Pharming Group NV

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Pharming Group N.V. develops and commercializes human therapeutic proteins for innovative therapies meeting important unmet patient needs

- Euronext: PHARM - market capitalization: €220-235 million
- HQ and manufacturing in Netherlands, R&D in France and US commercial operations in New Jersey with approximately 140 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

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Corporate Highlights

RUCONEST® commercialisation

- Re- acquisition of US commercialisation rights from Valeant in Dec 2016
- H1 2017 revenues: €30.6 million (H1 2016 revenues: €5.3 million)
- Q1 2017 operating profit €4.2 million (H1 2016 operating loss €6.2 million)
- US data exclusivity granted until July 2026

RUCONEST® development

- Prophylaxis of HAE Phase 2 study (DBPC) met all endpoints – as good as any
- Next stage being discussed with FDA
- Improving convenience; low volume vial for faster IV and for testing in SQ/IM treatment

Building a pipeline beyond RUCONEST®

- New pre-clinical programs for Factor VIII, Fabry and Pompe diseases
- Uses rabbit founder technology
- Combined market potential $4 billion+
- Other new programs under review

Solid Financial Base

- Re- financed debt structure with a $100 million 4 year debt facility with OrbiMed Advisors in July 2017
- Cash balance at year end 2016 : €31 million/ €25 million in 30 June 2017

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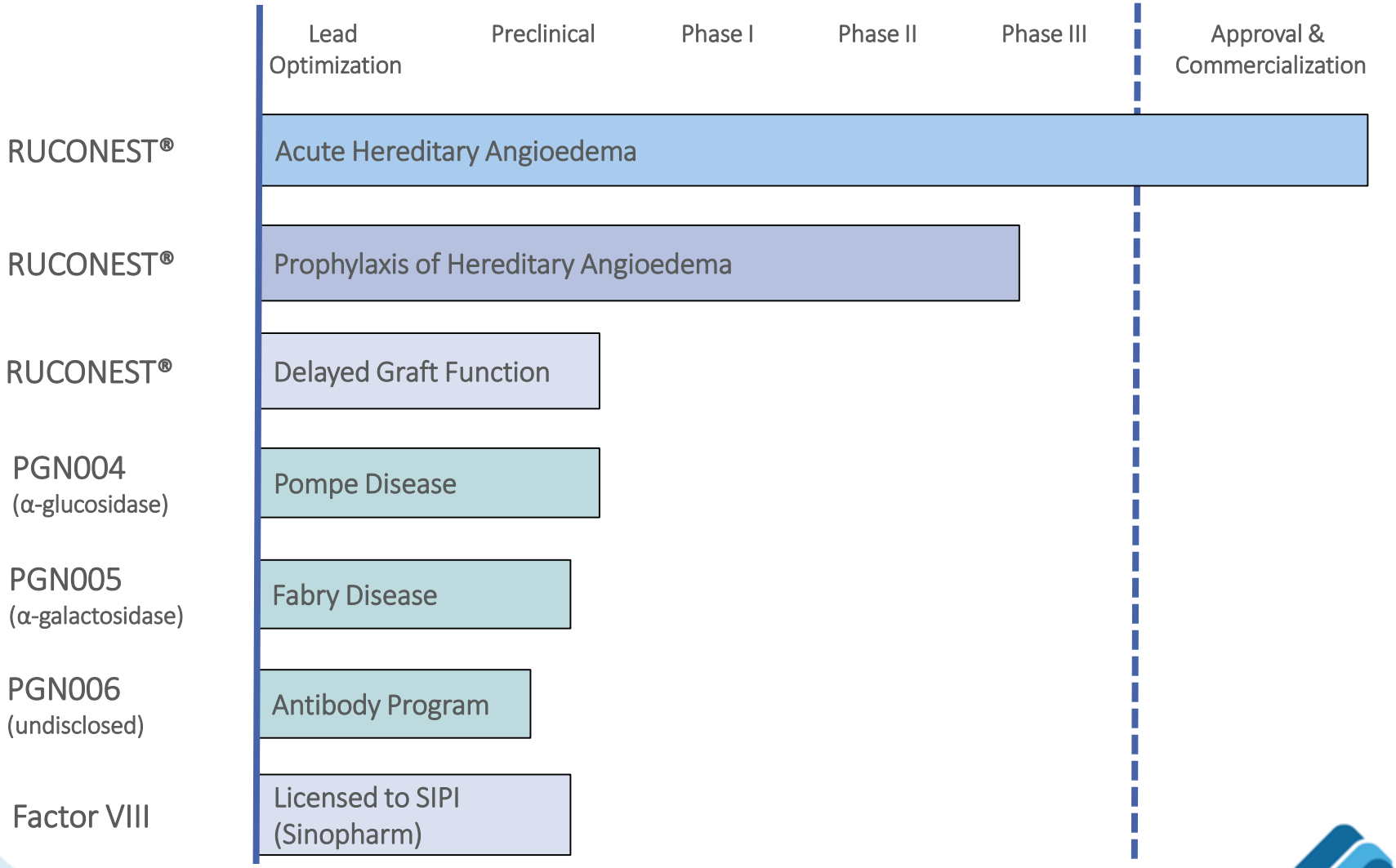
Pipeline



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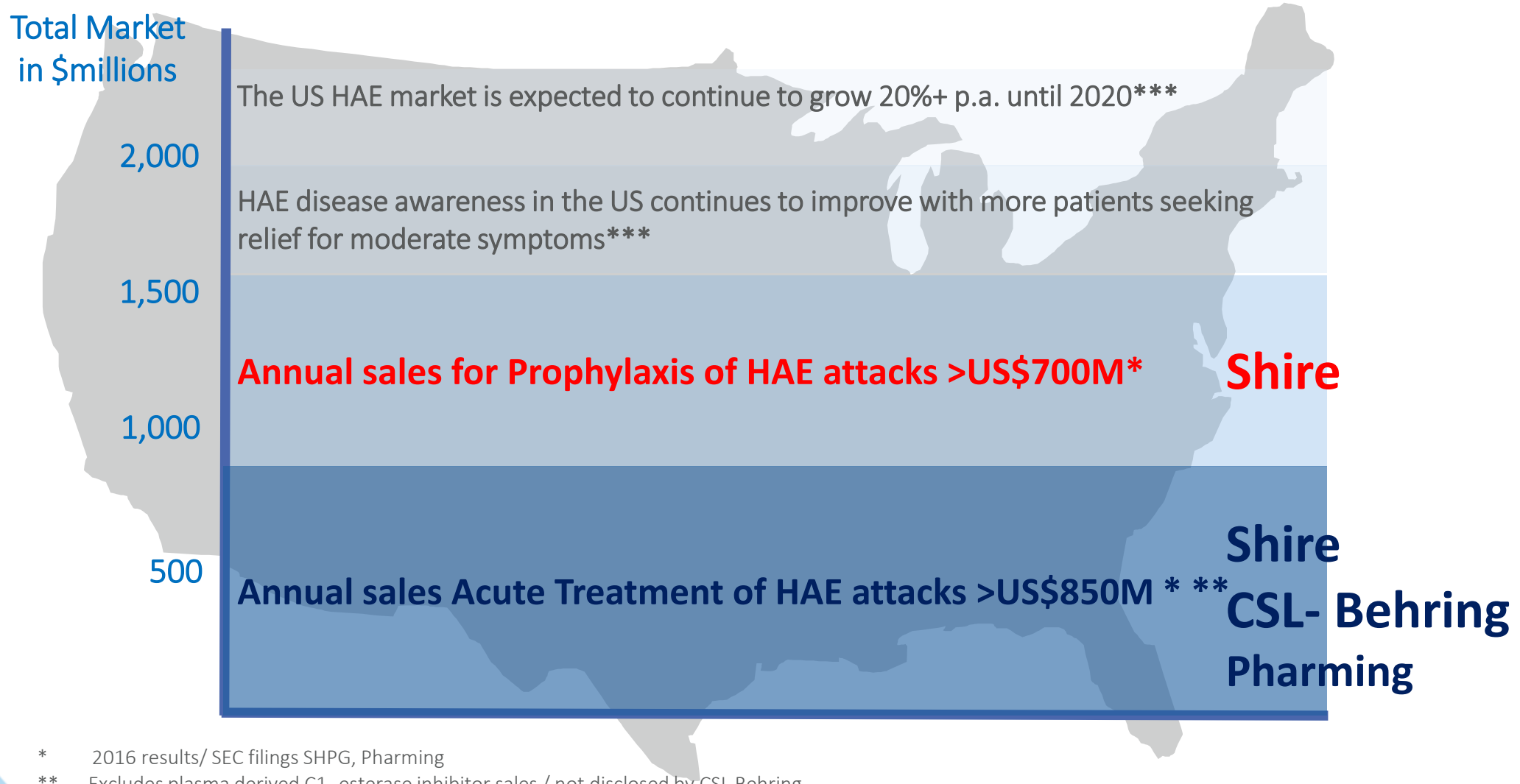
US HAE Market: Rapid Growth, Significant Potential, Very Competitive



* 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

- HAE is rare (between 1 in 10- 50,000) and unpredictable and swelling attacks strike in random anatomical locations and if untreated patients end up in the ER frequently and these attacks can be lethal
- The first generation treatments were inadequately effective
- Prophylaxis with plasma derived protein 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
- Prophylactic therapy is cumbersome and expensive (104 injections/year) and it exposes patients to significant amounts of blood plasma and in addition the cost and burden of the treatment of breakthrough attacks

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HAE Treatment Options Based on Published Results

- RUCONEST was launched as a late entrant and 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 generally stop the attack from developing and will protect for up to three days against subsequent attacks (up to 97%)
- Bradykinin/ kallikrein pathway inhibitors suppress symptoms but have limitations in response rates and suffer from break- through events, necessitating in up to 31% of cases additional dosing for one attack
- Plasma derived ERT do not have breakthroughs but are sub- optimally dosed and feature lower response rates

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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 may now be much better for many patients that are 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 case where an attack develops further
- Costly and cumbersome prophylaxis + acute rescue therapy combinations can then be limited to patients suffering from very frequent attacks
- Being in control and not swelling anymore will give patients perspective for increased QOL and save many prophylaxis injections, rescue therapy and significant $$$

*On the basis of comparing published data

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Clinical Trial Results in Prophylaxis of HAE

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RUCONEST® - Prophylaxis of HAE

- Phase II (double blind, placebo controlled, cross- over design) results meet primary endpoints for once and twice weekly regimen and show that twice-weekly prophylaxis treatment significantly (-73%) reduces attack frequency and features a 96% response rate (>50% reduction of attack frequency)
- The only approved product, a blood plasma derived C1- inhibitor concentrate dosed twice weekly, reduces attacks by 52% and has a 50% response rate*
- RUCONEST® is also approved for acute attacks, hence it can become its own rescue therapy

		Placebo	RUCONEST®	RUCONEST®
Intent –to-Treat Analysis			Once/ week	Twice/ week
(n=32)	Primary: Mean number of attacks	7.2	4.4	2.7
	Reduction in attacks	-	39%	63%
	p-value		*0.0004*	*p<0.0001*
(n=31)	Secondary: % Patients with more than 50 % reduction in attack frequency		42%	74%
Per Protocol Analysis				
(n=23)	Mean number of attacks	7.5	3.8	2
	Reduction in attacks	-	49%	73%
	p-value		*p<0.0001*	*p<0.0001*
(n=23)	% Patients with more than 50 % reduction in attack frequency		57%	96%

*Zuraw et al; Nanofiltered C1- inhibitor concentrate for the treatment of HAE: NEJM 363;6 (August 2010): pp 513-522

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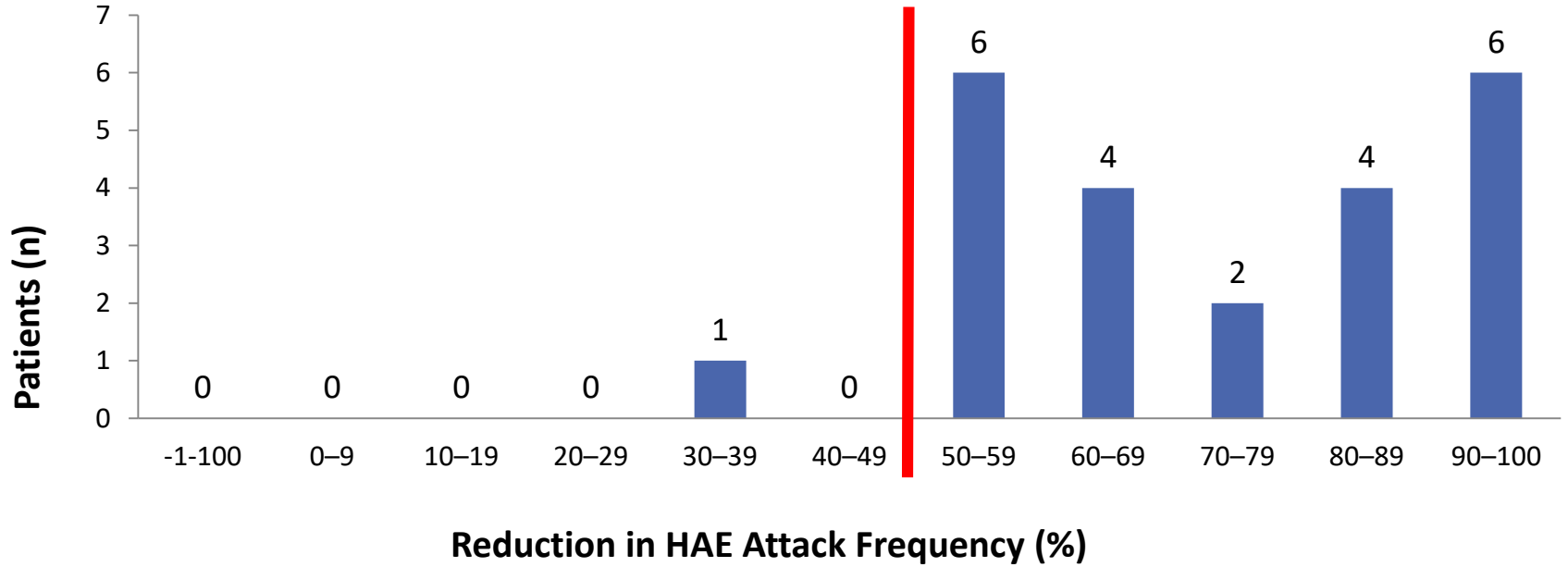
rhC1INH Prophylaxis: Clinical Response With Twice Weekly Dosing

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



HAE = hereditary angioedema; rhC1INH = recombinant human C1 esterase inhibitor.

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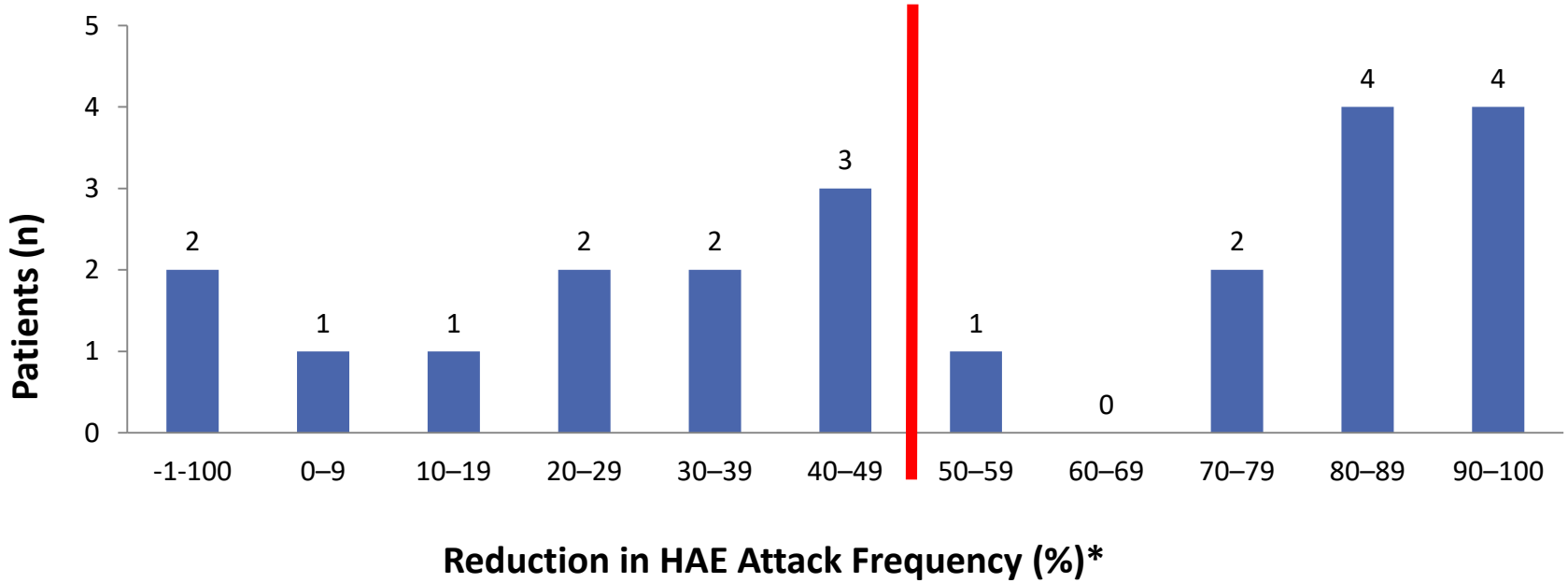
pdC1INH Prophylaxis: Clinical Response with Twice Weekly Dosing

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



***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.htm. Published May 2008. Accessed July 26, 2016.

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Next Generation RUCONEST®

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Next Generation RUCONEST

- 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 1H2018
- Investigating delivery technologies for alternative routes of administration

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US Commercialisation
Re-acquisition of North American Commercialisation Rights for RUCONEST®

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Re-acquisition of the North American Commercial Rights to RUCONEST® from Valeant on 7 December 2016

- Original licensing deal in 3Q 2010 with NASDAQ-listed Santarus for $50 million in upfront and regulatory milestones and profitable supply for 30% of US net sales, with a $45 million in future sales milestones
- December 2013; Salix announces acquisition of Santarus
- July 2014 - FDA approval, and Salix launches RUCONEST in November 2014
- March 2015: Valeant announces acquisition of Salix
- December 2016: Re-acquisition deal closed: upfront payment of $60 million
  - Additional self-funding milestones on sales up to a maximum of $65 million

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Building a US Infrastructure

- Acquired entire Valeant sales team as part of transaction (11 people),
- Expanded sales team and management, led by former senior HAE commercial executive as VP Commercial Operations
- Medical Science Liaison (MSL), Patient Services, Market Access and Managed Care teams in place from mid 2Q
- Major overhaul of Positioning, Messages and Business Rules/ SOPs and re-installment of full service patient care program RUCONEST SOLUTIONS
- Commercial Advisory Board to determine and monitor strategy in US, chaired by former CEO of a NASDAQ 100 Biotech and including former leading senior HAE commercial executives

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Attractive Growth Proposition

- Pharming has an excellent reputation in the HAE space, and strong support from the patients‘ associations
- RUCONEST is the one and 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 oral and/or patch versions
- This commercial infrastructure can be expanded through in-licensing/ acquisition of additional products
- Our pipeline products are expected to come online from 2021 onwards, providing additional scope for expansion of sales

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Financial Information and Outlook 2017

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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
  - Replaces original $40 million of debt and remainder ($40 million) of 18 months $49 million amortising convertible bonds
  - Cash burn to be reduced by €16m in 2017, and €8m in 2018, due lower repayments on debt and amortising bonds and lower cash interest
  - Recovery of 115 million shares (24% of outstanding shares) which would otherwise have been issued at prices below the current share price
- €11.6 million of unsecured tradeable 5 year (2016-2021) 8.5% convertible bonds with a conversion price of €0.284
- Additional recovery of shares (15.6 million) as result of cashless exercise of warrants
  - Remaining warrants (50.5 million) reduced to under 10% of outstanding shares (518 million)

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1H 2017 Results

6 months to 30 June

Amounts in €m except per share data	*2017*	*2016*	*% Change*
Income Statement			
Revenue from product sales	30.1	4.1	*617%*
Other revenue	0.5	1.1	*(67%)*
Total revenue	30.6	5.3	*477%*
Gross profit	27.0	3.2	*763%*
Operating result	4.2	(6.2)	
Net result	(30.2)	(6.7)	*(350%)*
Balance Sheet			
Cash & marketable securities	25.0	21.4	*17%*
Share Information			
Earnings per share before dilution (€)	(0.063)	(0.016)	(293%)

* For H1 2017 results release, please see www.pharming.com

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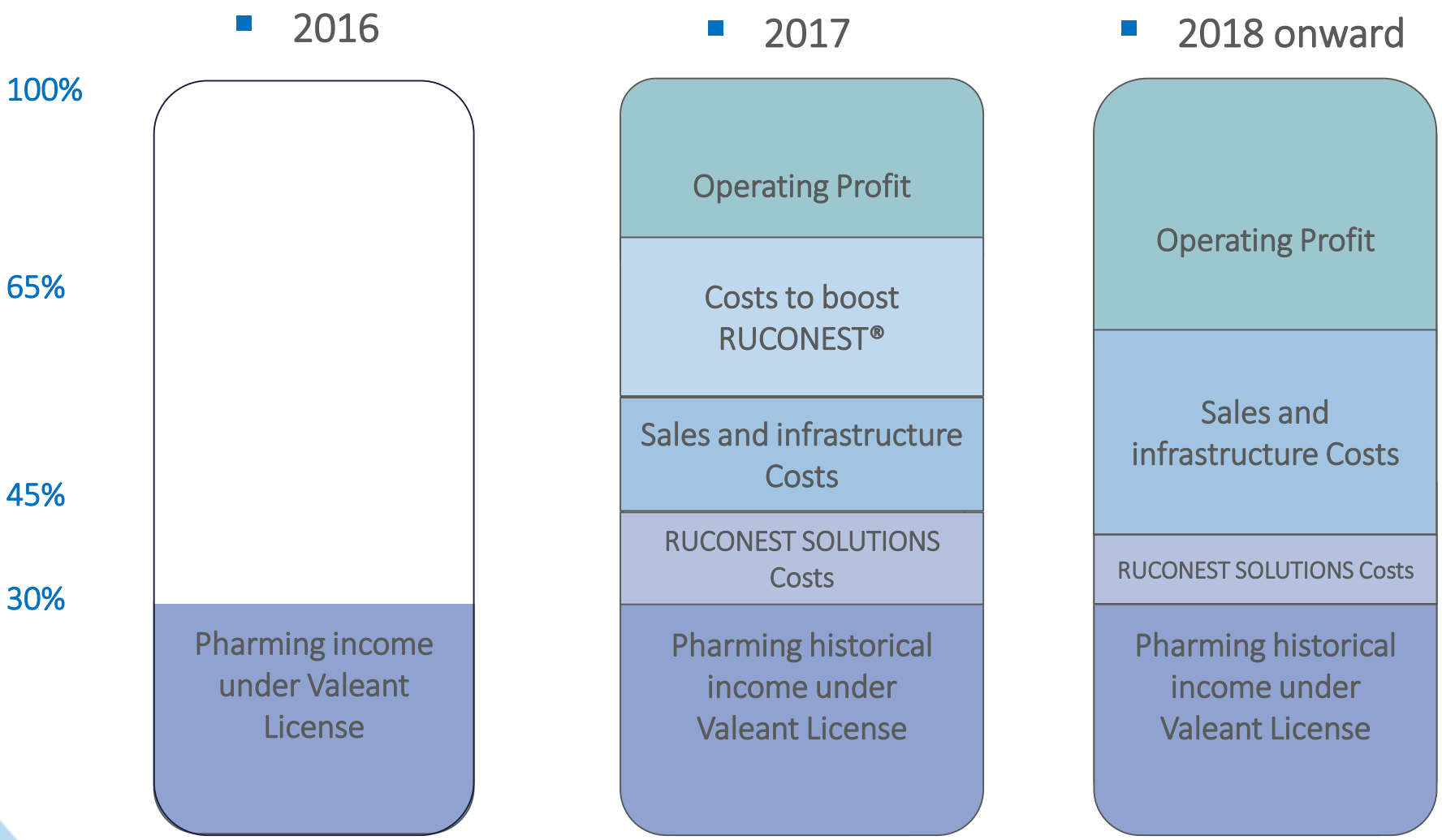
Financial Impact of Reacquisition of North American Rights for RUCONEST®



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Outlook for Remainder 2017

- Increasing sales and continued positive operating results
- Investment in the production of RUCONEST® in order to ensure continuity of supply.
- Assessment of the clinical trial results for RUCONEST® in prophylaxis of HAE by the US FDA and the development of other versions of RUCONEST®
- Increasing marketing activity where this can be profitable for Pharming, in addition to our current territories of Austria, France, Germany, United Kingdom and the Netherlands
- Continue to support our marketing partners in order to maximize the sales and distribution potential of RUCONEST® for patients in all territories, as we continue to believe that RUCONEST® represents a fast, effective, reliable and safe therapy option for HAE patients
- Continue to invest in the new pipeline programs in Pompe Disease and Fabry Disease

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Tickers:

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