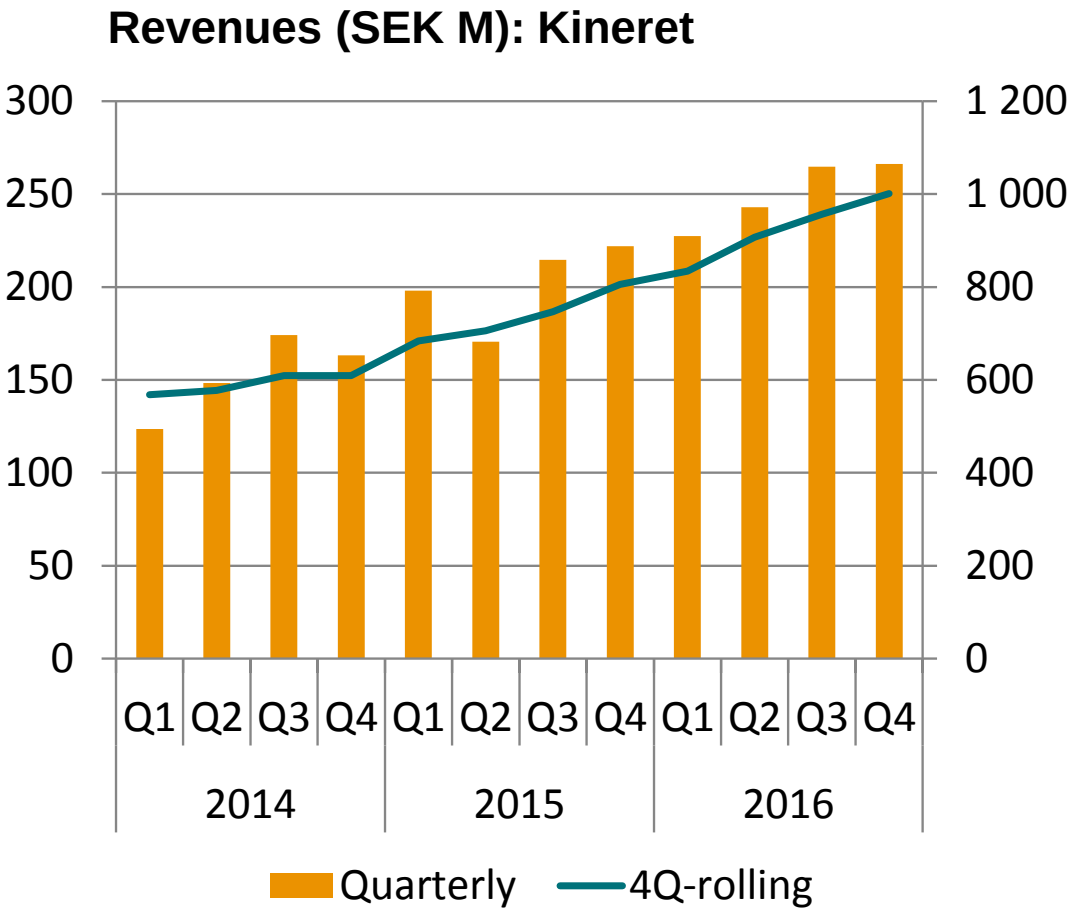
- Ongoing project to continuously improve the energy efficiency of our site
- Regular monitoring of operating costs of the buildings





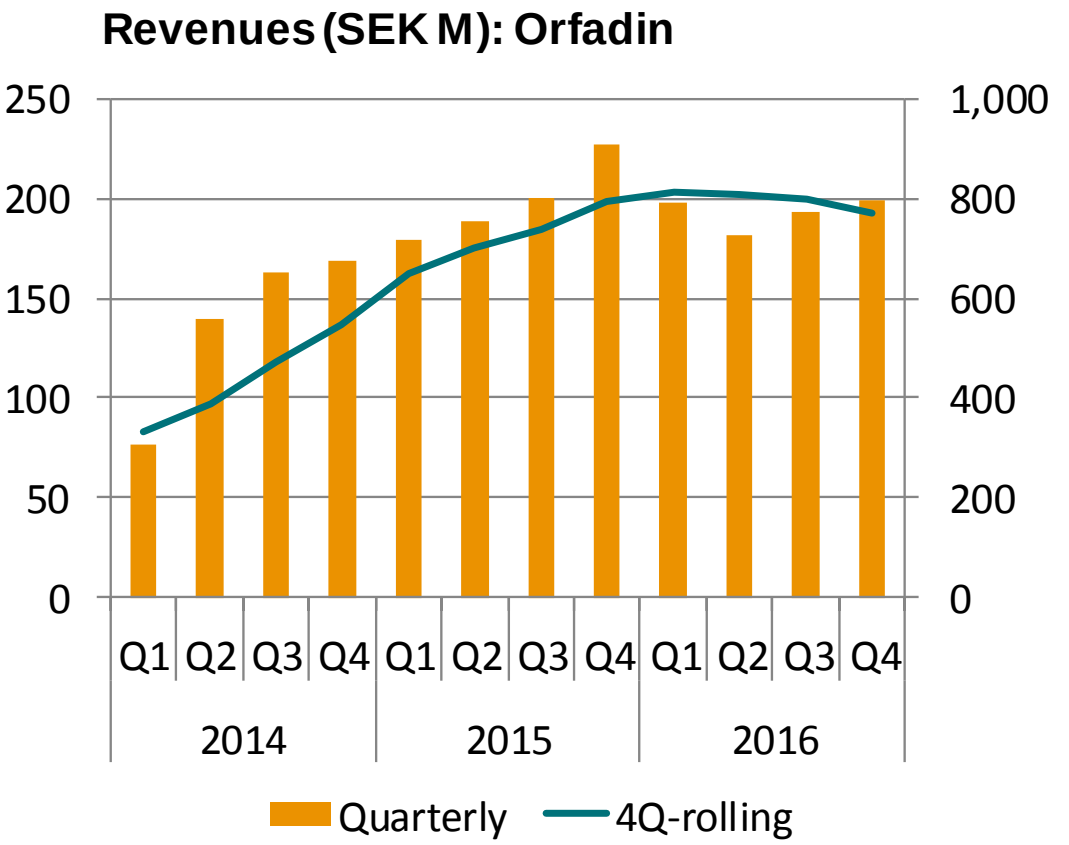
Kineret

- FY revenue SEK 1,001 M (805)
  - Increase of 24%
- US distribution model and patient support programme is driving growth
- Clinical programmes on track



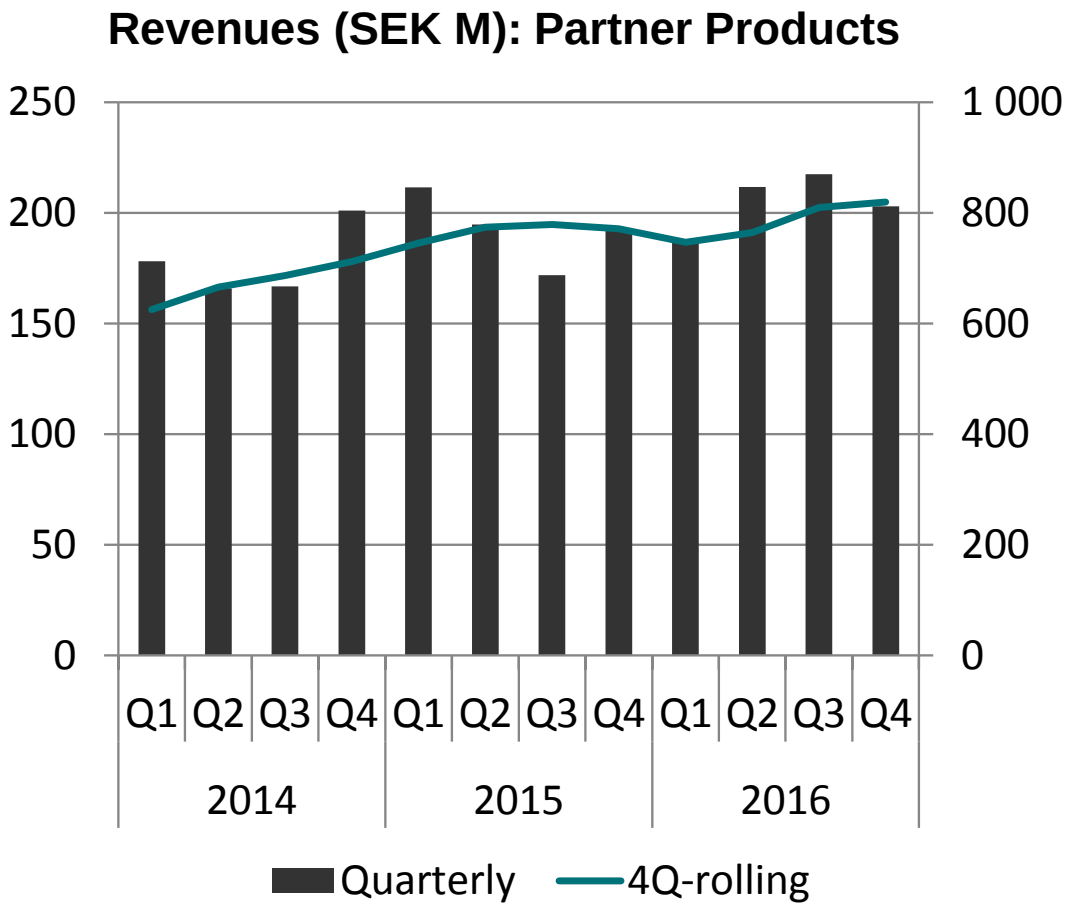
Orfadin

- FY revenue SEK 770 M (796)
  - Decrease of 3%
- North America
  - Performance in the US supported by 20 mg and oral suspension launches
  - Orfadin approved in Canada
  - Revenue in Canada negatively impacted by generic approvals
  - Revenue in EMENAR was negatively impacted by the approval of a generic formulation in Turkey



Partner Products

- FY revenues SEK 820 M (771)
  - Increase of 6%
- Revenue growth supported by new partnership with PharmaSwiss and Xiapex
- Sobi in discussions regarding a possible sale of Partner Products



Partner Products
– a business model based on sustainable access

- The European market consists of nearly 40 countries
- A cost-efficient platform for the provision of products for a limited number of patients is important, and has a direct impact on the availability of niche medications to European patients.

PARTNERS

Get access to strategic thinking, management and cost-effective solutions via a mature and experienced platform spanning the region.

PHYSICIANS

Meet a reliable and experienced partner in Sobi with a dedicated patient-centric approach.

PATIENTS

Receive access to innovative, new treatments.

BUDGET-HOLDERS & HEALTHCARE SYSTEMS

Provided with robust dossiers in order to evaluate treatments and include them in local healthcare systems.

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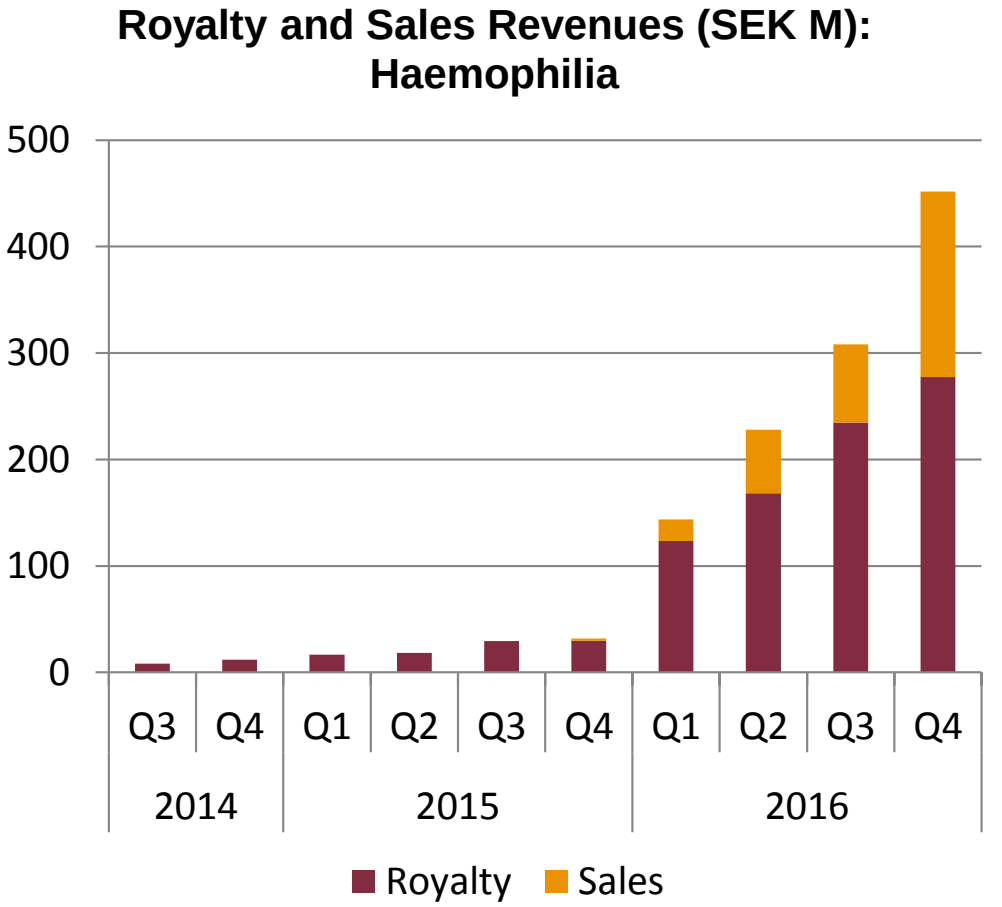
Lower risk and spread investments.

Haemophilia

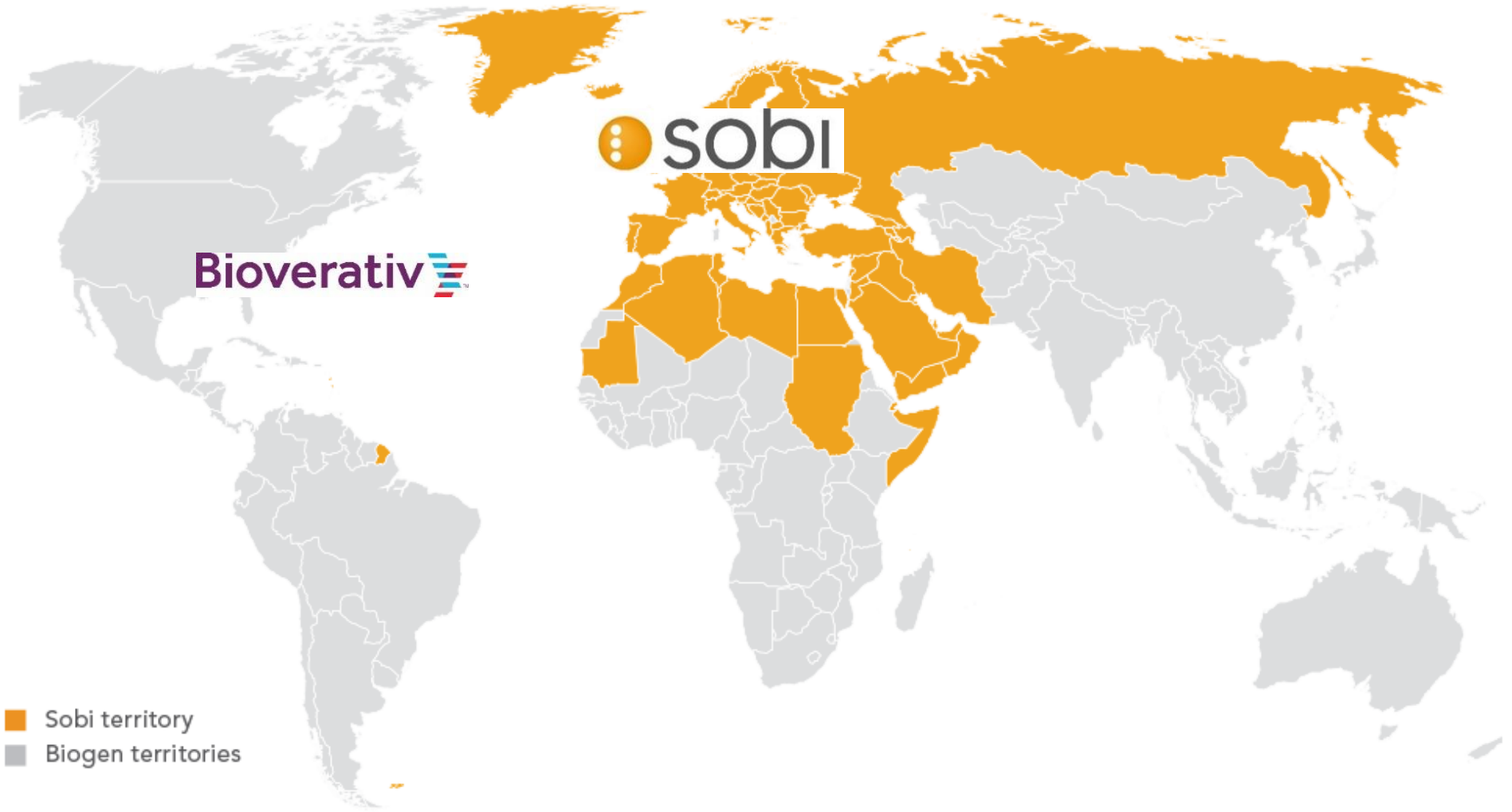
Haemophilia

- FY revenue of SEK 1,853 M (96)
  - SEK 327 M (2) in sales
  - SEK 803 M (95) in royalty revenue
  - SEK 708 M in one-time credits
  - SEK 14 M in milestone revenue

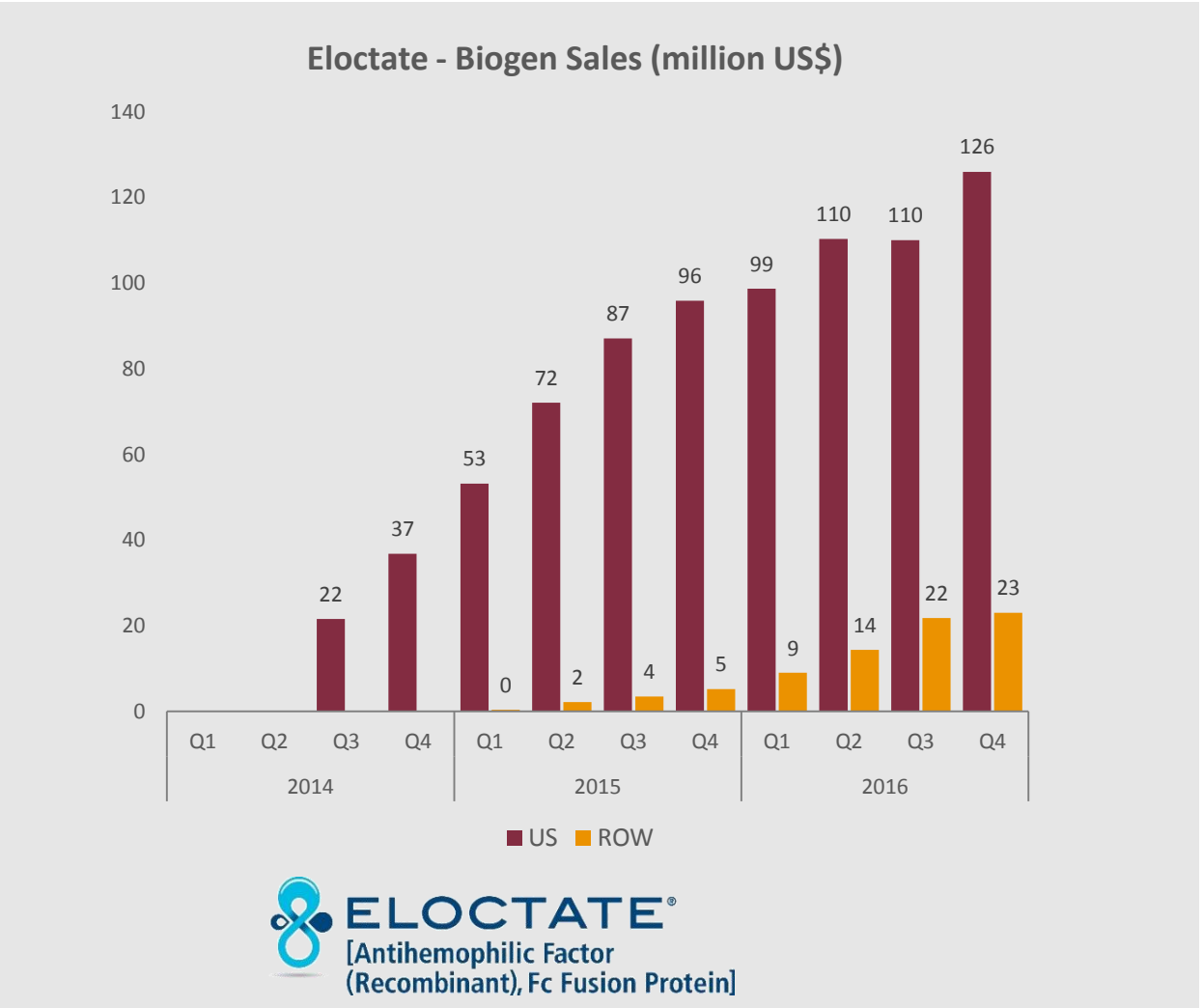


* Excluding one-time royalty credits and milestone revenues

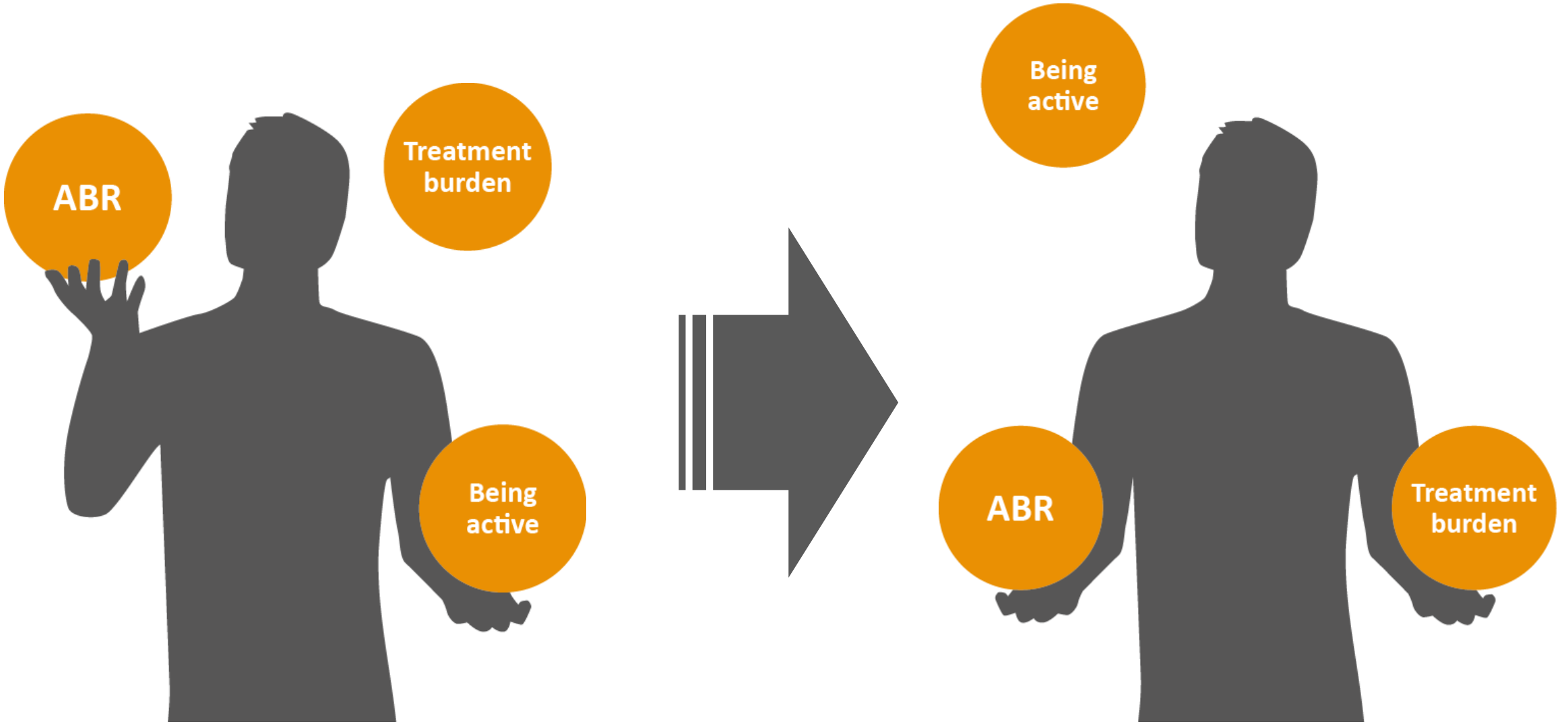
The Sobi and Bioverativ collaboration



Haemophilia – Bioverativ revenues

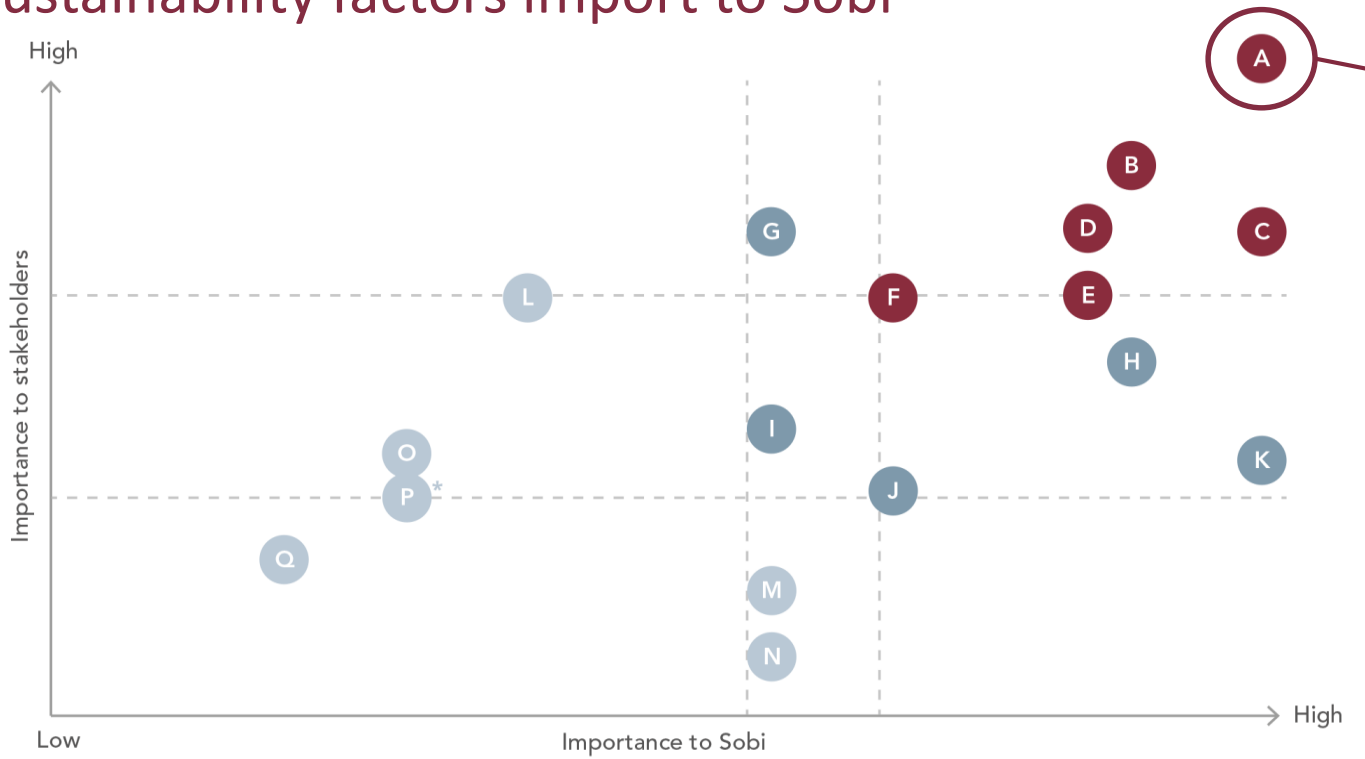


Redefining protection in haemophilia



Top Priority: Access to medicines + human rights

Sustainability factors import to Sobi



- **A** Access to healthcare and medicines
- **B** Sustainable and secure supply-chain
- **C** Responsible marketing and sales activities
- **D** Ethics in research
- **E** Engagement with patient groups
- **F** Research and development incl pipeline-programs
- **G** Regulatory and legal environment
- **H** Product safety and quality
- **I** Anti-corruption
- **J** Diversity and equal opportunity
- **K** Sustainable workforce
- **L** Environmental management regarding water, chemicals and pharmaceuticals
- **M** Anti-competetive practices
- **N** Protecting personal information
- **O** Counterfeit drugs prevention
- **P** Greenhousegas emissions
- **Q** Tax transparency and responsibility

"Persons living with a rare disease tend to remain a marginalised and invisible population, with little information available about their diseases and very few treatment options. They suffer inequality in accessing healthcare services and treatment, and in the prices they have to pay, due to their social status or their country of origin."

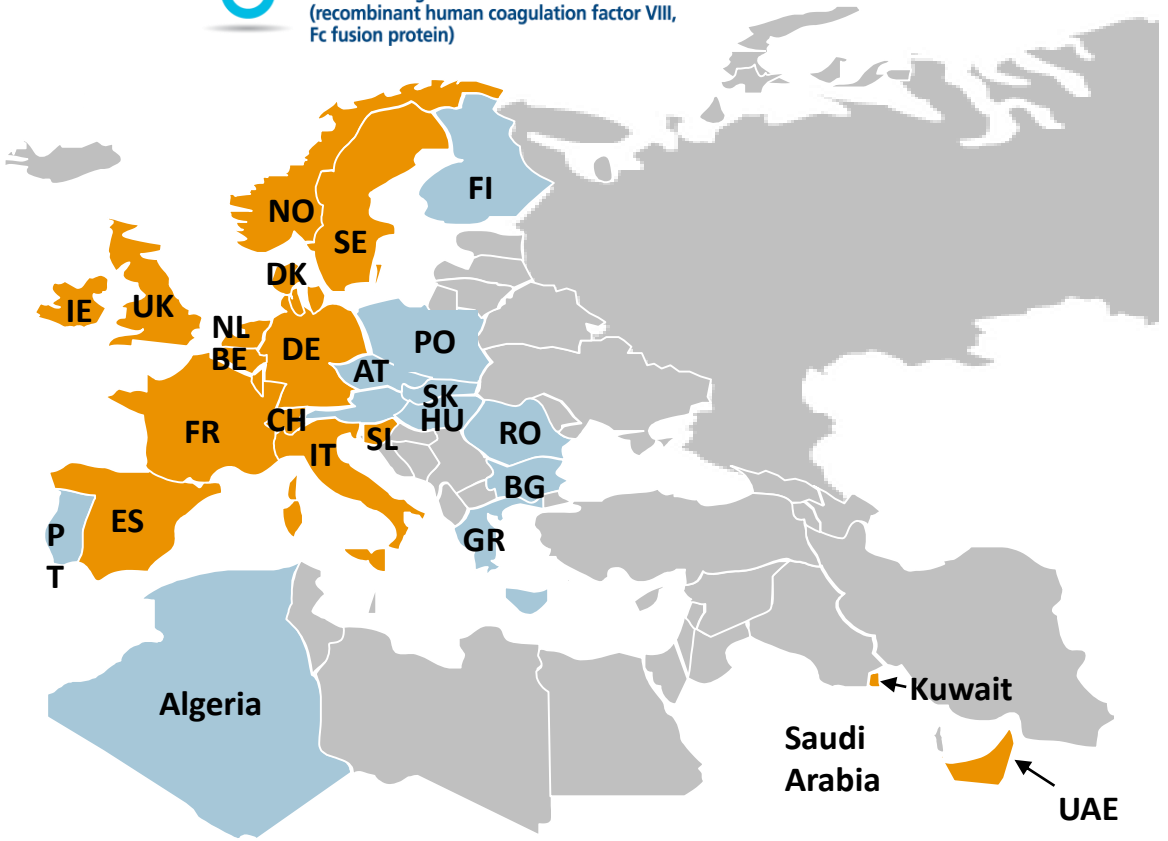
The NGO Committee for Rare Diseases (United Nations, New York)

Dialog with stakeholders to ensure sustainable access

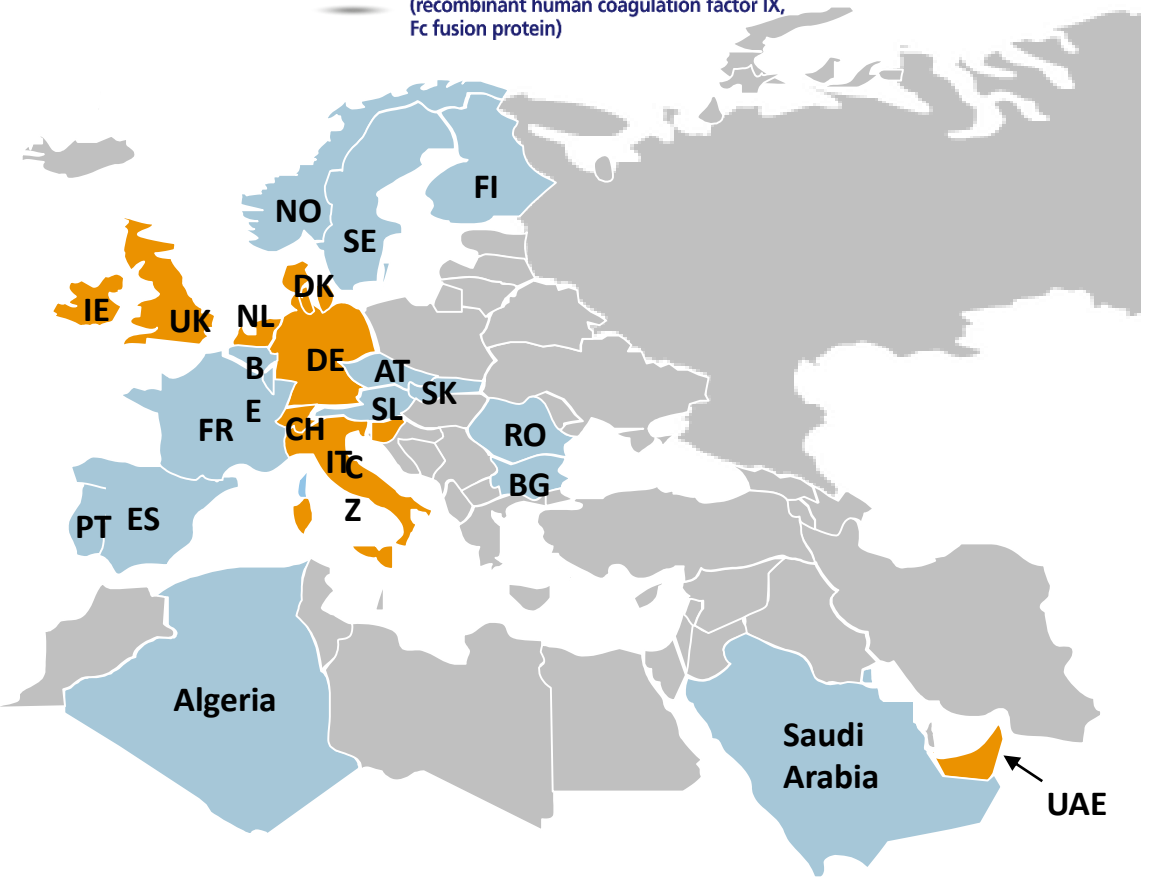
- Annual pricing is comparable to existing recombinant factors; often lower based on consumption savings
- Average time to reimbursement of 4.5 months across first 8 markets

Elocta & Alprolix reimbursement status Q1 2017

ELOCTA®
efmoroctocog alfa
(recombinant human coagulation factor VIII, Fc fusion protein)



ALPROLIX®
eftrenonacog alfa
(recombinant human coagulation factor IX, Fc fusion protein)

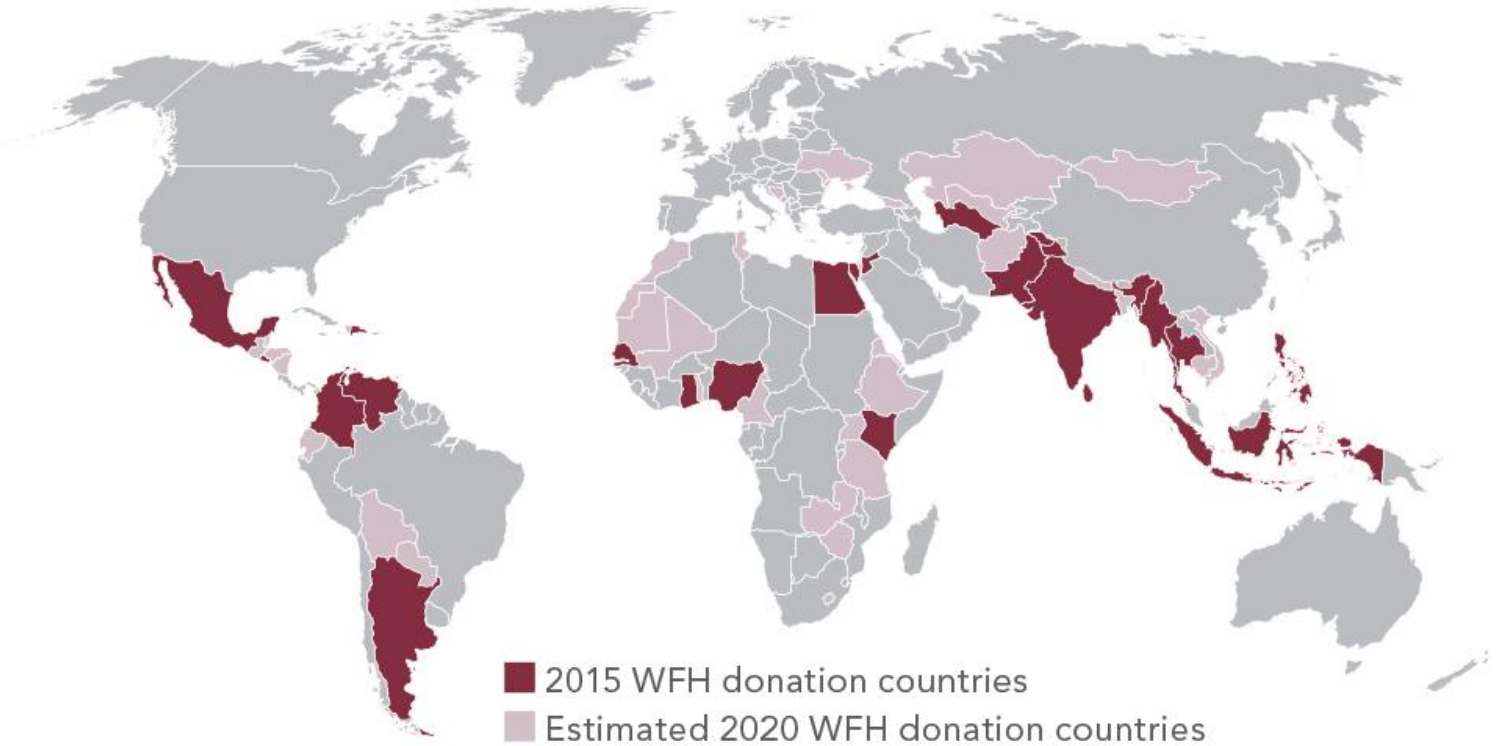


Reimbursement granted – treatment available

Pricing & reimbursement decision-making in process

Commitment to the global Haemophilia community

- Supporting leadership programmes, youth fellowship, and new technologie workshops in collaboration with the European Haemophilia Consortium (EHC)
- To provide predictable, sustainable access to people with haemophilia in developing healthcare systems
- Regular donations through WFH
- Emergency aid through Direct Relief



One billion international units over ten years*

*Donation programme started in July 2015

Reaching patients in 40 countries

Donation statistics July 2015 to December 2016

Humanitarian Aid reaching patients worldwide

188 MIU
IUs of clotting factor donated

14,800
People treated in 40 countries

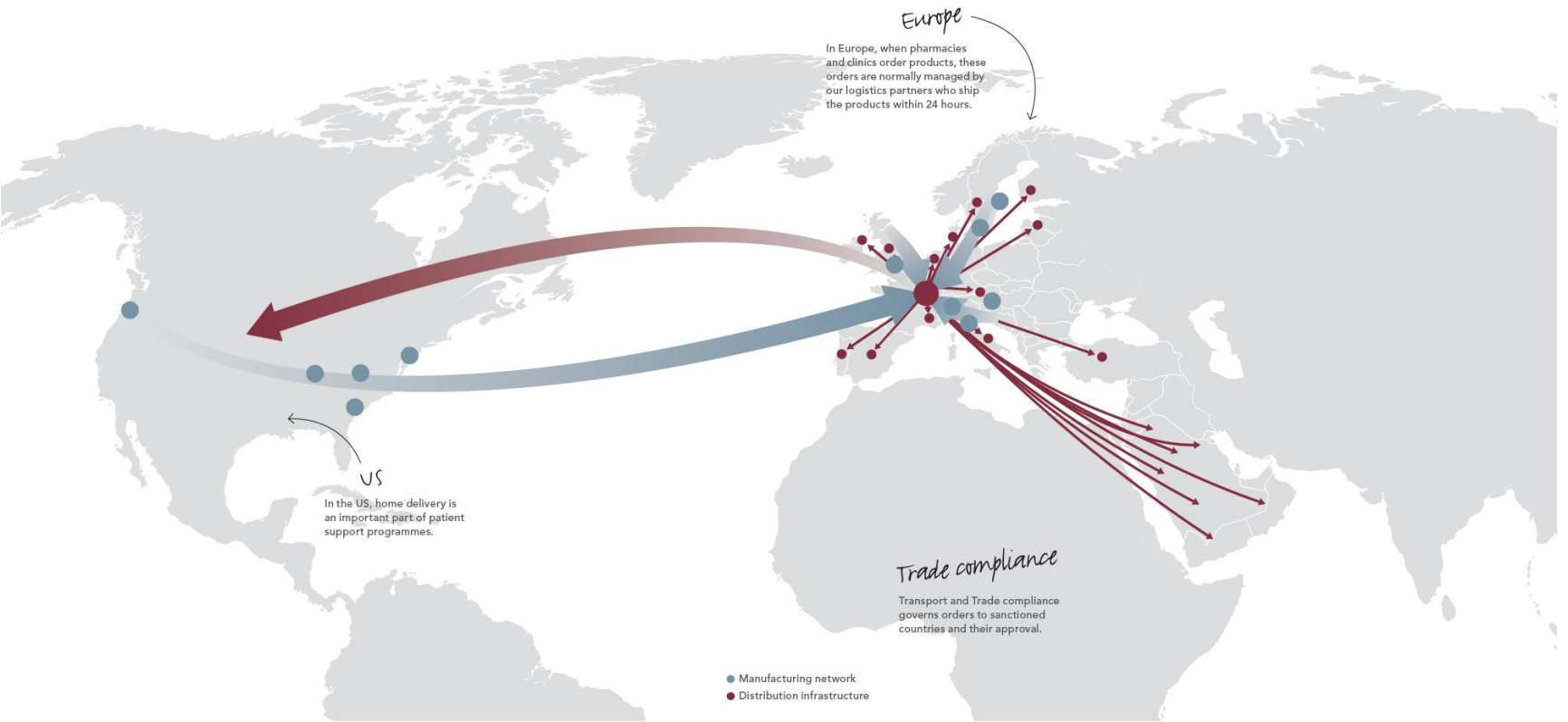
12,757
Acute bleeds
893 life-saving

695
Surgeries
70 life-saving

Percentage of paediatric patients receiving treatment in these countries has nearly doubled
(from 14% to 28%)

Supply chain security and sustainability

- We distribute to 60 counties world wide
- Serialisation project to ensure that products are not falsified
- Sobi has implemented due diligence-processes to reduce the risk for corruption and bribery



Pipeline

Our pipeline projects

Therapeutic area/Indication	Product/ Project	Pre-clinical	Phase 1	Phase 2	Phase 3
Haemophilia A	Elocta/A-SPIRE*				
Haemophilia A	Elocta/PUP[1]*				
Haemophilia B	Alprolix/B-YOND*				
Haemophilia B	Alprolix/PUP[1]*				
Acute gout	Kineret/anaGO				
Still's disease	Kineret/anaSTILLS				
Alkaptonuria	Orfadin/SONIA2				
MPSIIIA	SOBI003				
Anti-C5	SOBI005				
Anti-IL-1	SOBI006				

*Extension trial for an already approved indication

[1] PUP = Previously untreated patients

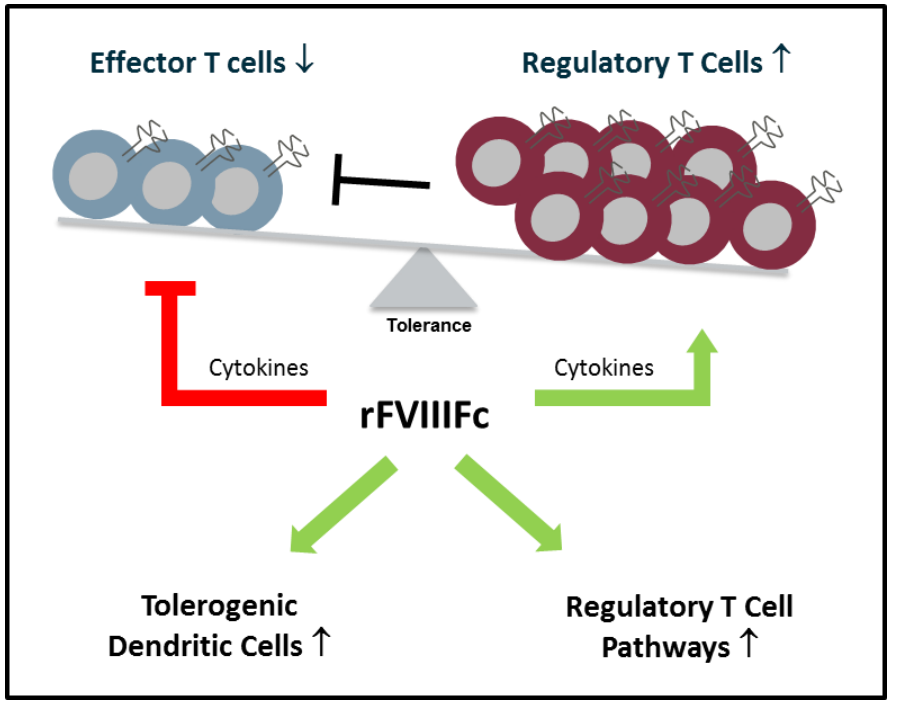
Fc fusion and immunoregulation

Initial preclinical data provides the scientific rationale for investigating rFVIIIFc in ITI[1]

- **Fc-fused antigens** have been implicated in the regulation of immune responses and Fc-fused proteins may have a role in promoting tolerance
- In mice, rFVIIIFc resulted in **significantly lower** antibody responses to rFVIII compared to other commercial rFVIII products
- rFVIIIFc at therapeutic levels appears to **induce tolerance to rFVIII** and up-regulates key tolerance-related proteins and cytokines in hemophilia A mice

1. Krishnamoorthy et al. Cell Immunol 2016

Working model for proposed mechanism of action of rFVIIIFc in induction of immune tolerance to rFVIII



Adapted from Krishnamoorthy et al. *Cell Immunol* 2016

Note: Immune Tolerance Induction is currently not an approved indication for Elocta

Sobi and Bioverativ will investigate Elocta/ELOCTATE for Immune Tolerance Induction in 2017

‘First ITI attempt’ Study

Objective: To investigate rFVIIIFc for ITI in patients with severe haemophilia A with inhibitors undergoing ITI for the first time

‘Rescue ITI’ Study

Objective : To investigate rFVIIIFc for ITI in patients with severe haemophilia A with inhibitors who have failed previous ITI therapies

Both studies expected to start in mid-2017

Note: Immune Tolerance Induction is currently not an approved indication for Elocta

Bioverativ to advance BIVV001 in H2 2017

Investigated to extend haemophilia A prophylaxis to once weekly or less frequent dosing

- Plans to advance to clinic in H2 2017
- Bioverativ development programme
- Sobi has opt-in right at EU filing

BIVV 001 has Potential to Extend Hemophilia A Prophylaxis to Once Weekly or Less Frequently

rFVIIIFc-VWF-XTEN

Technology

Uniquely engineered factor VIII molecule with a region of Fc dimer, VWF, and XTEN polypeptides

- Fc monomer, like Elo, enables recycling to extend time in circulation
- D'D3 inhibits binding to VWF which limits the ceiling for current FVIII products
- XTEN insertions increase half-life by protecting from clearance/proteolysis

Potential Clinical Profile

Potential to enable prophylaxis intervals in Hemophilia A of **once weekly or less frequent dosing**

Competitive Positioning

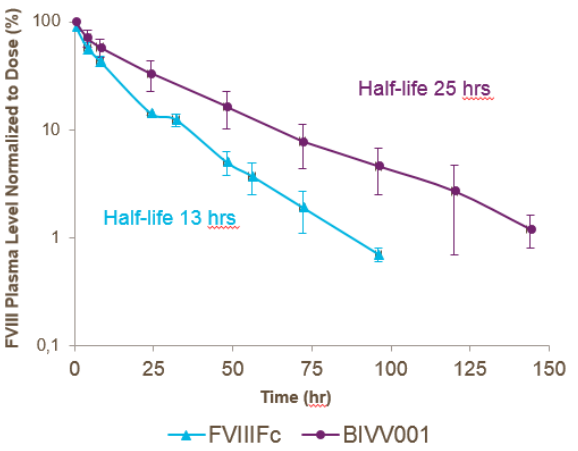
Only next gen FVIII molecule that can potentially achieve goal of weekly prophy dosing as it removes the 1/2 life limit found with all other EHL products caused by their binding to VWF. Known biology, MOA can potentially achieve full correction to stop bleeds and use in surgery

Other novel MOAs will require bypass agents or factor to treat bleeds

Timing

Intend to move into the clinic in H2 2017

Improved PK Profile of Intravenously Delivered BIVV 001 in Cynomolgus Monkeys



BIVV 001 showed 2-fold improvement in pharmacokinetic property compared to rFVIIIFc in cyno monkeys

Note: BIVV 001 is currently BIIB073, XTEN technology licensed from Amunix

Bioverativ | Integrated Capabilities | Talented Team | Strong Hemophilia Franchise | Well Capitalized to Advance Pipeline | Confidential and proprietary | 1

Bioverativ to advance BIVV002

Investigated to enable subcutaneous administration

- Working to advance programme towards the clinic
- Bioverativ development programme
- Sobi has opt-in right at EU filing

BIVV 002 Designed to Enable Subcutaneous Administration

rFIXFc-XTEN

Technology

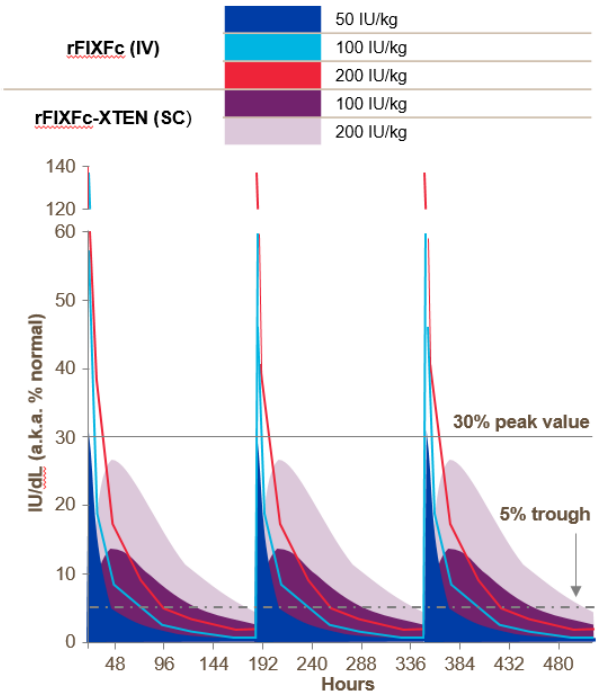
Combines Fc dimer and XTEN technology along with R338L Padua Factor IX variant in the treatment of Hemophilia B

Potential Clinical Profile

Potential to provide increased levels of protection with **subcutaneous dosing**. Leverages natural pathway with Fc fusion

Competitive Positioning

Potentially the only subcutaneous rFIX and enables a minimum of once weekly prophylactic dosing

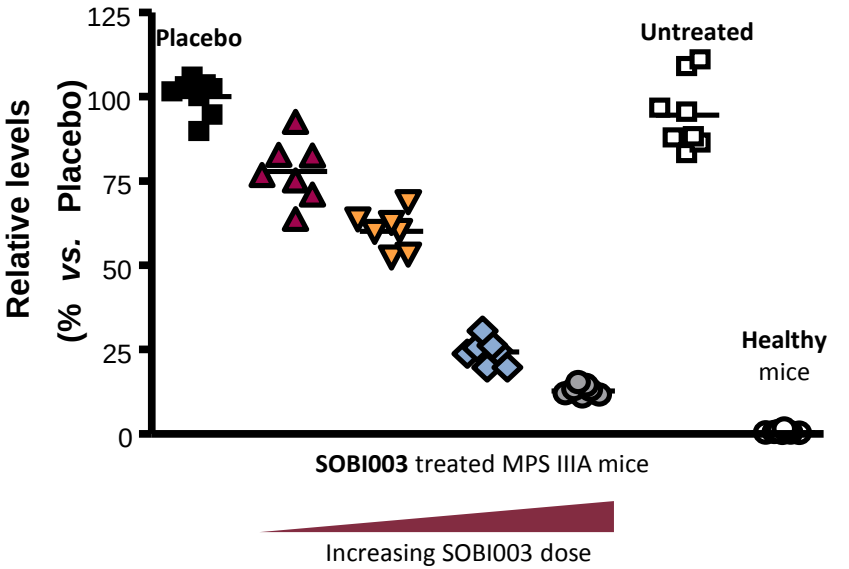


Note: BIVV 002 is currently BIIB085, XTEN technology licensed from Amunix
Source: Preliminary modeling Arjan van der Flier & Qin Weng, DMPK

Bioverativ | Integrated Capabilities | Talented Team | Strong Hemophilia Franchise | Well Capitalized to Advance Pipeline | Confidential and proprietary | 3

SOBI003 for MPS IIIA
- Systemic ERT with CNS penetration

- MPS IIIA is a Lysosomal Storage Disease – heparan sulfate accumulates and causes significant CNS morbidity and mortality
- SOBI003 is a recombinant human sulfamidase enzyme which has been modified to facilitate CNS uptake
- Orphan designation for SOBI003 was granted by the EC (Oct 2016)

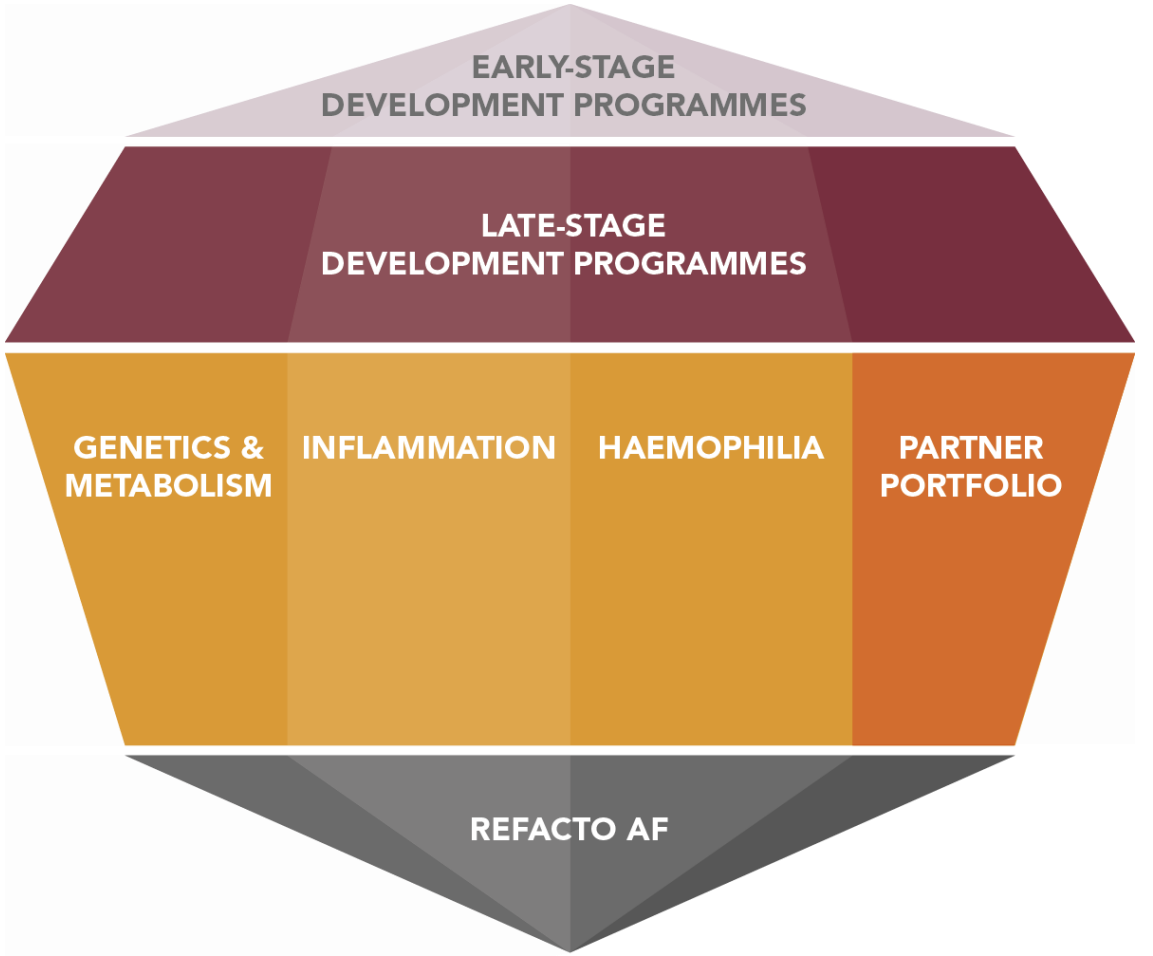


Dose dependent reduction of tetrasaccharide storage in the brain of MPS IIIA mice following treatment with SOBI003

Building our future

Strong focus on our business and capabilities within rare diseases

1. Diverse, growing, and profitable base business in Europe and North America focused on rare diseases
2. Launching first-to-market long-acting haemophilia factors in Sobi territory – providing forward cash flow to continue to build company
3. Growing the business organically with new partner products, and with a pipeline of early stage rare disease biologics

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