Q3 2020 results

rare **strength**

October 22, 2020

Forward looking statements

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Guido Oelkers | CEO

Henrik Stenqvist | CFO

Ravi Rao | Head of R&D and CMO

Presenters

Ongoing transformation towards innovative leadership

From…		To…
In-licensing/distributor agreements	1	Thoughtful M&A and outright ownership
Dependence of haemophilia	2	Diversified our portfolio into Haematology and Immunology
Few big bets in early R&D	3	5 late stage assets in our pipeline
Europe centric approach	4	Extended International footprint - entrenched position in the US - building organisations in Russia, Japan and China

Becoming a global leader in rare diseases

Operational highlights – *Solid quarter in challenging times*

Strong financial performance

	Q3 2020	Jan-Sep 2020
Total sales growth (CER)	6%	14%
Total adjusted EBITA growth	-15%	10%
Adjusted EBITA % of sales	(31%)	(39%)
Haematology sales (CER)	16%	17%
Immunology sales (CER)	11%	25%

Launch products delivering

Doptelet (avatrombopag) tablets	58% growth Q3 vs Q2 (CER)*	-
gamifant emapalumab-lzsg	83% (CER)	-4%

*Excluding milestone of SEK 87 M in Q2

Pipeline - key assets in solid conditions

Haematology

BIVV001 – in phase 3

Phase 1/2a results - published in NEJM - Potential to become first choice among factor VIII replacement products

Doptelet – launched in CLD and ITP

Phase 3 CIT study of avatrombopag - missed primary endpoint due to placebo effect

Immunology

Gamifant – launched in pHLH

Phase 2 study of emapalumab in acute graft failure (aGF) to be initiated in Q1 2021

SEL-212 – in phase 3

Phase 3 initiated and ongoing.
Phase 2 study shows SEL-212 potentially more potent than pegloticase.

Nirsevimab

Phase 3 study ongoing

*"…may become the **first choice among factor VIII replacement** products…"*

NEJM Editorial Prof Mannucci[1]

Peak sales** guidance **remains unchanged

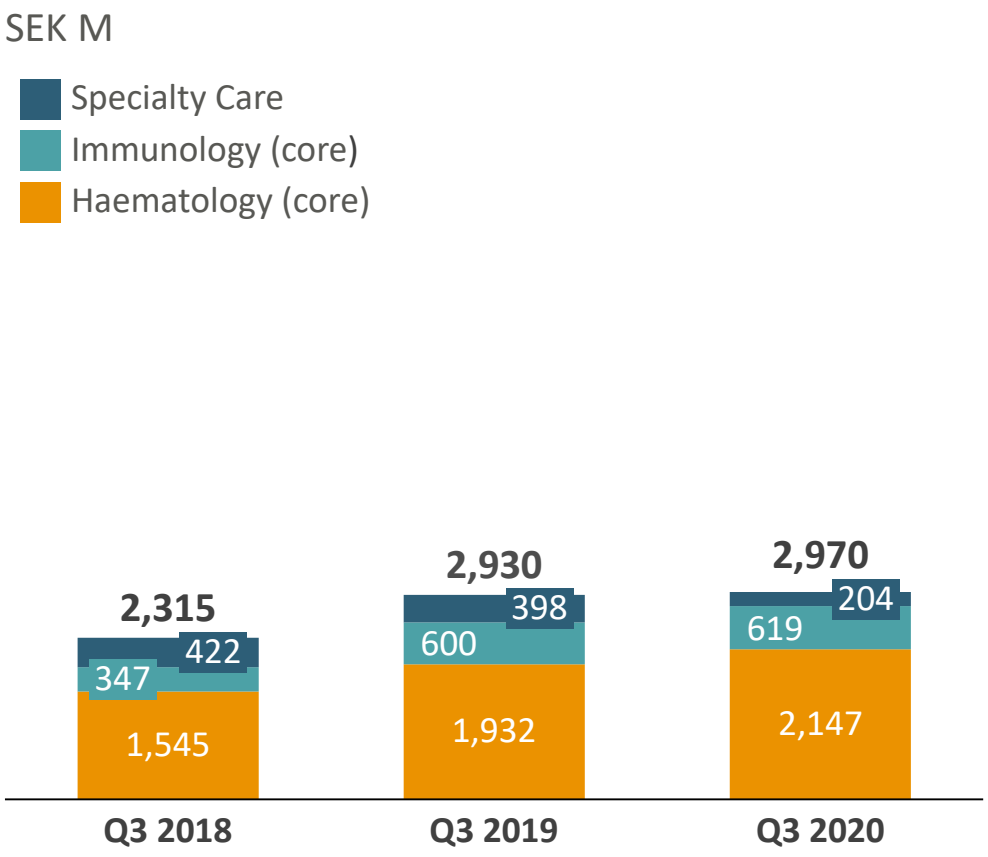
***Expanding** beyond pHLH*

***Differentiated treatment** in tophi patients*

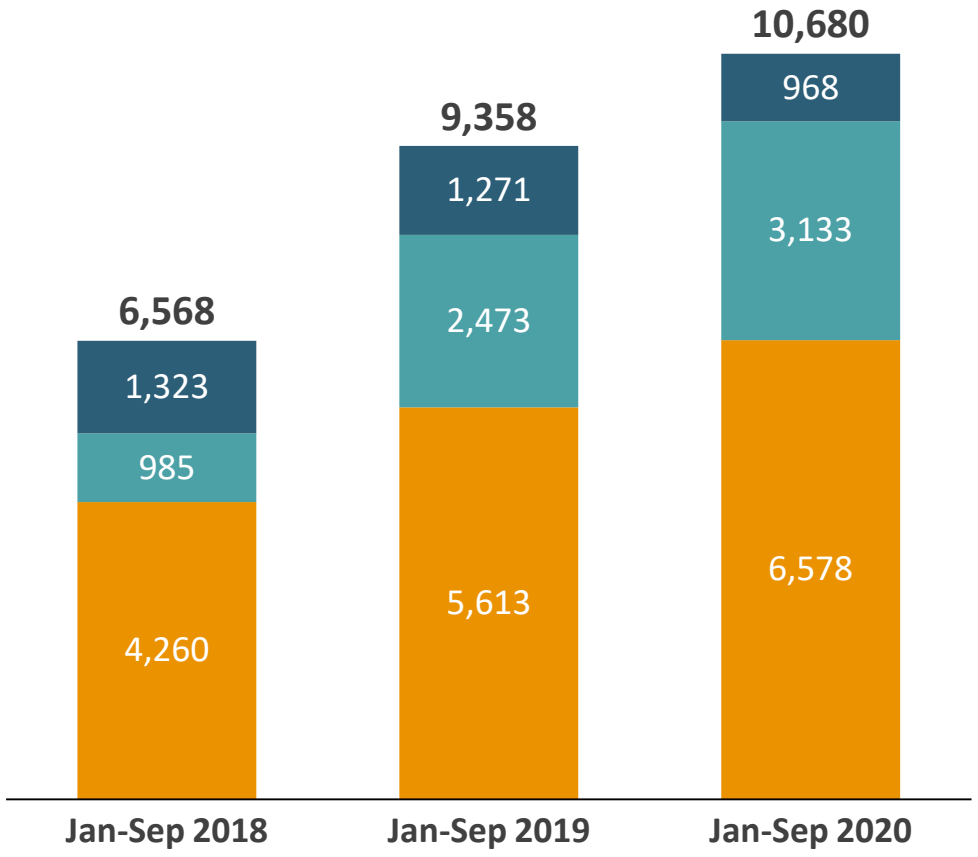
1. Mannucci PM. N Engl J Med. 2020

Continued solid top line growth

Q3 sales development



Jan-Sep sales development

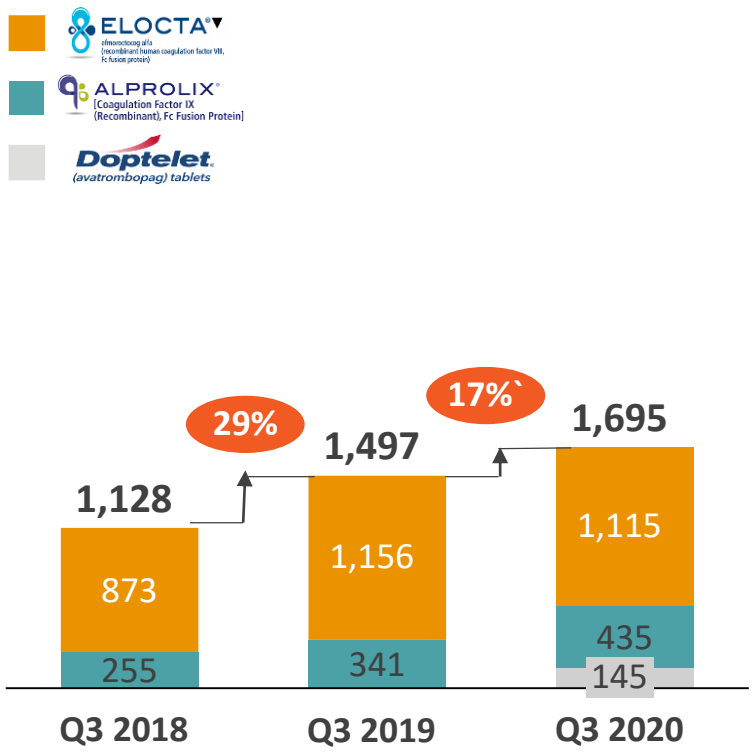


Business Review

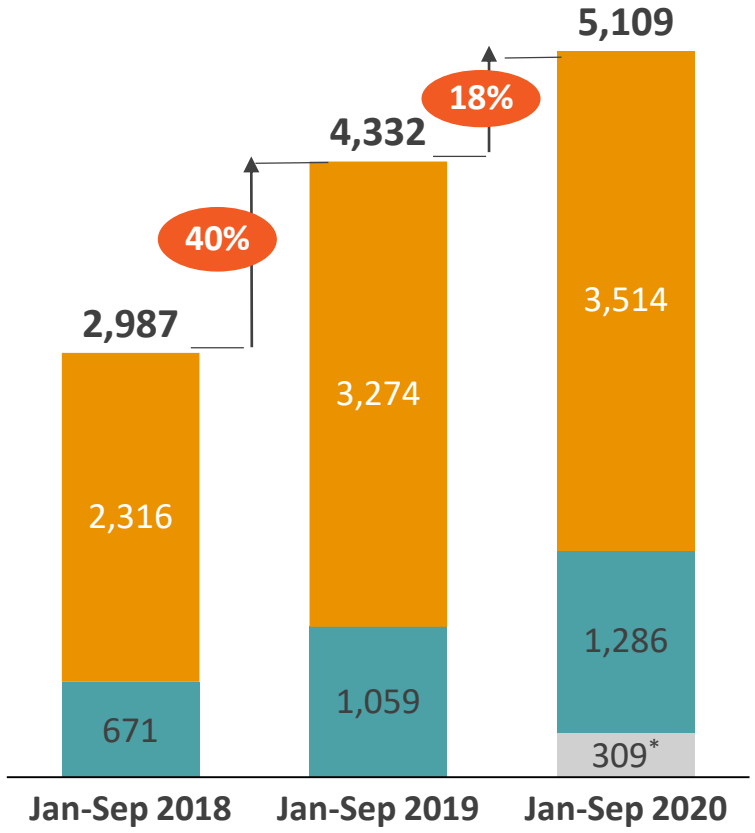
Continued double-digit growth for product sales in Haematology

Q3 product sales development

SEK M
Growth at CER



Jan-Sep product sales development



*Excluding milestone of SEK 87 M in Q2

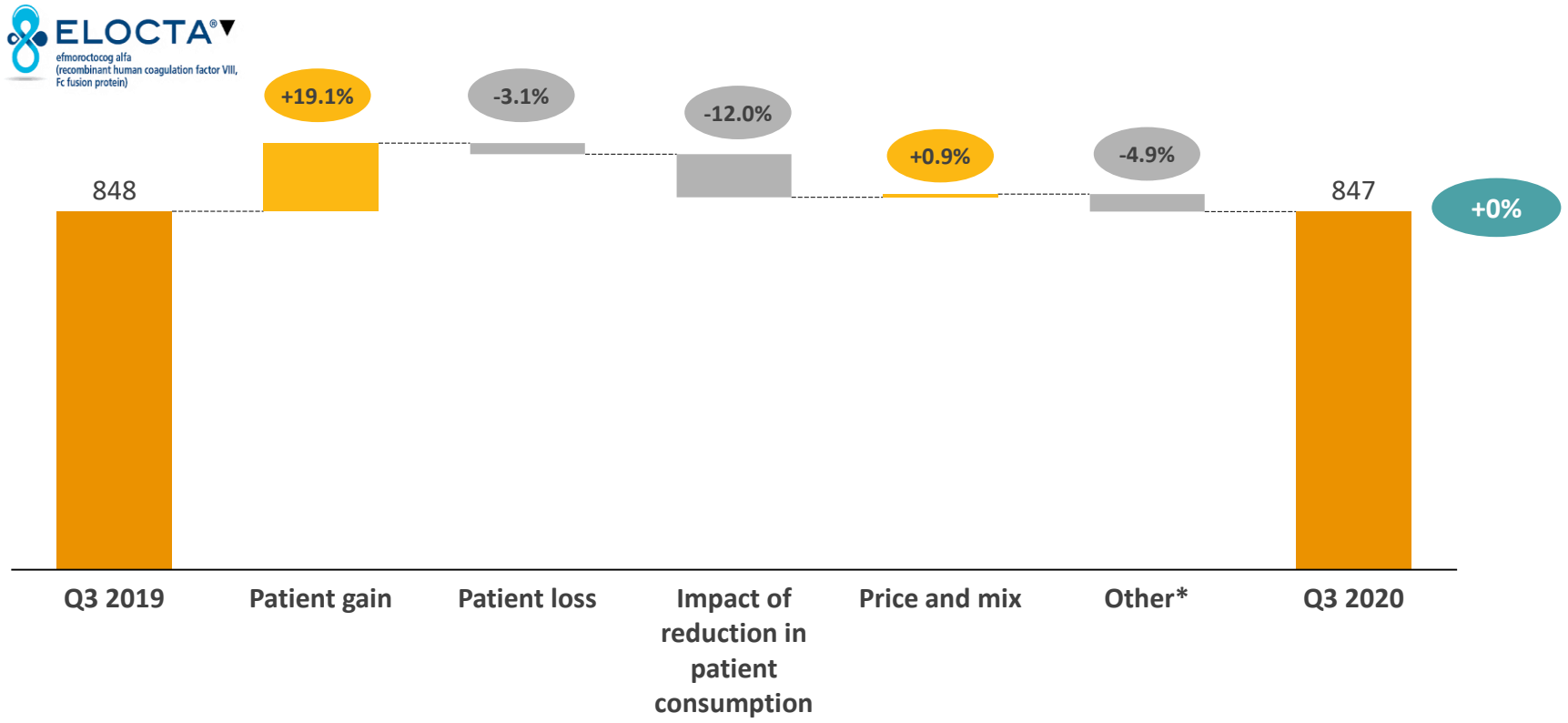
Highlights

- Strong top-line growth in Q3 particularly driven by a +33% increase in Alprolix
- Despite challenging environment strong growth of 18% for the first nine months
- Launch of Doptelet contributing to top line growth with SEK 145 M in the quarter

Elocta continued share gain offset by reduction in consumption per patient

Elocta sales – EU5 countries – Q3 2019 to Q3 2020

SEK M
growth at CER



* Mostly clawback adjustments

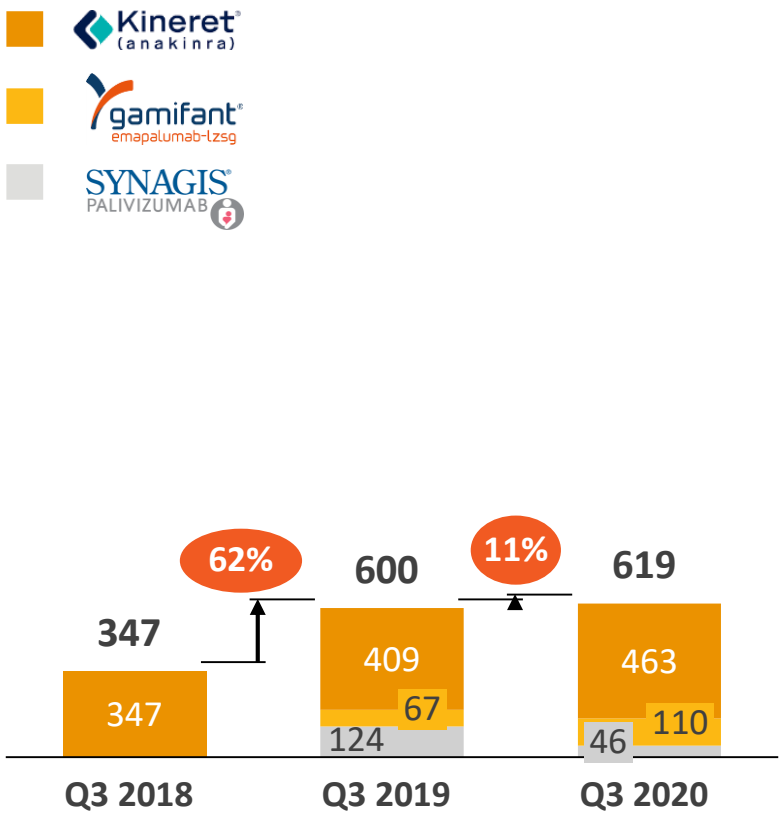
Highlights

- Continued patient gain
- Reduction in consumption per patient impacted by COVID-19

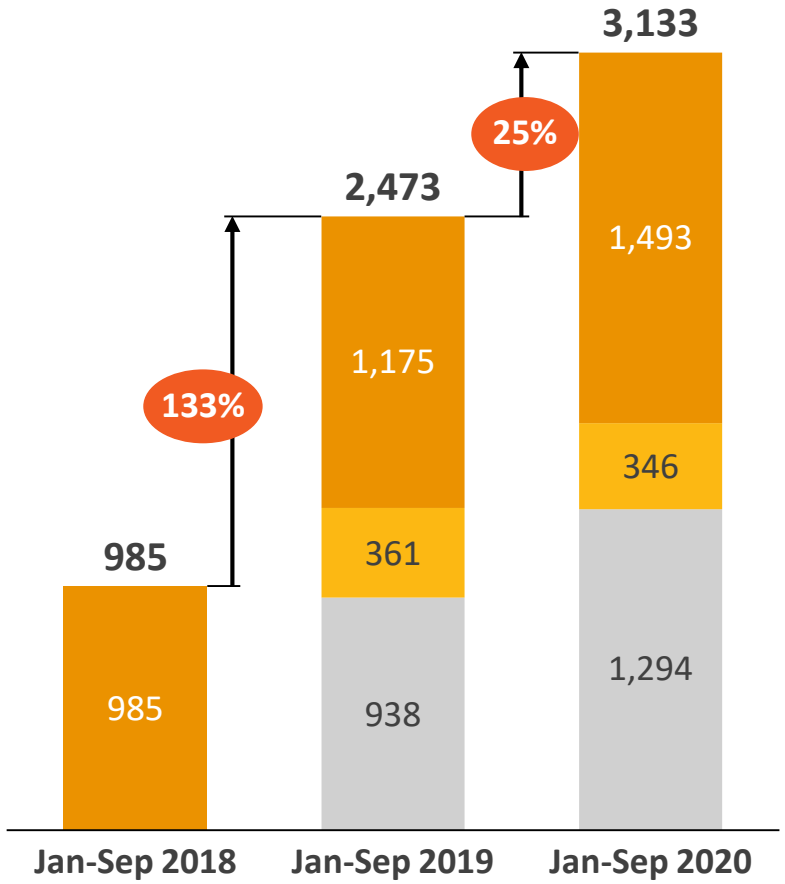
Strong top line revenue growth Immunology

Q3 product sales development

SEK M
Growth at CER



Jan-Sep product sales development



Highlights

Sales growth in Q3 driven by

- Strong double digit top-line growth for Kineret
- Gamifant sales grew by 83% at CER in Q3 with a positive patient trend
- Synagis sales decline due to higher stocking levels in 2019
- In Q3 29% growth at CER excluding Synagis

Pipeline

A brief introduction …

Ravi Rao

Head of R&D and CMO

Academic career

Imperial College London
Consultant Rheumatologist
and Senior Lecturer

Imperial College London
PhD Vascular Biology

University of Cambridge
MB BCh, Medicine
and Surgery

Industry experience

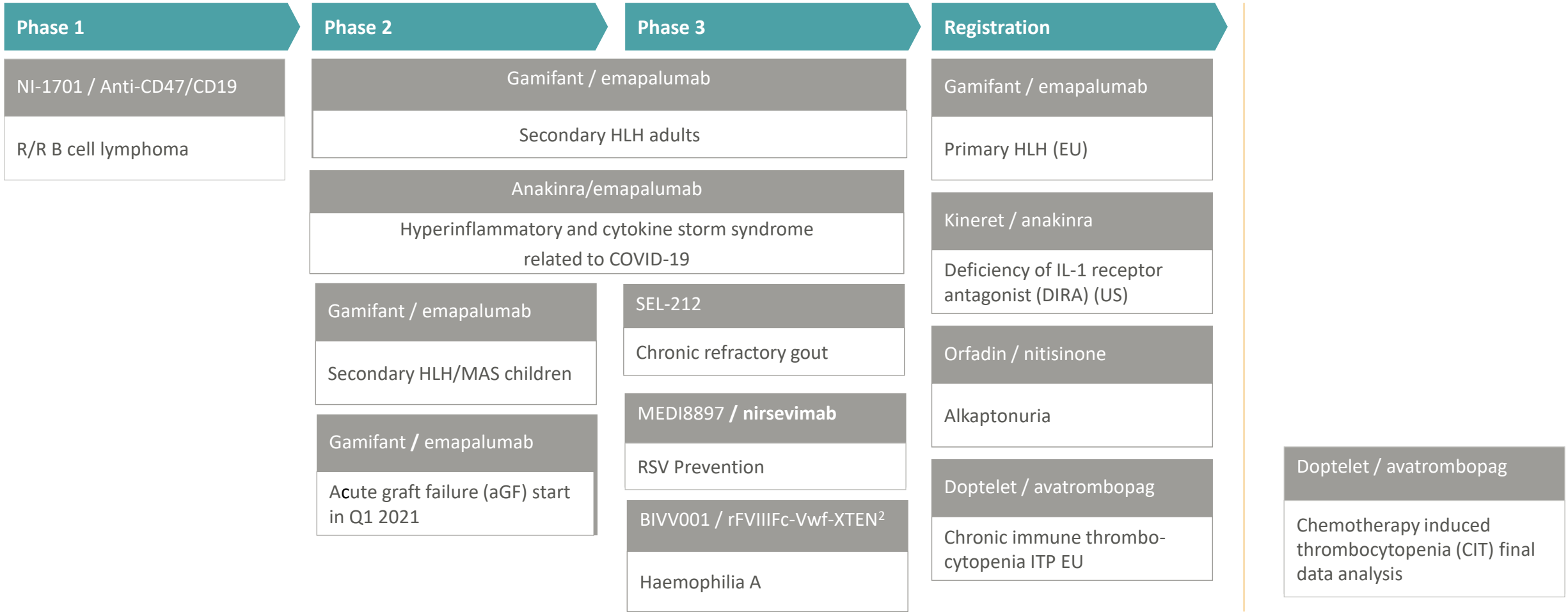
Chief Medical Officer, Aeglea
BioTherapeutics

Vice President, Global Medical Head,
Immunology-inflammation and Future
Pipeline Franchise at GSK

Group Medical Director, Immunology
Clinical Development at Roche

Q3 has been a mixed quarter but strong pipeline with five key assets in phase 2 and 3

Product pipeline



The read-out of the phase 3 study for avatrombopag in CIT did not turn out as expected and we are analysing the data in order to evaluate options

Avatrombopag CIT – Phase 3 study result

- No difference in response rate between placebo and avatrombopag groups

Primary endpoint (ITT)	Placebo	Avatrombopag
Avoidance of platelet transfusions, chemo-therapy dose reductions by ≥15%, dose delays by ≥4d	72.5%	69.5%

- Avatrombopag **consistently raised platelet** counts in patient with CIT compared to placebo
- **Safety profile** of avatrombopag was comparable to placebo

Ongoing work

- We believe an unmet medical need clearly exists in CIT
- Currently reviewing the full study dataset with further analyses ongoing
- Evaluating aspects regarding patient selection and trial design
- Working with clinical experts to define next steps

SEL-212 Phase 2 results show the potential for greater reductions in Serum Uric Acid compared to pegloticase

Study setup

129
patients with chronic refractory gout with different race and ethnicity

96.1% of all study patients are male with a mean age of **52 years**

Study results

Patients with <6 mg/dl SUA (Serum Uric Acid) for at least 80% of the evaluation time

		SEL-212	Pegloticase	p-value
Month 3	PP	70%	51%	0.0019
	ITT	70%	51%	0.0017
Month 3 and 6 combined	PP	59%	46%	0.056
	ITT	53%	46%	0.181

Primary study endpoint

Reduction in mean Serum Uric Acid (mg/dL)
all patients

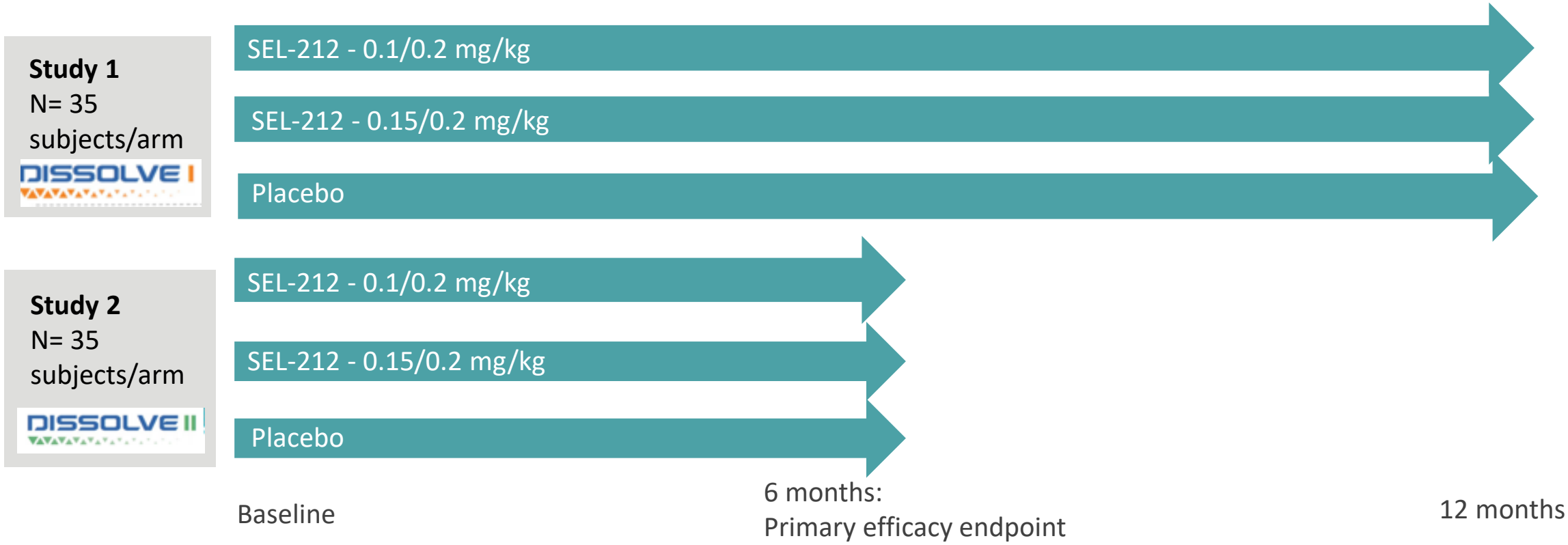
		SEL-212	Pegloticase	p-value
Month 3 and 6 combined	PP	-6.68	-4.51	0.003
	ITT	-6.79	-4.86	0.003

Reduction in mean Serum Uric Acid (mg/dL)
patients with tophi

		SEL-212	Pegloticase	
Month 3 and 6 combined	PP	-7.42	-4.64	0.016
	ITT	-7.32	-4.89	0.019

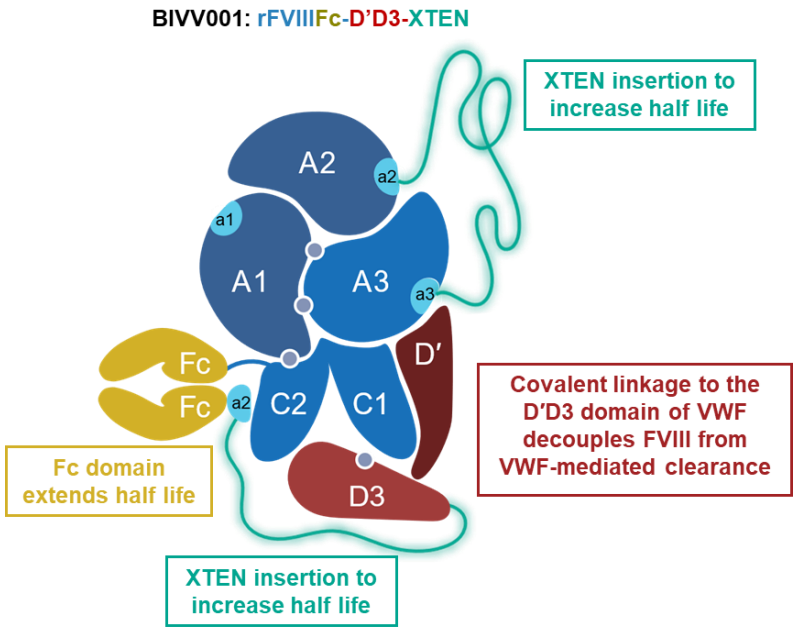
SEL-212 phase 3 study has commenced

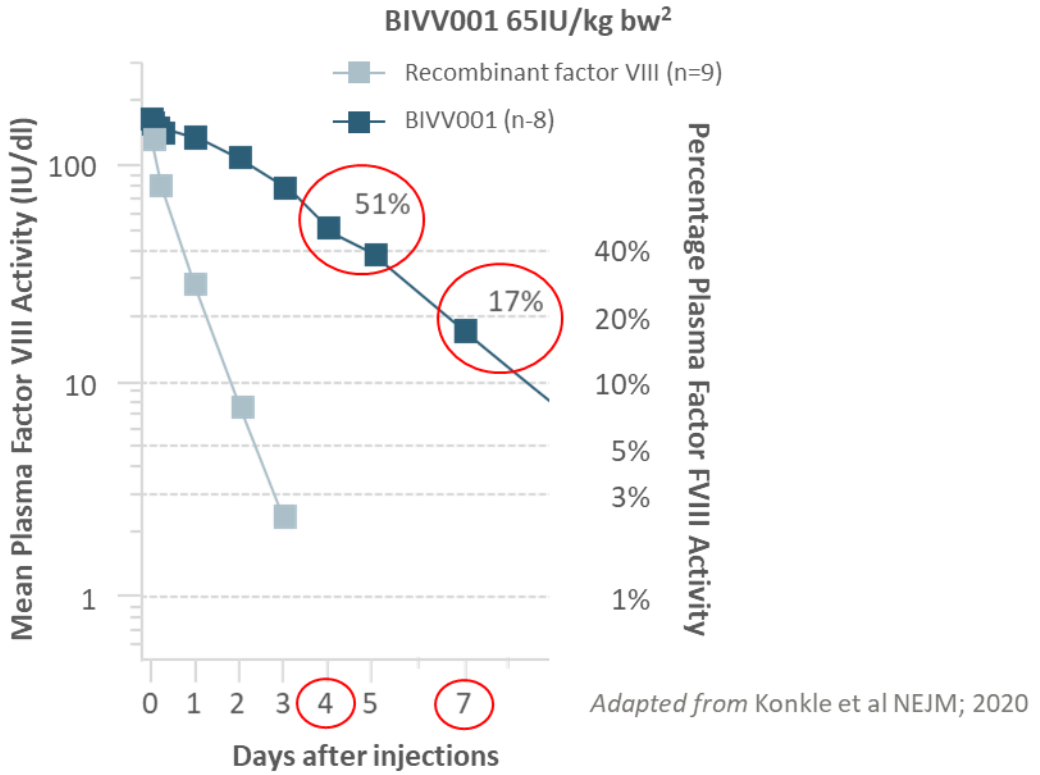
SEL-212 is being evaluated in a pivotal phase 3 programme, first patient randomised in September 2020 with topline data expected in 2H 2022



BIVV001 phase 1/2 results show high sustained factor activity levels

Structure of the BIVV001 Fusion Protein





Adapted from Konkle et al NEJM; 2020

"…it may become the **first choice among factor VIII** replacement products…" [2]

"…an authentic factor VIII is **more attractive than a procoagulant-activity mimetic** product…" [2]

BIVV001 is developed and commercialised in collaboration with Sanofi

XTEND-1: ongoing phase 3 study in severe hemophilia A

- 150 previously treated patients, >12y
- 1 year open label, interventional study
- Prophylactic and on-demand dose of 50 IU/kg QW
- Commenced December 2019

SOURCE: 1. Konkle BA et al., BIVV001 Fusion Protein as Factor VIII Replacement Therapy for Hemophilia A. N Engl J Med. 2020 Sep 10;383(11):1018-1027. doi: 10.1056/NEJMoa2002699. PMID: 32905674.
2. Pier M. **Mannucci**, M.D.. September 10, 2020. N Engl J Med 2020; 383:1068-1070. DOI: 10.1056/NEJMe2024545.

Clinical development of Gamifant for graft failure (start Q1 2021)

Large potential patient pool

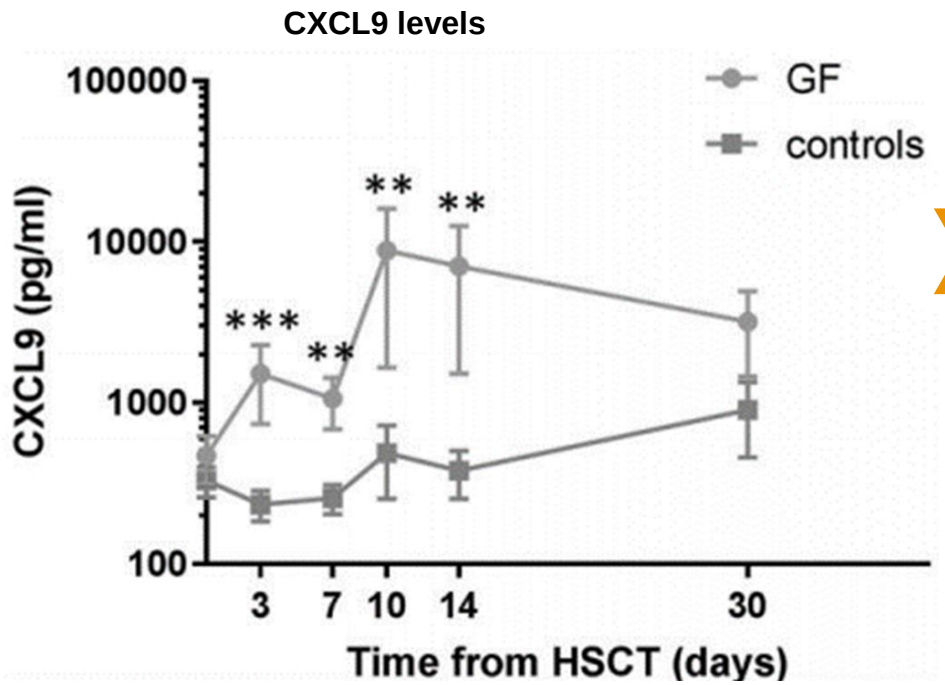
Total of ~24,000 patients (children and adults) undergoing hematopoietic stem cell transplantation (HSCT) every year with ~10% experiencing acute graft failure (aGF)

Strong scientific evidence

Increased serum levels of IFNγ and CXCL9 are predictors of acute graft failure

By day 3 after transplantation, patients developing graft failure had significantly elevated serum levels of CXCL9

Serum CXCL9 could be an early biomarker for the risk of graft failure



Two studies planned for Q1 2021

- **Interventional PoC study** to confirm appropriate emapalumab dose post HSCT to control IFNγ activity
- **Observational prospective study** to validate CXCL9 as a biomarker of GF in HSCT patients to enable phase 3 development

SOURCE: Merli et al., 2019. Haematologica. Volume 104(11):2314-2323

Research collaboration with bioMerieux to develop a CoDx for Gamifant in Graft Failure post HSCT

Research collaboration for a CXCL9 companion diagnostic assay

VIDAS™ CXCL9 for the prevention of graft failure post HSCT received **breakthrough device designation by FDA** in May 2020

- Fast turnaround (~1h)
- Easy to run (hands off)

Sobi/bioMérieux partnership for the development and commercialisation of **CXCL9 as a companion diagnostic assay on VIDAS™ platform**

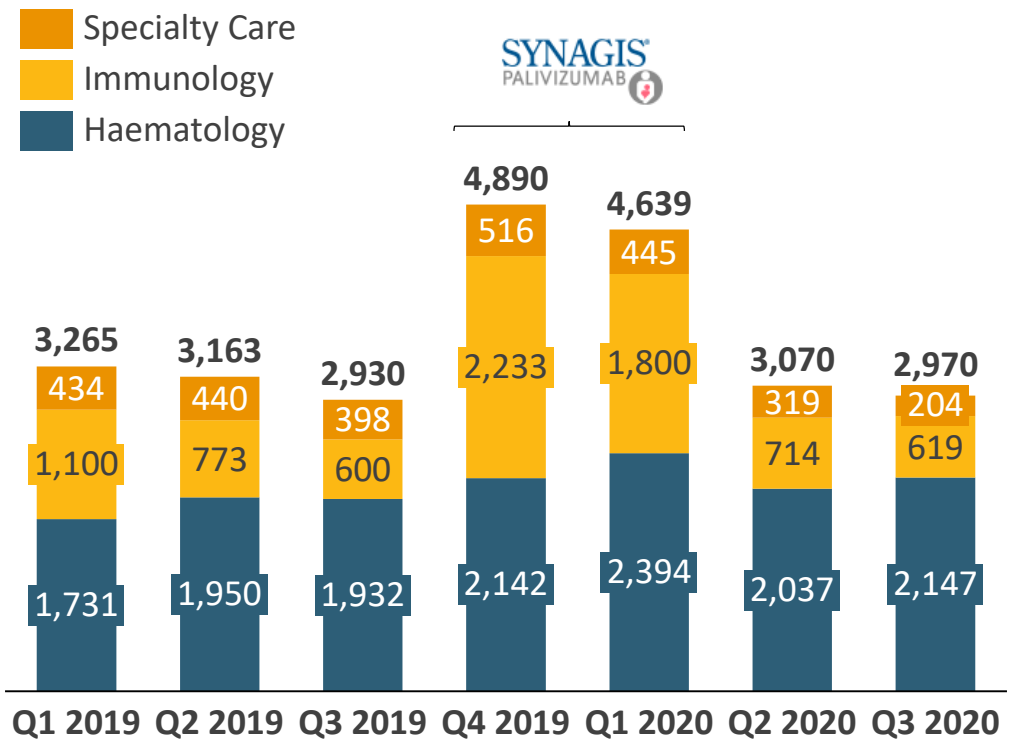
Could predict **graft failure following HSCT** as well as **future indications**

Note: CoDx = companion diagnostics; HSCT = Hematopoietic stem cell transplantation

Q3 2020 financial results

Q3 2020: Financial results

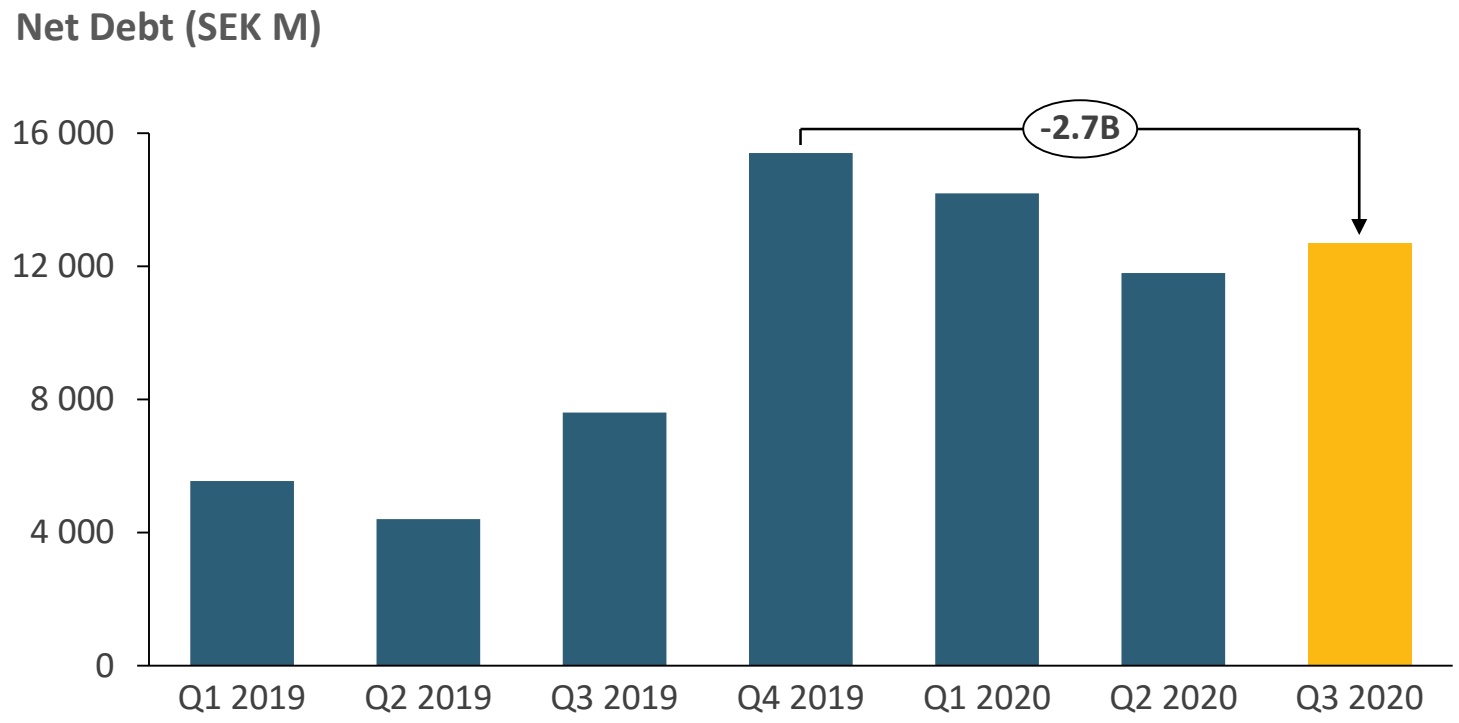
Total revenue (SEK M)



Amounts in SEK M	Q3 2020	Q3 2019	Change	Jan – Sept 2020	Jan – Sept 2020	Change	Full-year 2019
Total revenue	**2,970**	2,930	1%	**10,680**	9,358	14%	**14,248**
Gross profit	**2,339**	2,173	8%	**8,318**	7,080	17%	**10,913**
Gross margin[1]	**79%**	74%		**78%**	76%		**77%**
EBITA adjusted[1,2]	**933**	1,099	-15%	**4,124**	3,764	10%	**6,145**
EBITA margin adjusted[1,2]	**31%**	38%		**39%**	40%		**43%**
Profit for the period	**278**	542	-49%	**1,743**	1,944	-10%	**3,304**
Earnings per share, SEK adjusted[1,2,3]	**0.94**	1.84	-49%	**5.92**	6.98	-15%	**11.89**
Operating cashflow	**443**	995	-55%	**4,356**	2,658	64%	**3,634**
Net debt (+)/net cash (-)	**12,703**	7,606		**12,703**	7,606		**15,404**

[1]Alternative Performance Measures (APMs)
[2]EBITA full-year 2019 excluding non-recurring items; transaction costs related to the acquisition of Dova in Q4 of SEK 92 M, restructuring costs of SEK 157 M in Q2 and gain from divestment of SOBI005 in Q1 of SEK 37 M.
[3]EPS full-year 2019 excluding impairment of intangible assets of SEK 18 M related to restructuring in Q2.

Seasonal impact on cash flow in Q3, leverage < 2.0x



Acquisitions SYNAGIS PALIVIZUMAB gamifant emapalumab-lzsg Dova PHARMACEUTICALS SEL-212

Q3 2020 update

- **Operating cash flows** of SEK 0.4B
- **Net debt** increased by SEK 0.9B due to SEL-212 acquisition and settlement of Alprolix debt, offset by operating cash flow
- **Leverage** remains below 2.0x
- **Available liquidity** of SEK 6.5B

Q3 2020 Summary

We have forcefully responded to the COVID-19 pandemic

1

Secure health and safety of all employees

We encourage work from home
We cancelled all business travel

2

Safeguard supply of medication

We assess whether COVID impacts the supply of our medicines
We introduce mitigation measures where needed

3

Ensure access to treatment

We focus on patient support activities (e.g., access to care, digital channels with health care professionals)

4

Explore efficacy of anakinra in COVID

We support the generation of clinical evidence for Kineret as a potential treatment for COVID-19 patients

Financial outlook 2020[1] – updated

Revenue for the full-year 2020 is expected to be in the range of SEK 15,000—15,500 M.

EBITA is expected to be in the range of SEK 5,700—6,200 M.

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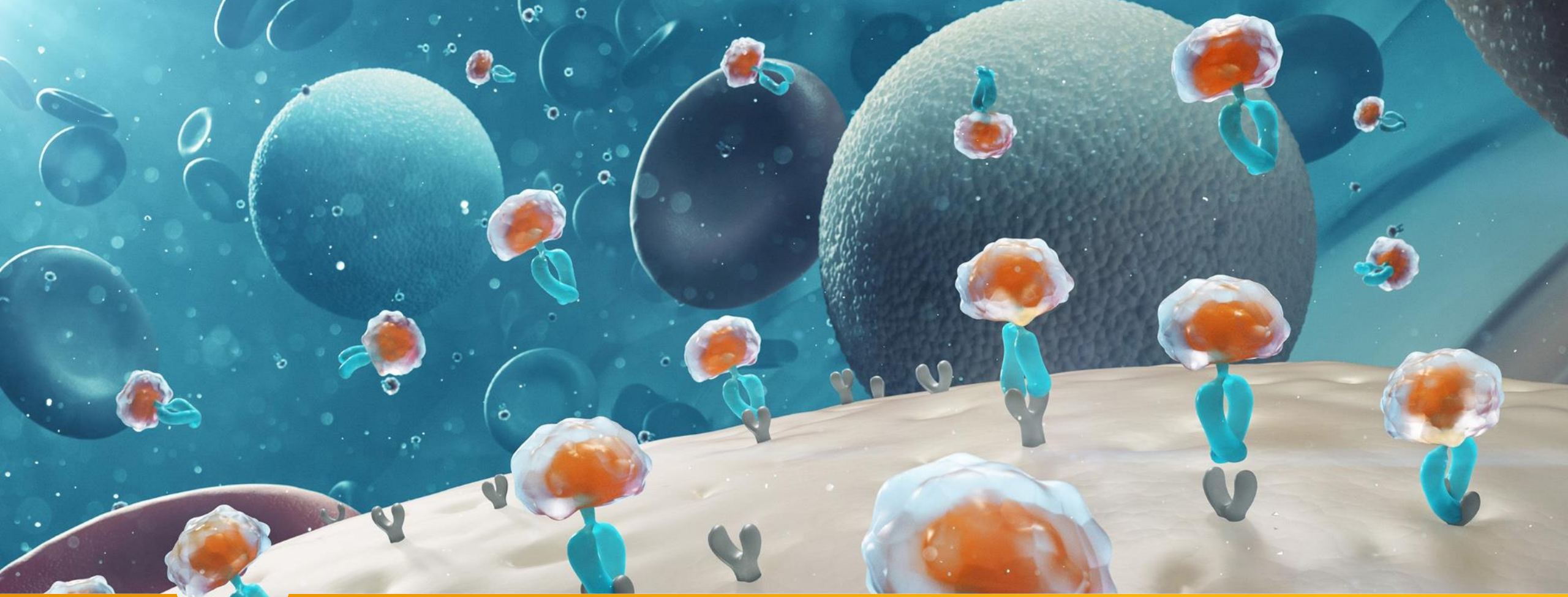
1. **Assuming exchange rates as of September 30 remain stable during the fourth quarter of 2020**



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