A strong platform for new and transformative treatments

Geoffrey McDonough
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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.
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.

Dedicated to rare diseases
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

With DX < 2 years old 2 year survival 1000 people tyro senemia growing up around the world our dream is that they one day become grand-parents.

29% 2 year survival with DX < 2 years old
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

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

2011

- Positive contributions from all parts of the business.
- Development pipeline setbacks as Kibrina phase 3 study did not meet primary endpoints.
- Orladin direct sales in the US.

2013

- Electo approved in the EU.
- Launch of two next-generation haemophilia products – Eloxanta and Alprolix in Europe and the Middle East.
- Pipeline strengthened with two candidate drugs in early stage development and new planned clinical programmes.

2014

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

2015

- Alprolix approved in the EU.
- Launch of Electo and Alprolix

2016

sobi established in the Middle East and the US

Expanded partner portfolio with Kibrina and Pharmawise portfolio

REVENUE SEK 2,177 M

REVENUE SEK 2,607 M

REVENUE SEK 3,228 M

REVENUE SEK 5,204 M

Launch of Eloxanta and Alprolix
Share development 2012 – 2 May 2017

- 2 May 2017: 142.90
- 30 Dec 2016: 106.70

Stocks compared:
- Swedish Orphan Biovitrum
- OMX Stockholm PI
- OMX Stockholm Pharmaceuticals & Biotechnology PI
- Nasdaq Biotech index
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

- SEK 770 M, USD 90 M -3%*
- SEK 1,106 M, USD 129 M +22%*
- SEK 1,852 M, USD 216 M >100%*
- SEK 820 M, USD 96 M 6%*
- SEK 656 M, USD 77 M -1%*

* Growth at Constant Exchange Rates
USD 1 = SEK 8,5613 (average year rate
• 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
- 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

![Revenues (SEK M): Orfadin](image)
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.

**Sobi**
Lower risk and spread investments.
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

Eloctate - Biogen Sales (million US$)

Alprolix - Biogen Sales (million US$)
Redefining protection in haemophilia
Top Priority: Access to medicines + human rights

Sustainability factors import to Sobi

“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

- Reimbursement granted – treatment available
- Pricing & reimbursement decision-making in process
Commitment to the global Haemophilia community

- Supporting leadership programmes, youth fellowship, and new technology 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

<table>
<thead>
<tr>
<th>Humanitarian Aid reaching patients worldwide</th>
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<tbody>
<tr>
<td><strong>188 MIU</strong> IUs of clotting factor donated</td>
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<td><strong>14,800</strong> People treated in 40 countries</td>
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<tr>
<td><strong>12,757</strong> Acute bleeds</td>
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<td><strong>695</strong> Surgeries</td>
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<td><strong>893 life-saving</strong></td>
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<td><strong>70 life-saving</strong></td>
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Percentage of paediatric patients receiving treatment in these countries has nearly doubled
(from 14% to 28%)
• We distribute to 60 counties worldwide
• 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

<table>
<thead>
<tr>
<th>Therapeutic area/Indication</th>
<th>Product/Project</th>
<th>Pre-clinical</th>
<th>Phase 1</th>
<th>Phase 2</th>
<th>Phase 3</th>
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<tbody>
<tr>
<td>Haemophilia A</td>
<td>Elocta/A-SPIRE*</td>
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<tr>
<td>Haemophilia A</td>
<td>Elocta/PUP1*</td>
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<td>Haemophilia B</td>
<td>Alprolix/B-YOND*</td>
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<td>Haemophilia B</td>
<td>Alprolix/PUP1*</td>
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<td>Acute gout</td>
<td>Kineret/anaGO</td>
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<td>Still’s disease</td>
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<td>Alkaptonuria</td>
<td>Orfadin/SONIA2</td>
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<td>MPSIIIA</td>
<td>SOBI003</td>
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<td>Anti-C5</td>
<td>SOBI005</td>
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<tr>
<td>Anti-IL-1</td>
<td>SOBI006</td>
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*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

- **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

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

Adapted from Krishnamoorthy et al. *Cell Immunol* 2016

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**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
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
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