Sobi Capital Markets Day 2020

rare **strength**

10 December 2020

Welcome

Paula Treutiger

Head of Communications and Investor Relations

Forward-looking statements

In order to utilise the 'Safe Harbor' provisions of the United States Private Securities Litigation Reform Act of 1995, Swedish Orphan Biovitrum AB (publ) 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 AB (publ), 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.

Today’s presenters

Guido Oelkers
CEO

Henrik Stenqvist
CFO

Ravi Rao
Head of R&D and CMO

Norbert Oppitz
Head of Immunology and International

Today's agenda

Strategy and realising opportunities	Guido Oelkers	13:05
Innovation management at Sobi	Ravi Rao	13:30
Break		14:30
Internationalisation strategy	Norbert Oppitz	14:45
Financial update	Henrik Stenqvist	15:00
Wrap up and Q&A	Guido Oelkers	15:15

Strategy and realising opportunities

Guido Oelkers

CEO

Five years ago, Sobi looked very different

Business largely characterised by **in-licensing/ distributor agreements**

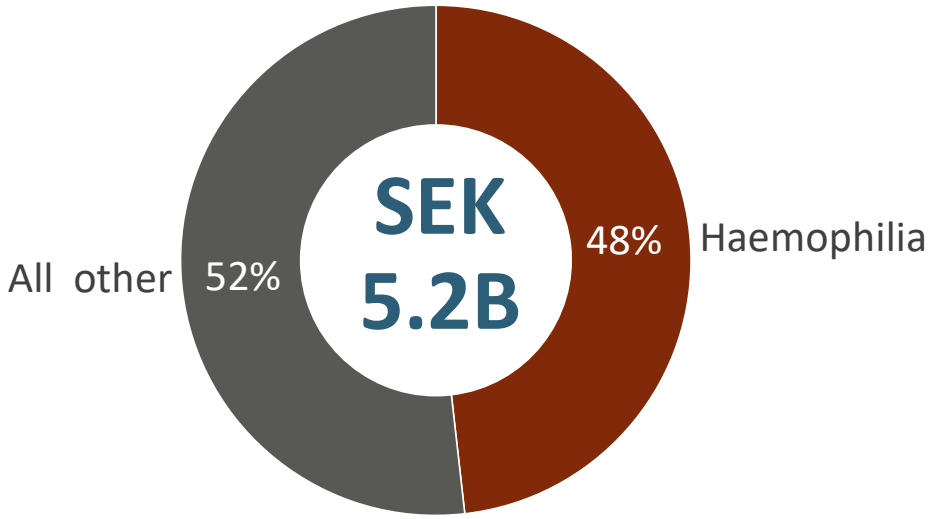
Significant **dependence on an emerging haemophilia portfolio**

Pipeline with **no big bets in rather early-stage R&D**

Europe-centric approach without significant footprint in the US

International business **sub-scale** in most geographies outside Europe

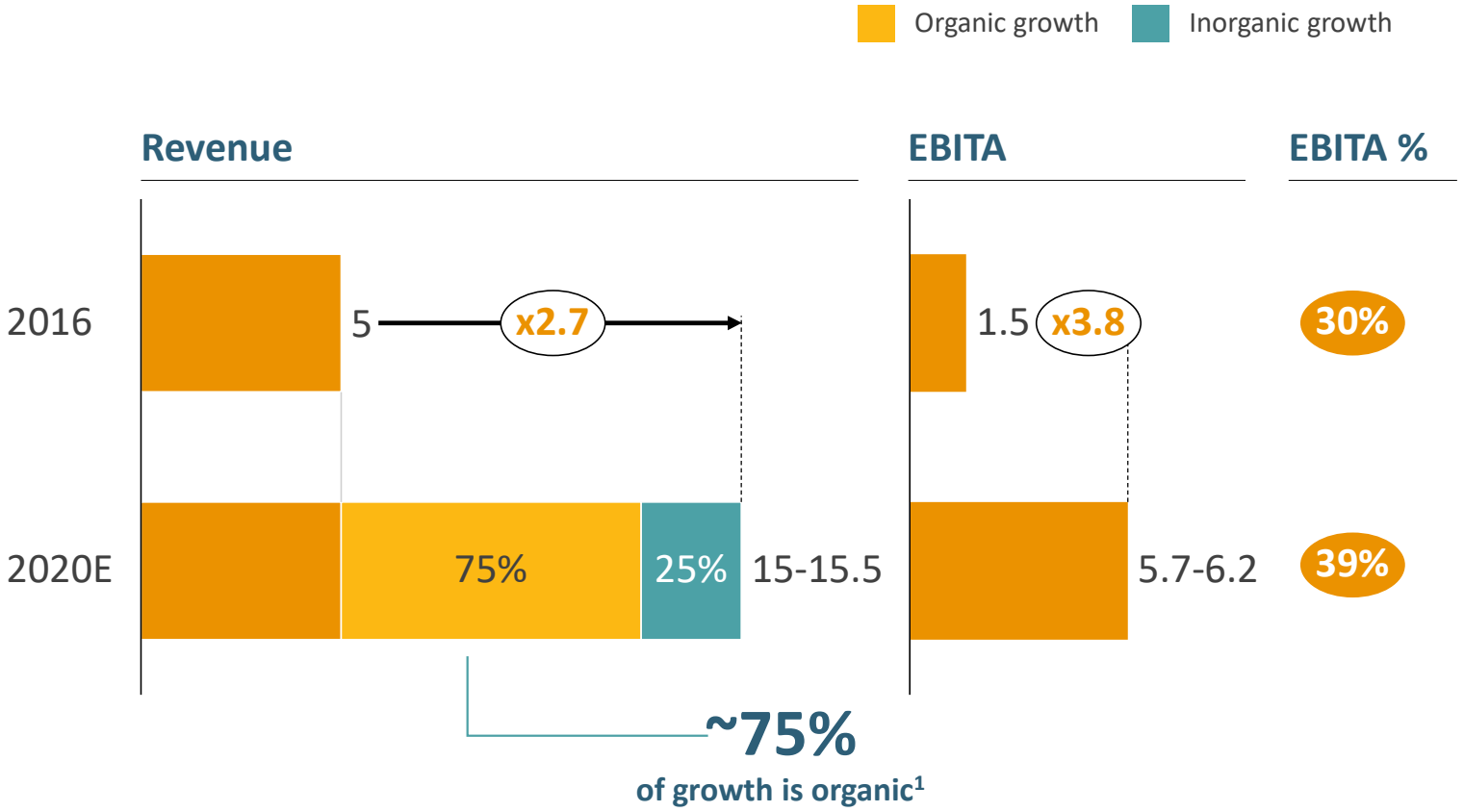
Revenue 2016



Source: Sobi

We have accomplished much in the past four years

SEK B



1. Inorganic growth defined as FY 2018 revenue for Synagis (USD 287M) and Doptelet (USD 10M)

Source: Sobi Annual Report 2016; 2020 guidance per Q3 2020 report

Committed to a continuous upgrade of our sustainability agenda

Sustainability strategy

Commitment to patients

- Our R&D is ethical and focused on medical need
- We expand access to treatment
- We are patientcentric & engage with our communities
- We contribute to knowledge to enhance the practice of medicine
- We focus on patient safety

Responsible behavior

- We have no tolerance for corruption
- We are transparent
- We source responsibly
- We develop our people and keep them safe and healthy
- We reduce our environmental footprint

Commitment to Agenda 2030 and the Paris Agreement

In 2017, we set out our strategy to build a regional leader in rare disease – a strategy on which we have forcefully delivered

Source: Sobi Annual Report 2017

1. We have made a significant impact in the haemophilia market

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

22%

Prophylaxis patient market share[1]

ALPROLIX™ [Coagulation Factor IX (Recombinant), Fc Fusion Protein]

35%

Prophylaxis patient market share[2]

#1

Ranked leader in haemophilia by healthcare professionals[3]

#1

in community and scientific engagement[4]

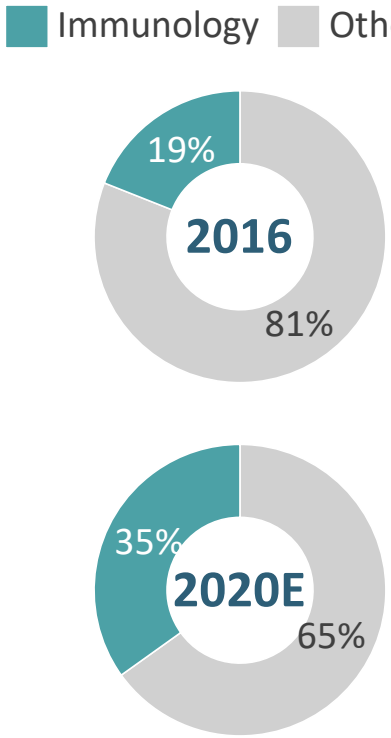
From **2023** we advance our leadership through BIVV001 as new normal

BIVV001 is developed and, if approved, will be commercialised in collaboration with Sanofi

1. Elocta FY sales for 2019 in relation to estimates based on MRB, Global Forecast of the Factor VIII Market by Region, Product Category and Company to 2022, November 2018; Data for Europe and Middle East and Africa 2. Internal data 3. Corporate Perception survey with HCPs treating haemophilia 2016 – 2019, N= 48 4. Internal data

2. Immunology: strong performance since establishment in 2016

Immunology revenue share

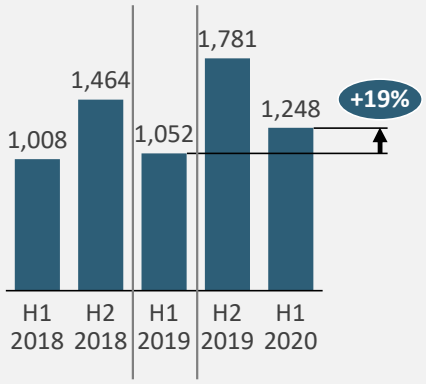


Driven by three major products

SYNAGIS® PALIVIZUMAB

Performing well under our ownership

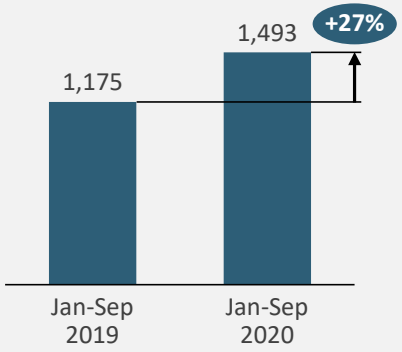
Revenue, SEK M



Kineret® (anakinra)

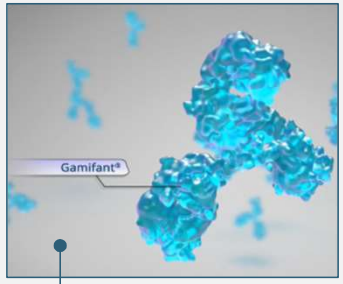
Growing strongly at 27% YTD

Revenue, SEK M



gamifant® emapalumab-lzsg

Established biology leading us into new fields

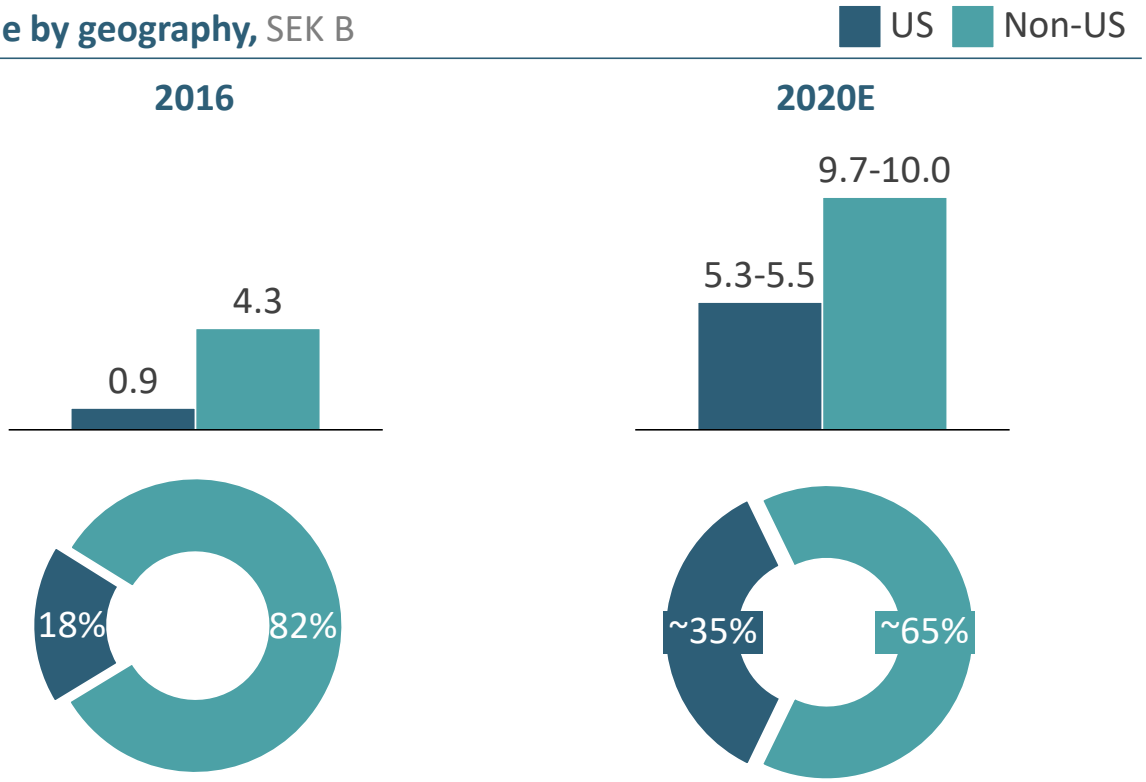


First targeted treatment for primary haemophagocytic lymphohistiocytosis (pHLH), a life-threatening hyper-inflammation disease

Source: Sobi

3. US business now a powerful platform for future growth

Revenue by geography, SEK B



Source: Sobi (2020E based on mid-point guidance provided by Sobi in Q3 2020 report)

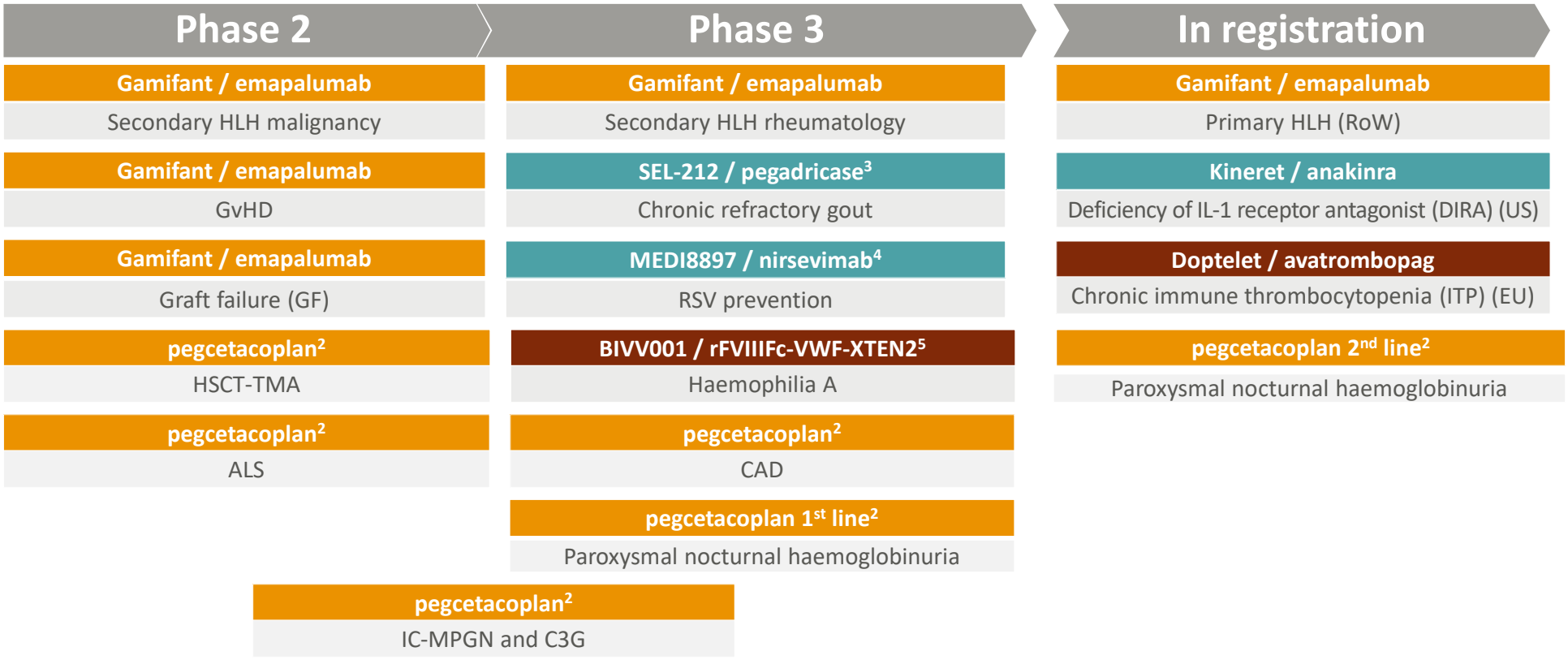
Background

Over the past five years, we have **increased our footprint in the US to ~35% of total sales**

Key drivers for increasing importance of US business were acquisitions of Synagis, Gamifant and Doptelet

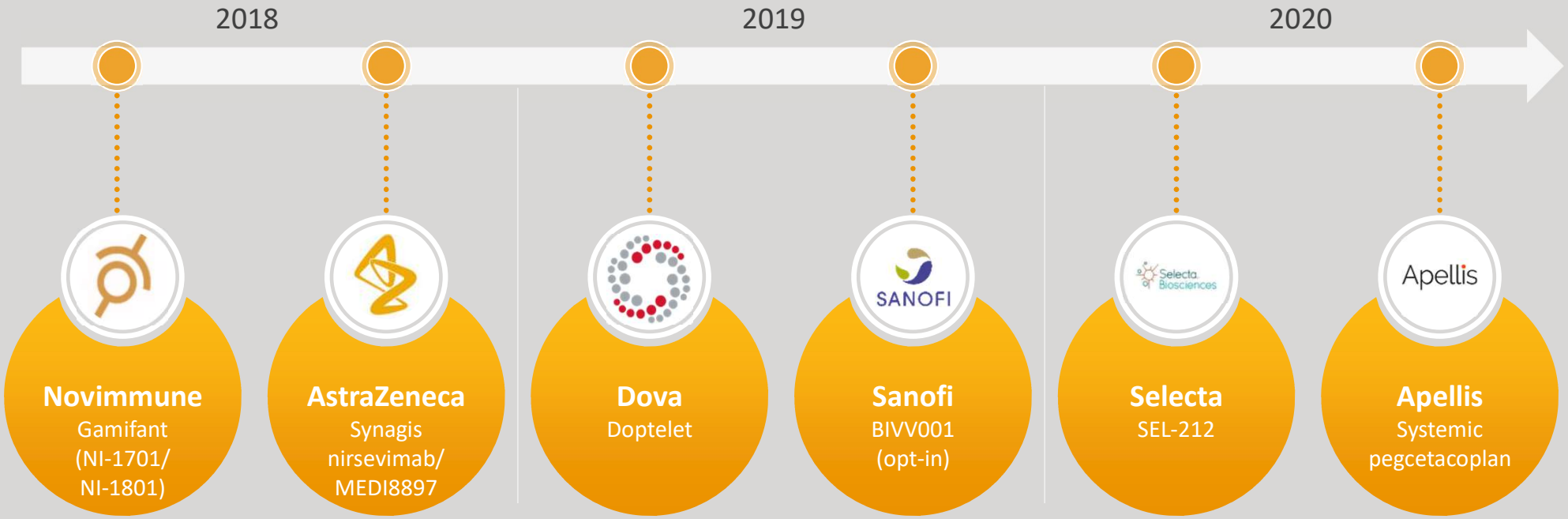
Going forward, **US will continue to grow** both in absolute and relative terms due to **Doptelet** and **Gamifant**

4. Capturing substantial value from our late-stage pipeline[1]



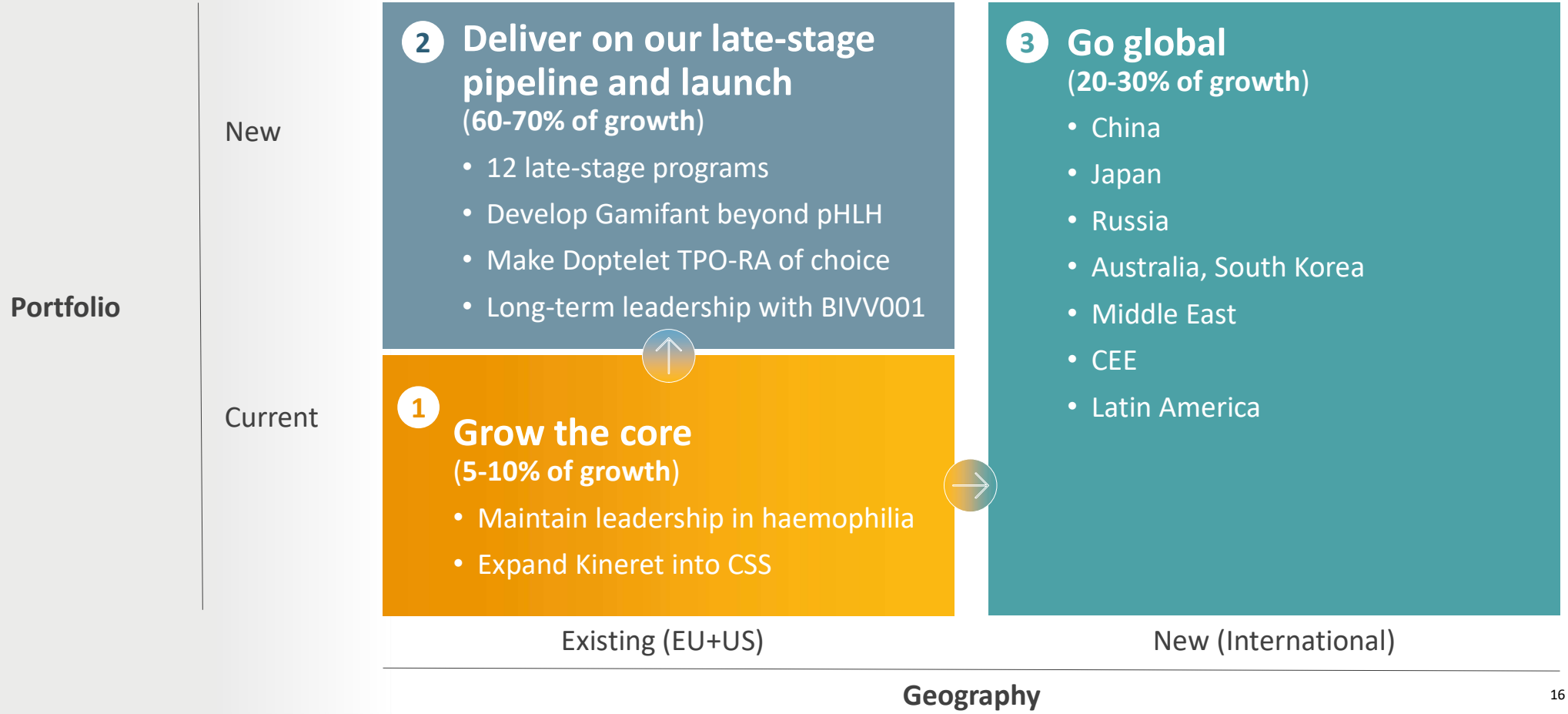
1. Not all programmes have been started 2. In collaboration with Apellis 3. Strategic licensing agreement with Selecta 4. Financial interest only, in collaboration with AstraZeneca 5. BIVV001 is developed and, if approved, will be commercialised in collaboration with Sanofi

M&A continues to be a key driver for our transformation



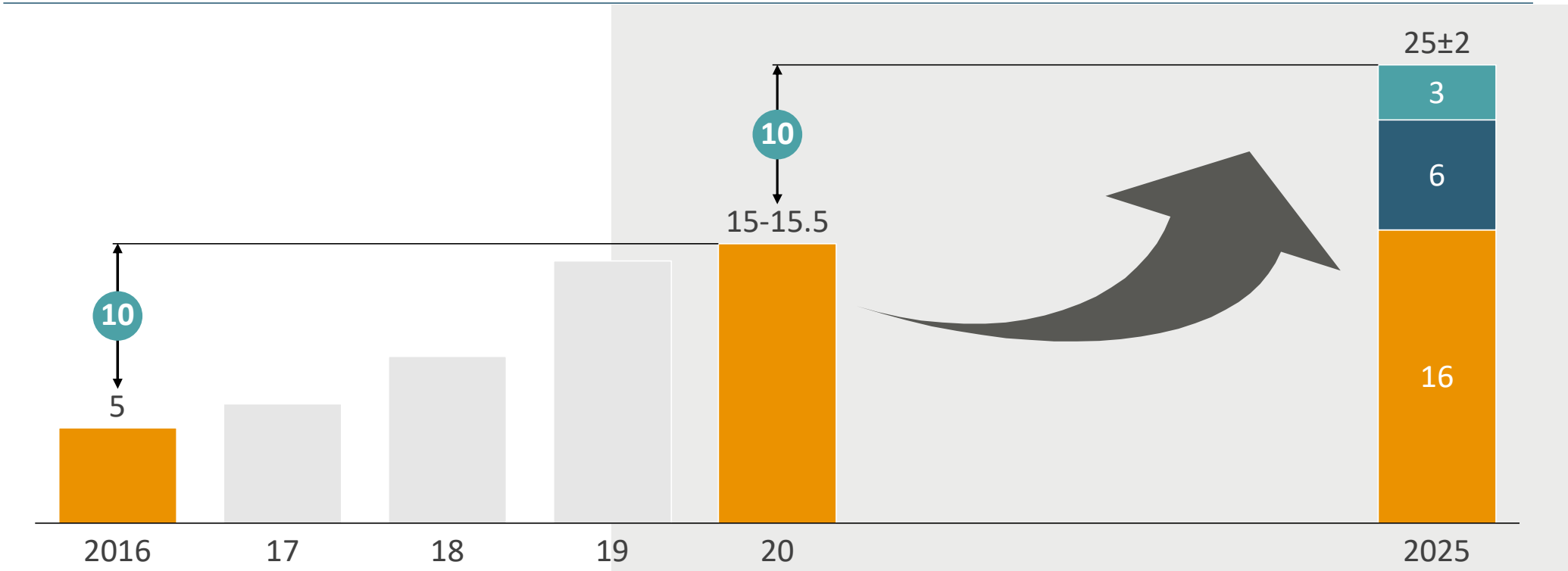
BIVV001 is developed and, if approved, will be commercialised in collaboration with Sanofi
Nirsevimab is being developed by AZ and commercialised by Sanofi. Sobi has an option to the rights to 50% of the profits and losses of the product in the US

How we will grow beyond today's core



For the coming five years, our new ambition is ‘25 by 25’

Revenue
in SEK B



1. Growth from core and pipeline in international markets
2. Growth from pipeline in current markets
3. Growth from core in current markets

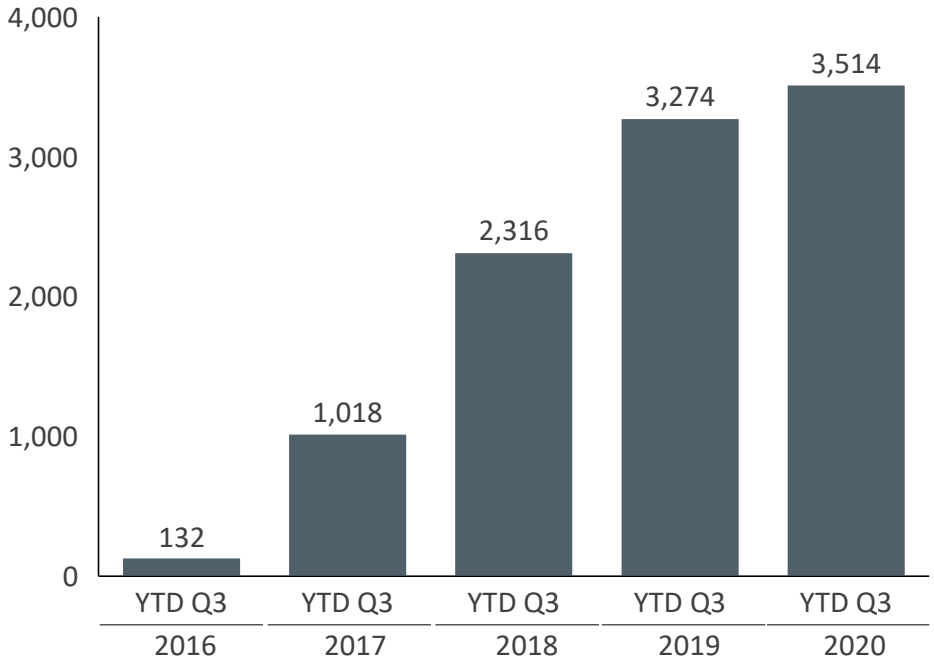
1. Committed to develop Elocta in a challenging environment

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

Opportunities

- New launches, such as Russia, opportunity of SEK 500 million
- Penetration upside in certain markets
- Large patient pool on SHL and plasma
- Clear long-term commitment to patients with BIVV001 and normalisation

YTD product sales[1]
SEK M



Headwinds

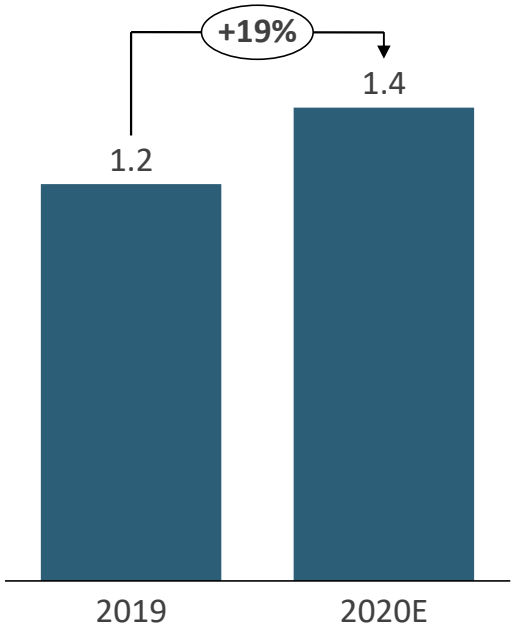
- Increasing competition from new EHLs and NFT
- Price pressure as a result of increasing competition and post COVID-19 cost constraints
- Access restrictions and reduction in per-capita consumption – COVID-19 prohibiting patients and pharma from visiting physicians and reducing surgical procedures

1. Excluding royalties

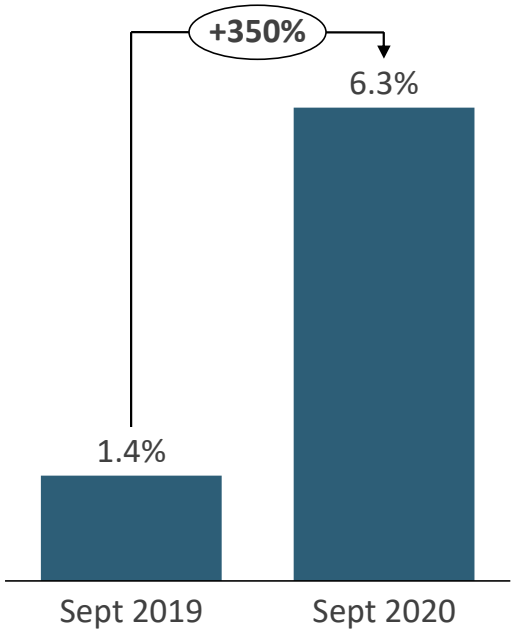
Source: Sobi

1. Creating material growth from a strong trajectory and clear differentiation

US TPO-RA[1] market
USD B



Sobi's market share in the US
Adult ITP only



1. Thrombopoietin receptor agonist 2. Approved for ITP in the US and awaiting approval in the EU. See prescribing information in the applicable territory for the approved indication. 3. Approved for patients with CLD and thrombocytopenia undergoing a procedure in US, Europe and China. See prescribing information in the applicable territory for the approved indication.

Source: Symphony Monthly Data (Sep 2020), RFT 11.4.20

See prescribing information in the applicable territory for the approved indication.

sobi

Doptelet® (avatrombopag) tablets

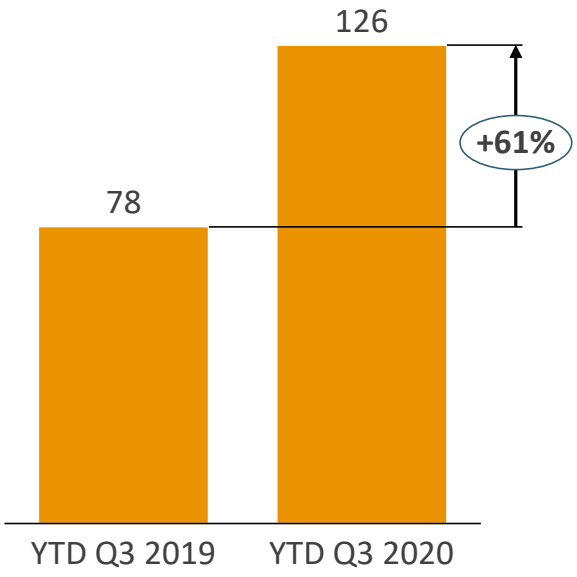
Doptelet is a small molecule **thrombopoietin receptor agonist** used to treat thrombocytopenia in **ITP[2]** and **CLD[3]**

As a **cost-effective oral treatment** with high efficacy and administered with food and without dietary restrictions, it is **clearly differentiated** against competitors and **well on the way to becoming the TPO-RA of choice**

We **enable broader access** to Doptelet through expansion into **new markets,** including in Europe and through our **international division** including Japan[2,3]

2. Developing Gamifant: a strong trajectory into the future

Development of Gamifant over time,
in thousands of mg



Source: Sobi

HLH

Maximise in pHLH

Expand geographically

Drive disease awareness

Expand into sHLH

Develop sHLH from rHLH and mHLH to iHLH

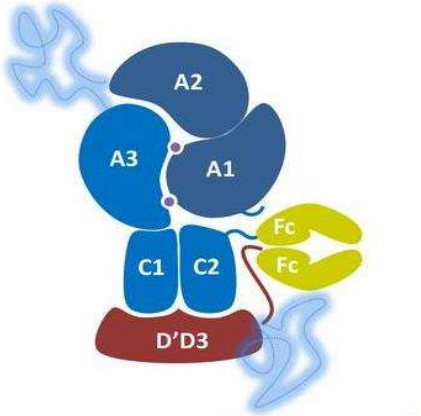
HSCT

Move to the next frontier: GF & GvHD

Develop GF with companion diagnostic

2. We opted in early[1] for **BIVV001** – to drive our leadership in haemophilia into the future

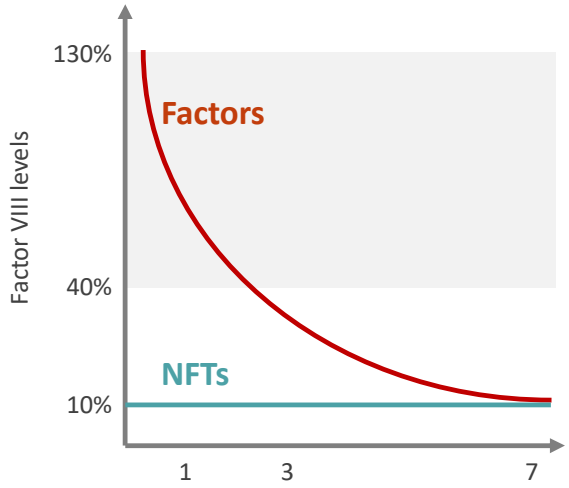
Extending half-life by factor of 3-4



Builds on the **Fc-fusion** technology – same as for Elocta – with added domains of **von Willebrand factor** and **XTEN®[2]** polypeptides

Improving patient QoL through once-weekly dosing

Illustrative



High sustained levels of factor VIII activity with **once-weekly dosing** with BIVV001

Overcoming current limitations in haemophilia treatments

BIVV001 used as monotherapy, whereas NFT require factor to treat breakthrough bleeds

BIVV001 has the potential to achieve more normal factor VIII activity allowing for active lives, whereas NFTs do not

BIVV001 is developed and, if approved, will be commercialised in collaboration with Sanofi

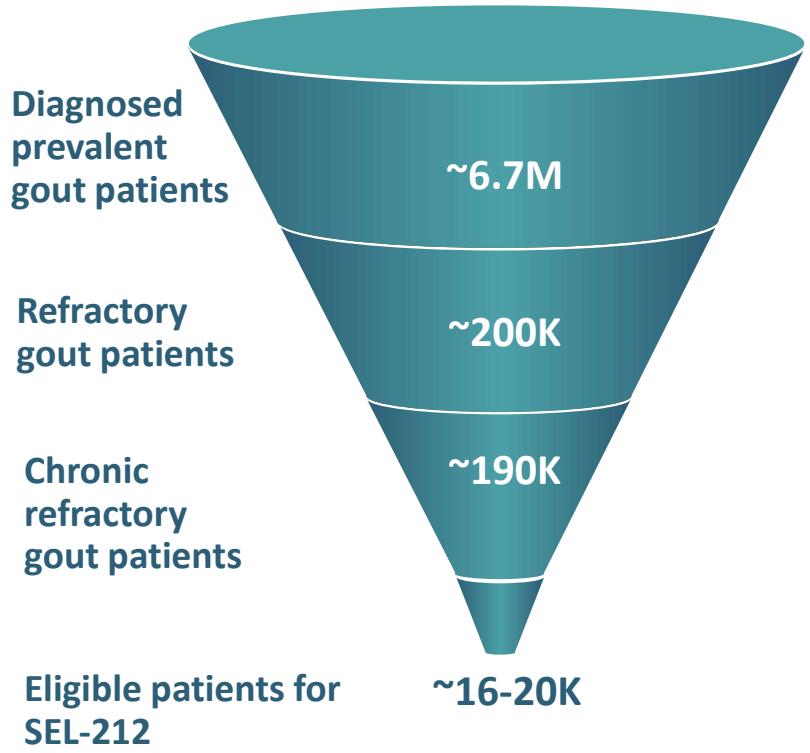
1. In September 2019, Sobi exercised early opt-in for the development and commercialisation of BIVV001, an investigational factor VIII therapy with the potential to provide extended protection from bleeds with once-weekly dosing for people with haemophilia A. Sobi and Sanofi also collaborate on the development and commercialisation of Alprolix and Elocta/ELOCTATE. Sobi has final development and commercialisation rights in the Sobi territory (essentially Europe, North Africa, Russia and most Middle Eastern markets). Sanofi has final development and commercialisation rights in North America and all other regions in the world excluding the Sobi territory and has manufacturing responsibility for Elocta/ELOCTATE and Alprolix. 2. XTEN is a registered trademark of Amunix Pharmaceuticals, Inc.

2. SEL-212 gives us a substantial opportunity in immunology

SEL-212's unique value proposition

- Reduced immunogenicity
- Sustained efficacy
- Greater reduction in SUA levels
- Better control in patients with tophi
- Monthly dosing

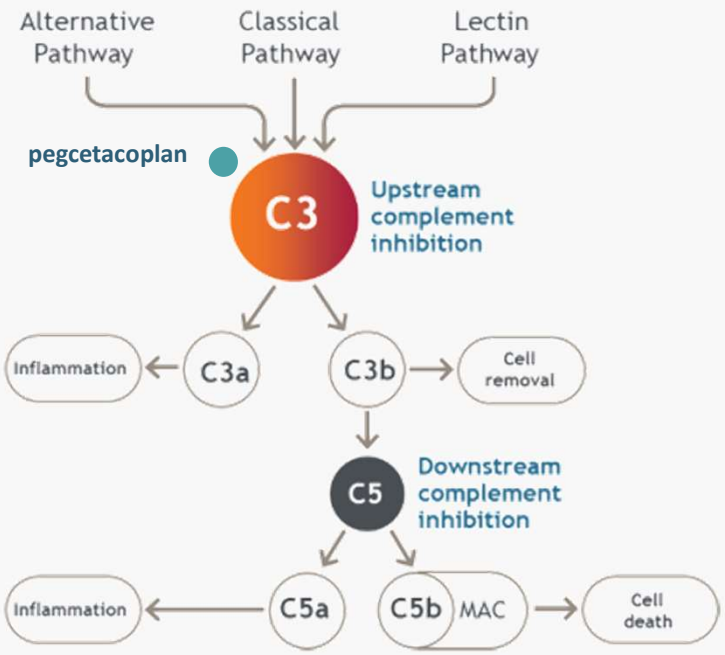
Patient population US (2030)



Source: Sobi

2. The partnership with Apellis is a great example of how we have expanded our late-stage pipeline with multiple opportunities

C3 has a central role in the complement cascade

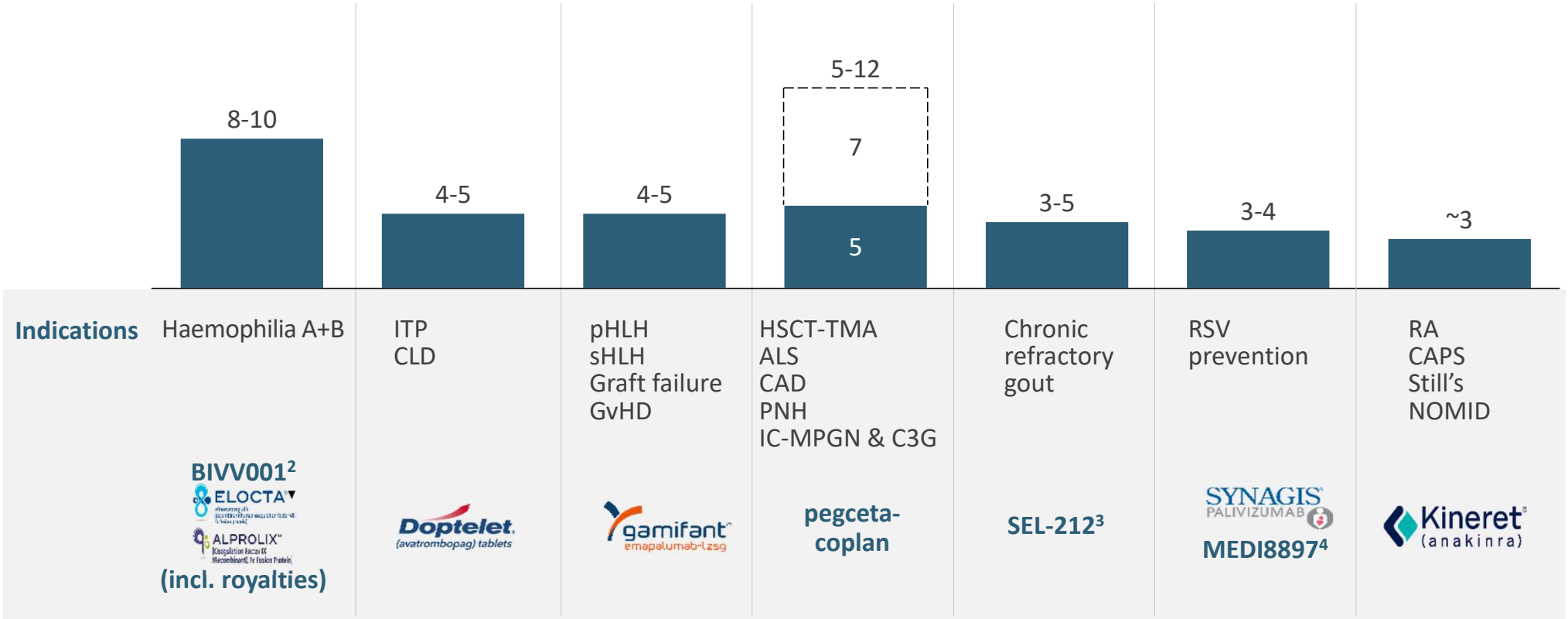


Indications	Patient opportunity ex-US (Sobi territory)
PNH	~9,000-11,000
CAD	~7,000-9,000
HSCT-TMA	~8,000-10,000
IC-MPGN/C3G	~6,000-8,000
ALS	~200,000

Source: Sobi

2. Estimated peak sales of our portfolio

Estimated peak sales[1], SEK B



BIVV001 is developed and, if approved, will be commercialised in collaboration with Sanofi

1. Peak sales relate to revenue potential from Sobi territories by product 2. BIVV001 is currently under clinical investigation and the safety and efficacy have not been evaluated by any regulatory authority 3. US and Europe 4. Sobi has only financial rights, peak sales based on analyst report

Source: Sobi

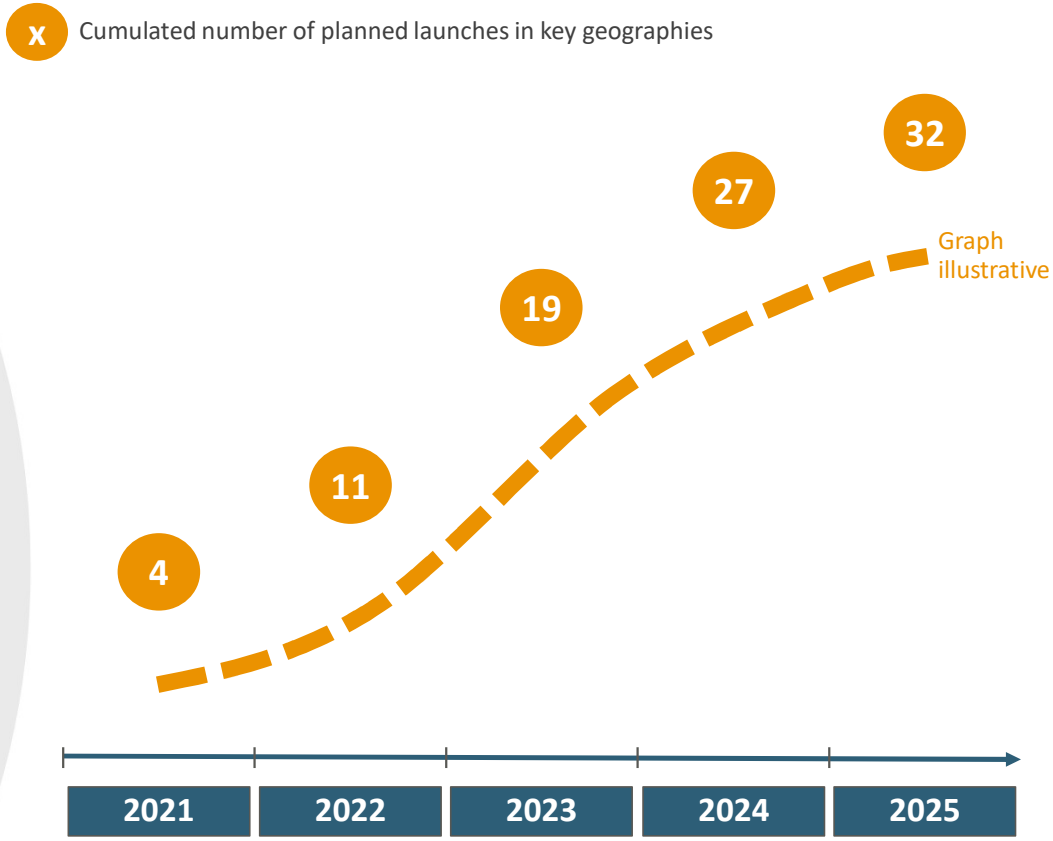
2. Capturing substantial value from our late-stage pipeline

5

Key geographies[1]

32

Launches in key geographies by 2025

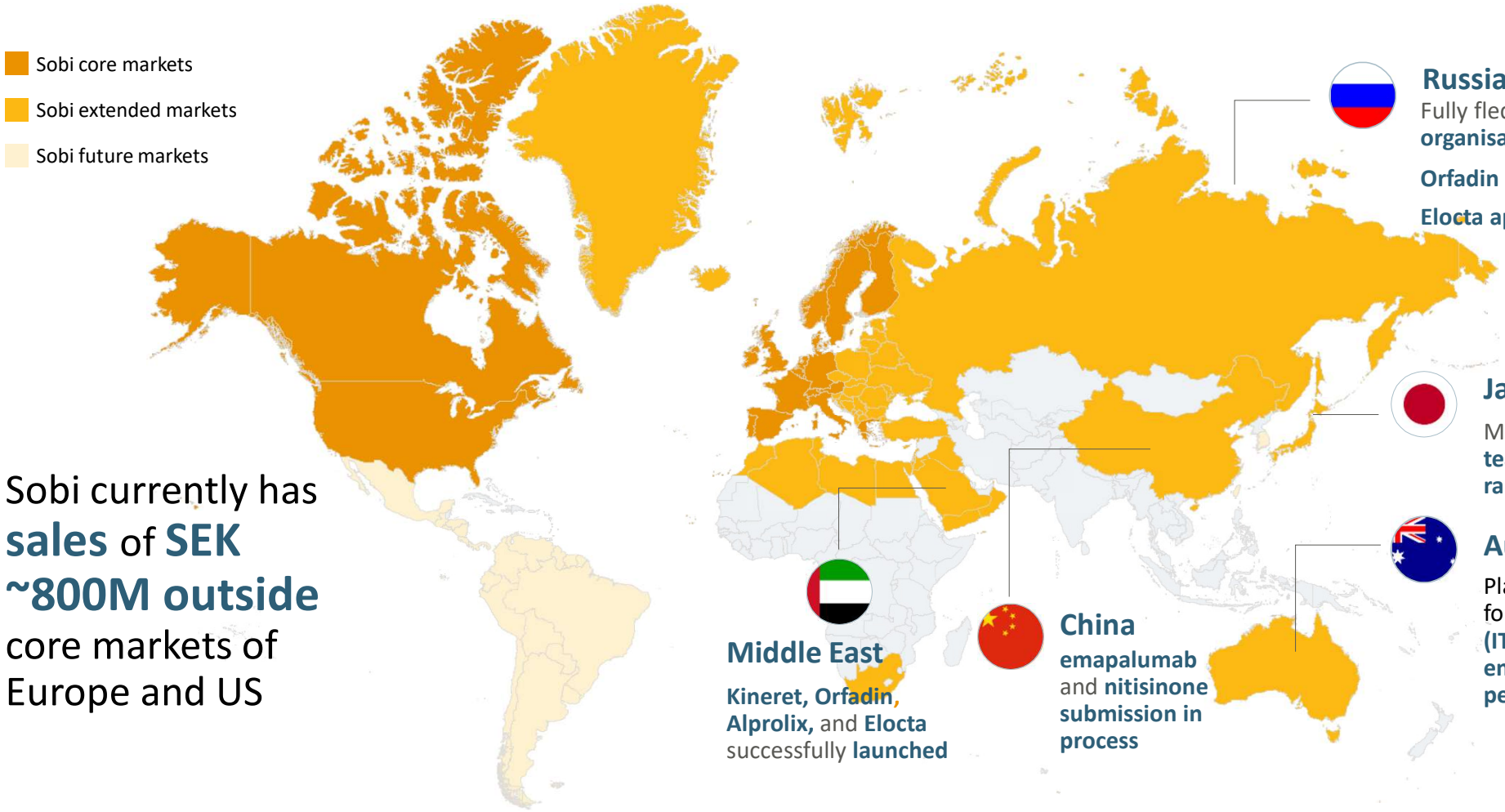


Examples of indications (illustrative)

- avatrombopag, ITP
- avatrombopag, CLD
- pegcetacoplan[2], PNH
- pegcetacoplan[2], CAD
- pegcetacoplan[2], IC-MPGN / C3G
- pegcetacoplan[2], HSCT-TMA
- emapalumab, GvHD
- emapalumab, pHLH & sHLH
- SEL-212[3], chronic refractory gout
- BIVV001[4], HA
- nirsevimab[5], RSV prevention

1. US, Europe, China, Japan, Russia 2. In collaboration with Apellis 3. Strategic licensing agreement with Selecta 4. In collaboration with Sanofi 5. Financial interest only, in collaboration with Astra Zeneca

3. We are building the next geographic growth platforms



Sobi currently has **sales** of **SEK ~800M outside** core markets of Europe and US

In summary: we are now transitioning Sobi from a regional to a global leader in rare disease

Innovation management at Sobi

Ravi Rao

Head of R&D and CMO

Building on Sobi's rare **strength** so we can transform patients' lives

Innovative and differentiated medicines

- Medicines with novel mechanisms of action: first in class or best in disease
- Enabling a step change in therapy for unmet medical need

At the intersection of haematology and immunology

- Lead haematology, build immunology and reap synergies between the two
- Understanding the needs of patients with rare disease across the world

Leadership in medicines development

- Multiple indications and integrated life cycle management
- Use of digital health, companion diagnostics and genetics

We currently have 6 products in development, with 12 programs[1]

Phase 2	Phase 3	in Registration
Gamifant / emapalumab Secondary HLH malignancy	**Gamifant / emapalumab** Secondary HLH rheumatology	**Gamifant / emapalumab** Primary HLH RoW
Gamifant / emapalumab GvHD	**SEL-212 / pegadricase[3]** Chronic refractory gout	**Kineret / anakinra** Deficiency of IL-1 receptor antagonist (DIRA) (US)
Gamifant / emapalumab Graft failure (GF)	**MEDI8897 / nirsevimab[4]** RSV Prevention	**Doptelet / avatrombopag** Chronic immune thrombo-cytopenia ITP EU
pegcetacoplan[2] HSCT-TMA	**BIVV001 / rFVIIIFc-VWF-XTEN2[5]** Haemophilia A	**pegcetacoplan 2nd line[2]** Paroxysmal nocturnal hemoglobinuria
pegcetacoplan[2] ALS	**pegcetacoplan[2]** CAD	
	pegcetacoplan 1st line[2] Paroxysmal nocturnal hemoglobinuria	
	pegcetacoplan[2] IC-MPGN and C3G	

1. Not all programs have been started 2. In collaboration with Apellis 3. Strategic licensing agreement with Selecta 4. Financial interest only, in collaboration with Astra Zeneca 5. BIVV001 is developed and, if approved, will be commercialised in collaboration with Sanofi

Six innovative and differentiated medicines to adress patient need across the spectrum of haematology and immunology

Lead in haematology

BIVV001

Bioengineered to allow for more normal factor levels with weekly dosage

Doptelet (avatrombopag) tablets

Convenient oral thrombopoietin Receptor Agonist

Overlap between haematology and immunology

gamifant emapalumab-lzsg

First-in-class treatment for IFNγ-driven diseases

pegcet-acoplan

First-in-class treatment for C3 driven diseases

Grow in immunology

SYNAGIS palivizumab

MEDI8897

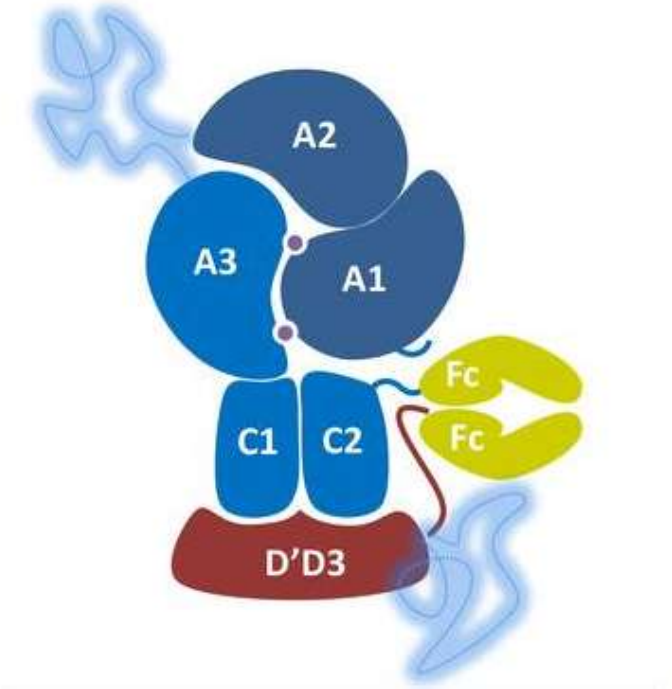
Single-injection prophylaxis for all infants in their first RSV season

SEL-212

Novel combination for the treatment of chronic refractory gout

BIVV001 is developed and, if approved, will be commercialised in collaboration with Sanofi
MEDI8897 is being developed by AZ and commercialized by Sanofi. Sobi has an option to the rights to 50% of the profits and losses of the product in the US

BIVV001 is designed for more normal FVIII levels in HA



Mode of Action

- Built on Fc fusion technology with added
  - von Willebrand factor domains
  - XTEN[1] polypeptides
- To result in high and sustained factor levels in circulation

Designed profile

First product with once weekly dosing and factor level in normal range of FVIII for a sustained period in the week

High levels of physical activity possible with low risk for bleed (e.g. basketball, rugby[2]) when factor level in normal range

No additional product needed: single administration for prophylaxis through to surgery

BIVV001 is developed and, if approved, will be commercialised in collaboration with Sanofi. BIVV001 is currently under clinical investigation and the safety and efficacy have not been evaluated by any regulatory authority.

1. XTEN is a registered trademark of Amunix Pharmaceuticals, Inc. 2. Broderick CR, Herbert RD, Latimer J, et al. Association Between Physical Activity and Risk of Bleeding in Children With Hemophilia. *JAMA*. 2012;308(14):1452–1459. doi:10.1001/jama.2012.12727

BIVV001 has the potential to normalise FVIII levels

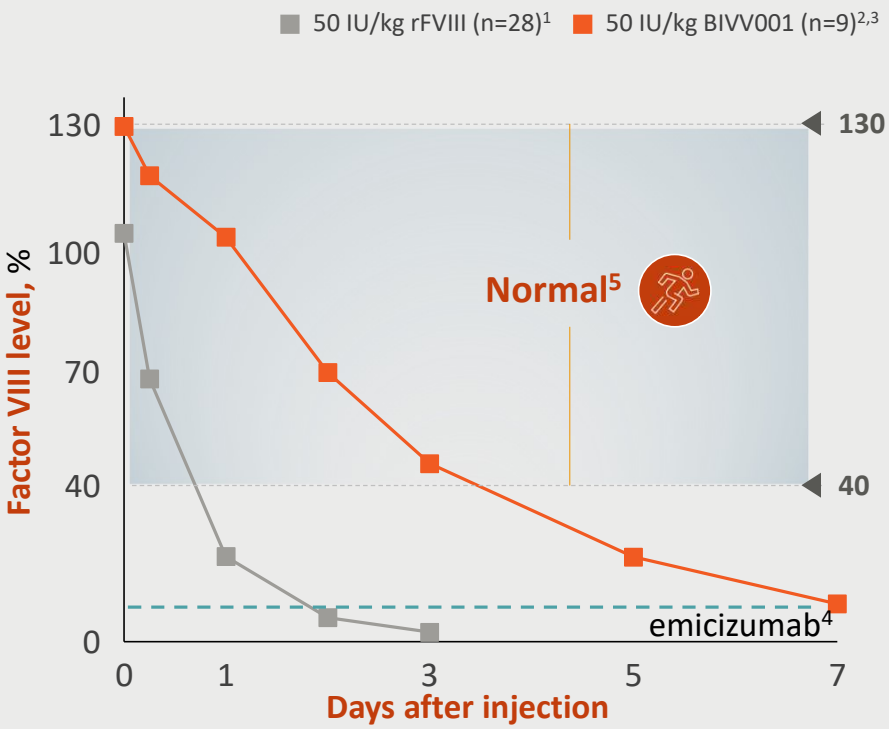
Disease Background:

- Haemophilia A is a genetic deficiency in clotting factor VIII
- Results in extensive bleeds, joint damage, low QoL

Differentiation:

- BIVV001 has the potential to enable more normal factor activity levels for the majority of the week
- Short-half life factor therapies do not deliver high sustained factor activity levels
- With emicizumab FVIII normalization is not possible
- Non-factor therapies, e.g., emicizumab, show no evidence of joint protection

Normalisation of factor VIII levels



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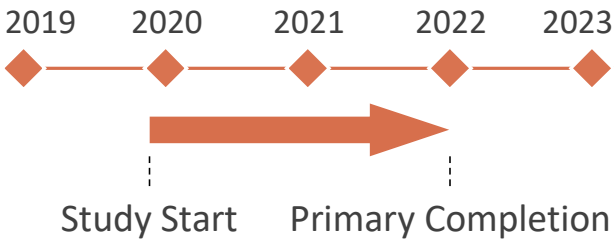
1. Mahlangu et al. Blood 2014 2. Lissitchkov T et al. Haemophilia 2020;26(S2):55 3. Lissitchkov T et al. Blood 2019;134(S1):625; 4. Equivalent FVIII level, based on Lenting P et al, ISTH 2019, Lenting P et al, Blood Adv. 2020 5. A. Srivastava, E. Santagostino, et al., WFH Guidelines for the Management of Hemophilia, 3rd edition. Haemophilia. 2020;26(Suppl 6):1–158

Two major phase 3 studies underway - BIVV001 expected to be submitted in US in 2022

Study timeline

Key design aspects

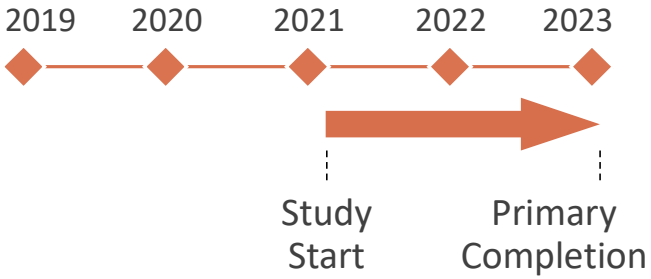
Phase 3 in adults & adolescents
N= 150
≥12 year old



Arms: Multi-centre, open-label, non-randomized 2-arm (prophylaxis and on-demand); previously treated patients with severe haemophilia A

Primary endpoint: Annualized bleeding rate (ABR) in prophylaxis treatment arm (time frame: baseline to 52 weeks)

Phase 3 in Paediatric
N= 65
<12 year old



Arms: Multi-centre, open-label 1-arm; previously treated patients with severe haemophilia A

Primary endpoint: Inhibitor development (time frame: baseline to 52 weeks)

Expected launch[1]

2023 Haemophilia A

1. Only territories relevant to Sobi displayed

BIVV001

Doptelet (avatrombopag) tablets

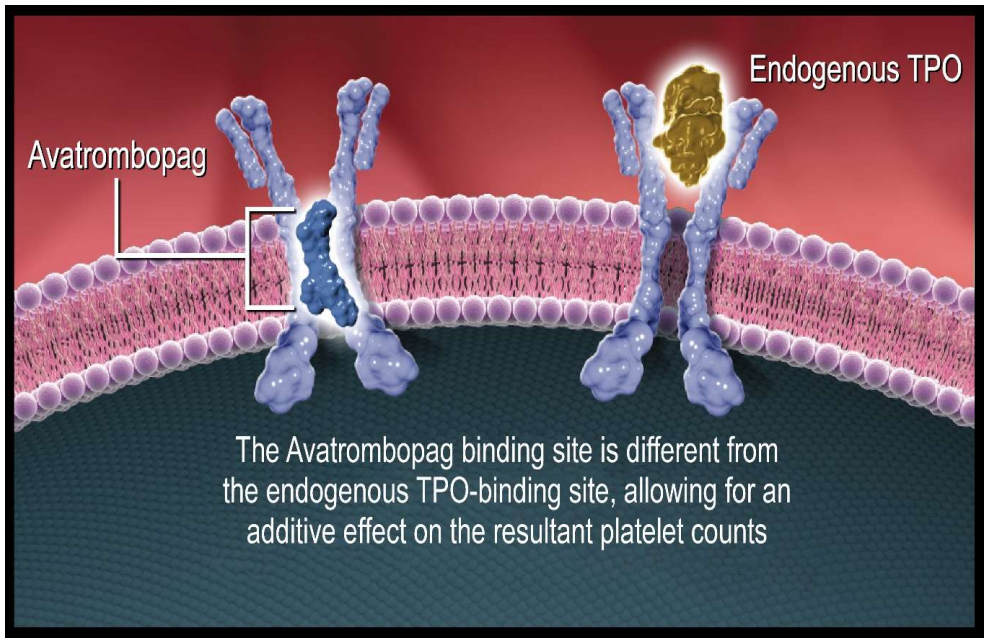
SEL-212

MEDI 8897

gamifant emapalumab-lzsg

pegceta-coplan

Doptelet could be a best-in-class treatment for CLD and ITP



Mode of Action

- Second-generation small-molecule thrombopoietin receptor agonist (TPO-RA) stimulating platelet production
- Stimulates proliferation and differentiation of megakaryocytes from bone marrow progenitor cells resulting in increased production of platelets

Profile

- Once daily oral dosing
- No dietary restrictions - convenient administration with food/regular meals
- No hepatotoxicity

Doptelet can increase platelet numbers to significantly reduce need for platelet transfusions

Disease background:

- Low levels of blood platelets can cause spontaneous bleeding
- Thrombocytopenia is caused by, e.g.:
  - Decreased production (e.g., chronic liver disease (CLD))
  - Increased destruction (e.g., immune thrombo-cytopenic purpura)

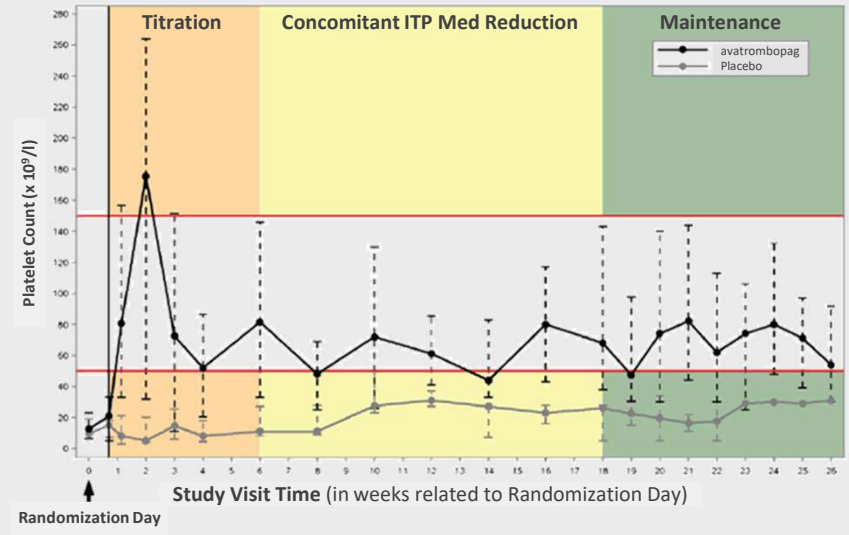
Current TPO-RA treatments:

- eltrombopag, romiplostim

Differentiation:

- Patient convenience: Doptelet as the only once daily oral TPO-RA approved for treatment of ITP without dietary restrictions[1]
- No hepatotoxicity

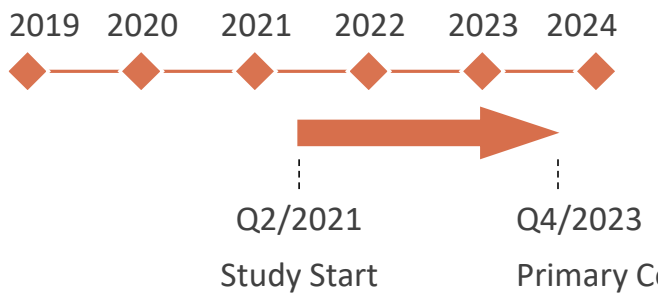
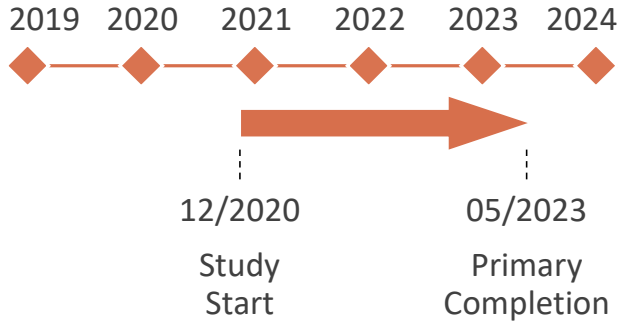
Median platelet counts during trial[2]



avatrombopag (n)	32	30	32	32	32	32	30	30	30	28	25	28	25	24	22	25	22	23	23	23	22
Placebo (n)	17	17	17	17	16	16	14	7	4	3	3	3	3	2	2	2	2	1	1	1	1

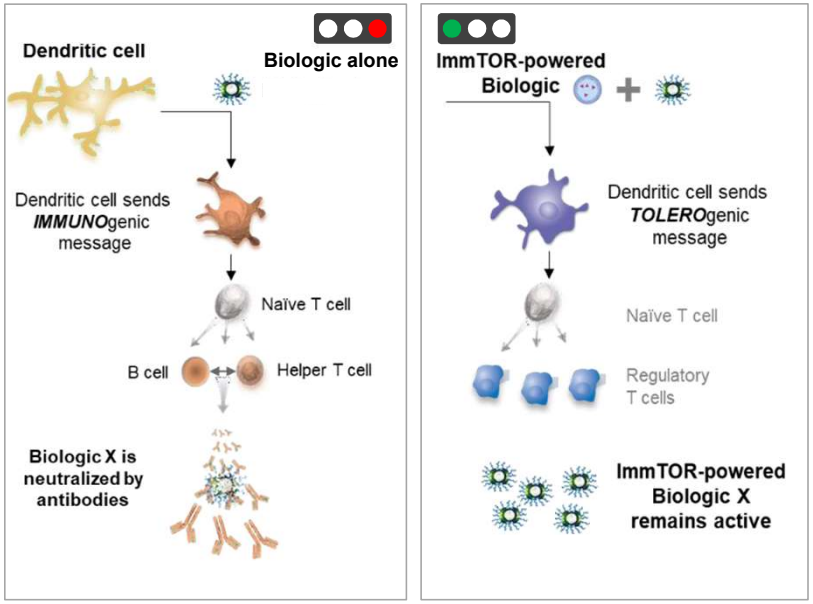
1. Approved for ITP in the US and awaiting approval in the EU See prescribing information in the applicable territory for the approved indication 2. Jurczak, et al. Br J Haematol. 2018;183(3):479-490

We have finished the majority of the development with focus on ongoing lifecycle studies

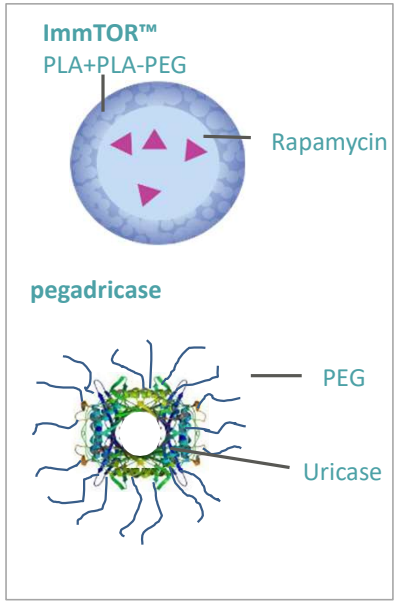
	Study timeline	Key design aspects	Expected launch
Early use study adults ITP N= 75		**Arms:** Pbo – 2:1 randomization with 6-month duration of treatment **Primary endpoint:** Continuous weeks of response (PC>50)	2021 ITP
Phase 3 in paediatric ITP N= 72 <18 year old		**Arms:** Randomized 3:1 to receive either avatrombopag or placebo (orally once daily for 12 weeks) **Primary endpoint:** Proportion of subjects achieving at least 6 out of 8 weekly platelet counts ≥50×10^9/L during the last 8 weeks of 12-week treatment, in the absence of rescue medication	

SEL-212 is a novel combination product candidate to improve treatment of chronic refractory gout

ImmTOR™[1] has the potential to enable sustained therapeutic activity of biologic therapies and unlock their potential



SEL-212: ImmTOR™ co-administered with pegadricase



Mode of Action

- ImmTOR™ co-administered with pegylated uricase to reduce serum uric acid (SuA) and inhibit antibody development

Designed profile

- Once-monthly infusion
- Reduced immunogenicity
- Sustained efficacy

1. ImmTOR™ is a registered trademark by Selecta Biosciences, Inc.

Phase 2 results show potential for greater reduction of SUA

Disease Background:

- Chronic gout: serum uric acid levels remain elevated and flares continue despite standard treatment
- Painful inflammatory arthritis: mono- or polyarticular
- Presence of subcutaneous tophi

Current Treatments:

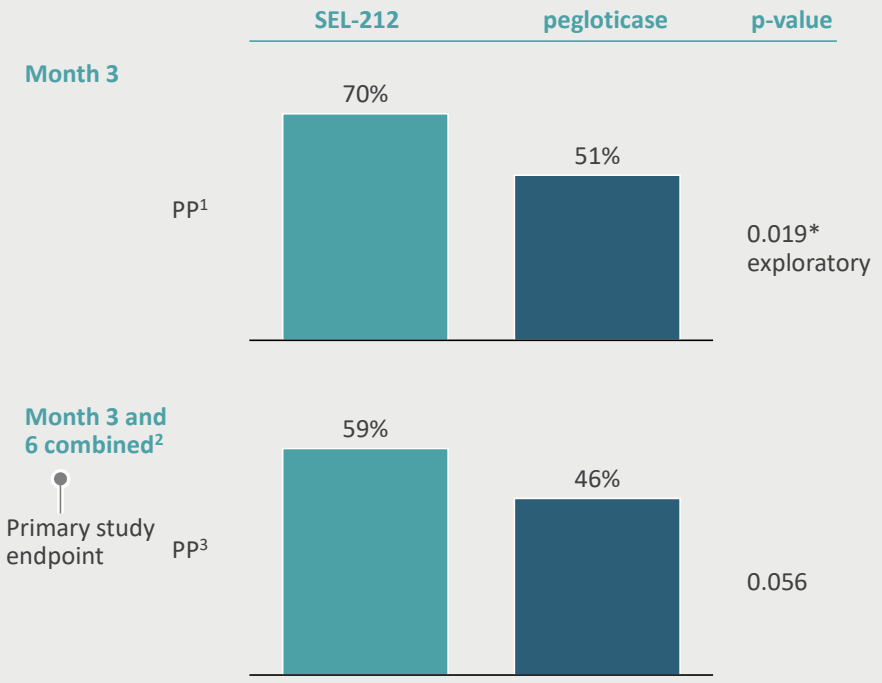
- pegloticase; efficacy rates of ~42%;
  - ~50% of patients can tolerate full treatment course (6m)

Differentiation:

- Less frequent dosing (monthly vs every other week)
- Statistically significantly greater overall reduction in mean serum uric acid (SUA) levels in SEL-212 versus pegloticase
- Higher efficacy rates in patients with tophi versus pegloticase
- No need for oral immunosuppression with MMF or MTX

COMPARE trial results (SEL-212 vs pegloticase)[1]

Patients with <6 mg/dl SUA for at least 80% of the evaluation time



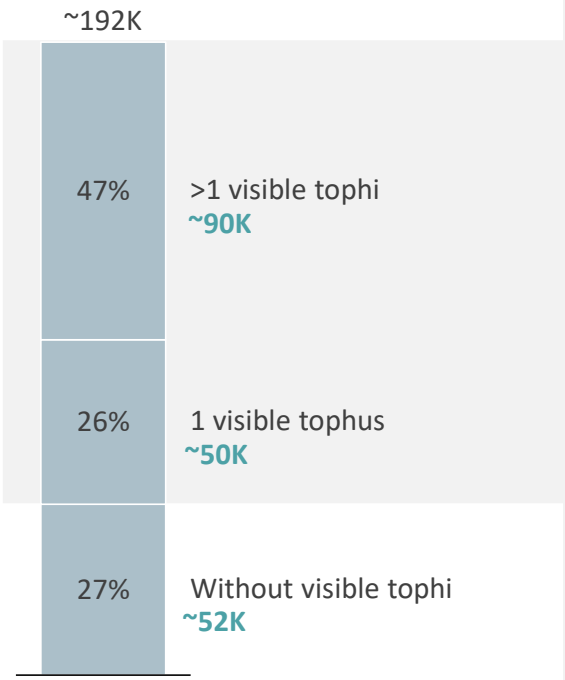
Note: Per FDA guidance on Statistical Considerations for Clinical Trials During the COVID-19 Public Health Emergency (June 2020), the statistical analysis plan was modified and submitted to FDA prior to database lock to address the potential impact of the COVID-19 pandemic on statistical analysis. This was necessary due to increased protocol deviations in the intention-to-treat (ITT) population observed during the ongoing COVID-19 pandemic. Data are therefore presented per protocol (PP) and ITT

1. Press release by Sobi and Selecta Biosciences on topline data of SEL-212 from phase 2 COMPARE study 2. SEL-212 showed a numerically higher response rate on the primary endpoint during months 3 and 6 combined, but did not meet the primary endpoint of statistical superiority 3. Presented per protocol

Patients with tophi have significantly higher responder rates for SEL-212 as compared to pegloticase based on phase 2 data

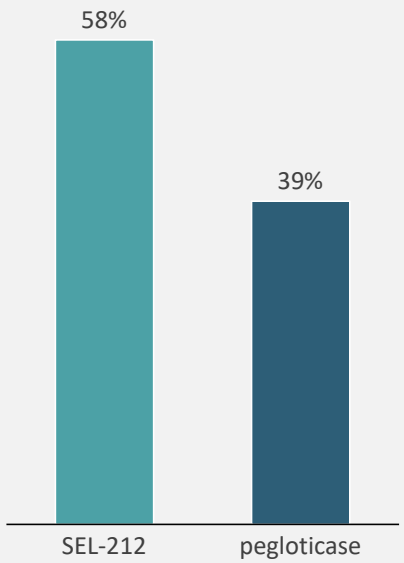
Chronic refractory gout patients

>70% have visible tophi US (2030)

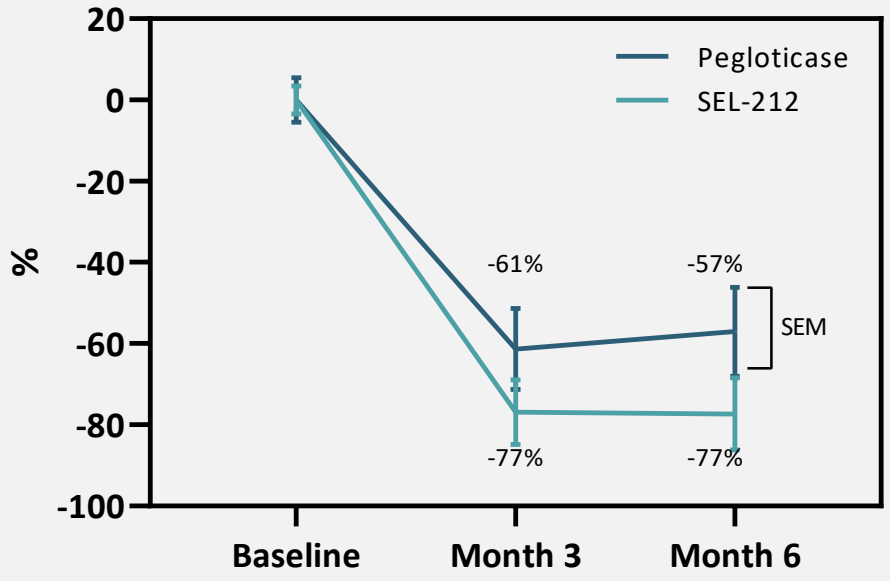


Study results for tophi patients[1]

Treatment response rates[2] (PP, months 3 and 6 combined)



Serum Uric Acid (SUA) percent reduction[3] (PP, baseline and months 3, 6)



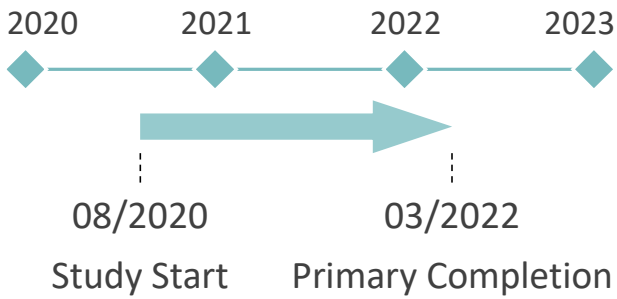
1. COMPARE study 2. Patients with <6mg/dl SUA for at least 80% of evaluation time 3. Computed as the mean SUA level during treatment period minus baseline SUA level divided by baseline SUA level multiplied by 100

Two phase 3 studies on SEL-212 have been launched this year

Study timeline

Key design aspects

Dissolve 1
Study 1
N= 105

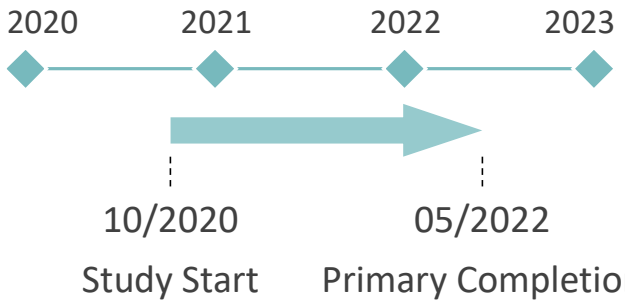


Arms: Randomized 1:1:1 to receive either one of two SEL-212 dose levels or placebo every 28 days for 6 months

Primary endpoint: Serum uric acid control during month 6

Extension: 6 months

Dissolve 2
Study 2
N= 105



Arms: Randomized 1:1:1 to receive either one of two SEL-212 dose levels or placebo every 28 days for 6 months

Primary endpoint: Serum uric acid control during month 6

Expected launch

2023 Chronic Refractory Gout

BIVV001

Doptelet (avatrombopag) tablets

SEL-212

MEDI 8897

gamifant emapalumab-lzsg

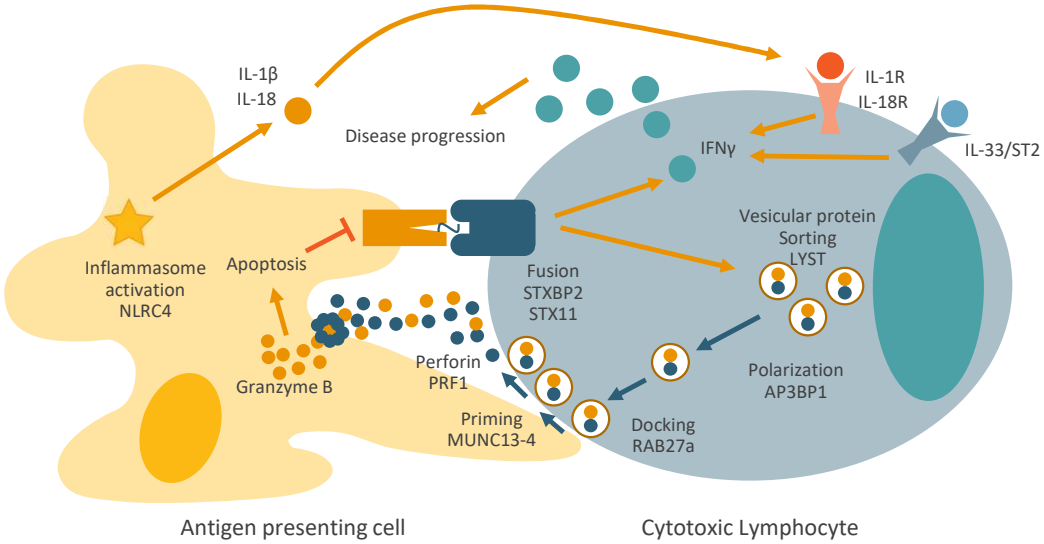
pegceta-coplan

About Synagis and MEDI8897

- The 2018 acquisition from AstraZeneca included rights to Synagis® (palivizumab) in the US as well as rights to participate in 50 per cent of the future earnings of the candidate drug MEDI8897 in the US
- Synagis is a medicine for the prevention of serious lower respiratory tract infections (LRTI) caused by respiratory syncytial virus (RSV) in high-risk infants and is the only approved preventative medicine for the condition
- MEDI8897 is a follow-on candidate to Synagis and a monoclonal antibody (mAb) being investigated for the prevention of LRTI caused by RSV in a broad infant population
- MEDI8897 derisks future revenue expectations for Synagis

MEDI8897 is being developed by AZ and commercialised by Sanofi. Sobi has an option to the rights to 50% of the profits and losses of the product in the US

Gamifant is the only treatment to target and neutralise IFNγ – a major proinflammatory cytokine



Mode of action

- Monoclonal antibody to neutralise interferon gamma (IFNγ)
- Aberrant IFNγ expression is associated with a number of autoinflammatory and autoimmune diseases such as HLH[1] – a severe, sometimes life-threatening systemic inflammatory syndrome

Profile

- Approved for primary HLH in US in Q4 2018[2]
- Three further indications in development: secondary HLH, Graft Failure and GvHD

1. Hemophagocytic lymphohistiocytosis. 2. Approved for the treatment of adult and pediatric patients with primary HLH with refractory, recurrent or progressive disease or intolerance with conventional HLH therapy, in US. See Gamifant prescribing information, FDA.

Sources: Jordan MB, et al.: How I treat hemophagocytic lymphohistiocytosis. Blood. 2011; Price B et al. Haemphagocytic lymphohistiocytosis: a fulminant syndrome associate with multiorgan failure and high mortality that frequently masquerades as sepsis and shock. S Afr Med J. 2014

pHLH is a life-threatening disease caused by genetic mutations

Disease Background[1]:

- Caused by multiple genetic mutations,
  - ~55% described, ~45% unknown
- Higher prevalence among children
- Mostly triggered by infection

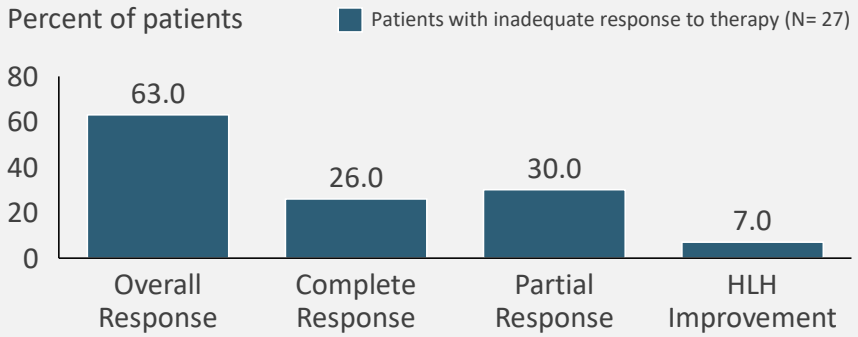
Current Treatments and differentiation:

- Gamifant is currently the only FDA approved therapy

Differentiation:

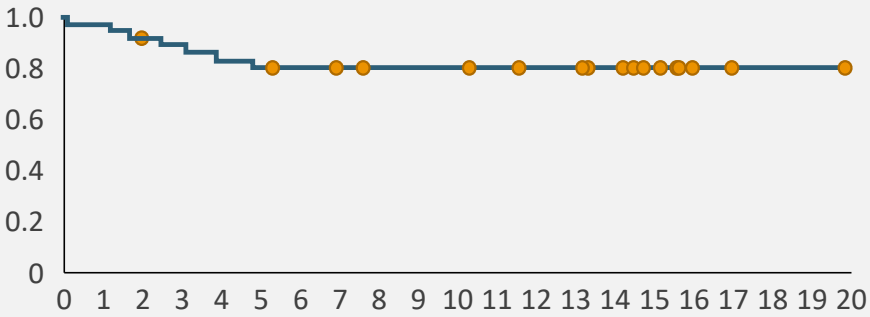
- Seeking approval in other countries (ex-EU)
- Further exploration of genetic databases to further inform patient care

NI-0501-04 Study: response at wk 8 (end of treatment)[2]



Overall response in previously treated patients was **significantly higher than the prespecified null hypothesis** (p=0.02)

NI-0501-05 Study: survival at 12m[2]



At last observation, 20/27 (74%) patients were alive with an **estimated probability of survival of 73.4% (95% CI 52.2–86.4) at 12 months**

1. Jordan et al Blood 2011; 118 (15): 4041 2. Locatelli et al N Engl J Med 2020; 382 (19): 1811

We are expanding emapalumab in HLH and beyond into diseases of high unmet need

Maximize pHLH

Expand geographically

Continue to drive disease awareness and **accelerate genetic understanding**

Expand into sHLH

Development of secondary HLH driven by **cytokine genetic** and **clinical data**:

Include rheumatologic HLH, malignant HLH and infectious HLH

Move to the next frontier: GF and GvHD post-HSCT

Precision medicine: Development in conjunction with a **companion diagnostic**

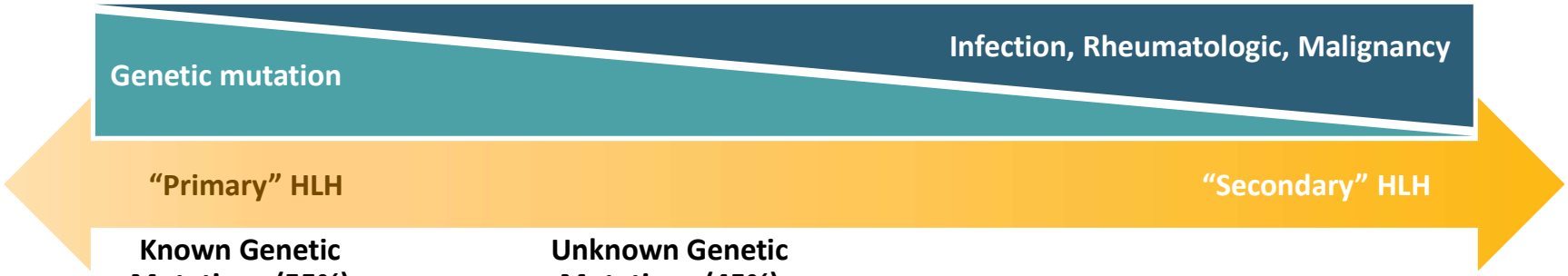
HLH **HSCT**

Expected launch

2021-2022	pHLH	
2023	sHLH	
2024	GvHD	

Many cases of HLH lie on a spectrum between sHLH and pHLH

We are gaining a deeper understanding of the impact of **unknown genetic mutations** and **infectious triggers** in the **pathogenesis of HLH**



	Known Genetic Mutations (55%)		**Unknown Genetic Mutations (45%)**	
	Perforin, Inflammasome, Immune deficiency			
Triggers	Infection: e.g., EBV, COVID-19, CMV, Iatrogenic, other			
Common phenotype	Fever Splenomegaly	Cytopenias Hyperferritinemia	Hypofibrinogenimia Hypertriglyceridemia	Low NK cell activity Elevated sCD25/sILRa

1. Source: Jordan et al Pediatr Blood Cancer 2019; 66 (11): e27929

Emapalumab could be a break-through treatment for **rheumatologic HLH**

Disease Background:

- Severe, potentially fatal condition associated with excessive activation of macrophages and T cells leading to an overwhelming inflammatory reaction[1]

Current Treatments:

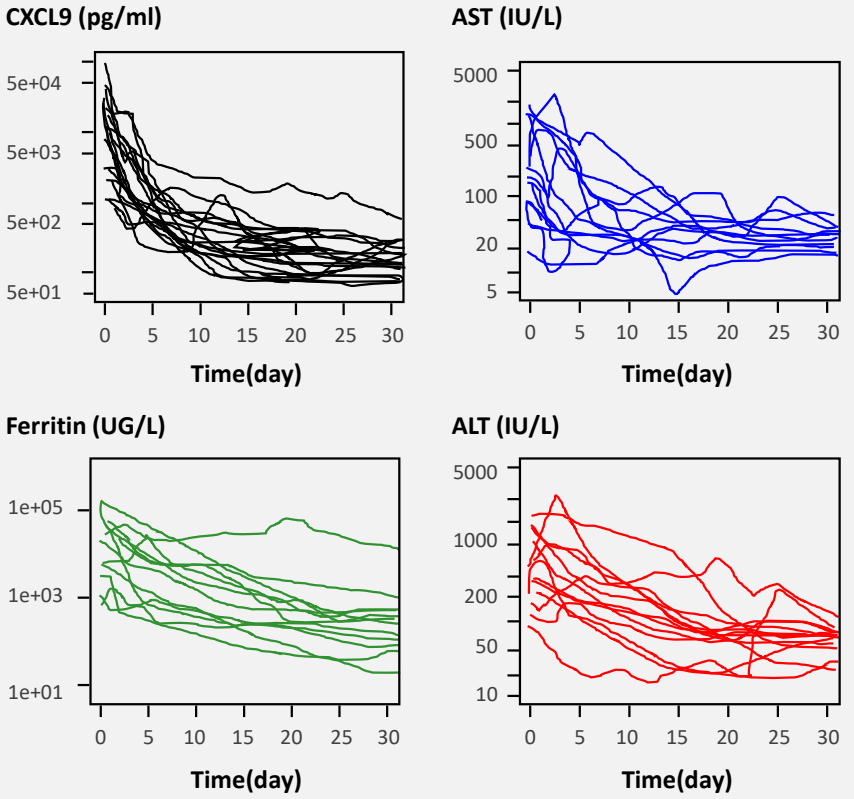
- No approved treatment to prevent or arrest rheumatological HLH (MAS in sJIA[2]) and AOSD

Differentiation:

- First IFN-targeted therapy for rheumatological HLH

Study NI-0501-06 MAS in SJIA/AOSD

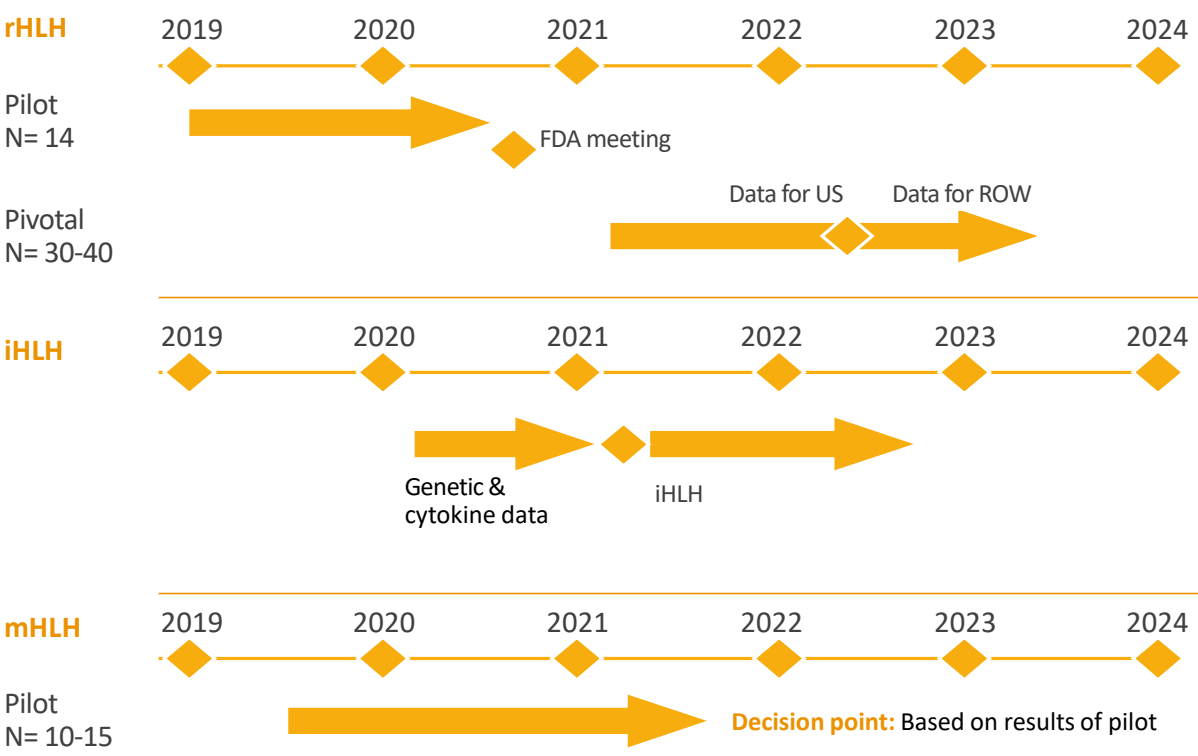
- Gamifant started on day 0:
  - 14/14 patients had a clinical response
  - Improvement in multiple lab parameters



1. Zhang et al. Arthritis Rheum. 2008 2. Macrophage Activation Syndrome in Systemic Juvenile Idiopathic Arthritis

We are carrying out an extensive development plan for sHLH

Timeline



Key design aspects

Arms: Single-arm, open label
Endpoints: MAS remission, duration of response

Arms: Single-arm, open label, observational run-in phase
Endpoints: MAS remission, duration of response

Genetic data mining/phenotype identification will guide design and patient selection criteria

Arms: Single-arm, open label, adaptive
Endpoints: ORR, survival, duration of response

Development of emapalumab for graft failure to start in Q1 2021

Disease Background:

- **Primary GF**: lack of engraftment of donor cells following Hematopoietic stem cell transplantation (HSCT); high mortality
- **Secondary GF**: loss of graft after initial engraftment

Current Treatments:

- Repeat graft

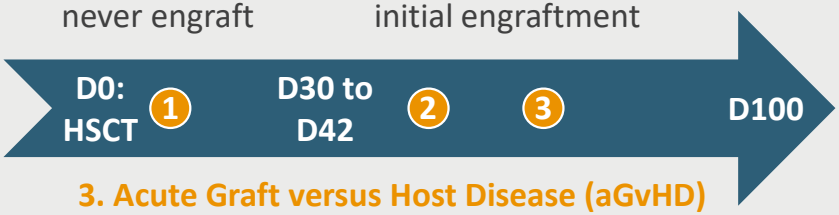
Differentiation:

- First IFN-targeted therapy for immune-related graft failure

Three possible complications following HSCT could be treated with emapalumab

1.Primary GF (pGF)
Transplanted cells never engraft

2. Secondary GF (sGF)
Loss of graft after initial engraftment



3. Acute Graft versus Host Disease (aGvHD)
Transplanted cells have immune reaction against the patient

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

Collaboration with bioMérieux to develop a Companion Diagnostic

Research collaboration for a CXCL9 companion diagnostic assay for Gamifant for GvHD

VIDAS™[1] CXCL9 for the prevention of graft failure post HSCT received breakthrough device designation by FDA in May 2020

Fast turnaround and hands off

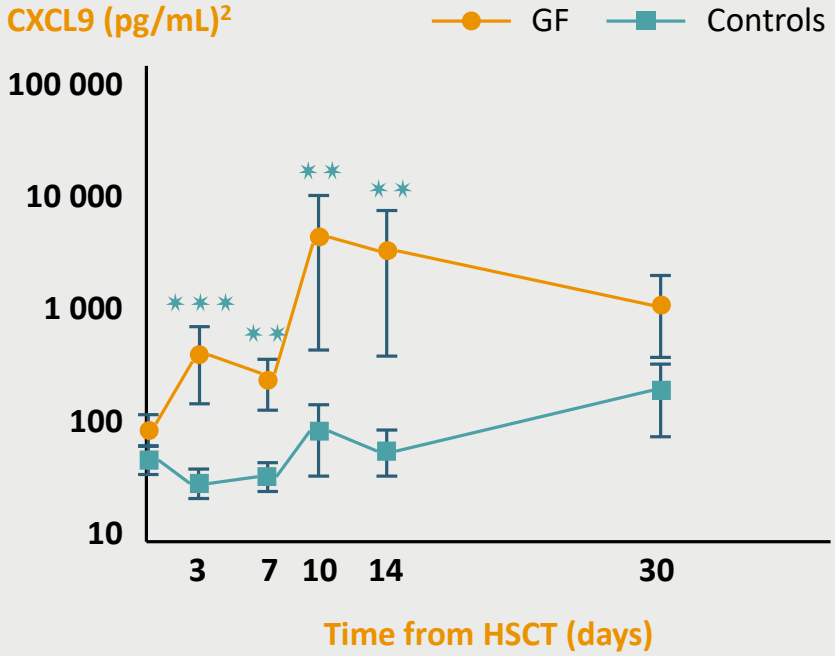
Could predict graft failure following HSCT as well as future indications

sobi rare strength

BIOMÉRIEUX

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

- As early as day 3 post-transplantation, patients developing GF had elevated serum CXCL9
- Serum CXCL9 appears to be an early biomarker for the risk of graft failure



1. Vidas™ is a registered trademark of bioMerieux 2. Merli et al., 2019. Haematologica
Note: CoDx = companion diagnostics; HSCT = Hematopoietic stem cell transplantation

Enabling development of emapalumab in GvHD to start in 2021

Disease Background:

- GvHD is a potentially serious complication of allogeneic HSCT
- Donor T cells attack the host which leads to multi-organ damage and can be life-threatening

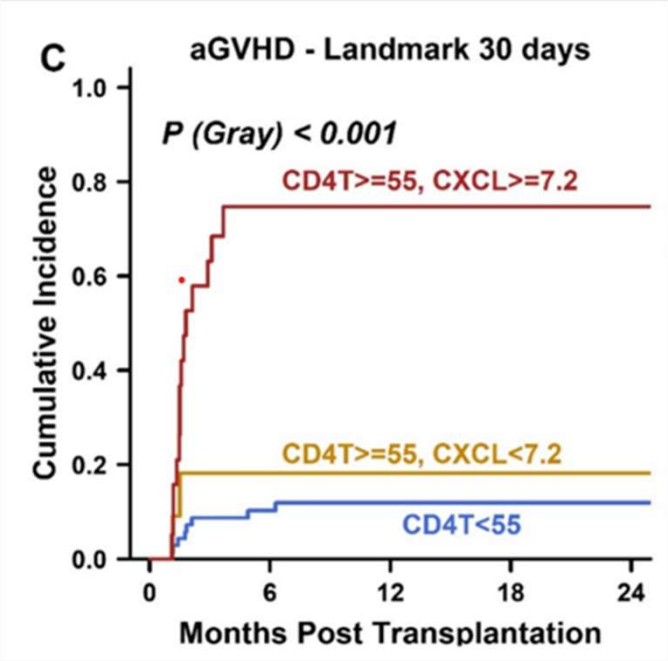
Current Treatments:

- Ruxolitinib currently approved

Differentiation:

- First IFN-targeted therapy for GvHD resistant to SOC

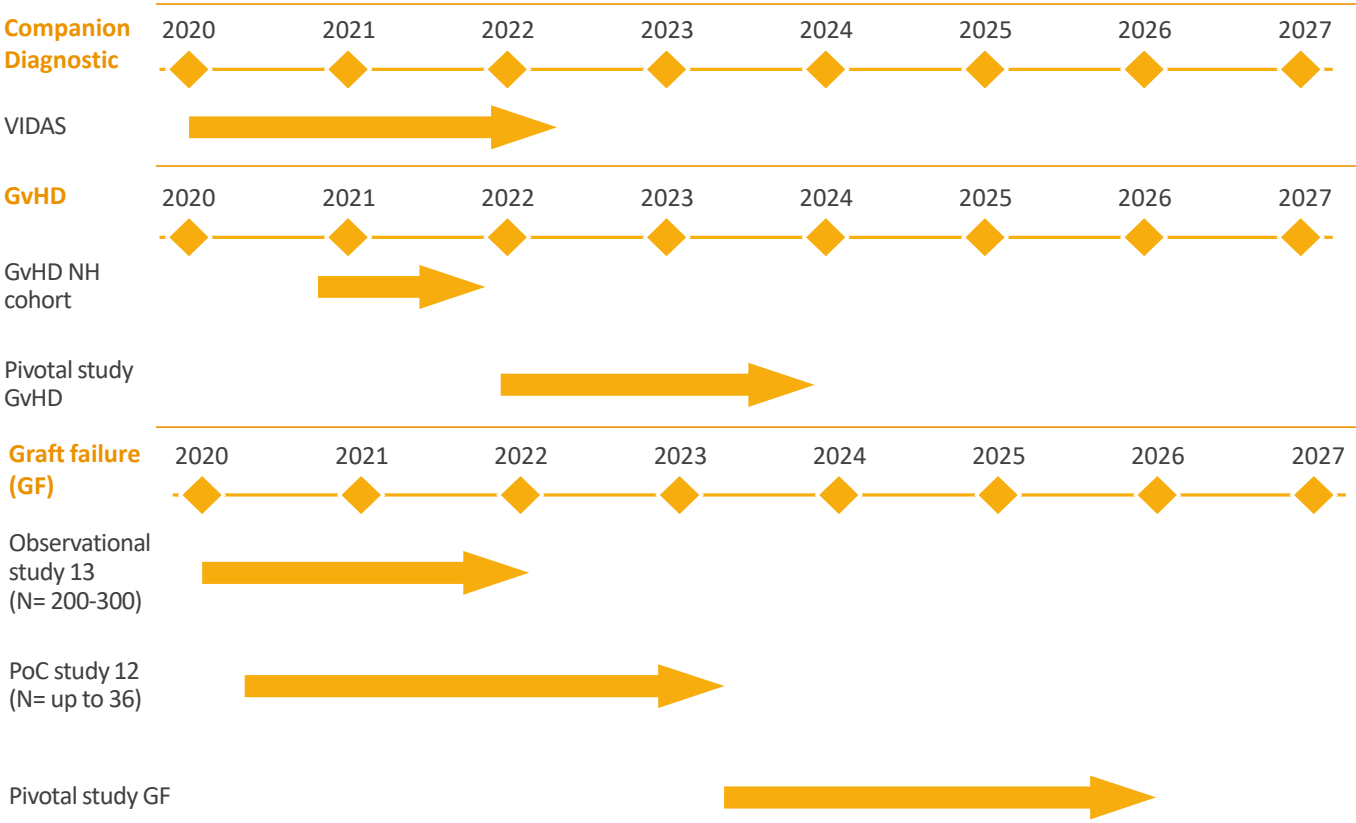
CXCL9 levels at d28 post transplant are predictive of 1 year cumulative incidence of aGvHD



Source: Merli et al., 2019. Haematologica. Volume 104(11):2314-2323, McCurdy et al., 2020

Development program for GvHD to start in 2021 with expected launch in 2024

Timeline



Key design aspects

Deliver **investigational use only device ready for clinical validation** prior to phase 3

Enabling activities to **inform study design and targeted population in aGvHD**

Registration study in 2nd and/or 3rd line in **steroid-resistant and/or ruxo-resistant patients**

Design: Allogeneic HSCT patients from HSCT to day 100 to support CoDx development, CXCL9 cut off for GF and GvHD pivotal studies

Endpoints: IFNG and CXCL9 levels

Design: Single-arm, open label, adaptive to confirm dose & efficacy in high-risk patients

Endpoints: PKPD, patient proportion with GF

RCT I in patients at high risk of primary and/or secondary GF to demonstrate reduction of GF rate

Anakinra & emapalumab have the potential to address key pathways in COVID-19 CSS[1]

Immuno-101 POC study of anakinra or emapalumab vs. SOC has been **stopped**

Ongoing clinical trial activity with **anakinra:**

- **18+ ongoing or planned RCT studies** of anakinra in moderate-severe disease
- **10 studies supported by Sobi** across US and EU; 1,000/~2,500 patients recruited

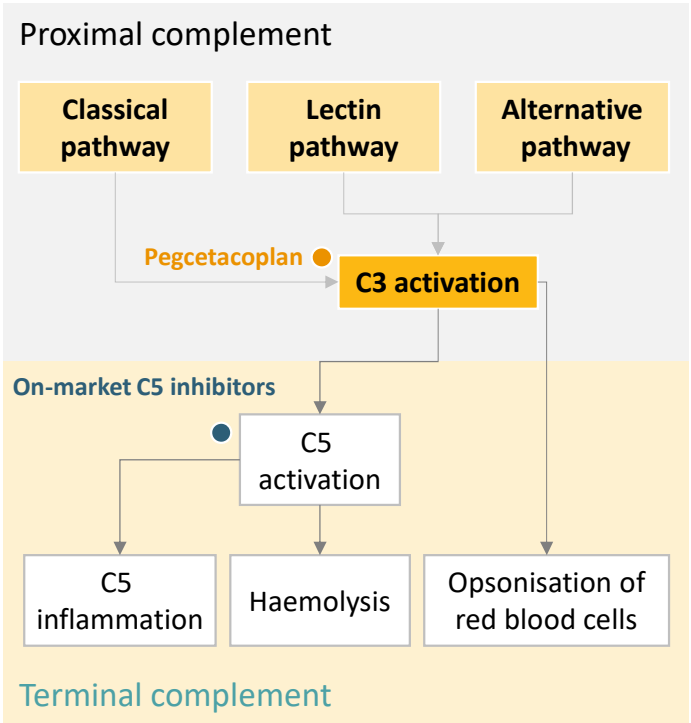
1. Fajgenbaum et al N Engl J Med 2020; 383: 2255 2. Soluble urokinase-type plasminogen activator receptor

Pegcetacoplan is a targeted C3 therapy – it is therefore modulating a key component of the complement system

Initiation

Amplification

Termination



Mode of Action

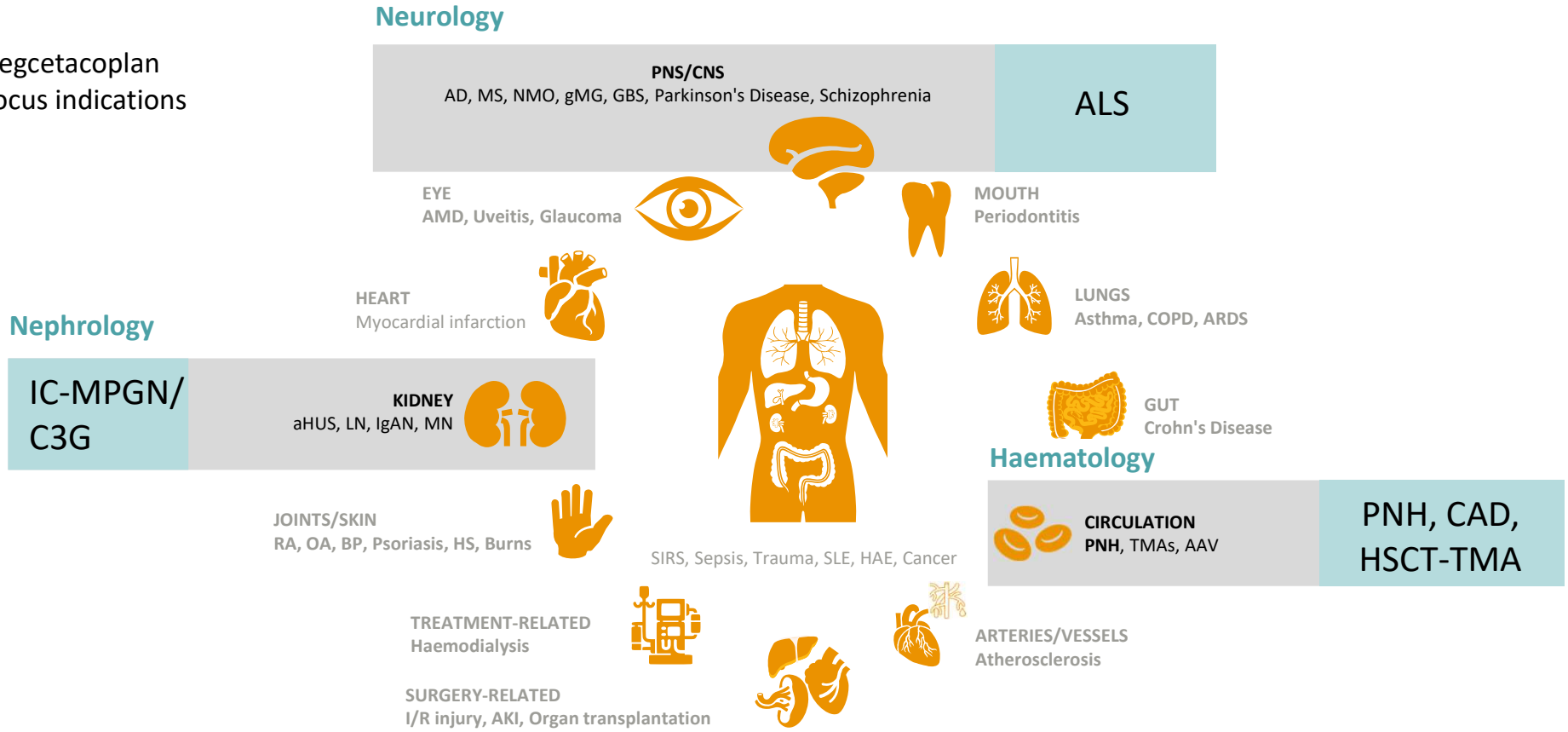
- Investigational, targeted C3 therapy designed to regulate excessive complement activation
- Uncontrolled complement activation is involved in the pathology of a broad range of disorders including various autoimmune and immune complement driven diseases

Designed profile

- Potential to elevate the standard of care in PNH
- Met primary endpoint and safety profile consistent with eculizumab in PEGASUS trial
- Subcutaneous administration

The complement system underlies many diseases; in addition to PNH, 3 others are part of the initial focus for pegcetacoplan

Pegcetacoplan focus indications



Source: Modified based on Zelek et al., Compendium of current complement therapeutics, Molecular Immunology 114 (2019) 341–352

Paroxysmal Nocturnal Haemoglobinuria (PNH): Superior improvement in Hb compared to eculizumab[1]

Disease Background (PNH):

- Acquired, rare, chronic, life-threatening blood disorder associated with abnormally low hemoglobin levels and transfusion dependence due to hemolysis
- Symptoms include severe fatigue, abdominal pain, difficulty breathing, and hemoglobinuria

Current Treatments:

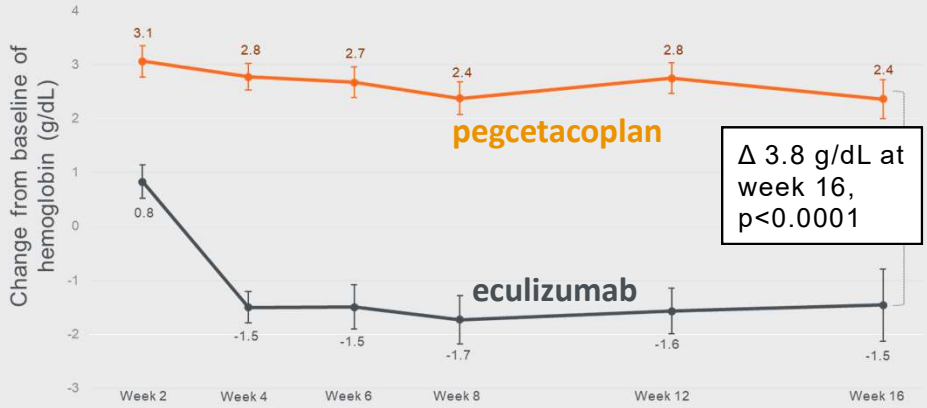
- C5 inhibitors

Differentiation:

- Superiority on improving Hb vs eculizumab due to effects on intra- and extra-vascular haemolysis
- Substantial improvement in other haematological and clinical parameters vs eculizumab

PEGASUS Study vs eculizumab: Week 16

Change from baseline hemoglobin in patients with a suboptimal response to eculizumab



1. Based on PEGASUS-study: 71% of pegcetacoplan-treated patients achieved LDH normalization vs. 15% of eculizumab-treated patients. LS Mean (+/- SE) plot of change from baseline in hemoglobin using MMRM model over time – randomized controlled period (ITT set)

Highly targeted treatment in Cold Agglutinin Disease (CAD)

Disease Background:

- Chronic and severe red blood disorder driven by extravascular hemolysis (IgM)
- Symptoms include anemia, transfusion requirements, and increased risk of thrombotic events like stroke or heart attack

Current Treatments:

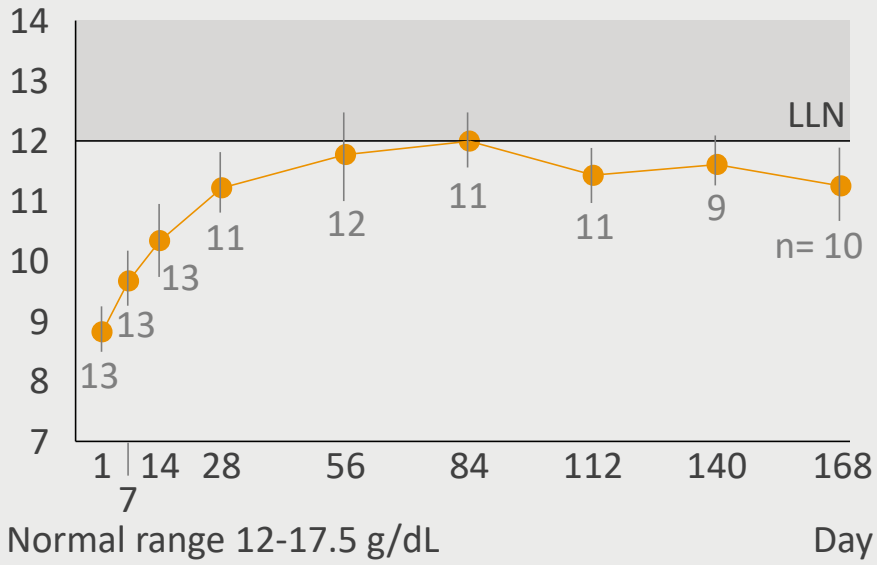
- No therapies approved or indicated to treat CAD

Differentiation:

- Expected to be first targeted C3 treatment for CAD

Interim Results: PLAUDIT Study, a phase 2 study n=24

Rapid, sustained, and durable increase in hemoglobin in response to pegcetacoplan



Source: 1. Catenion using physician and literature consensus.. 2. Fattizzo B, et al. European Hematology Association. June 13-16, 2019.

One of the first targeted therapies in HSCT-associated Thrombotic Microangiopathy (TMA)

Disease Background:

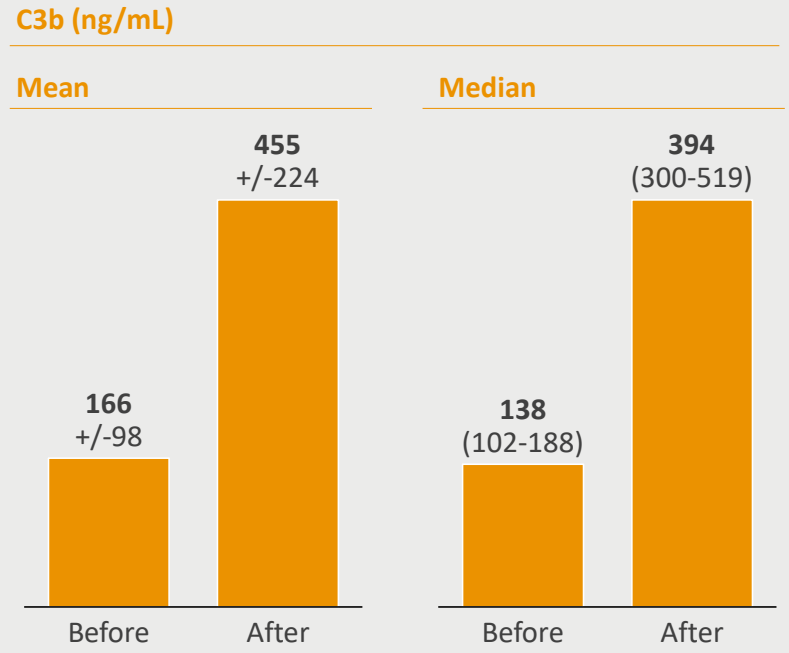
- HSCT-TMA is a rare inflammatory and thrombotic condition characterized by hemolytic anemia, thrombocytopenia, and evidence of multiorgan damage, particularly renal dysfunction[1]
- C3 is believed to play a critical role in TMA based on proinflammatory and procoagulant properties of C3a and C3b[2]

Current Treatments:

- No approved therapies

Differentiation:

- First targeted C3 treatment for HSCT-TMA



Elevated C3b levels in patients developing TMA post-HSCT

Qi et al. 2017

1. Dvorak C, et al. Frontiers in Pediatrics. 2019, Vol 7, article 133 2. Noris M, et al. Nature Reviews Nephrology. 2012, 8: 622–633

First line treatment in IC-MPGN and C3 Glomerulopathy[1]

Disease Background:

- Rare kidney diseases caused by excessive complement activation that can lead to kidney failure
- C3G is associated with a need for recurrent transplant in as many as 85%
- Classical and alternative pathways implicated in IC-MPGN

Current Treatments:

- No approved therapies

Differentiation:

- First targeted C3 treatment for C3G

DISCOVERY Study: Week 48*

	Baseline Mean (SE)	Week 48 Mean (SE)	Difference
24-hour uPCR, mg/mg	3.48 (0.82)	0.93 (0.27)	(73.3%)

*In five C3G patients; three patients were excluded from the analysis due to self-reported non-compliance or study drug interruption

1. Immune Complex Membranoproliferative Glomerulonephritis (IC-MPGN) and C3 Glomerulopathy (C3G)

Potentially registrational phase 2 study design laid out for ALS[1]

Disease Background:

- Neurodegenerative disease that results in progressive muscle weakness and paralysis due to the death of nerve cells in the brain and spinal cord
- High levels of C3 throughout motor system of patients may contribute to neuroinflammation and death of motor neurons

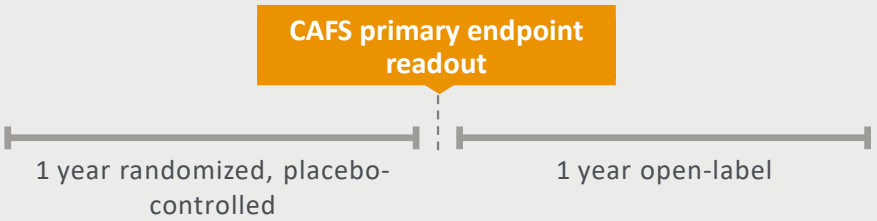
Current Treatments:

- No approved therapies have been shown to stop or reverse disease progression

Differentiation:

- First targeted C3 treatment for ALS

Potentially Registrational Phase 2 Study Design



Primary endpoint: Combined Assessment of Function and Survival at Week 52

Secondary endpoints: Measures of lung function, muscle strength, and quality of life

Design: Double blinded, randomized 2:1

Sample size: ~200 patients with sporadic ALS

Duration: 2 years

1. Amyotrophic Lateral Sclerosis (ALS)

Source: Arthur K et al., Nat Commun, 2016, Vol 7, article 12408

Our clinical development program is on the way to launch pegcetacoplan in 5 indications by 2024

Sobi leads development

Category	Disease	Phase 2	Phase 3
Haematology	PNH	→	→
	CAD	→	⇢ Initiate phase 3 study in 2021
	HSCT-TMA	⇢ Initiate phase 2 study in 2021	
Nephrology	IC-MPGN / C3G	→	⇢ Initiate phase 3 study in 2021
Neurology	ALS	→	

Expected launch

2021 PNH

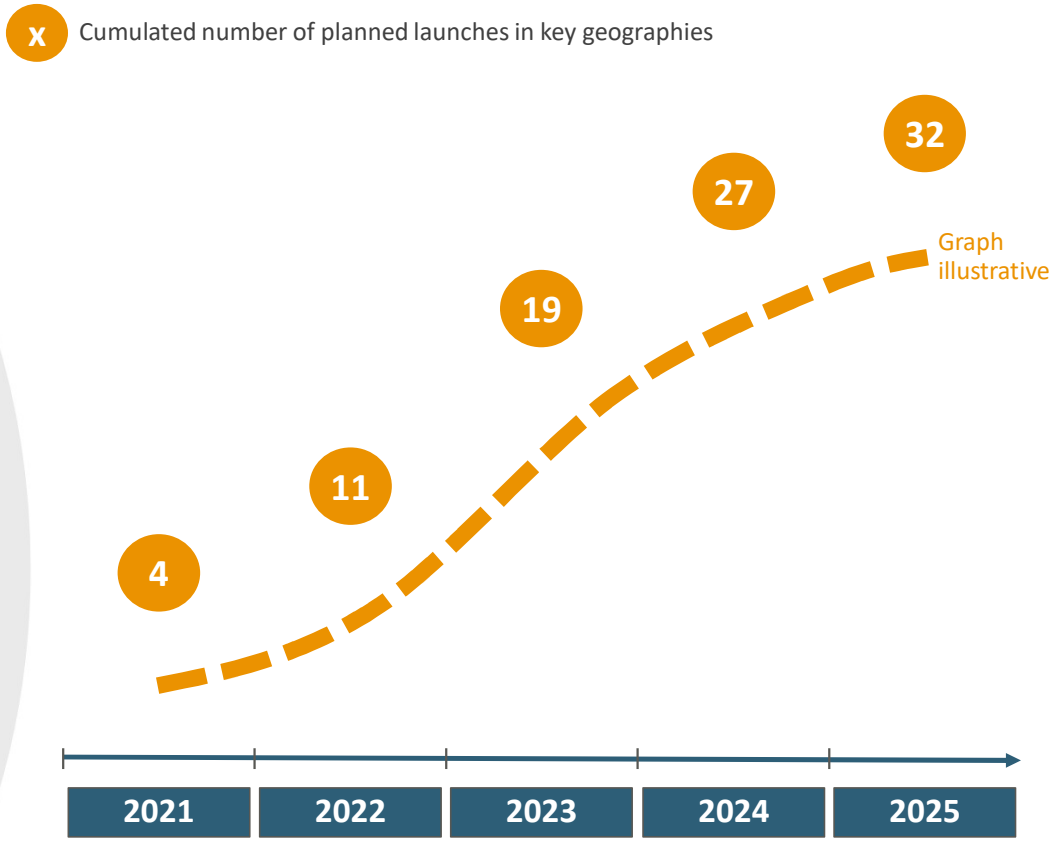
Capturing substantial value from our late-stage pipeline

5

Key geographies[1]

32

Launches in key geographies by 2025



Examples of indications (illustrative)

- avatrombopag, ITP
- avatrombopag, CLD
- pegcetacoplan[2], PNH
- pegcetacoplan[2], CAD
- pegcetacoplan[2], IC-MPGN / C3G
- pegcetacoplan[2], HSCT-TMA
- emapalumab, GvHD
- emapalumab, pHLH & sHLH
- SEL-212[3], chronic refractory gout
- BIVV001[4], HA
- nirsevimab[5], RSV prevention

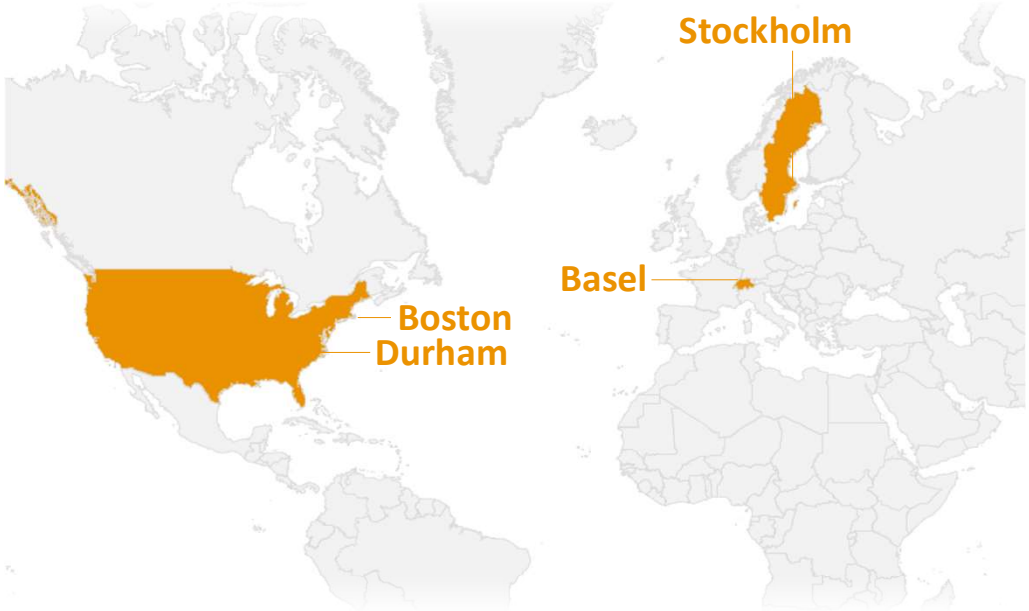
1. US, Europe, China, Japan, Russia 2. In collaboration with Apellis 3. Strategic licensing agreement with Selecta 4. In collaboration with Sanofi 5. Financial interest only, in collaboration with Astra Zeneca

Building our rare strength in R&D

- A portfolio across a broad range of rare haematologic and immunologic diseases
- Innovative and differentiated medicines
- Asset development in multiple indications and utilising leading enabling technology
- Deep experience with collaboration and partnership
- Sobi leadership in areas of expertise:
  - *emapalumab in sHLH, GF, GVHD (HSCT)*
  - *pegcetocoplan in CAD and TMA (HSCT)*
  - *anakinra in COVID19*

4 Centers of excellence with ~250 team members

- deepening experience of haematology and immunology



Internationalisation Strategy

Norbert Oppitz

The rare disease market represents compelling opportunities outside Europe and US

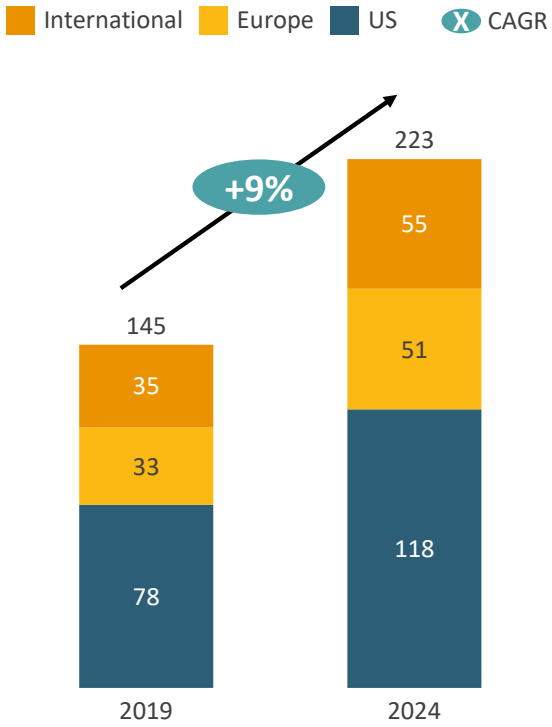
Key facts

7,000+
More than **7,000 designated rare diseases** exist

~85%
85% are serious / **life-threatening**

500
Only approximately 500 **approved drugs** to date

Rare disease market, USD B



Strong double-digit market growth for rare diseases **outside** current Sobi core **territories**

>250 million people affected by rare diseases excluding US/EU, thereof 60% children

Sobi committed to **reach 80% of affected patients** with Sobi rare disease products

Source: Profound

Our Mission 2025 goal is to go global and make our products available to twice as many patients as today

2017-2020

Becoming a regional leader in rare disease

Establish **US** as a **second pillar** next to Europe

Started to further internationalise, e.g. **Middle East**

Mission 2025

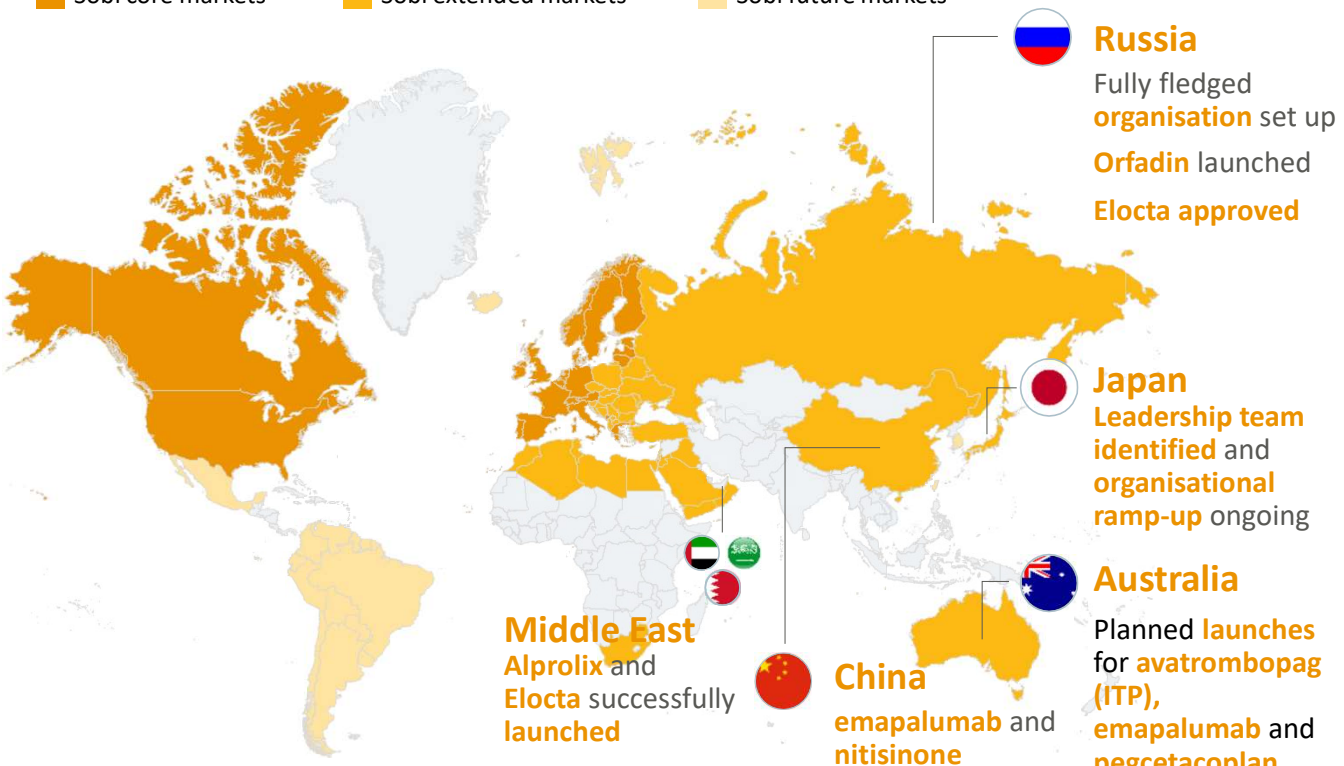
Global leader in rare disease

Presence established in key markets **China, Japan, Russia and Australia**

Partnerships to **serve underserved regions**

We have already expanded our market presence outside Europe and the US

Sobi core markets | Sobi extended markets | Sobi future markets



Russia
Fully fledged **organisation** set up
Orfadin launched
Elocta approved

Japan
Leadership team identified and **organisational ramp-up** ongoing

Australia
Planned **launches** for **avatrombopag (ITP)**, **emapalumab** and **pegcetacoplan**

Middle East
Alprolix and **Elocta** successfully **launched**

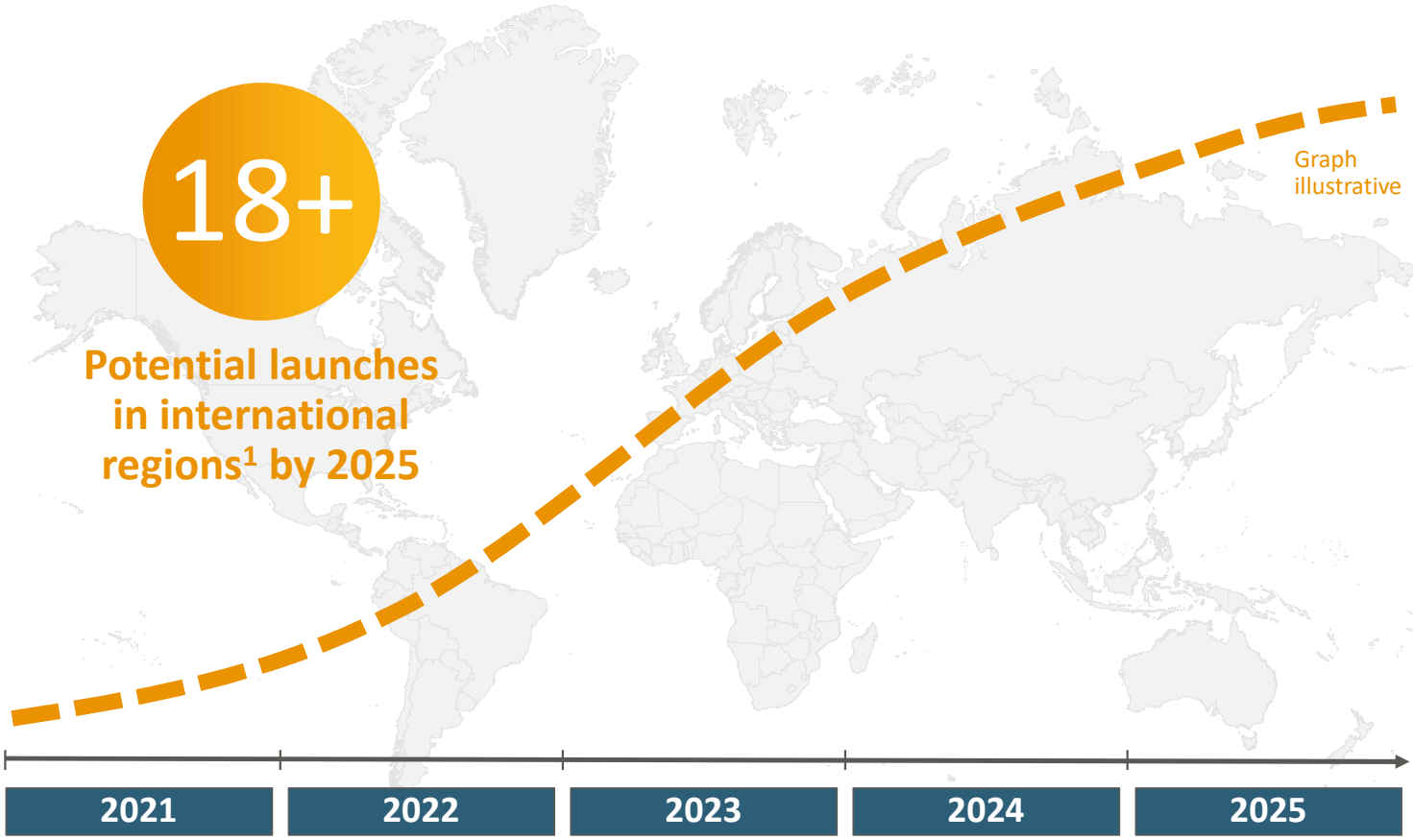
China
emapalumab and **nitisinone submission in process**

Sobi has already **established** presence in **Middle East, Russia** and **China**

Current sales of **SEK ~800M outside** its **core** markets (Europe and US)

Second wave now focuses on **Japan, Australia, South-East Asia** and **Latin America**

International will ramp up value with a high cadence of launches



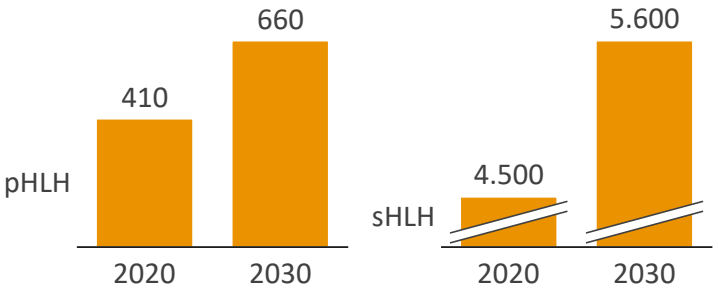
Examples of indications (illustrative)

- eloctate, HA
- nitisinone, HT-1
- emapalumab, pHLH
- emapalumab, sHLH
- emapalumab, GvHD
- avatrombopag, ITP
- avatrombopag, CLD
- pegcetacoplan[2], PNH
- pegcetacoplan[2], CAD
- pegcetacoplan[2], C3G
- SEL-212[3], chronic refractory gout

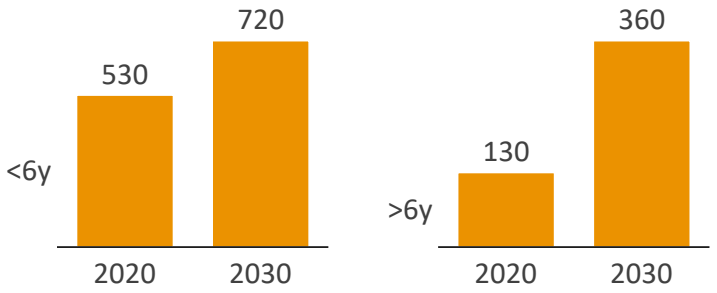
1. Regions outside NA and Europe 2. In collaboration with Apellis 3. Strategic licensing agreement with Selecta

Example: China – we are fast-tracking to enable patients to access emapalumab and nitisinone in 2022

HLH patients in China



HT-1 patients in China



emapalumab progress

Mar 20
China adboard on HLH

Jun 20
CDE meeting on emapa-lumab

Dec 20
NDA submission emapa-lumab

Q3-4 21
Potential approval (priority review)

nitisinone progress

Jun 20
China adboard on HAT-1

Jun 20
CDE meeting on nitisinone

Dec 20
NDA submission nitisinone

Q3 21
Potential approval (priority review)

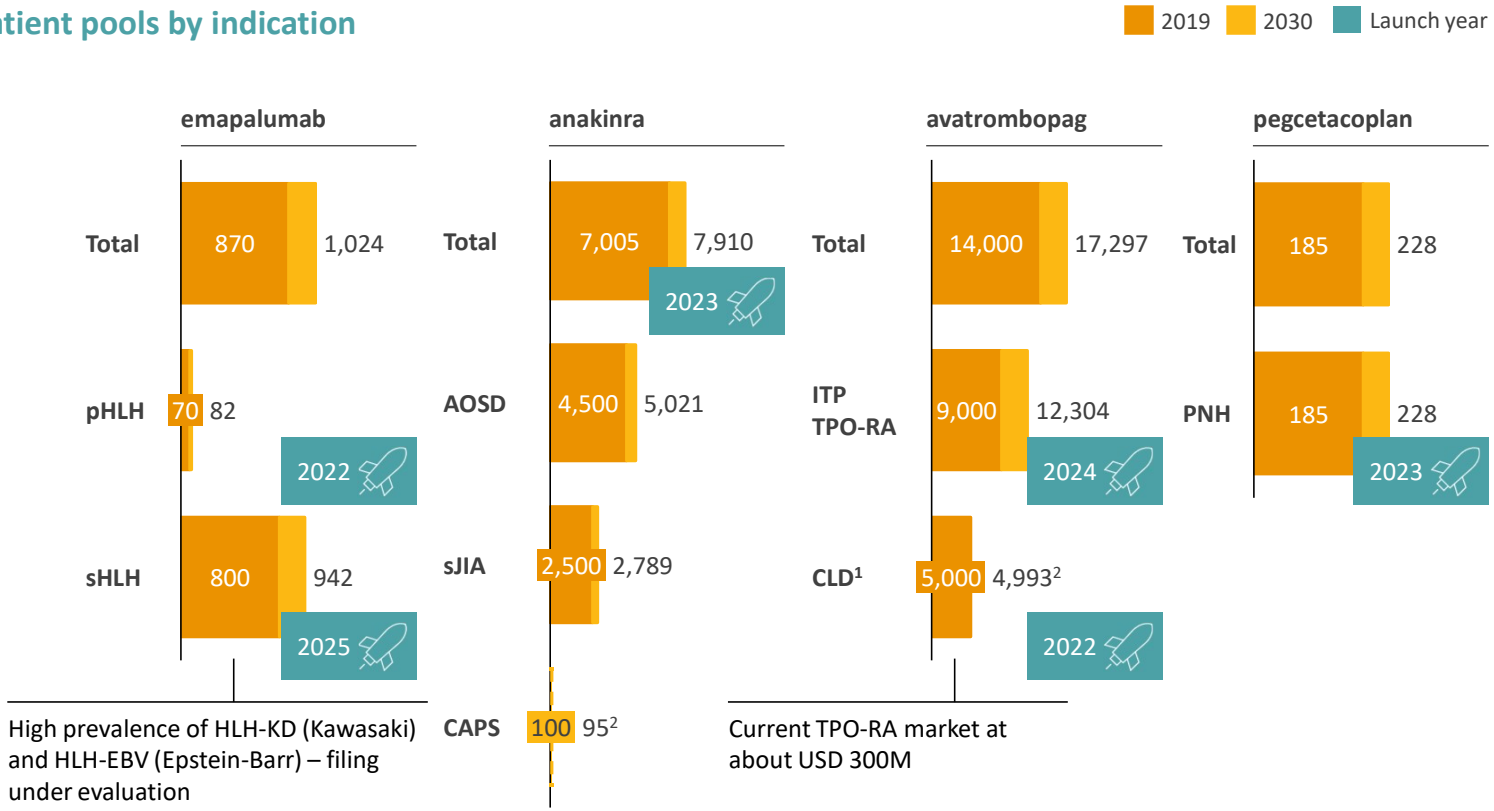
sobi

- **Legal entity** established
- **GM** with **20+ years** of experience
- **Medical support established**
- **Supply chain designed**
- **Regulatory** expertise **onboarded**
- Launch of anakinra in CAPS expected 2023

Source: Sobi

Example: Japan – en route to introduce emapalumab, anakinra, avatrombopag and pegcetacoplan

Patient pools by indication



1. CLD patients receiving surgery 2. Decrease in line with population expected

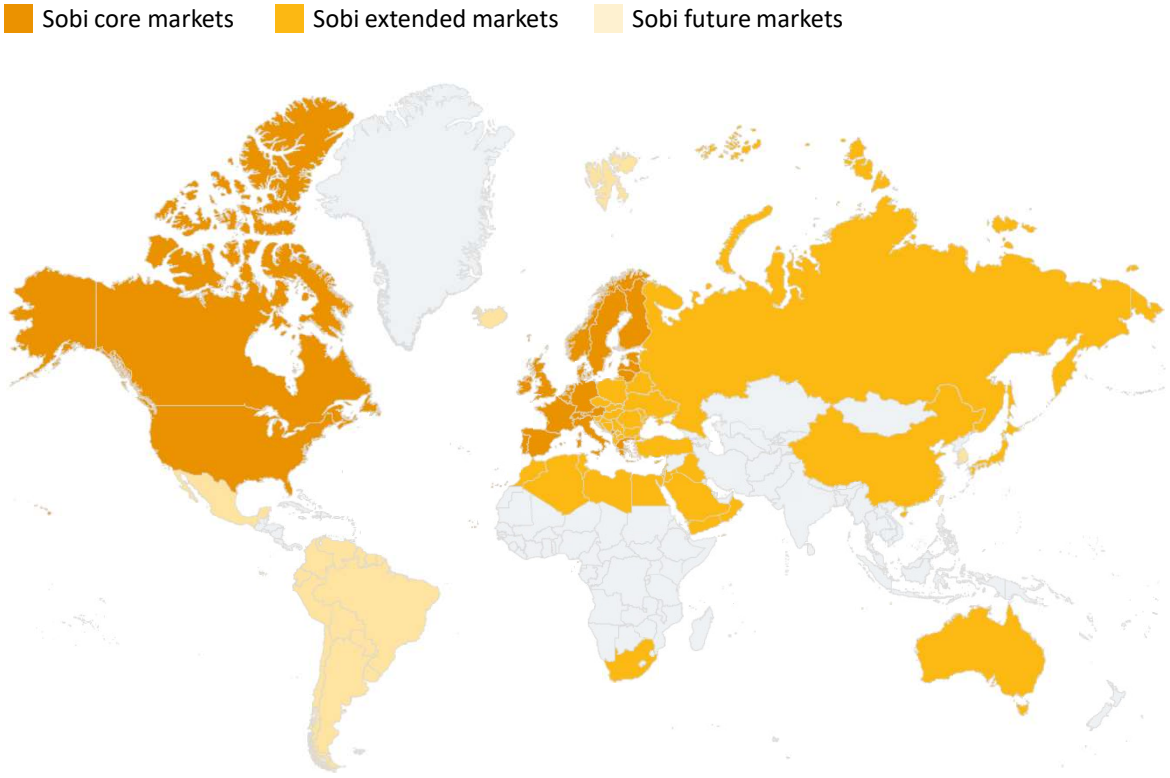
Source: Scientific research, journals, government publications, patient surveys, practice guidelines, publications from associations, expert interviews, claims data

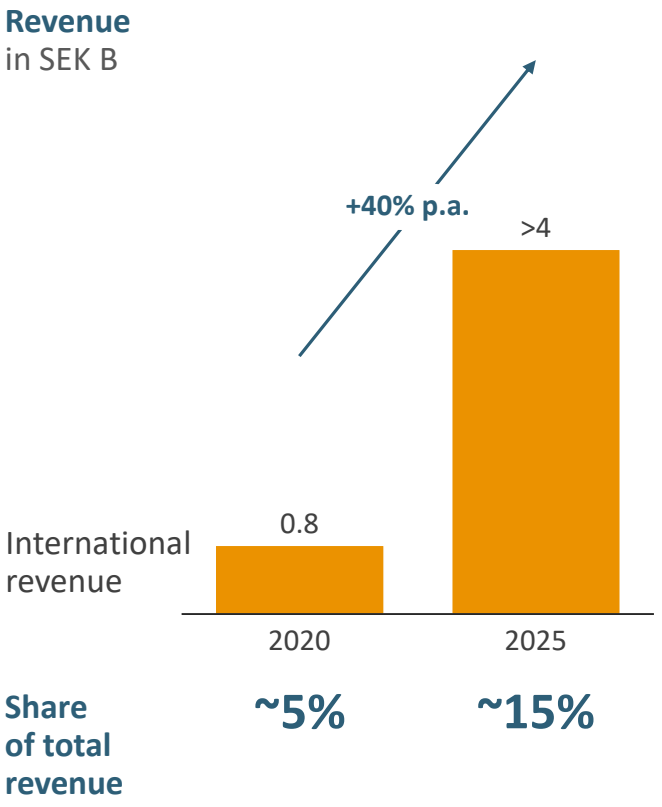
sobi

Well-established markets already

Build **experienced leadership team** and **scaled-up organisation**

By 2025, International is expected to contribute 15% of Sobi's revenue





Source: Sobi

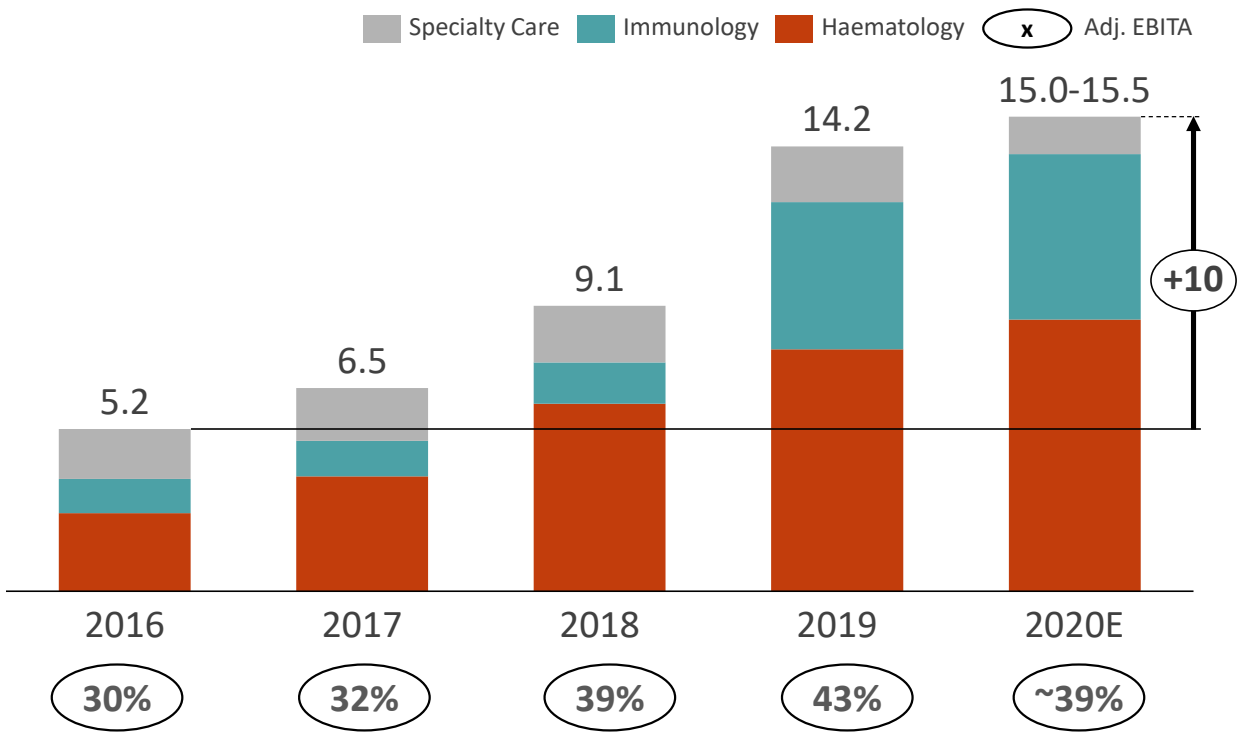
Financial update

Henrik Stenqvist

CFO

Operational performance in 2016 – 2020 has generated strong revenue, EBITA and cash flow

Total Revenue, SEK B



2020E based on mid-point guidance provided by Sobi in Q3 2020 report

Haematology

Achieved continued growth in Haemophilia

Realised successful launch of Doptelet

Immunology

Impact from Synagis acquisition and successful Gamifant launch

Realised growth in Kineret

Specialty Care

Declining revenues due to partner portfolio terminations

Orfadin decline due to generics

EBITA

Maintained stable ~40% margins over the last 3 years

Cash flow

Continued strong cash generation

Sobi is in a new stage as a company

Mission 2025

Global leader in rare disease

Competitive situation within **Haemophilia**

We now have a significant **pipeline of late-stage assets** to be brought to approval

Commercial success will be driven by **realizing the potential of launches on a global scale**

We will continue to make **strategic M&A**

Bringing assets to approval and realising commercial potential on a global scale

R&D spend – pipeline of late-stage assets to be brought to approval

- Moving forward in 2021-2022, **will increase to 13-15% of sales** driven by investments in:
  - **SEL-212**
  - **Pegcetacoplan** across several indications
  - **Emapalumab** indication expansion into GF and sHLH

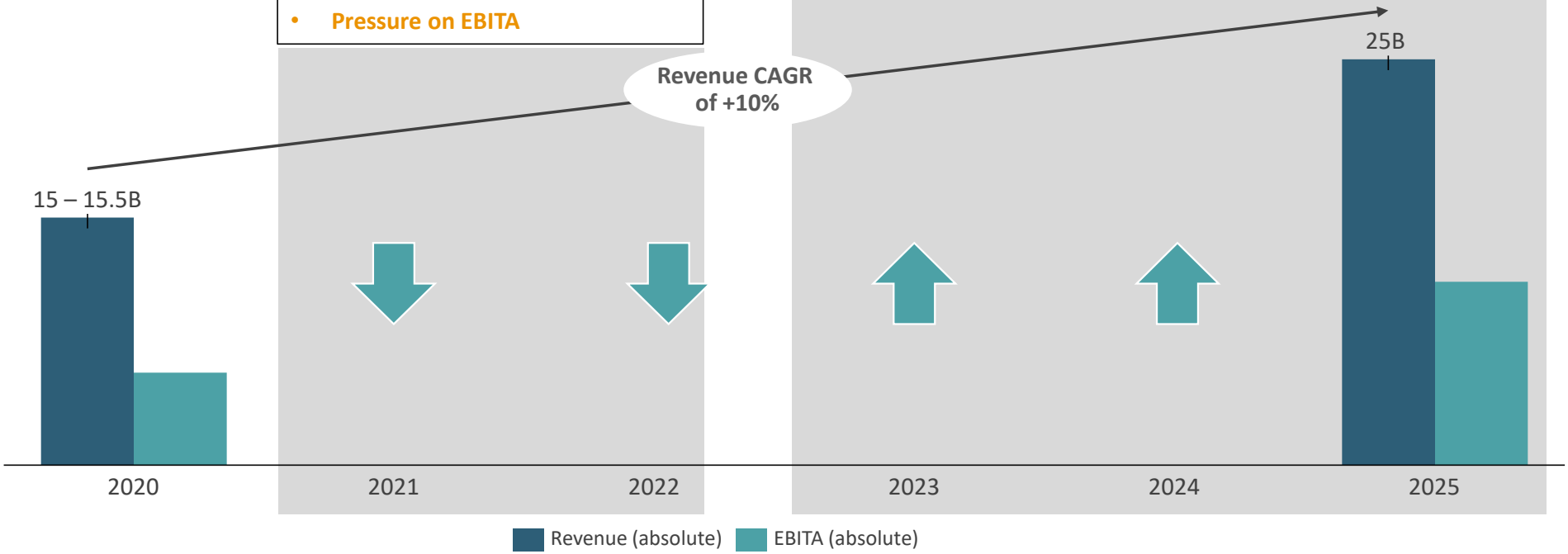
SG&A spend – realising the potential of launches on a global scale

- Continued investments in **Doptelet** and **Gamifant**
- Launch of **SEL-212, pegcetacoplan** and **BIVV-001**
- Further investments in **infrastructure in international markets**

Growth acceleration from 2023 onwards

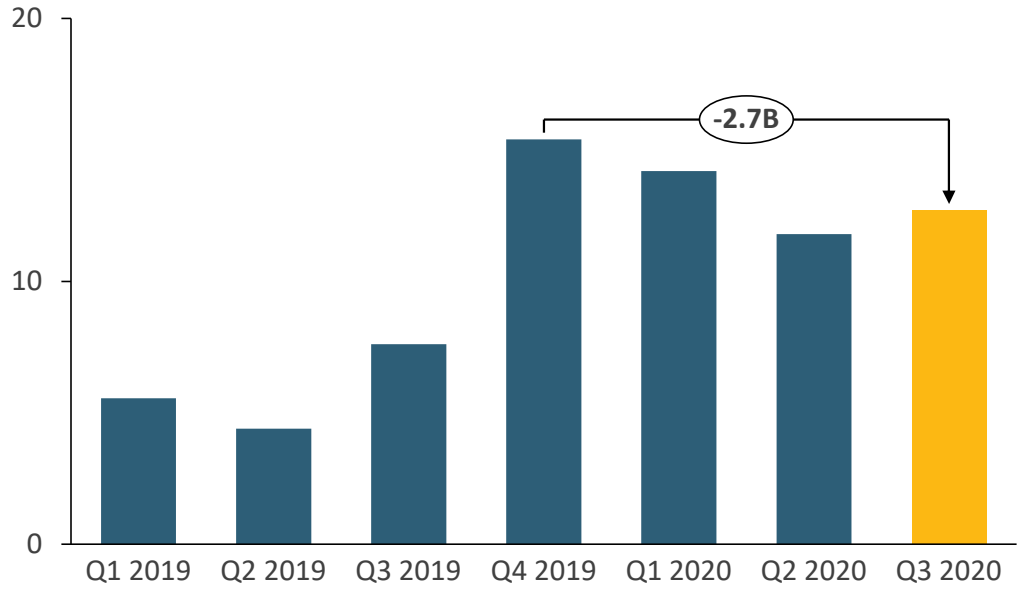
2021 – 2022:
- **Single-digit revenue growth**
- **Investment into the future** related to R&D, launches and internationalisation
- **Pressure on EBITA**

2023 – 2025:
- **Continuous potential double-digit revenue growth**
- Mid-term revenue aspiration of **25B SEK by 2025**
- **EBITA margin acceleration**



A continued strong cash flow will support growth through M&A

Net debt, SEK B



Acqui-sitions

SYNAGIS PALIVIZUMAB

gamifant emapalumab-lzsg

Dova PHARMACEUTICALS

SEL-212

Continued **strong cash generation** with seasonality driven by Synagis

Net debt / EBITDA will likely be **below 3** in short-term

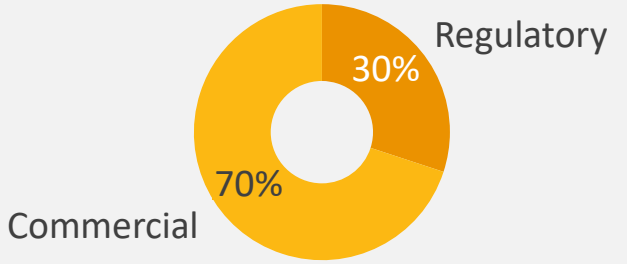
Ability to lever up to 3-4x range

Focus M+A in strategic areas – *derisk where possible*

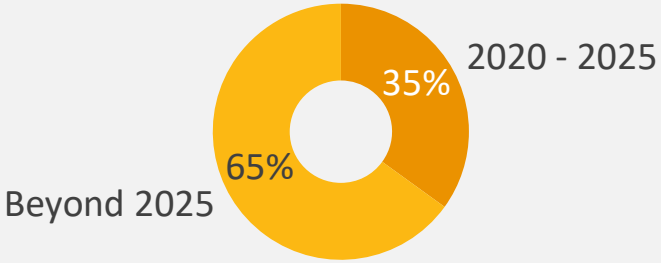
Example actions to de-risk

Pegceta-coplan	Limited upfront payment; future payments contingent on R&D and commercial success
SEL-212	Limited upfront payment; future payments contingent on R&D and commercial success
Dova PHARMACEUTICALS	3 indications, bulk of purchase value from CLD and ITP
SYNAGIS® PALIVIZUMAB	Financial stake in follow-on MEDI8897
gamifant® emapalumab-lzsg	Included FDA priority review voucher with high re-sale value de-risked by multiple potential indications

Majority of obligations relate to commercial milestones



Majority of obligations estimated due after 2025



Wrap-up and Q&A

Guido Oelkers

CEO

Conclusion

Proven track record - since 2016 we have created a strong foundation with regard to revenue, EBITA and cash flow, whilst building the organisation and our pipeline

Next phase requires investment into development, pre-launch and launches of our pipeline to **propel mid- to long-term growth**

Our ambition is to reach **SEK 25B revenues in 2025**

Further M+A will be explored