Pharming Group NV

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Kempen Healthcare Conference
Amsterdam,18-19 April 2018

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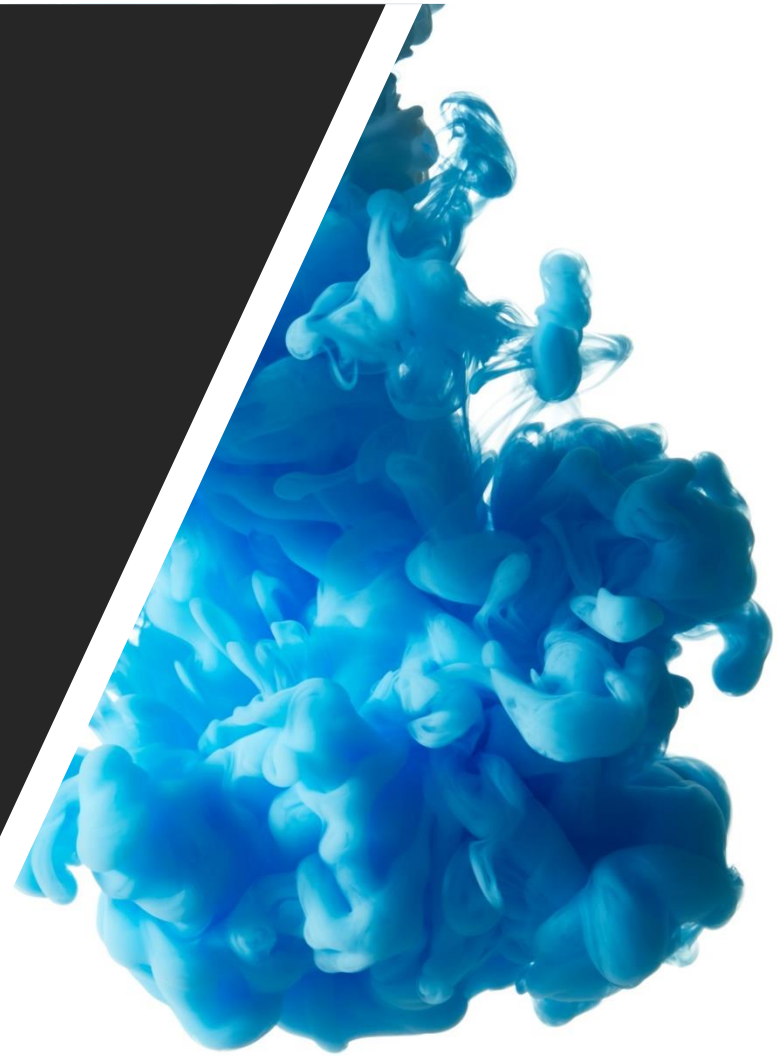

Overview

- Euronext: PHARM - market capitalization: ~€780 million ($960 million)
- HQ and western -EU commercialisation from NL, R&D in France and US commercial operations in New Jersey with approximately 145 employees
- 1st product approved and marketed: RUCONEST®
  - Recombinant human C1-esterase inhibitor (enzyme replacement therapy)
  - For acute angioedema attacks in patients with hereditary angioedema (HAE)
  - Marketed in USA, EU and Israel
  - US data exclusivity until 2026
- Platform technology and new Enzyme Replacement Therapies (ERT) for rare genetic conditions in late pre-clinical stage

We develop and commercialize human therapeutic proteins for innovative therapies meeting important unmet patient needs

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Corporate Highlights

RUCONEST® Commercialisation

- Re-acquisition of US commercialization rights from Valeant in Dec 2016
- 2017 revenues: €89.6 million (2016 revenues: €15.9 million)
- 2017 operating profit €21.9 million (2016 operating loss €11.5 million)
- Temporary supply issues during Q4 2017 at a competitor now resolved

RUCONEST® Franchise Development

- Prophylaxis of HAE Phase 2 study (DBPC) met all endpoints – as good as any
- sBLA accepted for review by FDA: Action date 21 September 2018
- Additional large (non-HAE-related) indications being assessed for RUCONEST®

Maturing pipeline beyond RUCONEST®

- Program for Pompe disease filing for IND end of this year, Fabry project following
- Uses same transgenic founder technology

Solid Financial Base

- Re-financed debt with a $100 million 4 year debt facility with OrbiMed Advisors in July 2017
- Positive cashflows: Cash balance at YE2017: €60.0 million (€38.6 million at 30 Sep 2017)

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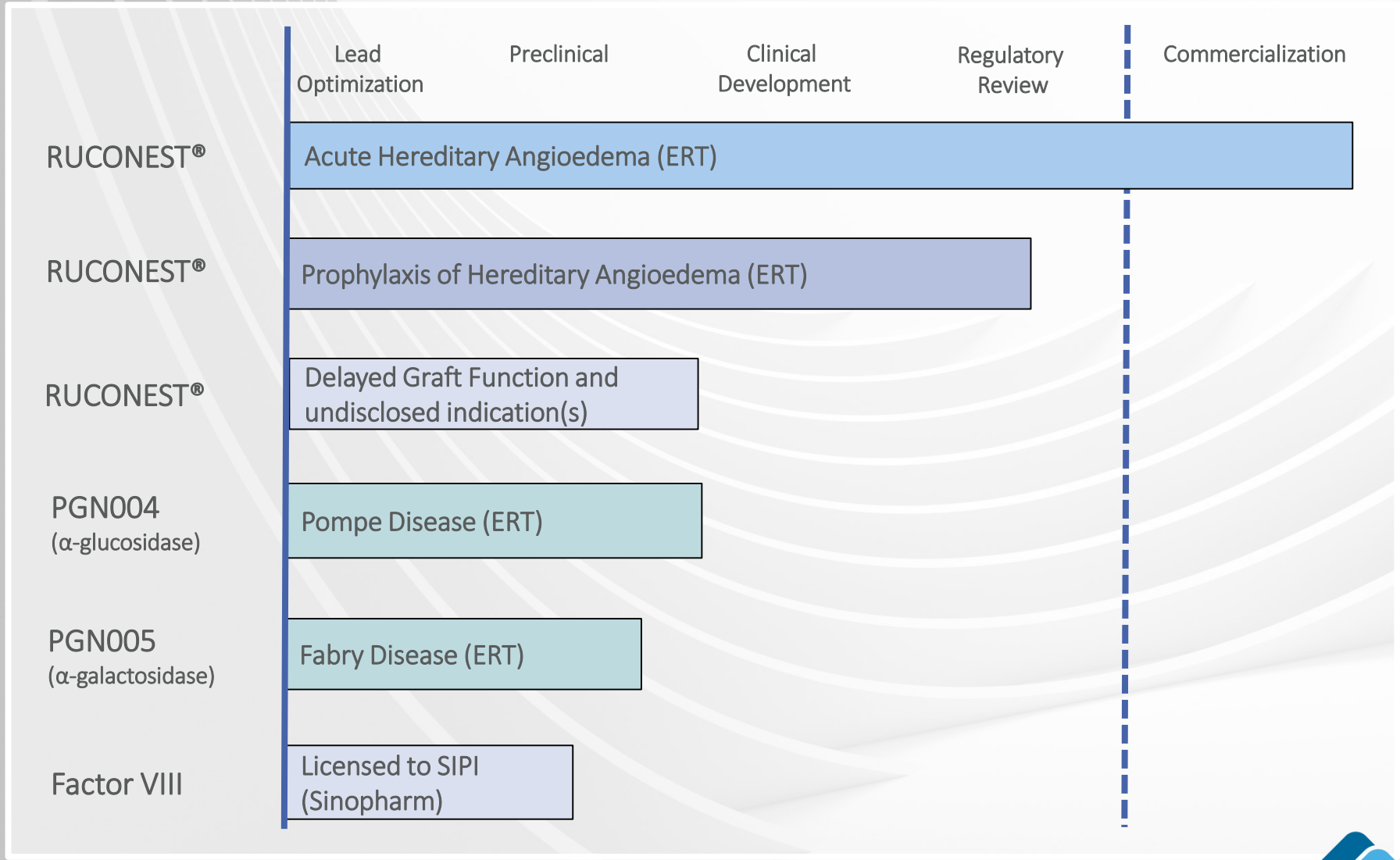
Building a franchise and pipeline



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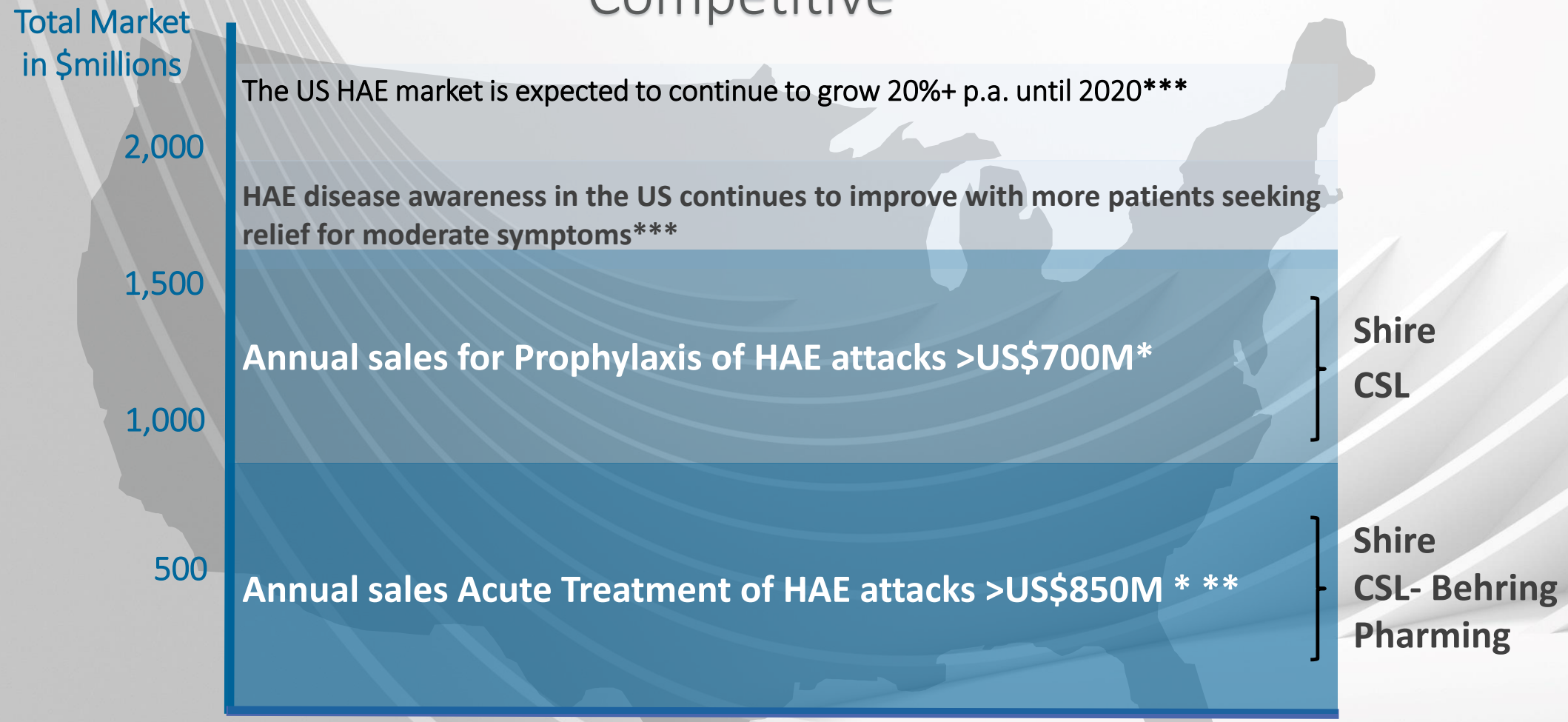
US HAE Market: Rapid Growth, Significant Potential, Very Competitive



* 2016 results/ SEC filings SHPG, Pharming

** Excludes plasma derived C1- esterase inhibitor sales / not disclosed by CSL Behring

*** Leerink Swann, competitor interviews, 13 September 2012

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US HAE Treatment Practices

- The first generation treatments were inadequately effective
- Prophylaxis with plasma derived enzyme replacement therapy therefore became a rational approach and was the first product approved in the US
- In the US this resulted in significant use of prophylactic treatments
- However: All of the currently available prophylactic treatments and all of the prophylactic treatments in development feature (frequent) break-through attacks (up to 50%), necessitating rescue medication for acute attacks to be at hand at all times, and/ or may have limitations for use in certain types of patients
- Current prophylactic therapy is cumbersome (currently at least 104 injections/year) and expensive and it exposes patients to significant amounts of blood plasma, significant supply risks and to the cost and burden of the treatment of breakthrough attacks

HAE is rare (around 1 in 30,000) and unpredictable: swelling attacks strike in random anatomical locations and, if untreated, patients frequently end up in the ICU. These attacks can be lethal.

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Quantity of plasma needed for various HAE plasma derived treatment options raises questions about cumulative exposure to blood plasma

Product	Dose	Source	Per dose	Required for 1 patient for a year	
			Human blood donations	Human donations (2 doses/week)	Total amount of plasma
Berinert®	20 IU/kg	Plasma	5	Varied	Varied
Cinryze®	1000-2500 IU	Plasma	3-8	300-750	0.2-0.6 tons
Haegarda®	60 IU/kg	Plasma	15	1500	1.2 tons
RUCONEST	50 IU/kg	Recombinant	0	0	0

"This is very powerful information. It's the first time that I've even grasped the magnitude of this issue. And I think this needs to be communicated with the rest of the medical community and even to the patients … I was hesitant to write more [prescriptions for C1-inhibitor products], because I worry with so many donations … " – Leading KOL

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HAE Treatment Options Based on Published Results

- RUCONEST was launched as first recombinant (non-plasma) Enzyme Replacement Therapy (ERT)
- Additional and recent data now show that RUCONEST is properly dosed ERT
- RUCONEST taken at the first signs of an attack will nearly always (up to 97%) stop the attack from developing and will protect in 93% of cases for at least three days against subsequent attacks
- Bradykinin/ kallikrein pathway inhibitors suppress symptoms, but have significant limitations in response rates and suffer from break-through events, necessitating additional dosing for the same attack in up to 31% of cases
- Plasma-derived ERTs do not generally have breakthrough attacks, but are sub-optimally dosed for acute treatment and so feature lower response rates and lower sustainability

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Opportunity for Rationalisation of Treatment

- With RUCONEST's recently extended dataset featuring proven and unsurpassed efficacy* and lasting effects (up to 3 days), individualised RUCONEST therapy thus becomes an alternative to many patients that are currently on a combination of prophylaxis and rescue therapies
- Treatment at the first signs of the attack with RUCONEST with the confidence of being able to dose additional RUCONEST for the rare occasions where an attack develops further
- Costly and cumbersome prophylaxis + acute rescue therapy combinations could then be limited to patients suffering from more frequent attacks
- Being in control and not swelling anymore will give patients perspective for increased quality of life and save many prophylaxis injections, rescue therapy and significant cost

*On the basis of comparing published data

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Building a RUCONEST franchise for HAE and beyond

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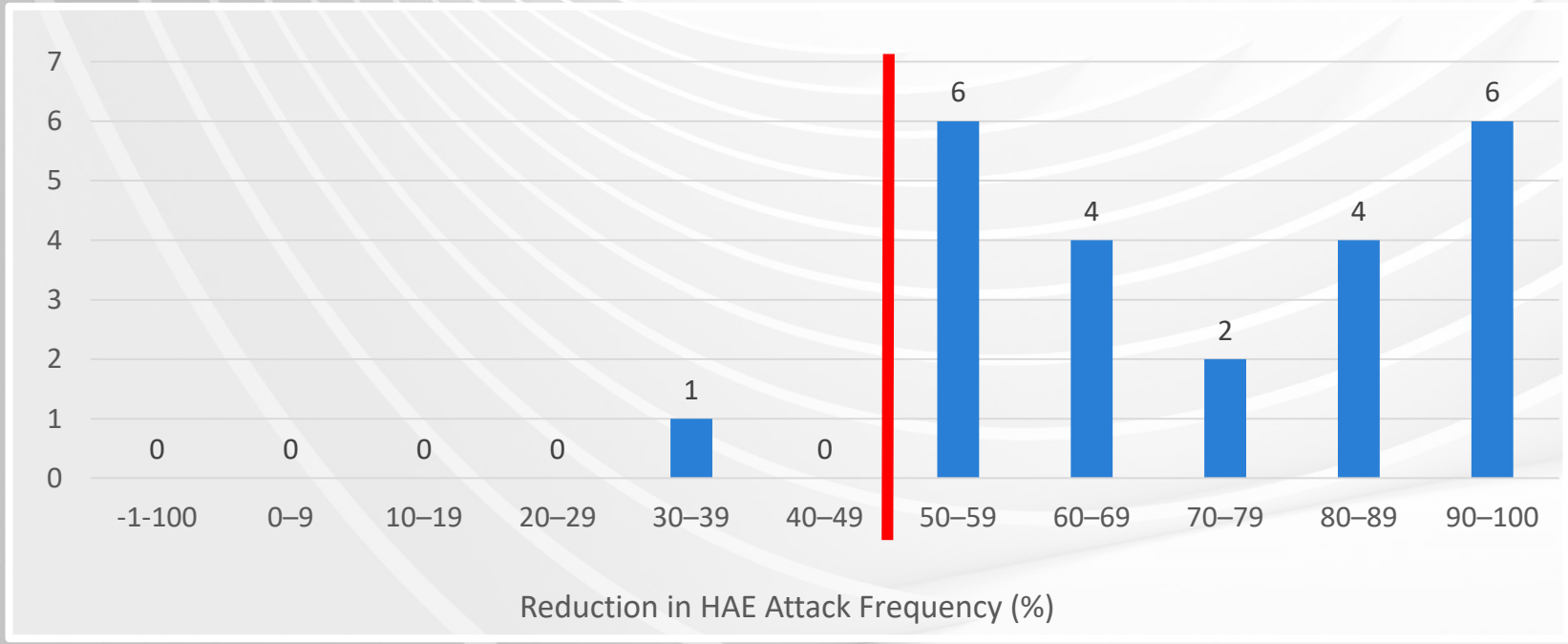
rhC1INH Prophylaxis: Clinical Response With Twice Weekly Dosing

Prophylaxis with Twice Weekly rhC1INH resulted in consistent reduction of HAE attack frequency (n=23)*



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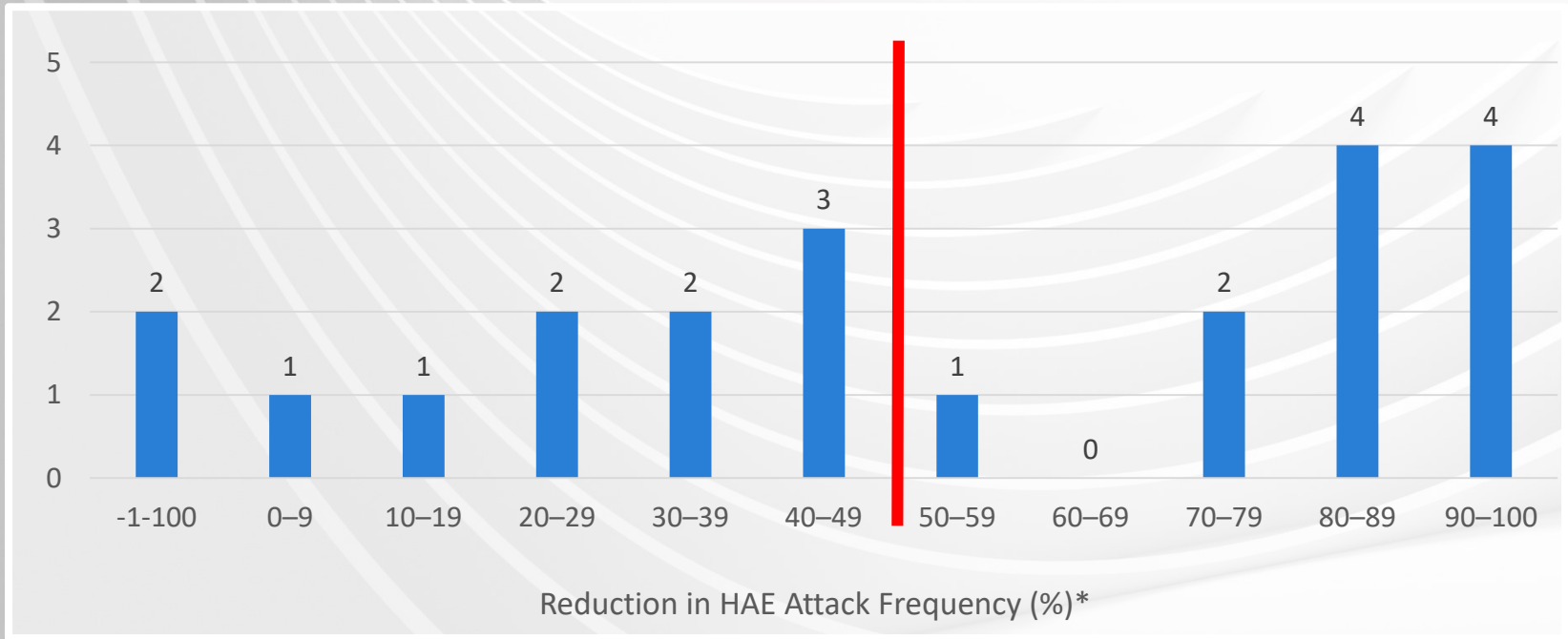
*Riedl et al; Recombinant human C1 esterase inhibitor for prophylaxis of Hereditary Angio-Edema: A Phase 2, multicentre, randomised, double blind, placebo controlled crossover trial; The Lancet; Vol 390, No. 10202, p1595-1602, 30 Sep 2017

pdC1INH Prophylaxis: Clinical Response With Twice Weekly Dosing

Prophylaxis with Twice Weekly Nano-filtered pdC1INH (n=22) resulted in varying reduction of HAE attack frequency



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*2 patients had an increase in HAE attack frequency while receiving nanofiltered C1INH prophylaxis: One patient an increase of 8% and one patient an increase of 85%.

C1INH = C1 esterase inhibitor; HAE = hereditary angioedema. FDA Briefing Document. Blood Products Advisory Committee Meeting. http://www.fda.gov/ohrms/dockets/ac/08/briefing/2008-4355B2-1b.html Published May 2008. Accessed July 26, 2016.

Next Generation RUCONEST for HAE

- RUCONEST efficacy and safety profile for the treatment of HAE attacks is unsurpassed (on the basis of comparing published literature and patient experience)
- Next step: Improving convenience of use
  - New highly concentrated vial in development for faster application of IV therapy (significantly lower volume and very rapid dissolution)
  - New vial will also enable clinical trials to test sub-cutaneous (SC) and intra- muscular (IM) injections for both **treatment and prophylaxis of HAE attacks**
- Clinical trials for SC and IM applications are planned to start in 2H2018
- Investigating delivery technologies for alternative routes of administration

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RUCONEST development beyond HAE

- RUCONEST as first and only recombinant (non-plasma) Enzyme Replacement Therapy (ERT) is based on Pharming's very scalable platform
- Several (undisclosed) Investigator Sponsored Studies in additional indications are underway and initial results expected to become available during 2018
- Company-driven clinical development plan for an undisclosed additional indication has been initiated and first patients to be treated are expected by YE2018
- A Research Day and analyst briefing on new indications and all development progress is planned for end of Q2 2018

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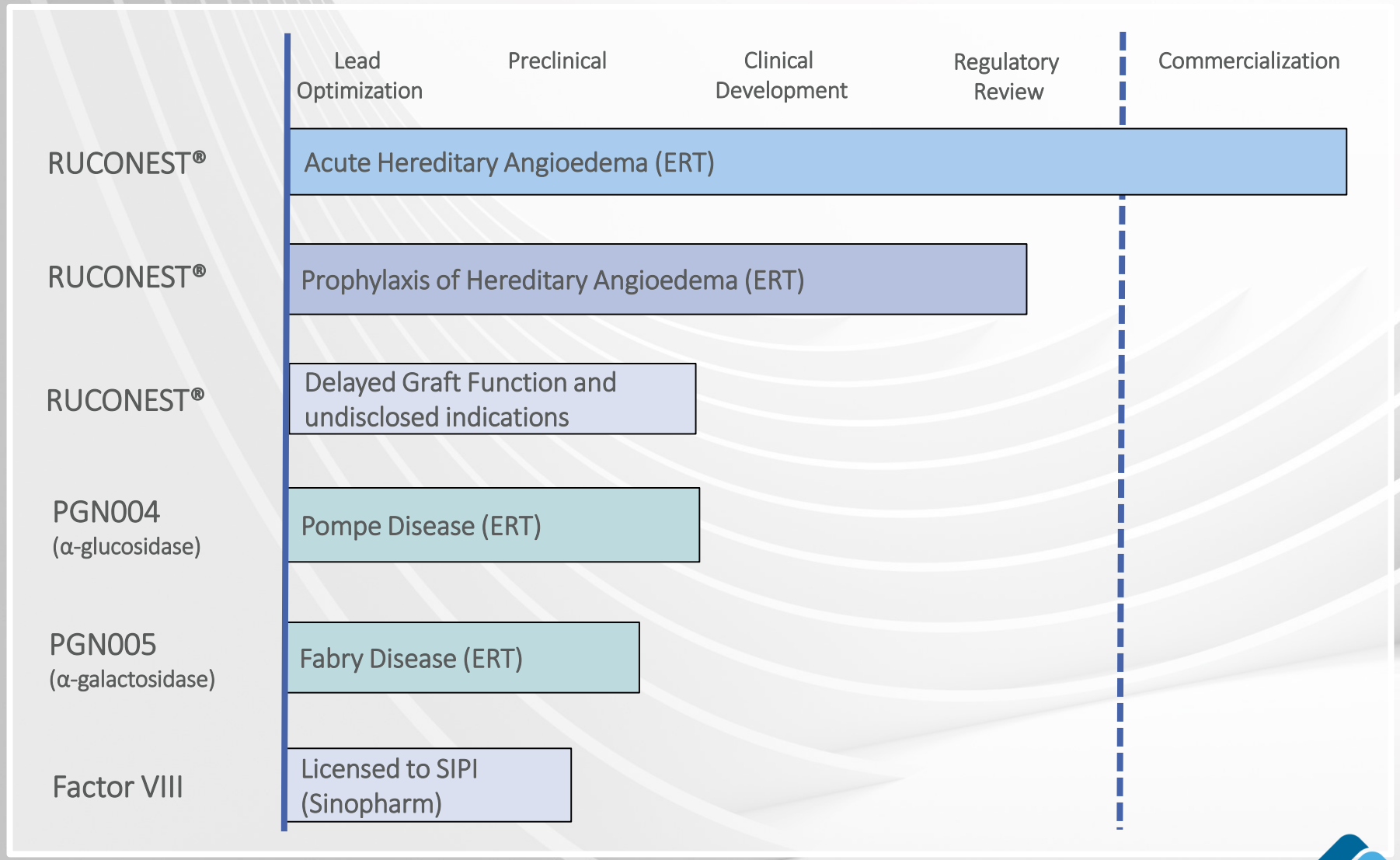
Building a franchise and pipeline



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rh-α-glucosidase (rhaGLU) for Pompe

- Current rhaGLU products risk/ benefit profile offers room for improvement
  - Boxed warnings for immunogenicity/ antibody formation and associated sub-optimal clinical results
- CHO cell bio- reactors derived highly glycosylated proteins such as rhaGLU and rhC1INH appear to reach “the limits” of capabilities of cell based reactors
- Our rhC1INH RUCONEST (equally highly glycosylated) from our transgenic (rabbit) platform does not generate relevant antibody response
- A (small n=4) 36 weeks clinical trial in infants with transgenic (rabbit) derived rhaGLU showed promising efficacy and did not report any safety concerns (2001)*
- De- novo proprietary constructs for our rabbit platform for rhaGLU were developed (2015) and rhaGLU is being produced; up- scaling for initial clinical trial supplies ongoing
- Plan to have IND/ CTA ready to file by YE2018

Pompe’s disease is a rare autosomal recessive lysosomal storage disease, caused by the lack of functional α-glucosidase (5-10k patients world-wide) and if untreated can be fatal in the first year of life

*Van den Hout et al; J. Inherit. Metab. Dis. 24(2001) 266-274

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Financial Information and Outlook 2018

Financing and Capital Structure

- A $100 million 4 year debt facility (July 2021) with OrbiMed Advisors
  - Interest approximately 12%, reducing to 11% if the company reaches $100m in sales
  - Recovery of 115 million shares (24% of outstanding shares) which would otherwise have been issued at prices well below the current share price - now worth around $200 million
- All convertible bonds now redeemed
- Almost all warrants exercised; 1.36 million remaining (0.23% of outstanding shares (596 million)
- Cash at 31 December 2017; €60.0 million ($72 million).
  - 30 September 2017: €38.6 million)

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Preliminary 2017 Results

Amounts in €m except per share data	*2017*	*2016*	*% Change*
Income Statement			
Product Sales	88.7	13.7	*547%*
License Revenue	0.9	2.2	*(59%)*
Total Revenue	89.6	15.9	*464%*
Gross profit	77.2	11.2	*589%*
Operating result	21.9	(11.5)	*290%*
Financial Income, expenses and adjustments	(101.9)	(6.0)	*n/a*
Net result	(80.0)	(17.5)	*(357%)*
Balance Sheet			
Cash & marketable securities	60.0	32.1	*87%*
Share Information			
Earnings per share before dilution (€)	(0.160)	(0.042)	*(492%)*

* For FY 2017 results release, please see www.pharming.com

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Outlook for Full Year 2018

Increasing sales and continued positive results

For the remainder of 2018, the Company expects:

- Continued growth in sales of RUCONEST®, mainly driven by the US and EU operations
- Continuation of positive trend in operating results and achievement of positive Net Earnings during the year
- Continued investment in the expansion of production of RUCONEST® in order to ensure supply
- Investment in further clinical trial programs for RUCONEST® in acute treatment and prophylaxis of HAE with the low volume i.v. version, and in research into new painless versions of RUCONEST®
- Investment in clinical trials to explore additional indications for RUCONEST®.
- Investment in development of the new pipeline programs in Pompe disease and Fabry's disease
- Increasing marketing activity for Pharming, such as opening new countries for RUCONEST®
- Continue to support all our marketing partners to maximize the sales and distribution potential of RUCONEST® for patients in all territories

No further financial guidance for 2018 is provided

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Attractive Growth Proposition

- Pharming has an excellent reputation in the HAE space, and strong support from the patients‘ associations, for further growth against current and new competition
- RUCONEST is the only non-blood-plasma-derived C1 inhibitor therapy and features unsurpassed efficacy and safety profile for treatments of attacks of HAE (comparing published data)
- Next generation RUCONEST: Improving convenience to allow for faster IV and SC/ IM treatment, and potentially other painless administration versions
- Clinical development of additional larger indications beyond HAE for RUCONEST has been initiated
- Maturing rare disease ERT pipeline, first product (Pompe ERT) expected to enter clinical stage development in 2019

Pharming has an excellent reputation in the HAE space, and strong support from the patients‘ associations

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www.pharming.com

Tickers:
ENXTAM: PHARM
Bloomberg: PHAR.AS

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