



Pharming Group NV

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Overview

- Euronext: PHARM - market capitalization: €260-290 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

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

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

The origin of Pharming

1980

1990

1995

2008



Use of transgenesis for the production of protein therapies

History of Pharming

1988 Business establishment: Production of proteins for pharmaceutical applications in the milk of transgenic animals

1990 Herman: The first transgenic bull

1994 First demonstration of the concept: Human proteins from cow's milk

1998 Transgenic rabbits produce rh- α -glucosidase and rhC1INH *

2000 Start clinical development rh- α -glucosidase (Pompe's disease)

2001 Start of clinical development rhC1INH

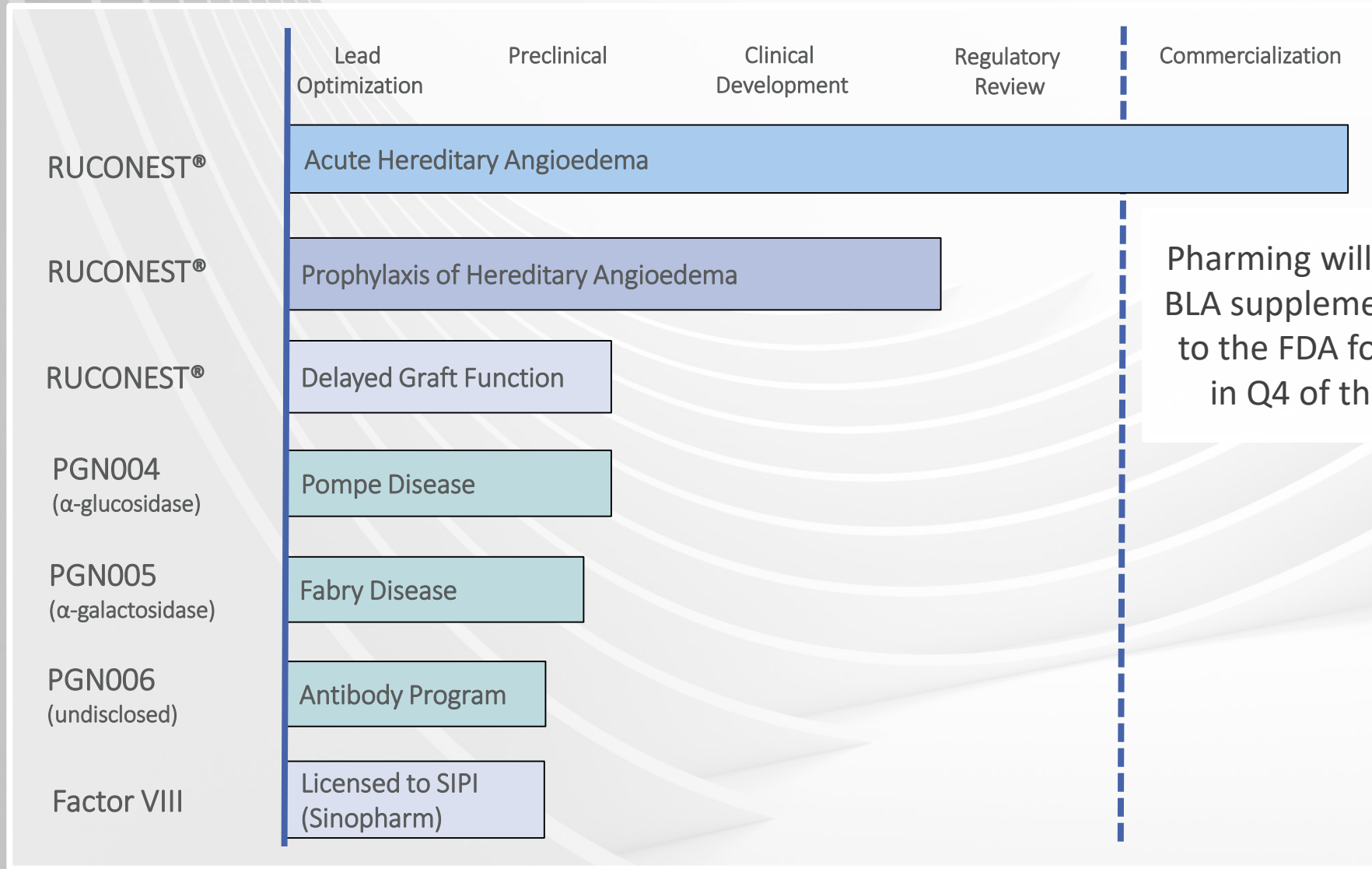
2010 rhC1INH EU Approval (EMA) **

2014 rhC1INH USA Approval (FDA) **

- *recombinant human C1-esterase inhibitor / conestat alpha
- ** for the treatment of angioedema attacks in patients with hereditary angioedema



Pipeline



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

Total Market
in \$millions

2,000

1,500

1,000

500

The US HAE market is expected to continue to grow 20%+ p.a. until 2020***

HAE disease awareness in the US continues to improve with more patients seeking relief for moderate symptoms***

Annual sales for Prophylaxis of HAE attacks >US\$700M*

Shire

Annual sales Acute Treatment of HAE attacks >US\$850M * **

Shire
CSL- Behring
Pharming

* 2016 results/ SEC filings SHPG, Pharming

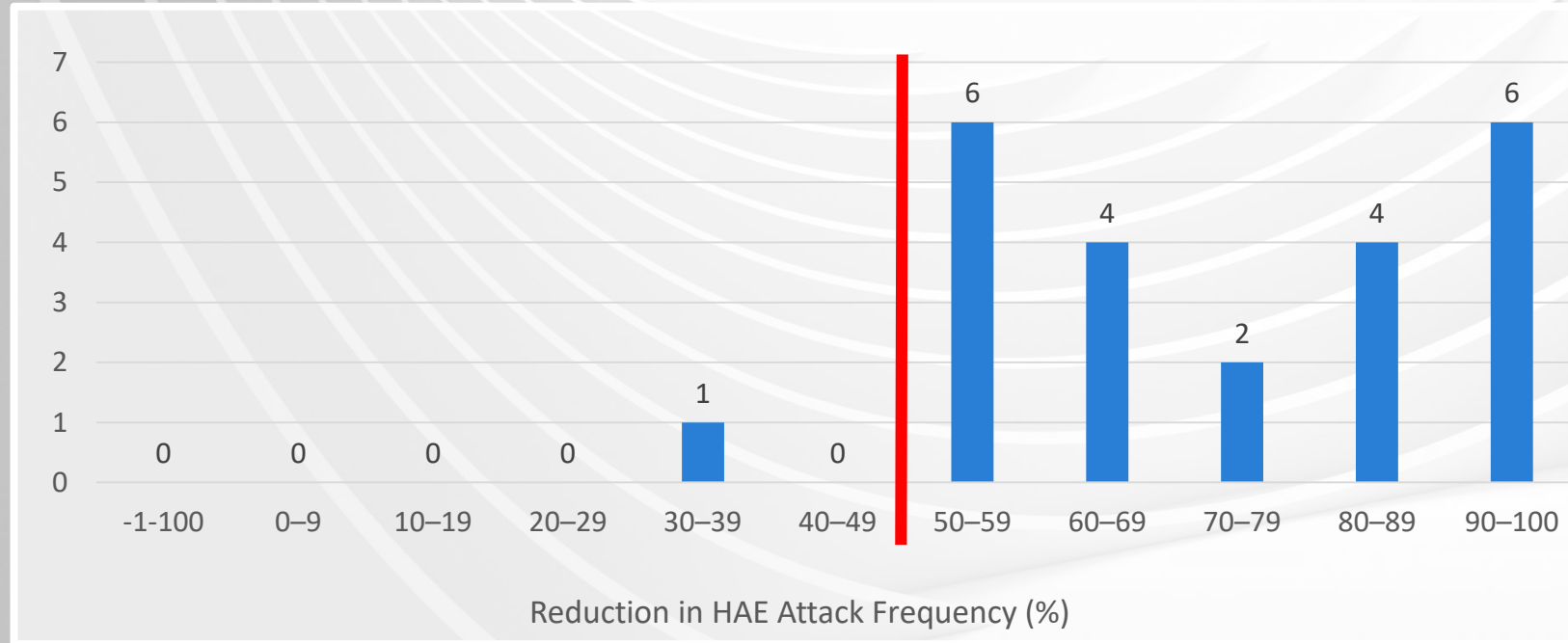
** Excludes plasma derived C1- esterase inhibitor sales / not disclosed by CSL Behring

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

Clinical Trial Results in Prophylaxis of HAE

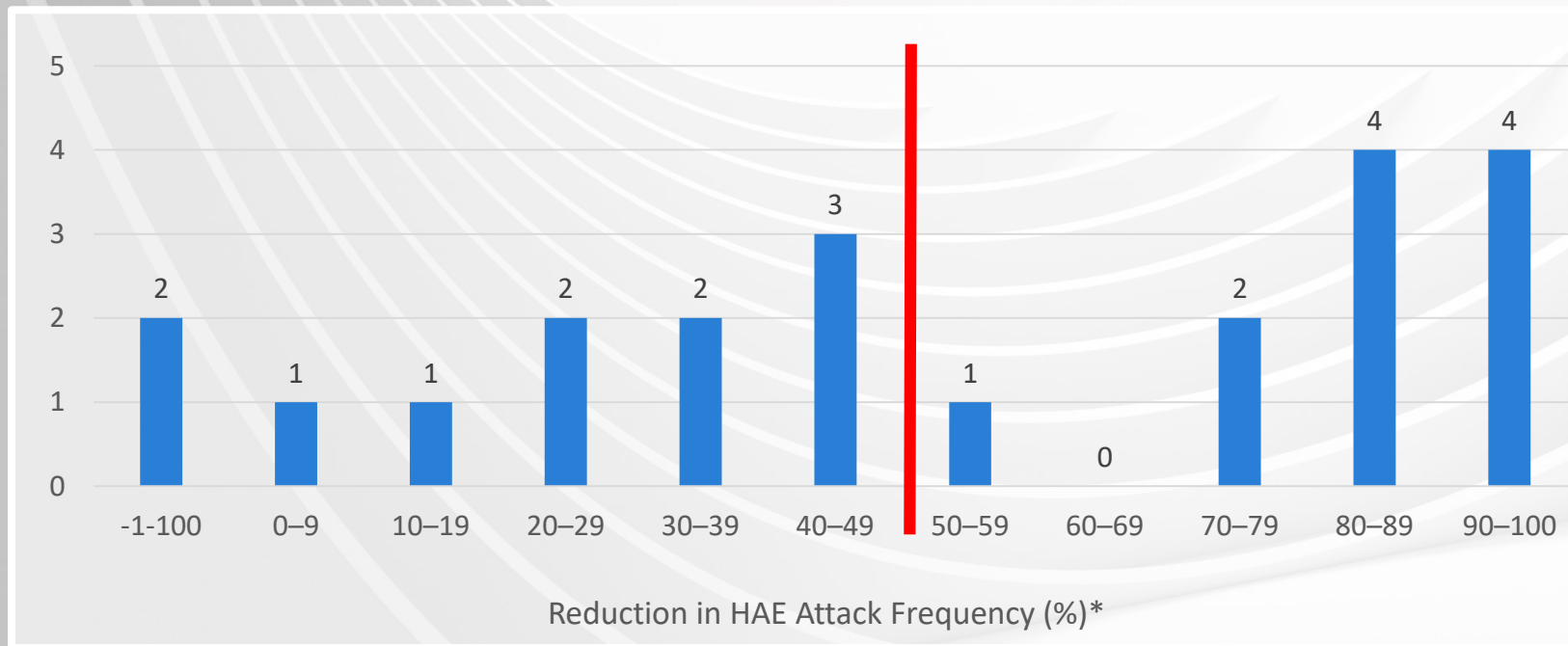
rhC1INH Prophylaxis: Clinical Response With Twice Weekly Dosing

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



rhC1INH 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.html> Published May 2008. Accessed July 26, 2016.

Next Generation RUCONEST®

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
- Investigating delivery technologies for alternative routes of administration



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

Re-acquisition overview

- 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

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

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



Attractive Growth Proposition

- 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

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

Financial Information and Outlook 2017

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)



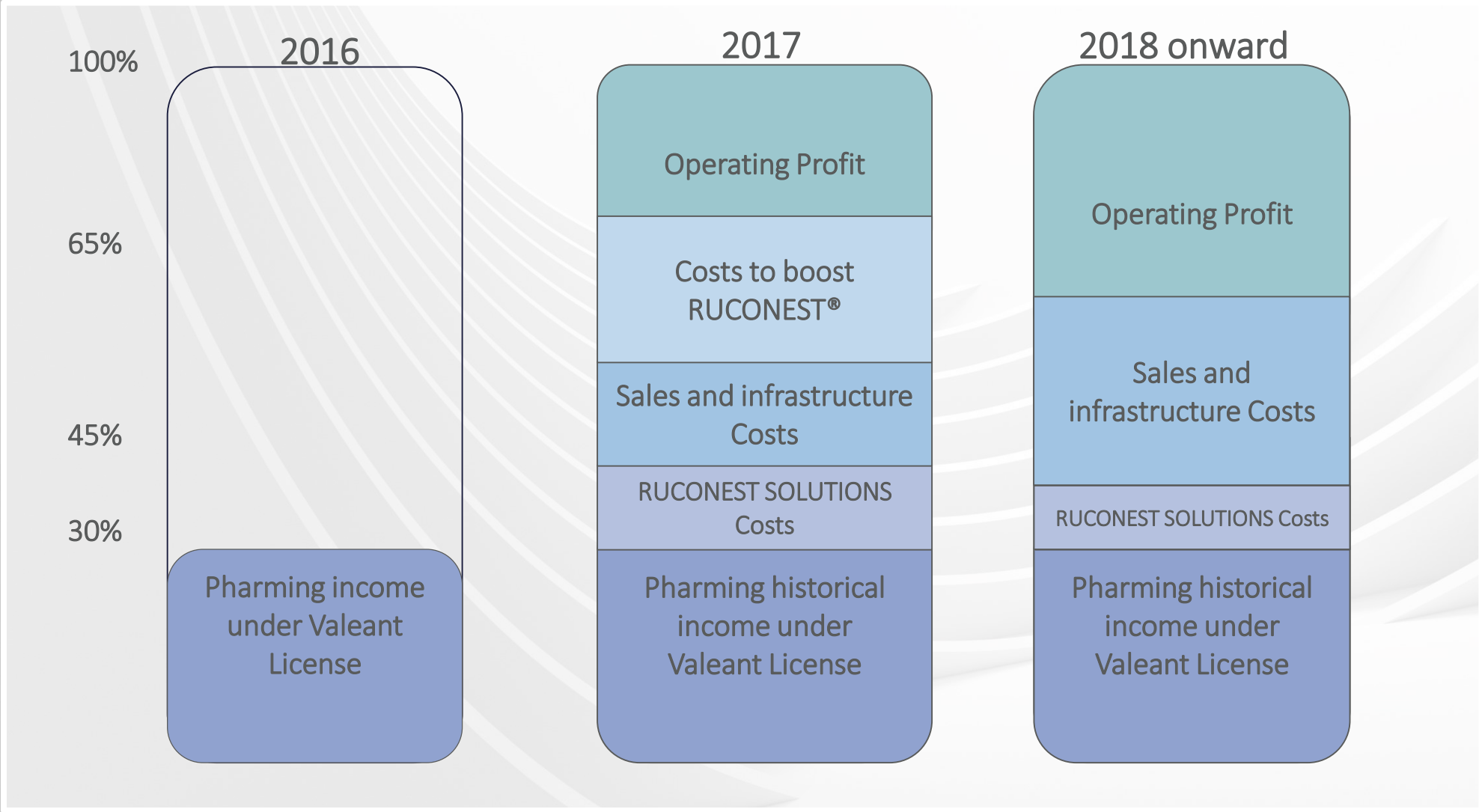
1H 2017 Results

6 months to 30 June

<i>Amounts in €m except per share data</i>	<i>2017</i>	<i>2016</i>	<i>% Change</i>
<i>Income Statement</i>			
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%)
<i>Balance Sheet</i>			
Cash & marketable securities	25.0	21.4	17%
<i>Share Information</i>			
Earnings per share before dilution (€)	(0.063)	(0.016)	(293%)

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

Financial Impact of Reacquisition of North American Rights for RUCONEST®



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

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

Increasing sales and
continued positive
operating results

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Tickers:

ENXTAM: PHARM

Bloomberg: PHAR.AS



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Questions?