Pharming Group NV

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Company Overview

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

- Euronext: PHARM - market capitalization: ~€800 million ($977 million) at €1.33 per share
- Headquarters in NL, R&D in France, EU and US commercial operations with approximately 145 employees in total
- 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, Korea and Israel with other territories coming
- Platform technology makes recombinant human molecules cleanly and efficiently
- New Enzyme Replacement Therapies (ERT) for other genetic conditions about to enter clinic

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Corporate Highlights

RUCONEST® Commercialisation	• Re-acquisition of US commercialization rights from Valeant in December 2016 • Q1 2018 revenues: €29.5 million (Q1 2017: €15.5 million) • Temporary supply issues during Q4 2017 at a competitor now resolved
RUCONEST® Franchise Development	• Prophylaxis of HAE with published data showing efficacy 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 filing for IND end of this year • Use same transgenic founder technology to target $1 billion+ markets where all existing products have boxed warnings
Solid Financial Base	• Financed with a $100 million 4yr facility with OrbiMed Advisors in July 2017 • Q1 2018 operating profit: €8.2 million (Q1 2017: €3.9 million) • Q1 2018 net profit: €3.3 million (Q1 2017: net loss €5.7 million) • Positive cashflows: Cash balance at Q1 2018: €60.0 million (Q1 2017 €27.6 million)

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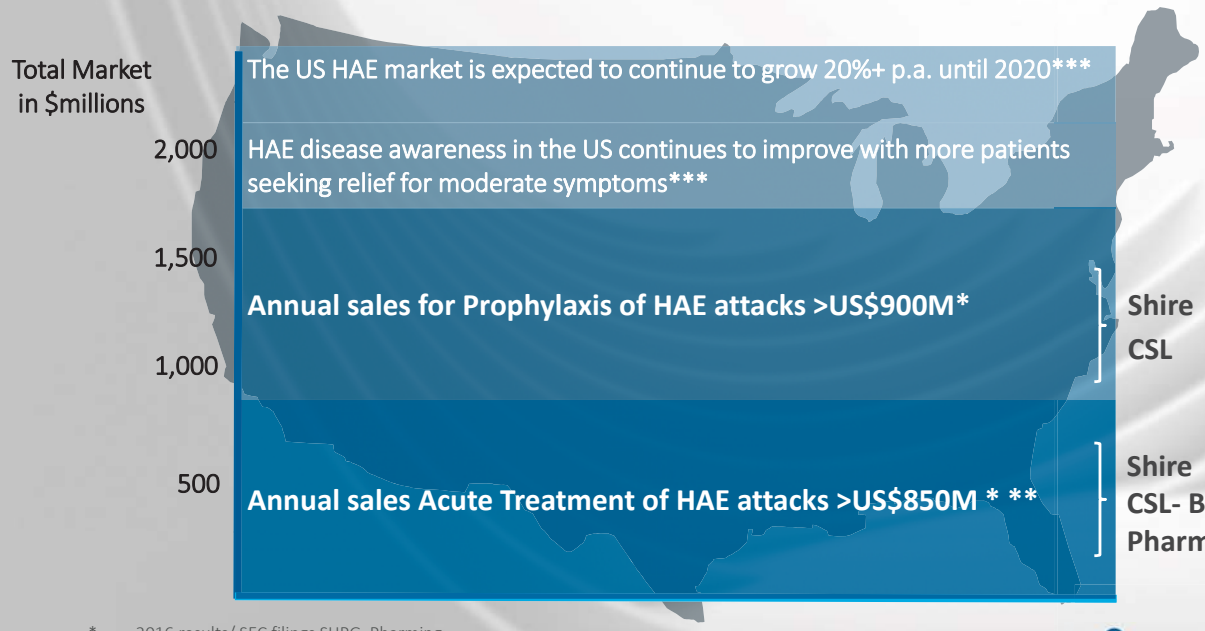
US HAE Market Overview – Rapid Growth, Significant Potential



* 2016 results/ SEC filings SHPG, Pharming
** Includes estimate for plasma-derived C1- esterase inhibitor sales / not disclosed by CSL Behring
*** Leerink Swann, competitor interviews, 13 September 2012 & analyst reports

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HAE Treatment Options Based on Published Results

RUCONEST

- Launched as 1st recombinant (non-plasma) Enzyme Replacement Therapy (ERT)
- Acts fast and stops attacks in up to 97% of cases
- Protects against subsequent attacks in 93% of cases for up to 3 days

Bradykinin / kallikrein pathway inhibitors

- Symptoms suppression, 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
- No clarity on long-term effects of shutting down this pathway

Plasma-derived ERTs

- No breakthrough attacks, but sub-optimally dosed for acute treatment, so feature lower response rates and lower sustainability

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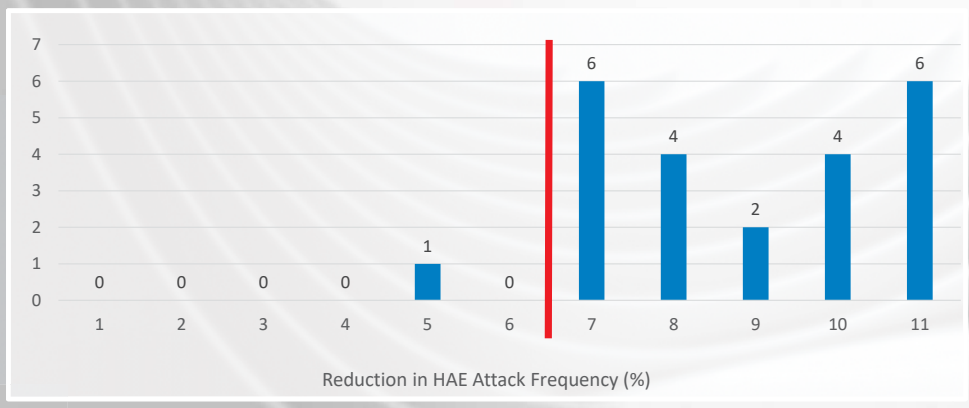
rhC1INH Prophylaxis: Clinical Response with 2x Weekly Dosing

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



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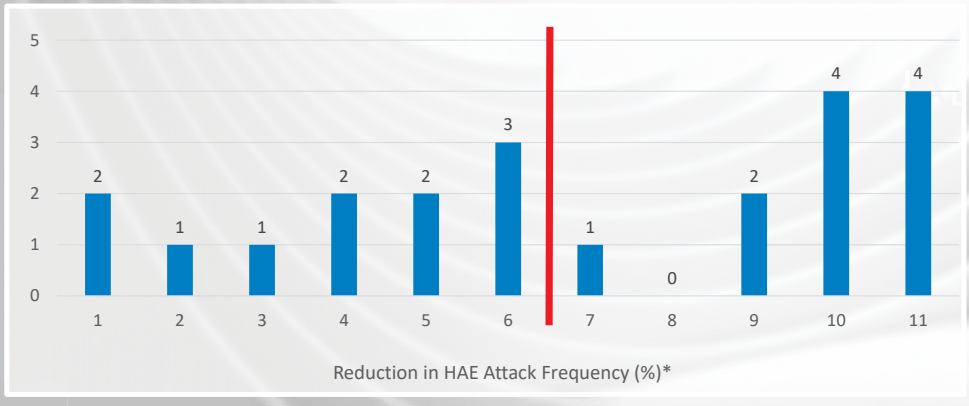
*Riedl et al; Recombinant human C1 esterase inhibitor for prophylaxis of Hereditary Angio-Edema: A Phase 2, multicentre, randomised, double blind, placebo controlled crossover trial; The Lancet; Vol 390, No. 10202, p1595-1602, 30 Sep 2017

pdC1INH Prophylaxis: Clinical Response with 2 x 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 plasma-derived 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.

Large quantities of blood plasma needed

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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Next Generation RUCONEST for HAE

Efficacy and safety profile for the treatment of HAE attacks is unsurpassed*

- Improve convenience of use
- Development of highly concentrated vial for faster application of intra-venous (IV) therapy (significantly lower volume and very rapid dissolution)
- New vial will enable clinical trials to test subcutaneous (SC), intracutaneous (IC) and intra-muscular (IM) injections for both treatment and prophylaxis of HAE attacks
- Clinical trials for SC, IC and IM applications are planned in 2H2018

*on the basis of comparing published literature and patient experience

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RUCONEST development beyond HAE

RUCONEST as first and only recombinant (non-plasma) ERT is based on Pharming's very scalable platform

Investigator Sponsored Studies in additional indications are underway and initial results from first of these is expected in Q3 2018

Company-driven clinical development plan for an undisclosed additional indication to be initiated and first patients are to be treated within 2018

Very difficult for plasma-derived C1 esterase inhibitors to tackle these indications because of the limitations on available donations

Capital Markets Briefing on the new indications and development progress on 21 June 2018 In New York City / live webcast

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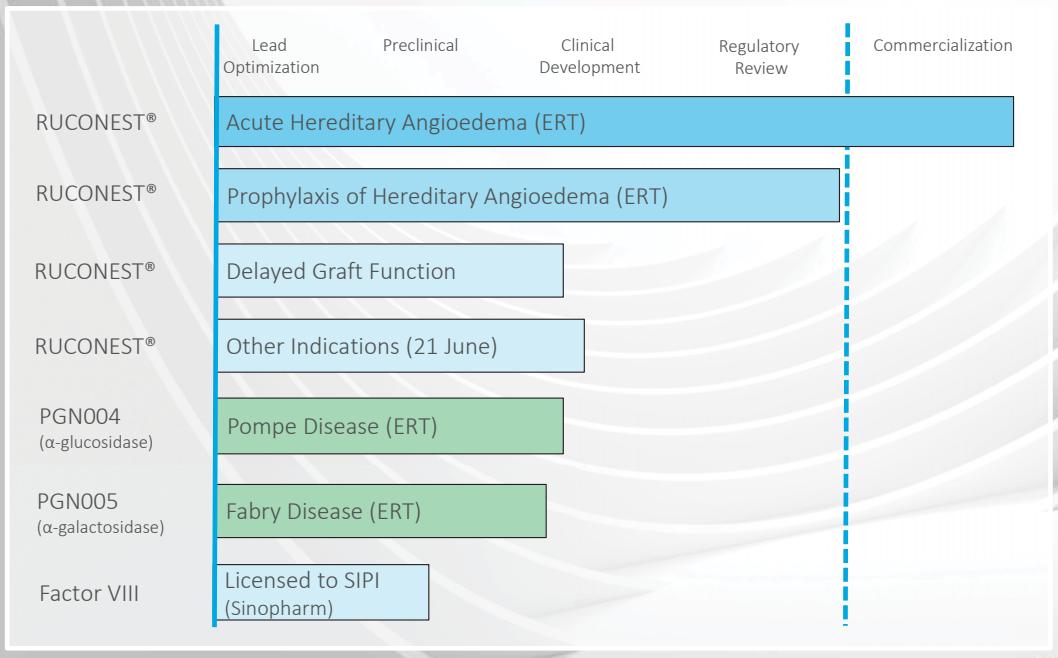
Building a multiproduct franchise and pipeline



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rh-α-glucosidase (rhaGLU) for Pompe

Pompe's disease

- Rare autosomal recessive lysosomal storage disease
- Caused by the lack of functional α-glucosidase (haGLU or GAA)
- 5-10k patients world-wide, with global market over $1 billion
- Usually fatal in the first year of life if untreated, can be fatal if diagnosed later

rhaGLU

- Risk/ benefit profile of existing products is poor, with limited penetration of the population as a result
- Boxed warnings for immunogenicity / antibody formation and associated sub-optimal clinical results
- Cell line-derived recombinant highly glycosylated proteins such as rhaGLU and rhC1INH appear to reach "the limits" of capabilities of cell-based reactors, with products usually highly immunogenic or with off-target effects

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

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rh-α-glucosidase (rhaGLU) for Pompe

rhaGLU

- rhC1INH RUCONEST (equally highly glycosylated) from our transgenic (rabbit) platform does not generate relevant antibody response
- A small 36-week clinical trial in infants with transgenic (rabbit-derived) rhaGLU showed good efficacy and did not report any safety concerns (2001)*
- De novo proprietary constructs for our rabbit platform for rhaGLU have been developed (2015) and rhaGLU is being produced initial clinical trial supplies ongoing
- Plan to have IND filed by YE2018

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

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Financing and Capital Structure

- $100 million 4 year debt facility with OrbiMed Advisors, maturing July 2021
  - Interest ~12%, reducing to **11% in 2019**
  - 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; **0.23%** of outstanding shares remaining
- Outstanding shares: **609 million**

- Cash at 31 March 2018: **€59.8m ($72m)**
- Cash at 30 September 2017 (i.e. After financing): **€38.6m**

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Q1 2018 Results

Financial summary - Euros

3 months to 31 March

Amounts in €m except per share data	*2018*	*2017*	*% Change*
Income Statement			
Revenue from product sales	29.3	15.2	*93%*
Other revenue	0.2	0.3	*(33%)*
Total revenue	29.5	15.5	*90%*
Gross profit	24.5	13.8	*78%*
Operating result	8.2	3.9	*110%*
Net result	3.3	(5.7)	*158%*
Balance Sheet			
Cash & marketable securities	59.8	27.6	*117%*
Share Information			
Earnings per share before dilution (€)	0.006	(0.012)	*150%*

* For Q1 2018 results release, please see www.pharming.com/News

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12 Months Outlook

- Continued growth in sales of RUCONEST® driven by the US and EU operations
- Continuation of positive trend in operating results
- Continuation of positive Net Earnings during the year
- Continued investment in the expansion of production of RUCONEST
- Research and (Clinical) Development investments:
  - RUCONEST® in HAE (SC/IC/IM) with low volume vial to start by end 2018
  - Additional indications for RUCONEST® to start by end of 2018
  - New pipeline: Clinical development Pompe disease early 2019
- Increasing marketing activity, 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

Increasing sales and continued positive results

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Excellent growth proposition

1. Excellent reputation in the HAE space and strong support from the patients' associations, with only product to potentially get approval for both acute and prophylaxis indications in near term
2. 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)
3. Improving convenience of the next generation RUCONEST to allow for faster IV and SC/IM treatment, with other painless administration versions under research
4. About to initiate clinical development of additional much larger indications beyond HAE for RUCONEST
5. Follow-on pipeline, α- glucosidase (ERT for Pompe's disease) expected to enter clinical development early 2019 with potential for lower immunogenicity compared to existing products

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www.pharming.com

Tickers:
ENXTAM: PHARM
Bloomberg: PHAR.AS

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