

#### **INTERIM REPORT**

JANUARY-SEPTEMBER 2018



### Continued strong growth in Q3

- Total revenues of SEK 2,315 M (1,601) in Q3 and SEK 6,568 M (4,636) YTD<sup>1</sup>
- 45 per cent sales growth in the quarter compared with Q3 2017 (34 per cent at constant exchange rates (CER))
- EBITA increased 74 per cent to SEK 933 M (536) in Q3 and 85 per cent to SEK 2,655 M (1,434) YTD<sup>1</sup>
- Net cash position of SEK 2,483 M (1,472 as of 31 December 2017)
- Revenues for Elocta® and Alprolix® were SEK 873 M (417) and SEK 255 M (98) respectively for Q3<sup>1</sup>
- Kineret® revenues were SEK 347 M (272) in Q3, an increase of 28 per cent
- Orfadin® revenues were SEK 217 M (202) in Q3, an increase of 8 per cent
- Sobi<sup>TM</sup> strengthened both the Specialty Care franchise and the US product portfolio by acquiring the global rights to emapalumab
- First patients dosed in the phase 1/2 study with SOBI003
- Outlook 2018 updated, see page 8

REVENUES

SEK **2,315** M

+45%

GROSS MARGIN<sup>1</sup>

**75**%

EBITA 1

**SEK 933** M

+74%

EARNINGS PER SHARE

**SEK 2.31** 

## Financial summary

	Q3	Q3		Jan-Sep	Jan-Sep		Full-year
Amounts in SEK M	2018	2017	Change	2018	2017	Change	2017
Total revenues	2,315	1,601	45%	6,568	4,636	42%	6,511
Gross profit	1,741	1,129	54%	4,830	3,320	45%	4,657
Gross margin <sup>1</sup>	75%	70%		74%	72%		72%
EBITA <sup>1</sup>	933	536	74%	2,655	1,434	85%	2,053
EBITA margin <sup>1</sup>	40%	33%		40%	31%		32%
EBIT (Operating profit/loss)	820	426	92%	2,320	1,092	113%	1,600
Profit for the period	623	324	93%	1,823	791	130%	1,149
Earnings per share, SEK	2.31	1.20	92%	6.77	2.94	130%	4.27

<sup>&</sup>lt;sup>1</sup>Alternative Performance Measures (APMs), see page 14 for further information.

<sup>&</sup>lt;sup>1</sup> Revenues as well as EBITA were positively affected by SEK 56 M, in Q3 2018, related to adjusted pharmaceutical taxes in France from 2017, whereof SEK 52 M relates to Elocta.

## **CEO** statement



The strong growth continued in Q3. Total revenues grew 45 per cent, totalling SEK 2,315 M for the quarter. Sales for Elocta and Alprolix continued to climb. Specialty Care showed solid growth, up 18 per cent year-on-year, driven by Kineret. During the quarter we embarked upon our external growth journey with the acquisition of the global rights to emapalumab, a potential treatment for the rare disease primary haemophagocytic lymphohistiocytosis (pHLH). The PDUFA (Prescription Drug User Fee Act) decision by the FDA is expected on 20 November (PDUFA date) and our North American team is preparing for launch.

Haemophilia

Elocta sales reached SEK 873 M (417) and Alprolix sales SEK 255 M (98), up 110 and 161 per cent respectively. The main countries generating this growth were France, Germany and Italy. Total Haemophilia revenues grew 63 per cent to SEK 1,545 M (948), including royalties and manufacturing revenues for ReFacto.

Our Haemophilia franchise continues to perform well. As expected in this market, uptake slows over the summer period. Despite this, Elocta had another strong quarter. Alprolix sales gained momentum and are growing quickly, with great progress particularly in France. Alprolix was approved for reimbursement in Sweden and Slovakia during the quarter.

Elocta and Alprolix are now recognised as the standard of care in many countries. This is based on the well-established safety and efficacy profile of both products, confirmed by several years of real-world experience from thousands of patients in all patient groups. Replacing the missing coagulation factor is fundamental in haemophilia treatment and feedback from the medical community strengthens our belief that factor replacement therapy will continue to be the mainstay of treatment in the future.

#### **Specialty Care**

Specialty Care also delivered a solid result, with year-on-year growth at 18 per cent and revenues of SEK 770 M (653). Growth was driven mainly by Kineret.

"The acquisition of the global rights to emapalumab is an excellent fit with our company strategy"

> Guido Oelkers, CEO and President

ELOCTA PRODUCT SALES

+110%

ALPROLIX PRODUCT SALES

+161%

Kineret continued to perform well: sales reached SEK 347 M (272), an increase of 28 per cent, with growth in both EMENAR and North America. For the EMENAR region, part of the sales increase can be attributed to the launch of Kineret for Still's disease.

Orfadin sales reached SEK 217 M (202), an increase of 8 per cent. Despite increasing competition we continue to hold our market share.

The launch of Ravicti® for the treatment of urea-cycle disorders (UCD) continues. Ravicti has been approved for reimbursement in several European countries and we see significant patient uptake across Europe, particularly in Denmark, Germany, the Netherlands, Spain and the UK. Several patients in Saudi Arabia have also switched to Ravicti.

#### Pipeline advances

In August, the first patient in the phase 1/2 study with SOBI003 for the potential treatment of MPS IIIA (Sanfilippo syndrome) received the first dose. The second patient has also been dosed since then. After the third patient has been dosed, and following a safety review, the study can progress to the next dose level.

The primary efficacy results from anaGO, the phase 2 study of anakinra in patients with acute gout, were released. For the primary endpoint of patient-assessed pain intensity in the most affected joint there was a substantial reduction from baseline, both following treatment with anakinra and the comparator triamcinolone. There was a clinically meaningful pain reduction with anakinra in line with expectations of IL-1 blockade in this disease. However, no statistically significant difference between the two treatments was obtained. Given the continued unmet medical need among patients who cannot tolerate or who do not respond to conventional therapy, and the well-established safety profile of Kineret, we will seek to discuss our options with the FDA for moving anakinra forward into phase 3 in this indication. We see further potential in the area of interleukin-1 (IL-1) blockade and are continuing to explore new indications for the benefit of patients.

#### **Delivering on our strategy**

Our recent acquisition of the global rights to emapalumab, a potential treatment for primary haemophagocytic lymphohistiocytosis (pHLH), from Novimmune SA is an excellent fit for our company strategy. It is expected to address a significant unmet medical need and matches our expertise and experience in rare diseases. Emapalumab is expected to contribute strongly to expanding our inflammation and auto-immune disease focus in our Specialty Care franchise and will be particularly important for our North American business. Whilst the decision on emapalumab's approval will be the responsibility of the respective regulatory bodies, we are optimistically looking forward to the next milestone - a regulatory decision by the FDA - on 20 November (PDUFA date). Based on this, we believe that emapalumab is an attractive near-term commercial opportunity for Sobi with sales potential from 2019 onwards.

As for EU, the application for emapalumab was submitted to the EMA in August and a regulatory decision is expected by the end of 2019.

Together with expanding revenues from the rest of our Specialty Care portfolio, and sustained strong growth in our Haemophilia franchise, we are well positioned for continued investment in both our own research and development, and in securing future opportunities for external growth.

Solna, Sweden, 31 October 2018

Guido Oelkers, CEO and President

Kineret sales +28%

> First patient dosed in the phase 1/2 study with SOBI003.

## **Business review Q3**

#### Haemophilia

During the quarter, Alprolix was approved for reimbursement in Sweden and Slovakia and is now reimbursed in a total of 18 countries. Elocta is reimbursed in 25 countries.

Sobi and Bioverativ, a Sanofi company, hosted a satellite symposium on "Protection in Haemophilia: The Evolving Science and Long-Term Outcomes of Fc Fusion Factors" at the ISTH's 64th Scientific and Standardization Committee (SSC) meeting in Dublin, Ireland. The symposium explored the interactions of Elocta with the immune system and its potential for immune modulation. Moreover, the pharmacokinetic characteristics of factor IX, including recombinant factor IX (rFIX) products such as Alprolix, were discussed, with a focus on extravascular distribution. Finally, the presentation of the long-term clinical outcome data for Elocta and Alprolix aimed to look beyond simply reducing annual bleeding rates (ABRs), considering other outcomes such as joint health, the resolution of target joints and improved quality of life.

#### **Specialty Care**

On 20 July, Sobi announced that the company had entered into an exclusive licence agreement for the perpetual global rights to emapalumab, a late-stage orphan drug candidate for the treatment of primary haemophagocytic lymphohistiocytosis (pHLH), developed by Novimmune SA. An upfront payment of CHF 50 M in cash was paid by Sobi for the global licence, with a total of CHF 400 M in additional payments over an eight-year period. The additional unpaid payments may, however, be accelerated by either party at any time after 1 July 2019. The acquisition was completed on 23 August.

The ongoing launch of Ravicti made good progress during the quarter, with reimbursement approved in four new markets: England, Ireland, Norway and Scotland. There has been solid uptake in the markets where reimbursement has already been approved.

#### **R&D** pipeline

The first patient in the phase 1/2 study with SOBI003 for the treatment of MPS IIIA was dosed in the beginning of August.

#### Corporate

Sobi appointed Anne Marie de Jonge Schuermans as Head of the new Technical Operations organisation, bringing together Sobi's Manufacturing Operations/Biologics Development & Supply, Quality, Supply Chain, Procurement and Environment & Safety operations. Anne Marie joined Sobi on 1 October, and is a member of Sobi's Executive Committee.



## Financial review Q3

#### **Total revenues**

Total revenues for the quarter amounted to SEK 2,315 M (1,601), up 45 per cent compared with the third quarter of 2017 (34 per cent at CER).

Revenues for the first nine months were SEK 6,568 M (4,636), an increase of 42 per cent (37 per cent at CER).

#### Revenues by business area

#### Haemophilia

Total revenues for the Haemophilia franchise reached SEK 1,545 M (948) for the quarter, up 63 per cent. Nine-month revenues amounted to SEK 4,260 M (2,568), an increase of 66 per cent. Revenues for Elocta were positively affected by SEK 52 M, in Q3 2018, related to adjusted pharmaceutical taxes in France from 2017.

Product sales rose 119 per cent to SEK 1,128 M (515) for the quarter. The year-on-year growth was mainly attributable to France, Germany and Italy. Elocta sales amounted to SEK 873 M (417) and Alprolix sales to SEK 255 M (98). Nine-month product sales totalled SEK 2,987 M (1,249) where Elocta accounted for SEK 2,316 M (1,018) and Alprolix for SEK 671 M (232).

Royalty revenues were SEK 338 M (298) for the quarter and SEK 974 M (880) for the first nine months.

ReFacto manufacturing revenues were SEK 79 M (135) for the quarter, down due to the lower year-on-year order pattern. Nine-month manufacturing revenues totalled SEK 299 M (440).

#### **Specialty Care**

Specialty Care delivered a strong performance across the portfolio with revenues of SEK 770 M (653) for the quarter, an increase of 18 per cent. Revenues for the first nine months were SEK 2,308 M (2,068), an increase of 12 per cent.

Kineret had continued solid growth across North America and EMENAR with revenues of SEK 347 M (272) for the quarter, an increase of 28 per cent. Revenues for the first nine months were SEK 985 M (835), an increase of 18 per cent. The EMENAR region, EU5 and Turkey accounted for most of this growth. The commercial launch of Kineret for Still's disease in the EU is ongoing. Sobi's increased share of voice and reach in the rheumatology market has led to strong double-digit growth in the US.

## Revenues by business area

	Q3	Q3		Change	Jan-Sep	Jan-Sep		Change	Full-year
Amounts in SEK M	2018	2017	Change	at CER <sup>1</sup>	2018	2017	Change	at CER <sup>1</sup>	2017
Haemophilia									
Elocta	873	417	110%	93%	2,316	1,018	128%	115%	1,557
Alprolix	255	98	161%	140%	671	232	190%	172%	363
Manufacturing	79	135	-41%	-41%	299	440	-32%	-32%	559
Royalty	338	298	13%	7%	974	880	11%	14%	1,203
Total	1,545	948	63%	52%	4,260	2,568	66%	60%	3,682
Specialty Care									
Orfadin	217	202	8%	-2%	678	639	6%	4%	862
Kineret	347	272	28%	17%	985	835	18%	15%	1,142
Other	205	179	14%	5%	645	594	9%	4%	825
Total	770	653	18%	8%	2,308	2,068	12%	9%	2,829
					-				
Total revenues	2,315	1,601	45%	34%	6,568	4,636	42%	37%	6,511

<sup>&</sup>lt;sup>1</sup>Constant exchange rates.

Orfadin had a solid performance in both EMENAR and in North America due to patient support programmes and new formulations. The first generics have entered the market, however the impact was not material. Revenues were SEK 217 M (202) for the guarter, an increase of 8 per cent. Revenues for the first nine months amounted to SEK 678 M (639), an increase of 6 per cent.

#### **Gross profit**

Gross profit for the quarter was SEK 1,741 M (1,129), representing a gross margin of 75 per cent (70). A favourable product mix, the positive one-time impact of pharmaceutical taxes in France (for further information see page 8, Other information) and currency effects were the main contributors.

Gross profit for the first nine months was SEK 4,830 M (3,320), representing a gross margin of 74 per cent (72).

#### **Operating expenses**

Sales and administrative expenses excluding amortisation and write-downs amounted to SEK 509 M (371) for the quarter. The increase was mainly driven by continued investments in Haemophilia. In Specialty Care, the increase was mainly driven by continued investments in Kineret and preparations for the anticipated launch of emapalumab in North America.

Sales and administrative expenses excluding amortisation and write-downs amounted to SEK 1,425 M (1,167) for the first nine months. The increase reflects activities in the Haemophilia franchise in EMENAR, including marketing and personnel increases as well as investments in the North American region.

Research and development expenses amounted to SEK 287 M (214) for the quarter, and SEK 762 M (679) for the first nine months. The expenses reflect increased spending on programmes for Kineret, SOBI003, phasing of Sobi's 50 per cent share of Bioverativ's ongoing development costs for haemophilia, as well as expenses related to the acquisition of the global rights to emapalumab.

During the quarter, total operating expenses were affected by costs of SEK 47 M related to the long-term incentive programmes. There is no cash flow impact from these programmes.

#### **Operating profit**

EBITA was SEK 933 M (536) for the quarter and SEK 2,655 M (1,434) for the first nine months, corresponding to a margin of 40 (33) and 40 per cent (31) respectively.

Amortisation and write-downs of intangible assets amounted to SEK 113 M (110) for the quarter and SEK 335 M (342) for the first nine months. Full-year 2017 included a

## Operating profit/loss

	Q3	Q3	Jan-Sep	Jan-Sep	Full-year
Amounts in SEK M	2018	2017	2018	2017	2017
Total revenues	2,315	1,601	6,568	4,636	6,511
Total cost of goods and services sold	-574	-473	-1,738	-1,316	-1,854
Gross profit	1,741	1,129	4,830	3,320	4,657
Gross margin	75%	70%	74%	72%	72%
Sales and administrative expenses before amortisation and write-downs	-509	-371	-1,425	-1,167	-1,644
Research and development expenses	-287	-214	-762	-679	-908
Total opex less amortisation and write-downs	-796	-585	-2,187	-1,846	-2,551
Other operating revenue/expenses	-12	-8	12	-40	-52
EBITA	933	536	2,655	1,434	2,053
Amortisation and write-down related to Sales and administrative expenses	-113	-110	-335	-342	-453
EBIT	820	426	2,320	1,092	1,600

The statement is a non-IFRS statement. For an IFRS income statement, please refer to the Consolidated statement of comprehensive income.

write-down for one of the early-stage pipeline programmes amounting to SEK 12 M.

EBIT amounted to SEK 820 M (426) for the guarter and SEK 2,320 M (1,092) for the first nine months. EBIT increased by SEK 394 M for the quarter and SEK 1,228 M for the first nine months.

#### Net financial items and tax

Net financial items amounted to SEK -13 M (-17) for the quarter, including exchange rate gains/losses of SEK 4 M (-2).

Net financial items for the first nine months amounted to SEK -17 M (-53), including exchange rate gains/losses of SEK 25 M (-4). The difference was mainly attributable to a lower interest expense for the debt to Bioverativ and higher exchange rate gains. For more information regarding the agreement with Bioverativ, see Note 17 in the 2017 Annual Report.

Tax amounted to SEK -183 M (-85) for the quarter and SEK -480 M (-247) for the first nine months, corresponding to an effective tax rate of 22.7 and 20.9 per cent respectively. On 14 June 2018, the corporate tax rate in Sweden was reduced to 21.4 per cent from 1 January 2019, and to 20.6 per cent from 1 January 2021. The Group's deferred tax was revalued in Q2. The revaluation resulted in a lower effective tax rate year-to-date.

Profit totalled SEK 623 M (324) for the guarter and SEK 1,823 M (791) for the first nine months.

#### **Cash flow and investments**

Cash flow from operations before change in working capital amounted to SEK 613 M (280) for the quarter and to SEK 1,800 M (964) for the first nine months.

Working capital impacted cash flow by SEK 98 M (300) for the quarter and SEK -248 M (112) for the first nine months.

Cash flow from investing activities was SEK -512 M (-7) for the quarter and SEK -547 M (-97) for the first nine months. The largest investment during the quarter was the acquisition of the global rights to emapalumab, whereof cash flow impact amounted to SEK 497 M.

## **Financial** calendar

O4 2018 20 February 2019 Q1 2019 25 April 2019 17 July 2019 Q2 2019 31 October 2019 Q3 2019

#### Cash

At the end of the quarter, cash and cash equivalents amounted to SEK 2,488 M, compared with SEK 1,478 M at 31 December 2017.

#### Net cash/debt

Sobi ended the quarter with a net cash position of SEK 2,483 M, compared with SEK 1,472 M at 31 December 2017.

#### Equity

At 30 September 2018, consolidated shareholders' equity was SEK 8,499 M compared with SEK 6,701 M at 31 December 2017.

#### **Parent Company**

In the third quarter of 2018, net sales for the Parent Company, Swedish Orphan Biovitrum AB (publ), amounted to SEK 2,141 M (1,545), of which SEK 1,262 M (821) referred to sales to Group companies. Nine-month sales amounted to SEK 5,871 M (4,151) of which SEK 3,241 M (1,970) referred to sales to Group companies.

Profit after financial items amounted to SEK 936 M (521) for the quarter and SEK 2,517 M (1,129) for the first nine months.

Investments in tangible and intangible assets affecting cash flows amounted to SEK 14 M (7) for the guarter and SEK 46 M (90) for the first nine months.

## Other information

#### Personnel

At 30 September 2018, the number of full-time equivalents was 881 (800 at 31 December 2017).

#### Pharmaceutical taxes update in France

The uncertainty regarding the provision for pharmaceutical taxes recorded at 31 December 2017, which was reported in Q1 and Q2 2018, has now been resolved following a final notification of 2017 from the French authorities during Q3. The outcome had a positive one-time impact of SEK 56 M in revenues as well as EBITA, whereof SEK 52 M relates to Elocta.

#### Significant events after the reporting period

Primary efficacy results from the anaGO phase 2 study were released, showing that, for the primary endpoint of patient-assessed pain intensity in the most affected joint, there was a substantial reduction from baseline, following treatment with anakinra as well as with the comparator triamcinolone. There was a clinically meaningful pain reduction with anakinra in line with expectations of IL-1 blockade in this disease. However, no statistically significant difference between the two treatments was obtained.

### Financial outlook 2018<sup>1,2</sup>— updated

Sobi expects total revenues for the full year to be in the range of SEK 8,900 - 9,000 M (8,600 - 8,800).

The gross margin is expected to be in the range of 73 - 74 per cent (at least 70).

Sobi expects EBITA for the full year to be in the range of SEK 3,400 - 3,500 M, including development and commercialisation costs for emapalumab of around SEK 200 M, which was not included in the previous outlook for 2018 (3,400 - 3,600).

Solna, Sweden, 31 October 2018

Guido Oelkers, CEO and President

#### **Forward-looking statements**

This report includes forward-looking statements. Actual results may differ from those stated. Internal factors such as the successful management of research programmes and intellectual property rights may affect future results. There are also external conditions such as the economic climate, political changes and competing research programmes that may affect Sobi's results.

This information is information that Swedish Orphan Biovitrum AB (publ) is obliged to make public pursuant to the EU Market Abuse Regulation and the Securities Markets Act. The information was submitted for publication, through the agency of Linda Holmström, Senior Communications Manager, at 08:00 CET on 31 October 2018.

<sup>&</sup>lt;sup>1</sup>At current exchange rates as of 31 October 2018.

<sup>&</sup>lt;sup>2</sup>The latest outlook was published on 18 July 2018.

## Auditor's review

#### Introduction

We have reviewed the condensed interim report for Swedish Orphan Biovitrum AB (publ) as of 30 September 2018, and for the nine-month period then ended. The Board of Directors and the Chief Executive Officer are responsible for the preparation and presentation of this interim report in accordance with IAS 34 and the Swedish Annual Accounts Act. Our responsibility is to express a conclusion on this interim report based on our review.

#### **Scope of review**

We conducted our review in accordance with the International Standard on Review Engagements, ISRE 2410 *Review of Interim Financial Statements Performed by the Independent Auditor of the Entity.* A review consists of making inquiries, primarily of persons responsible for financial and accounting matters, and applying analytical and other review procedures. A review is substantially less in scope than an audit conducted in accordance with International Standards on Auditing and other generally accepted auditing standards in Sweden.

The procedures performed in a review do not enable us to obtain assurance that we would become aware of all significant matters that might be identified in an audit. Accordingly, we do not express an audit opinion.

#### Conclusion

Based on our review, nothing has come to our attention that causes us to believe that the interim report is not prepared, in all material respects, in accordance with IAS 34 and the Swedish Annual Accounts Act regarding the Group, and in accordance with the Swedish Annual Accounts Act regarding the Parent Company.

Stockholm, 31 October 2018

Ernst & Young AB

Björn Ohlsson

Authorised Public Accountant

# Financial statements – Group Statement of comprehensive income

	Q3	Q3	Jan-Sep	Jan-Sep	Full-Year
Amounts in SEK M	2018	2017	2018	2017	2017
Total revenues <sup>1</sup>	2,315	1,601	6,568	4,636	6,511
Total cost of goods and services sold	-574	-473	-1,738	-1,316	-1,854
Gross profit	1,741	1,129	4,830	3,320	4,657
Sales and administrative expenses <sup>2</sup>	<b>C22</b>	401	1 700	1 500	2.006
Research and development expenses	-622 -287	-481 -214	-1,760 -762	-1,509 -679	-2,096 -908
Other operating revenue/expenses	-287 -12	-214 -8	-762 12	-679 -40	
Operating profit	820	426	2,320	1,092	-52 <b>1,600</b>
Financial income/expenses <sup>3</sup>	-13	-17	-17	-53	-68
Profit before tax	807	409	2,303	1,038	1,532
Income tax expenses	-183	-85	-480	-247	-384
Profit for the period	623	324	1,823	791	1,149
All earnings are attributable to Parent Company shareholders					
Other comprehensive income					
Items that will not be reclassified to profit/loss					
Remeasurements of post-employment benefit obligations	_	0	3	2	-1
Items that may be reclassified subsequently to profit/loss					
Translation difference	-6	-5	16	-7	-1
Cash flow hedge (net of tax)	16	55	-75	173	150
Comprehensive income for the period	634	375	1,766	958	1,296
<sup>1</sup> See page 5 for split by business area.					
<sup>2</sup> Amortisation and write-downs of intangible assets included in Sales and administrative expenses.	-113	-110	-335	-342	-453
<sup>3</sup> Including financing costs amounting to:	0	0	-1	-1	1
Earnings per share, SEK	2.31	1.20	6.77	2.94	4.27
Earnings per share after dilution, SEK	2.30	1.20	6.74	2.93	4.25

## **Balance** sheet

	Sep	Dec	Sep
Amounts in SEK M	2018	2017	2017
ASSETS			
Non-current assets			
Intangible assets <sup>1,2</sup>	10,242	6,445	6,535
Tangible assets	135	134	122
Financial assets	227	167	155
Total non-current assets	10,604	6,746	6,812
Current assets			
Inventories	1,174	1,053	1,095
Accounts receivable	1,511	1,129	941
Current receivables, non-interest bearing	488	496	469
Cash and cash equivalents	2,488	1,478	1,758
Total current assets	5,662	4,157	4,263
Total assets	16,266	10,903	11,075
EQUITY AND LIABILITIES			
Shareholders' equity	8,499	6,701	6,352
Long-term liabilities			
Long-term liabilities <sup>3</sup>	4	5	503
Long-term liabilities, non-interest bearing	1,213	1,832	1,880
Total long-term liabilities	·	1,838	
Total long-term liabilities	1,217	1,030	2,383
Current liabilities			
Current liabilities	1	2	2
Current liabilities, non-interest bearing <sup>2</sup>	6,548	2,363	2,339
Total current liabilities	6,550	2,365	2,341
Total equity and liabilities	16,266	10,903	11,075

<sup>&</sup>lt;sup>1</sup>Including goodwill of SEK 1,554 M.

## Changes in equity

	Jan-Sep	Full-year	Jan-Sep
Amounts in SEK M	2018	2017	2017
Opening balance <sup>1</sup>	6,701	5,365	5,365
Share-based compensation to employees	32	40	28
Comprehensive income for the period <sup>2</sup>	1,766	1,296	958
Equity at end of period	8,499	6,701	6,352

 $<sup>^{1}</sup>$ Adjustment of deferred tax affected the 2017 opening balance by SEK 11 M.

<sup>&</sup>lt;sup>2</sup>The increase is related to the acquisition of the global rights to emapalumab for CHF 450 M. The liability is classified as short-term since the additional payments may be accelerated by either party any time after 1 July 2019.

<sup>&</sup>lt;sup>3</sup>External bank loan of SEK 500 M was repaid in 2017.

<sup>&</sup>lt;sup>2</sup>Whereof changes in cash flow hedges amounted to SEK -75 M (173).

## Cash flow statement

	Q3	Q3	Jan-Sep	Jan-Sep	Full-year
Amounts in SEK M	2018	2017	2018	2017	2017
Profit for the period	623	324	1,823	791	1,149
Adjustment for non-cash items <sup>1</sup>	-10	-45	-23	173	283
Cash flow from operations before change in working capital	613	280	1,800	964	1,431
Change in working capital	98	300	-248	112	-98
Cash flow from operations	712	580	1,553	1,076	1,333
Investment in intangible assets	-503	-3	-523	-72	-92
Investment in tangible assets	-10	-5	-29	-27	-48
Divestment of tangible assets	1	1	2	1	1
Investment in financial assets	0	0	3	1	-1
Cash flow from investing activities	-512	-7	-547	-97	-139
Loans - Raising/Amortisation	_	_	_	_	-500
Net finance lease	-1	_	-1		
Cash flow from financing activities	-1	-	-1	_	-500
Change in cash and cash equivalents	198	573	1,005	979	694
Cash and cash equivalents at the beginning of the period	2,306	1,189	1,478	786	786
Translation difference in cash flow and cash and cash equivalents	-16	-5	6	-7	-1
Cash and cash equivalents at the end of the period	2,488	1,758	2,488	1,758	1,478
<sup>1</sup> Adjustment for non-cash items:					
Depreciation of tangible assets	9	8	27	25	33
Amortisation and write-downs of intangible assets	113	110	335	342	453
Deferred tax	-23	25	-77	89	164
Other, whereof SEK -319 M (-219) in Q3 2018 and SEK -438 M in full-					
year 2017 reflect Elocta and Alprolix non-cash transactions, see also	110	100	200	202	267
Note 17 in Sobi's 2017 Annual Report for more information about	-110	-188	-308	-283	-367
the agreement with Bioverativ					
Non-cash items	-10	-45	-23	173	283

## Key ratios and other information

	Q3	Q3	Jan-Sep	Jan-Sep	Full-year
Amounts in SEK M	2018	2017	2018	2017	2017
Profit measures					
Gross profit	1,741	1,129	4,830	3,320	4,657
EBITDA <sup>1</sup>	942	544	2,682	1,459	2,086
EBITA <sup>1</sup>	933	536	2,655	1,434	2,053
EBIT (Earnings before interest and tax)	820	426	2,320	1,092	1,600
Profit/loss	623	324	1,823	791	1,149
Per share data (SEK)					
Earnings per share	2.31	1.20	6.77	2.94	4.27
Earnings per share after dilution	2.30	1.20	6.74	2.93	4.25
Shareholders' equity per share <sup>1</sup>	31.1	23.3	31.1	23.3	24.6
Shareholders' equity per share after dilution <sup>1</sup>	31.0	23.2	31.0	23.2	24.5
Other information					
Gross margin <sup>1</sup>	75%	70%	74%	72%	72%
EBITA margin <sup>1</sup>	40%	33%	40%	31%	32%
Equity ratio <sup>1</sup>	52%	57%	52%	57%	61%
Net cash (-)/debt (+) <sup>1</sup>	-2,483	-1,253	-2,483	-1,253	-1,472
Number of ordinary shares <sup>2</sup>	273,322,117	272,507,708	273,322,117	272,507,708	272,507,708
Number of ordinary shares (in treasury)	3,429,430	3,383,739	3,429,430	3,383,739	3,249,870
Number of ordinary shares (excluding shares in treasury)	269,892,687	269,123,969	269,892,687	269,123,969	269,257,838
Number of ordinary shares after dilution	274,159,143	273,388,248	274,159,143	273,388,248	273,458,932
Average number of ordinary shares (excluding shares in treasury)	269,681,071	269,123,969	269,398,916	268,970,953	269,020,363
Average number of ordinary shares after dilution (excluding shares in treasury)	270,601,711	269,930,337	270,630,624	269,852,041	270,003,546

<sup>&</sup>lt;sup>1</sup>Alternative performance measures (APMs), see next page for further information.

<sup>&</sup>lt;sup>2</sup>The increase in the number of shares results from an issue of 814,409 class C shares which have been converted to ordinary shares.

## Financial measures not defined according to IFRS

Sobi uses certain financial measures in the interim report that are not defined according to IFRS. The company considers these measures to provide valuable supplementary information for investors and company management, as they enable an assessment and benchmarking of the company's reporting. Since not all companies calculate financial measures in the same way, these are not always comparable to measures used by other companies. These financial measures should not, therefore, be regarded as substitutes for measures defined according to IFRS. The following metrics are not defined according to IFRS: All amounts in SEK M unless otherwise stated.

	Q3	Q3	Jan-Sep	Jan-Sep	Full-year
	2018	2017	2018	2017	2017
Total revenues	2,315	1,601	6,568	4,636	6,511
Cost of goods and services sold	-574	-473	-1,738	-1,316	-1,854
Gross profit	1,741	1,129	4,830	3,320	4,65
Gross margin, %	75%	70%	74%	72%	72%
Gross profit - Total revenues less cost of goods and	services sold.				
<b>Gross margin</b> - Gross profit as a percentage of total					
Operating profit	820	426	2,320	1,092	1,60
Plus amortisation and write-downs of intangible	113	110	335	342	45:
assets EBITA	933	536	2,655	1,434	2,05
Plus depreciations of tangible assets	9	8	27	25	3:
EBITDA	942	544	2,682	1,459	2,08
	40%	33%	40%	31%	32%
EBITA margin, %  EBITA - Earnings before interest, tax and amortisation  EBITDA - Earnings before interest, tax, depreciation  EBITA margin - EBITA as a percentage of total revenue	on. and amortisation.	3370	.070	3170	32/1
EBITA - Earnings before interest, tax and amortisation EBITDA - Earnings before interest, tax, depreciation EBITA margin - EBITA as a percentage of total revent	on. and amortisation.	3370	.57	31/6	JEN
EBITA - Earnings before interest, tax and amortisation EBITDA - Earnings before interest, tax, depreciation EBITA margin - EBITA as a percentage of total revenuable. Liabilities to credit institutions	on. and amortisation. ues.	503	4	503	
EBITA - Earnings before interest, tax and amortisation EBITDA - Earnings before interest, tax, depreciation EBITA margin - EBITA as a percentage of total revent	on. and amortisation.				
EBITA - Earnings before interest, tax and amortisation EBITDA - Earnings before interest, tax, depreciation EBITA margin - EBITA as a percentage of total revenue Liabilities to credit institutions - Long-term - Current	on. and amortisation. ues. 4	503	4	503	
EBITA - Earnings before interest, tax and amortisation EBITDA - Earnings before interest, tax, depreciation EBITA margin - EBITA as a percentage of total revenue Liabilities to credit institutions - Long-term	on. and amortisation. ues. 4 1	503 2	4 1	503 2	
EBITA - Earnings before interest, tax and amortisation EBITDA - Earnings before interest, tax, depreciation EBITA margin - EBITA as a percentage of total revenutiabilities to credit institutions - Long-term - Current Interest-bearing liability Cash and cash equivalents	on. and amortisation. ues.  4 1 6	503 2 <b>505</b>	4 1 6	503 2 <b>505</b>	1,47
EBITA - Earnings before interest, tax and amortisation EBITDA - Earnings before interest, tax, depreciation EBITA margin - EBITA as a percentage of total revenue Liabilities to credit institutions - Long-term - Current Interest-bearing liability	on. and amortisation. ues.  4 1 6 2,488 -2,483	503 2 <b>505</b> 1,758 <b>-1,253</b>	2,488 -2,483	503 2 <b>505</b> 1,758	1,47
EBITA - Earnings before interest, tax and amortisation EBITDA - Earnings before interest, tax, depreciation EBITA margin - EBITA as a percentage of total revenue Liabilities to credit institutions - Long-term - Current Interest-bearing liability  Cash and cash equivalents Net debt (+)/Net cash (-) Interest-bearing liability - Credit facilities and other Net debt(+)/Net cash(-) - Interest-bearing long-term	on. and amortisation. ues.  4 1 6 2,488 -2,483	503 2 <b>505</b> 1,758 <b>-1,253</b>	2,488 -2,483	503 2 <b>505</b> 1,758	1,47 <b>-1,47</b>
EBITA - Earnings before interest, tax and amortisation EBITDA - Earnings before interest, tax, depreciation EBITA margin - EBITA as a percentage of total revenue Liabilities to credit institutions - Long-term - Current Interest-bearing liability Cash and cash equivalents Net debt (+)/Net cash (-) Interest-bearing liability - Credit facilities and other	on. and amortisation. ues.  4 1 6 2,488 -2,483 liabilities to credit in and current liabilities	503 2 505 1,758 -1,253 nstitutions. ies less cash at bar	2,488 -2,483	503 2 <b>505</b> 1,758 <b>-1,253</b>	1,47. - <b>1,47</b> .
EBITA - Earnings before interest, tax and amortisation EBITDA - Earnings before interest, tax, depreciation EBITA margin - EBITA as a percentage of total revenue  Liabilities to credit institutions  - Long-term  - Current  Interest-bearing liability  Cash and cash equivalents  Net debt (+)/Net cash (-)  Interest-bearing liability - Credit facilities and other  Net debt(+)/Net cash(-) - Interest-bearing long-term  Equity  Total assets	on. and amortisation. ues.  4 1 6 2,488 -2,483 liabilities to credit in and current liabilit	503 2 505 1,758 -1,253 nstitutions. ies less cash at bar 6,352	4 1 6 2,488 -2,483 nk.	503 2 <b>505</b> 1,758 <b>-1,253</b>	1,47 - <b>1,47</b> 6,70 10,90
EBITA - Earnings before interest, tax and amortisation EBITDA - Earnings before interest, tax, depreciation EBITA margin - EBITA as a percentage of total revenue  Liabilities to credit institutions - Long-term - Current Interest-bearing liability  Cash and cash equivalents Net debt (+)/Net cash (-)  Interest-bearing liability - Credit facilities and other Net debt(+)/Net cash(-) - Interest-bearing long-term  Equity Total assets Equity ratio, %	on. and amortisation. ues.  4 1 6 2,488 -2,483  liabilities to credit in and current liabilit 8,499 16,266	503 2 505 1,758 -1,253 nstitutions. ies less cash at bar 6,352 11,075 57%	4 1 6 2,488 -2,483 nk. 8,499 16,266	503 2 <b>505</b> 1,758 <b>-1,253</b> 6,352 11,075	
EBITA - Earnings before interest, tax and amortisation EBITDA - Earnings before interest, tax, depreciation EBITA margin - EBITA as a percentage of total revenue  Liabilities to credit institutions  - Long-term  - Current  Interest-bearing liability  Cash and cash equivalents  Net debt (+)/Net cash (-)  Interest-bearing liability - Credit facilities and other  Net debt(+)/Net cash(-) - Interest-bearing long-term  Equity  Total assets	on. and amortisation. ues.  4 1 6 2,488 -2,483  liabilities to credit in and current liabilit 8,499 16,266	503 2 <b>505</b> 1,758 -1,253 nstitutions. ies less cash at bar 6,352 11,075	4 1 6 2,488 -2,483 nk. 8,499 16,266	503 2 <b>505</b> 1,758 <b>-1,253</b> 6,352 11,075	1,47: - <b>1,47</b> : 6,70 10,90

**Equity ratio** - Shareholders' equity as a proportion of total assets. **Equity per share** - Equity divided by the number of shares.

## Financial statements – Parent Company Income statement

	Q3	Q3	Jan-Sep	Jan-Sep	Full-year
Amounts in SEK M	2018	2017	2018	2017	2017
Total revenues	2,141	1,545	5,871	4,151	5,756
Total cost of goods and services sold	-603	-493	-1,672	-1,351	-1,861
Gross profit	1,538	1,052	4,199	2,799	3,895
Sales and administrative expenses <sup>1</sup>	-329	-306	-977	-947	-1,400
Research and development expenses	-249	-204	-698	-645	-855
Other operating revenue/expenses	-12	-5	9	-28	-40
Operating profit	948	537	2,533	1,180	1,600
Result from participation in Group companies <sup>2</sup>	_	_	_	_	-1,000
Financial income/expenses	-11	-16	-15	-51	-65
Profit/loss after financial items	936	521	2,517	1,129	535
Appropriations	_	_	_	_	-911
Profit/loss before tax	936	521	2,517	1,129	-376
Income tax expenses	-181	-41	-482	-102	-132
Profit/loss for the period	755	480	2,035	1,028	-508

## Statement of other comprehensive income

	Q3	Q3	Jan-Sep	Jan-Sep	Full-year
Amounts in SEK M	2018	2017	2018	2017	2017
Profit/loss for the period	755	480	2,035	1,028	-508
Items that may be subsequently reclassified to profit/loss					
Cash flow hedge (net of tax)	17	55	-75	173	150
Comprehensive income for the period	772	535	1,960	1,201	-358
<sup>1</sup> Amortisation and write-downs of intangible assets included in Sales and administrative expenses.	-74	-71	-217	-225	-296

<sup>&</sup>lt;sup>2</sup>The Parent Company wrote down the value of the shares in the subsidiary Swedish Orphan Biovitrum International AB in 2017 by SEK 1,000 M.

## **Balance** sheet

	Sep	Dec	Sep
Amounts in SEK M	2018	2017	2017
ASSETS			
Non-current assets			
Intangible assets	3,862	4,058	4,109
Tangible assets	112	114	102
Financial assets	2,915	2,915	3,882
Total non-current assets	6,889	7,087	8,093
Current assets			
Inventories	994	894	939
Current receivables, non-interest bearing	2,717	1,779	1,632
Cash and cash equivalents	2,340	1,381	1,685
Total current assets	6,050	4,054	4,256
Total assets	12,939	11,140	12,349
EQUITY AND LIABILITIES			
Shareholders' equity	7,428	5,436	6,984
Untaxed reserves	2,124	2,124	1,154
Long-term liabilities			
Long-term liabilities <sup>1</sup>	_	_	498
Long-term liabilities, non-interest bearing	546	1,159	1,272
Total long-term liabilities	546	1,159	1,770
Current liabilities			
Current liabilities, non-interest bearing	2,841	2,421	2,442
Total current liabilities	2,841	2,421	2,442
Total equity and liabilities	12,939	11,140	12,349

 $<sup>^{1}\</sup>mbox{External}$  bank loan of SEK 500 M was repaid 2017.

## Change in shareholders' equity

	Jan-Sep	Full-year	Jan-Sep
Amounts in SEK M	2018	2017	2017
Opening balance <sup>1</sup>	5,436	5,755	5,755
Share-based compensation to employees	32	40	28
Comprehensive income for the period <sup>2</sup>	1,960	-358	1,201
Equity at end of period	7,428	5,436	6,984

 $<sup>^{1}\!\</sup>mbox{Adjustment}$  of deferred tax affected the opening balance 2017 by SEK 11 M.

 $<sup>^2\</sup>mbox{Whereof}$  changes in cash flow hedges amounted to SEK -75 M (173).

## Financial notes

## Note 1 – Accounting policies and measurement bases and other information

#### Significant accounting policies

This report has been prepared in accordance with IAS 34 and the Swedish Annual Accounts Act. The consolidated financial statements for the period January-September 2018 have been prepared in accordance with International Financial Reporting Standards (IFRS) and the International Financial Reporting Interpretations Committee (IFRIC) interpretations as adopted by the EU and the Swedish Annual Accounts Act.

The Parent Company applies the Annual Accounts Act and the Swedish Financial Reporting Board's Recommendation RFR 2 Accounting for Legal Entities.

The consolidated financial statements have been prepared using the historical cost convention, except in the case of financial assets and certain financial assets and liabilities (including derivative instruments) that are measured at fair value through profit or loss.

The accounting policies applied, except for the changes listed below, are in accordance with those described in the 2017 Annual Report. More detailed information about the Group's accounting policies and measurement bases can be found in the 2017 Annual Report, available at www.sobi.com.

#### Changes in accounting policies

The new accounting standards IFRS 9 Financial Instruments and IFRS 15 Revenue from Contracts with Customers came into effect on 1 January 2018. Preparations continue for the new accounting standard IFRS 16 Leases, which will apply for financial periods beginning on or after 1 January 2019.

IFRS 9 Financial Instruments replaces IAS 39 Financial Instruments: Recognition and Measurement.

The standard contains rules for the classification and measurement of financial assets and liabilities, impairment of financial instruments and hedge accounting. One of the changes relates to liabilities measured at fair value. The part of the change relating to fair value of liabilities due to changes in own credit risk should be reported in other comprehensive income instead of in profit or loss, unless this

causes inconsistency in the accounting. Sobi has no liabilities measured at fair value and is therefore not affected by the change.

Another change relates to hedge accounting and requires increased disclosure of risk management and the effect of hedge accounting. Sobi's hedge accounting is made in accordance with IAS 39 with disclosures in accordance with IFRS 9; the new hedge requirements have no material impact on current hedge activities. Finally, new principles have been introduced regarding impairment of financial assets, where the model is based on expected losses. Sobi has applied the retrospective transition method which has no material impact on either earnings or financial position. In accordance with IFRS 9, Sobi has chosen not to recalculate comparative figures.

IFRS 15 contains a comprehensive accounting model for revenues from customer contracts and replaces the existing standards for revenue accounting, such as IAS 18.

Sobi has conducted a thorough analysis of the effects that the introduction of IFRS 15 may have on the Group's financial statements, and these have no material impact on either earnings or the financial position. To reach this conclusion, agreements and transactions have been reviewed and tested against the standard's five-step model for revenue recognition. Consequently, revenue recognition according to IFRS 15 has been applied in its entirety and remains unchanged from the present standard. As a transition method, Sobi has chosen full retrospective application, which means that the company applies IFRS 15 prospectively for contracts in place on the transition date. Revenue recognition remains unchanged on transition to the new standard.

IFRS 16 replaces IAS 17 Leases, with new accounting requirements for lessees. All leasing contracts, except short-term and low-value assets, must be reported as assets with the right-of-use, and as a corresponding liability in the lessee's balance sheet. Leasing fees will be replaced by depreciation and interest expenses. Underlying contracts recognised as a right-of-use asset consist mainly of rental agreements and cars. A modified retrospective approach has been chosen as the transition method with the right-of-use asset being the same as the lease liability at transition date 1 January 2019.

The main impact will be seen in the non-IFRS measurement EBITDA (Earnings before interest, tax, depreciation, amortisation). Together with a minor increase in total assets/financial liabilities, key ratios containing these parameters will be affected.

#### **Operating risks**

All business operations involve risk. Managed risk-taking is necessary to maintain good profitability. Risk may be due to events in the external environment and may affect a certain industry or market. Risk may also be specific to a certain company.

Sobi is exposed to three main risk categories:

- Operational risks, e.g. due to the capital-intensive and risky nature of new drug development, dependence on external partners in various collaborations, product liability claims and laws, and rules on the treatment of hazardous materials.
- External risks, such as patent infringements, competition within product concepts and decisions by authorities regarding product use and prices.
- Financial risks, such as currency risk, interest risk, credit risk and liquidity risk.

A more detailed description of the Group's risk exposure and risk management is included in Sobi's 2017 Annual Report (see the Directors' Report). There are no major changes in the Group's risk exposure and risk management in 2018 compared with the previous year.

#### Note 2 – Fair value of financial instruments

The Group carries derivatives (see the 2017 Annual Report for a narrative description of the purpose of the holdings). The derivatives (under the heading "current assets/ liabilities") are all categorised within Level 2 of the fair value hierarchy in the IFRS 13 standard (inputs other than quoted prices that are observable for the instruments, either directly or indirectly, are used in the fair value measurement). All derivatives are measured at fair value based on market data in accordance with IFRS. At 30 September 2018, the net reported value of derivatives on the balance sheet was SEK 1 M (-6).

At 30 September 2018, all other financial instruments on the balance sheet had reported values that are in all material aspects equivalent to fair value.

## Definitions and glossary

A recombinant, EHL clotting factor IX therapy approved in the EU, Iceland, Kuwait, **Alprolix** Liechtenstein, Norway, Saudi Arabia and Switzerland, as well as in Australia, Brazil, (eftrenonacog alfa)

Canada, Japan, New Zealand, the United States and other countries, for the treatment of

haemophilia B, which can be used by people of all ages.

**Acute gout** An autoinflammatory disease and an intensely painful and disabling inflammatory arthritis

> involving one or several joints. Gout is also a disease associated with multiple comorbidities, which may limit the use of some conventional treatment regimens.

**AnaGO** A randomised double-blind, multicentre phase 2 study being conducted in North America

studying two dose levels of anakinra in comparison to intramuscular triamcinolone for the

treatment of acute gout.

Constant exchange rate. **CER** 

Earnings per share The portion of a company's profit allocated to each outstanding share of common stock.

Extended half-life, which means that the circulation in the body is prolonged. Sobi's **EHL** 

haemophilia treatments, Elocta and Alprolix, are EHL products.

A recombinant, EHL clotting factor VIII therapy approved in the EU, Iceland, Kuwait, **Elocta** 

> Liechtenstein, Norway, Saudi Arabia and Switzerland for the treatment of haemophilia A, which can be used by people of all ages. It is also approved in Australia, Brazil, Canada,

Japan, New Zealand, the United States and other countries, where it is known as

ELOCTATE®.

European Medicines Agency. **EMA** 

Emapalumab is an anti-interferon-gamma (IFN-y) monoclonal antibody (mAb), currently emapalumab

> under FDA and EMA review for the treatment of primary haemophagocytic lymphohistiocytosis (pHLH), a life-threatening syndrome of immune activation.

Abbreviation for business region including Europe, Middle East, North Africa and Russia. **EMENAR** 

France, Germany, Italy, Spain and the UK. **EU5 markets** 

The US Food and Drug Administration. **FDA** 

Unit that indicates the workload of an employed person in a way that makes workloads **Full-time equivalents** 

Haemophagocytic

(efmoroctocog alfa)

the disease (pHLH, inherited) mainly occurs in infants and young children and the lymphohistiocytosis (HLH)

secondary form of the disease (sHLH, acquired) is acquired from or associated with

A rare and life-threatening syndrome of extreme immune activation. The primary form of

## Definitions and glossary

#### Haemophilia

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 haemophilia B occurs in about one in 25,000 male births annually. Both occur more rarely in females. People with haemophilia experience bleeding episodes that may cause pain, limited mobility, irreversible joint damage and life-threatening haemorrhages.

## Hereditary tyrosinemia type 1 (HT-1)

People with HT-1 have problems breaking down an amino acid called tyrosine. Toxic by-products are formed and accumulate in the body, which can cause liver, renal and neurological complications.

#### Kineret (anakinra)

A recombinant protein drug that blocks the biological activity of interleukin-1 a and b (IL-1a and IL-1b) by binding to IL-1 type 1 receptors (IL-R 1), expressed in a variety of tissues and organs, thereby blocking the IL-1 signalling. IL-1 is a key mediator of inflammation and a significant contributor to autoinflammatory diseases.

#### Mucopolysaccharidosis (MPS) type IIIA (Sanfilippo A syndrome)

A progressive, life-threatening and rare inherited metabolic disorder affecting children from a young age. Belongs to a group of diseases called lysosomal storage disorders (LSDs).

#### Orfadin (nitisinone)

A drug used to treat hereditary tyrosinaemia type 1 (HT-1). It blocks the breakdown of tyrosine, thereby reducing the amount of toxic tyrosine by-products in the body. Patients must maintain a special diet in combination with Orfadin treatment as

## PDUFA date (Prescription Drug User Fee Act)

A date when the FDA is expected to authorise drug applications.

## Ravicti (glycerol phenylbutyrate [GPB])

A drug indicated for use as adjunctive therapy for chronic management of adult and paediatric patients aged two months or older with urea cycle disorders (UCDs).

## (glycerol phenylbutyrate [GPB] SOBI003

A product candidate and a chemically modified variant of a recombinant human sulfamidase, using Sobi's proprietary glycan modification technology Modifa™, intended as an enzyme-replacement therapy in the lysosomal storage disease MPS IIIA, aimed at reducing heparan sulfate storage materials in affected cells.

#### Still's disease

An autoinflammatory disease that affects both children and adults, characterised by persistent high spiking fevers, recurring rashes and arthritis. Still's disease is also known as systemic-onset juvenile idiopathic arthritis (SJIA) or adult-onset Still's disease (AOSD).

#### **Urea cycle disorders (UCD)**

Inborn errors of metabolism comprising a group of inherited deficiencies of one of the enzymes or transporters involved in the urea cycle, which converts ammonia to urea. They are rare, serious and life-threatening disorders since absence or severe dysfunction of the enzymes or transporters results in the accumulation of toxic levels of ammonia in the blood and brain of affected patients.

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 transform lives 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. For more information visit www.sobi.com



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