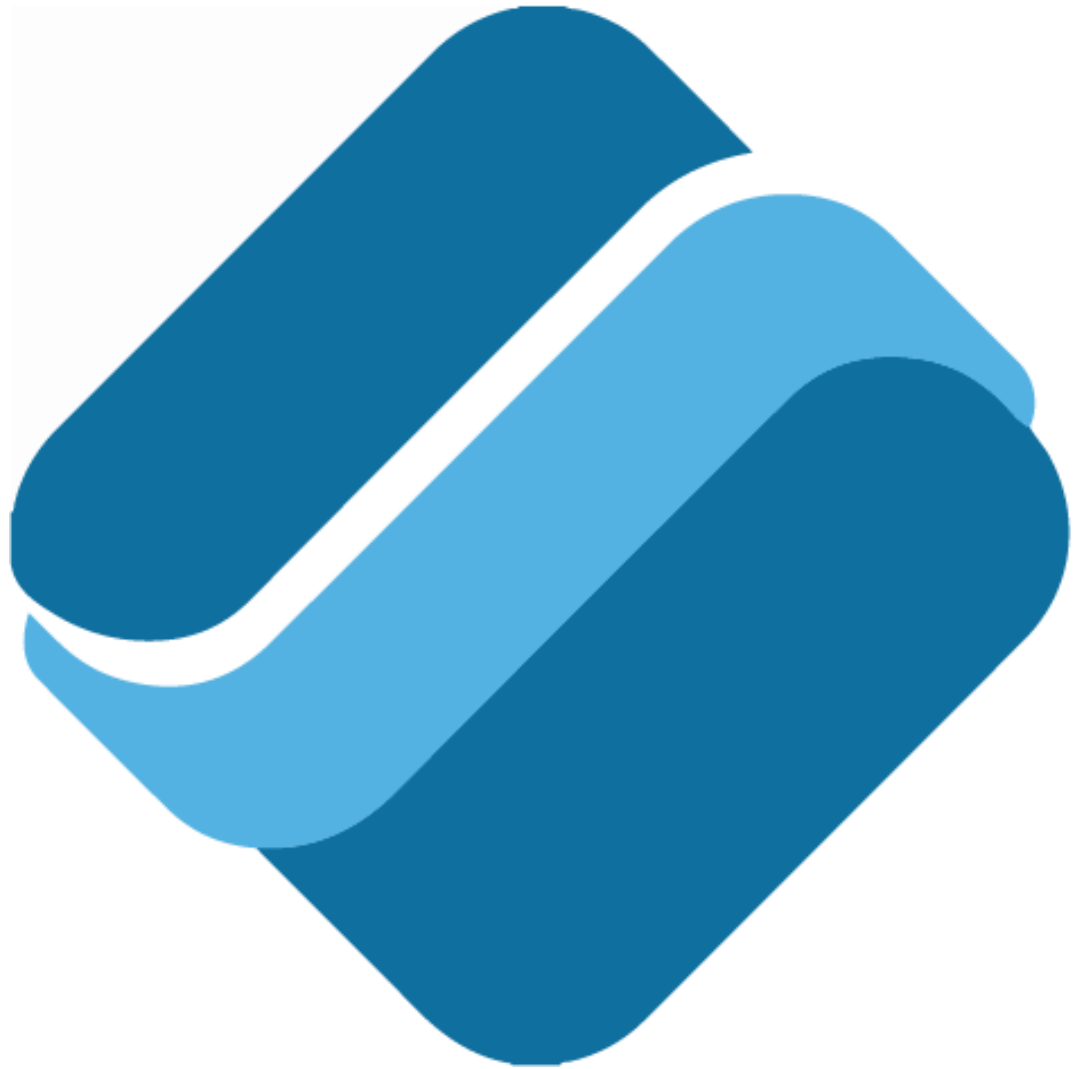
Pharming



Pharming Group NV

Sijmen de Vries
Chief Executive Officer

Investor Tour
Zwolle

4th October 2018

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Agenda

19:00 uur - Receptie

19:25 uur - Welkomstwoord

19:30 uur - Korte corporate presentatie

20:00 uur - Sijmen de Vries Q&A met Leon Melens

20:30 uur - Audience Q&A

21:00 uur - Aanvang borrel

22:00 uur - Einde

Company Overview

- Euronext: PHARM: ~€585 million (~$678 million) at €0.95 per share
- Headquarters in NL, R&D in France, EU and US: 170 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
  - Now being explored for larger indications: pre-eclampsia, CIN, Cardiac Protection, DGF
- Platform technology makes recombinant human molecules cleanly and efficiently
- New Enzyme Replacement Therapies (ERT) for other genetic conditions about to enter clinic

Three horizons of growth

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Making RUCONEST® a better HAE product

- Low volume IV
- Subcutaneous
- Intramuscular
- Painless intradermal
- Prophylaxis for HAE

Add more HAE sales

Meeting other unmet medical needs with the same product

- Pre-eclampsia
- Others such as Contrast-induced Nephropathy, Cardiac Protection, Delayed Graft Function and Hypovolemic Shock

Add more RUCONEST® sales

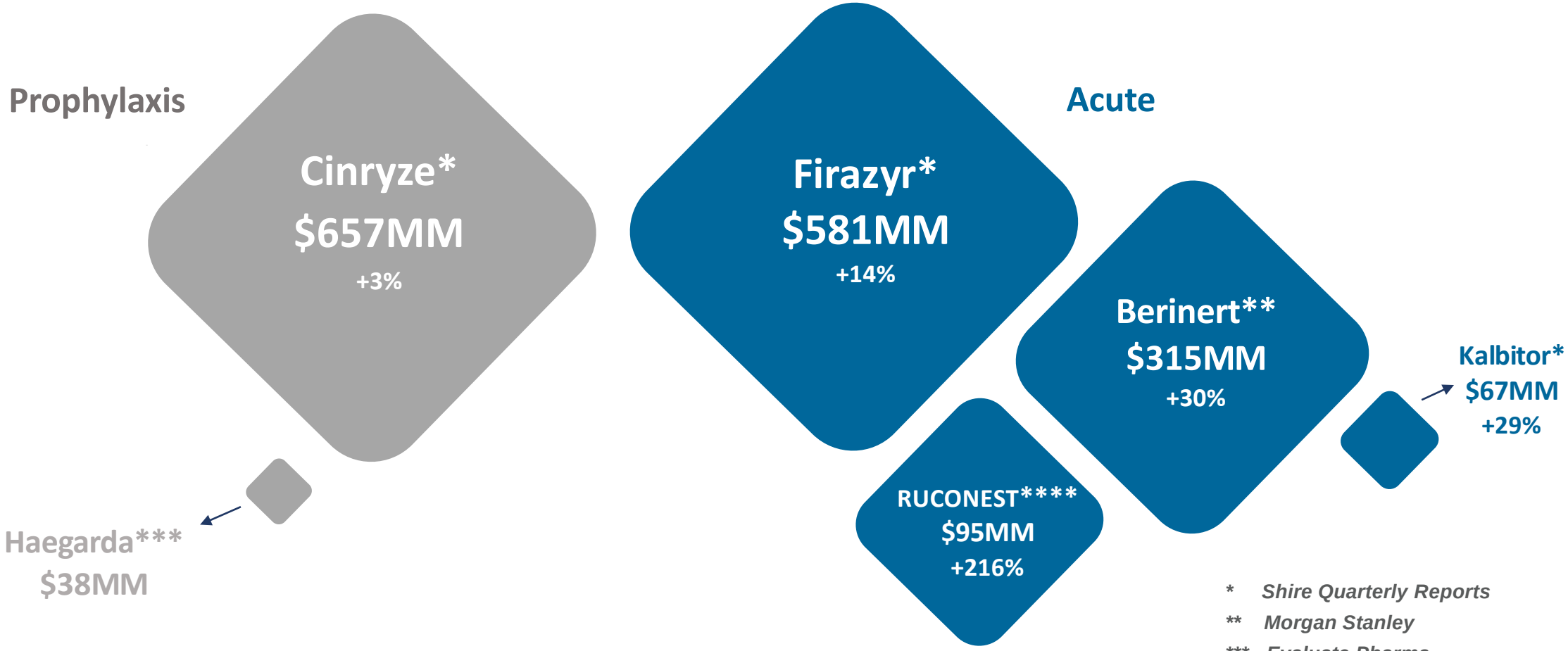
Meeting other unmet medical needs with another products

- α-glucosidase (Pompe)
- α-galactosidase (Fabry)
- Others

Add more products to sell

US Market for HAE drugs

~$1.7 billion Sales in 2017 (Year-on-year growth 17%)



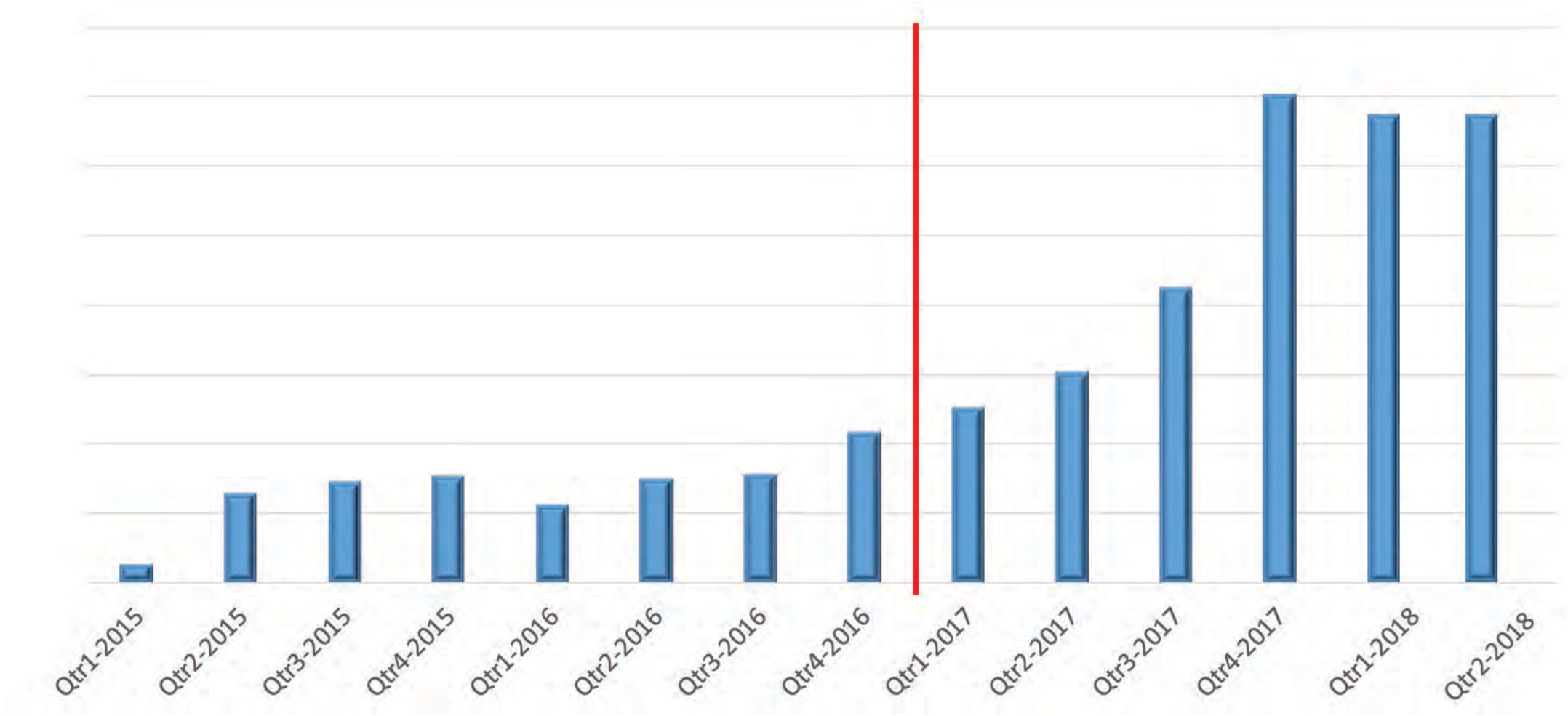
* *Shire Quarterly Reports*

** *Morgan Stanley*

*** *Evaluate Pharma*

**** *Pharming Reported Net Sales*

US quarterly sales development in volumes



What patients want when living with HAE

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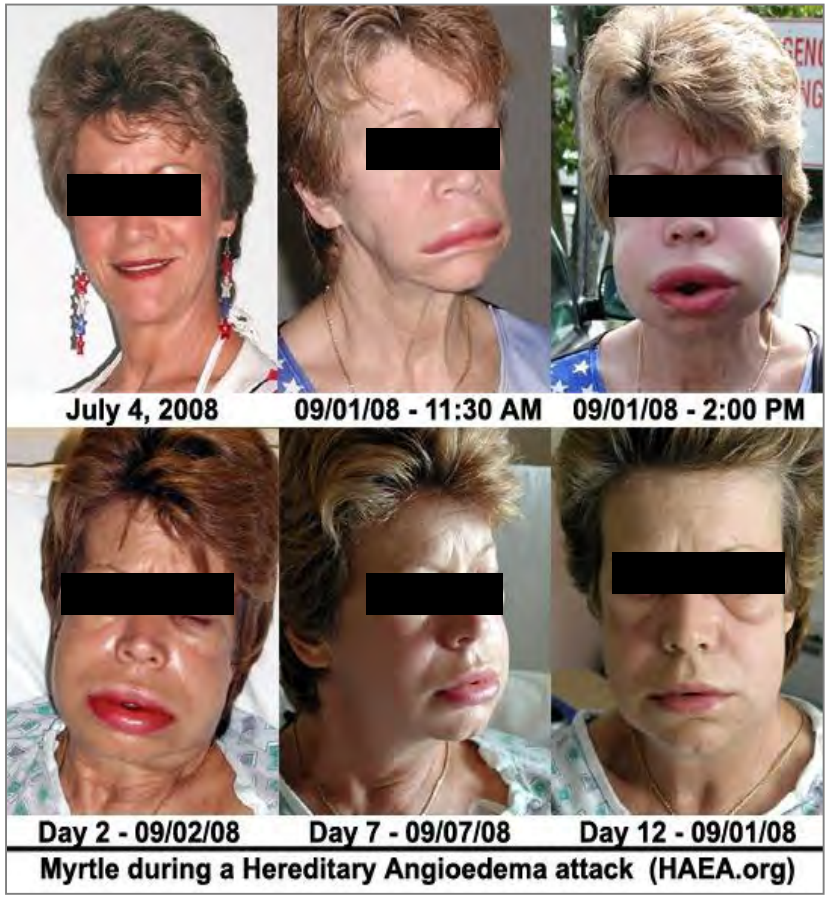
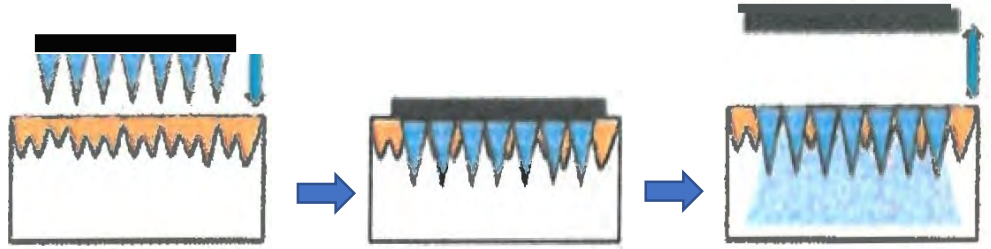
Patients want a therapy that is convenient and pain-free



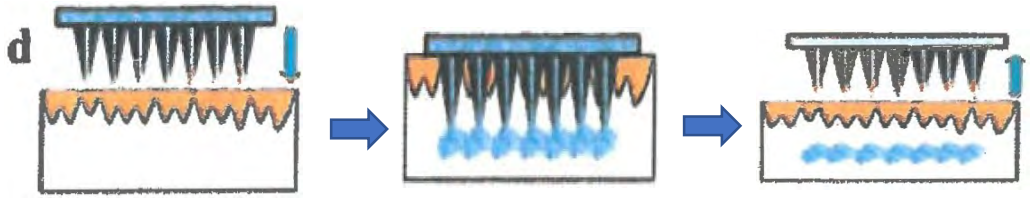
"My veins are shot, so I inject my medicine in my stomach. It is so painful and my stomach has bruises everywhere. I wish I had a medicine that didn't hurt and was quick and easy to use."

Formulation development of RUCONEST®

- The "RUCONEST® liquid" formulation (14ml → 3ml) can be used as starting material for the generation of subcutaneous, intra-muscular & intradermal application systems
- New proprietary 'painless' intradermal delivery applications are being developed:
- Dissolving point device:



- Reservoir device:



- These painless versions should differentiate RUCONEST® from competitors, all of whom have painful injections

New Activities with rhC1INH (RUCONEST®)

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Initial Therapeutic Indications selected:

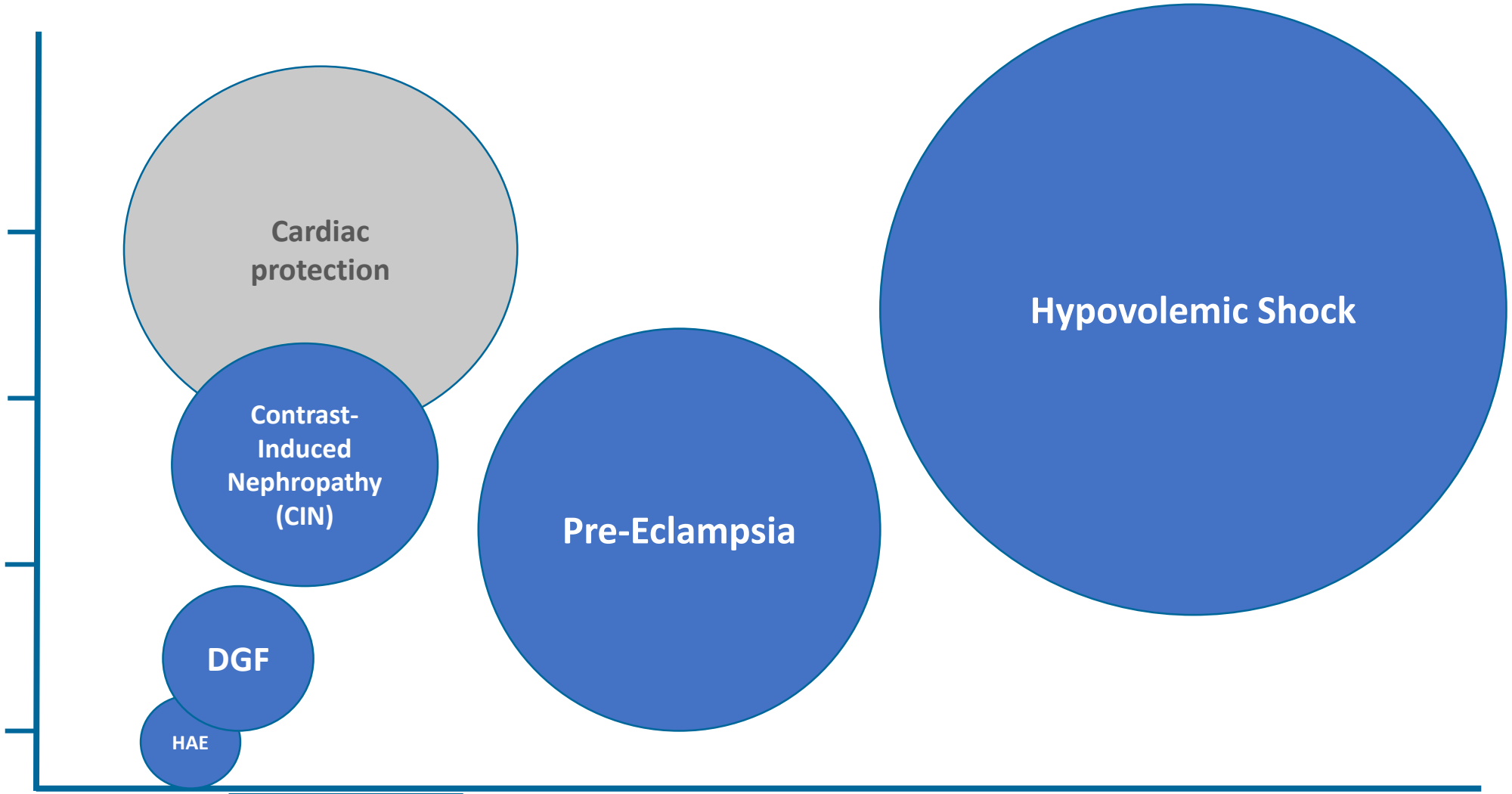
New Potential Indications using existing formulation

- Organ damage after contrast media application:- Contrast-induced Nephropathy (CIN) (ongoing Investigator initiated study)
- Tissue Damage after Toxic Event :- Pre-Eclampsia (Zwangerschapsvergiftiging) (new Pharming)
- Tissue Damage after Hypoxic Event :- Delayed Graft Function (new Investigator initiated study)
- Vascular/cardiac damage due to investigation/operation:- Cardiac protection (depends on data from above study)
- Shock response after trauma:- Hypovolemic Shock – ongoing preclinical research projects with US Army and US Air Force

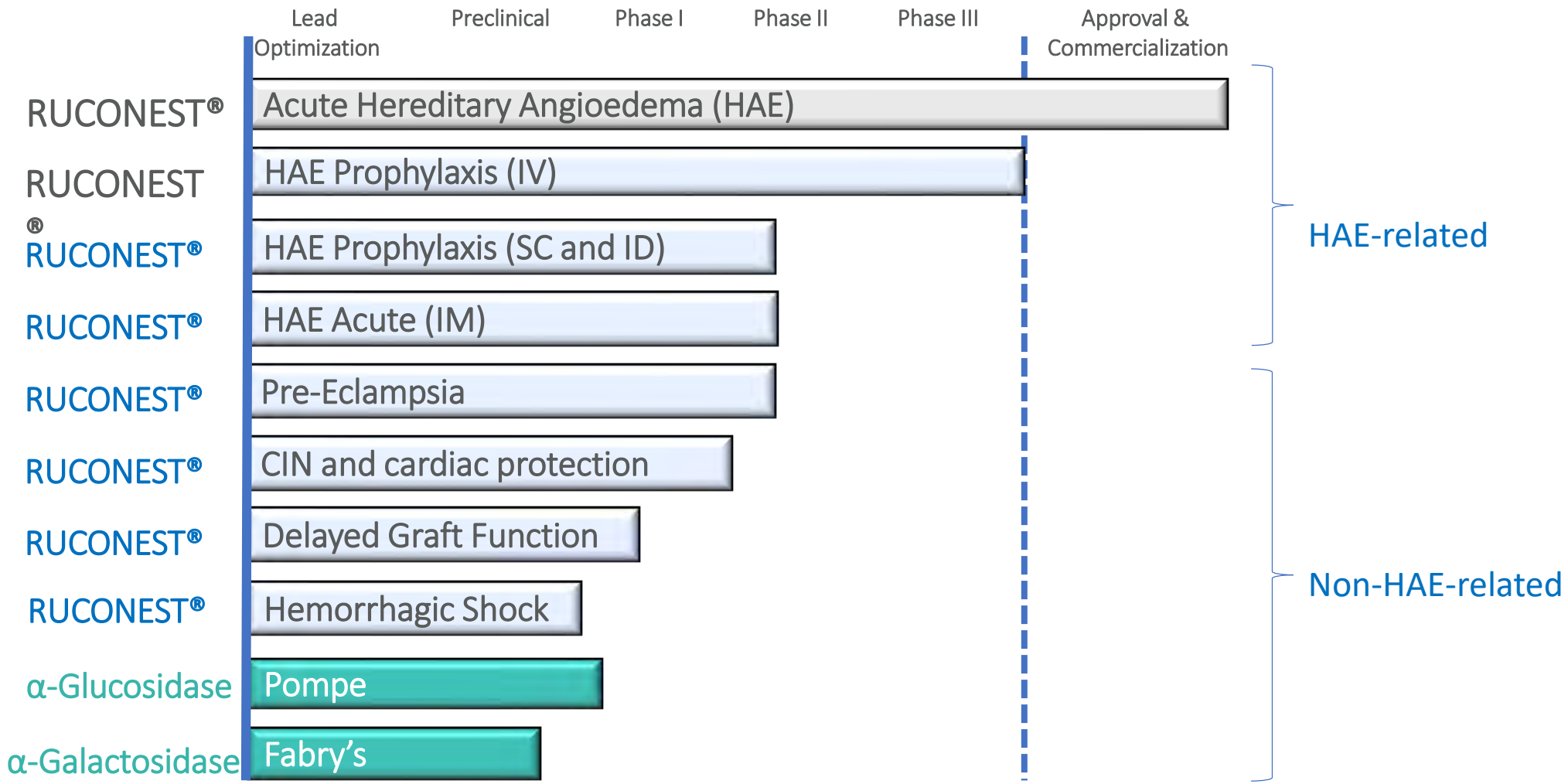
Brand New IP:

- New Pharming patents filed in 2018 covering the new indications
- Patents cover all forms of rhC1INH

Potential addressable future markets

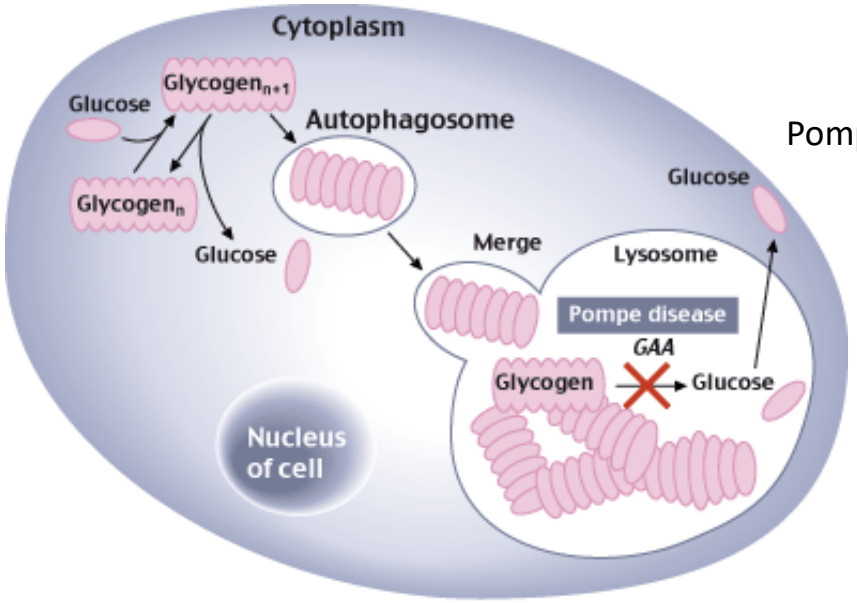


Expansion of pipeline to multiple products and markets

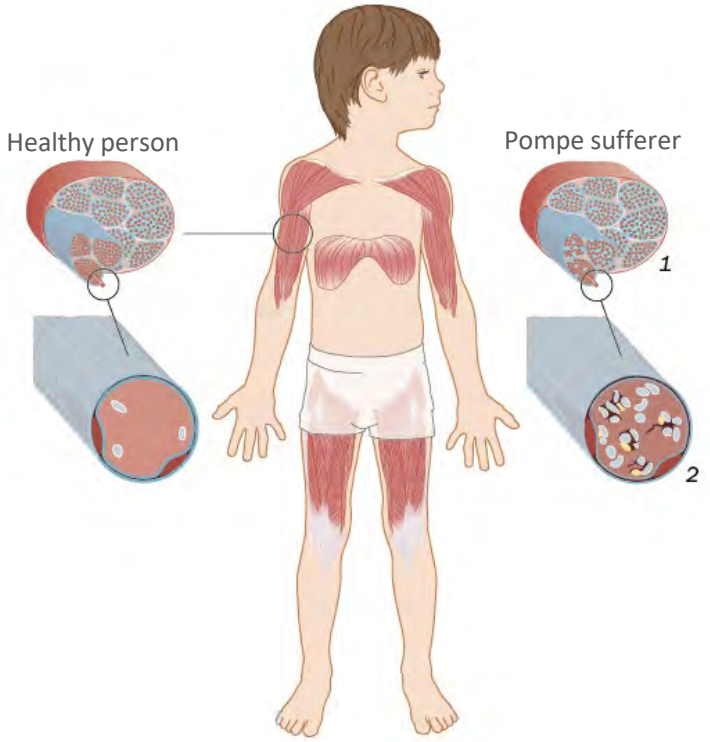


Pompe diseases

Normal situation



Pompe disease



- Progressive decrease in muscle strength starting with the legs and moving to smaller muscles in the trunk and arms, such as the diaphragm and other muscles required for breathing
- Cardiac failure and respiratory failure are the most common causes of death

Pompe diseases

α-glucosidase

Market potential for Pharming is over $1 billion per year in each indication

- Attractive market:
  - All current products have severe shortcomings and boxed warnings, but together sell for over $1 billion
  - Second generation products have their own shortcomings
  - Many patients are not on therapy because of antibody formation or adverse reactions
  - rhC1INH RUCONEST (equally highly-glycosylated) from our transgenic (rabbit) platform does not generate relevant antibody responses
  - A small 36-week clinical trial in infants with previous transgenic (rabbit-derived) rhaGLU showed good efficacy and did not report any safety concerns (2001)*

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

Pompe diseases

α-glucosidase

De novo proprietary constructs for our rabbit platform for rhaGLU have been developed and a new recombinant rhaGLU is being produced for initial clinical trial supplies

  - New version is closer to natural human GAA than previous Pharming version, and also believed to be much closer than any other tested recombinant version to date

- α-glucosidase for Pompe disease now finalising last parts of manufacturing file and upscaling production to produce clinical trial material
- IND expected to begin 1H2019
- α-galactosidase for Fabry in mid preclinical development; expected to reach IND filing stage in 2020

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Add more products to sell

Potential for significant newsflow over the coming years

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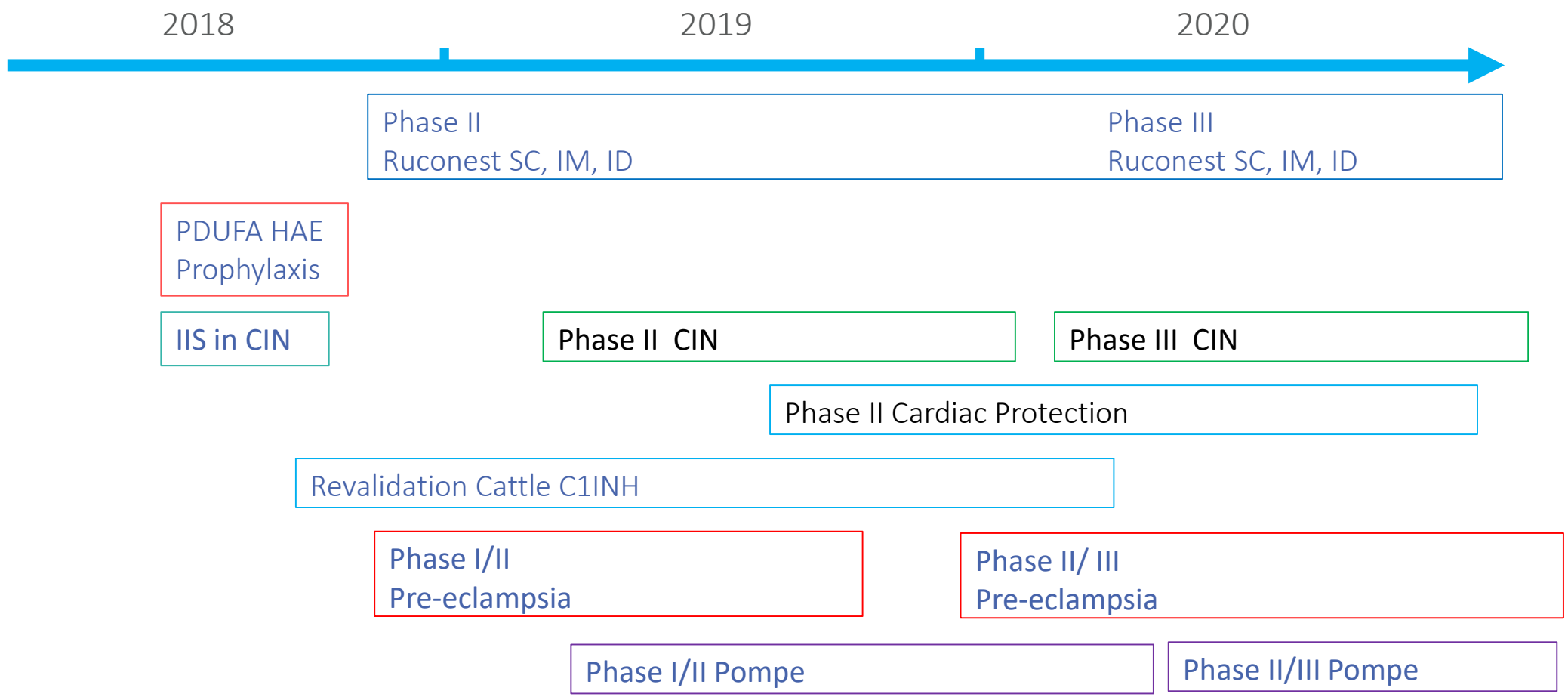
Assumed solely for purpose of diagram: positive results of studies



12 Month 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/ID/IM) with low volume vial trials
  - Clinical trials for additional indications for RUCONEST®
  - 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

Q&A with Leon Melens

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Questions?

Sign up for our press releases, or ask further questions
at info@pharming.com

www.pharming.com

Tickers:
ENXTAM: PHARM
Bloomberg: PHAR.AS

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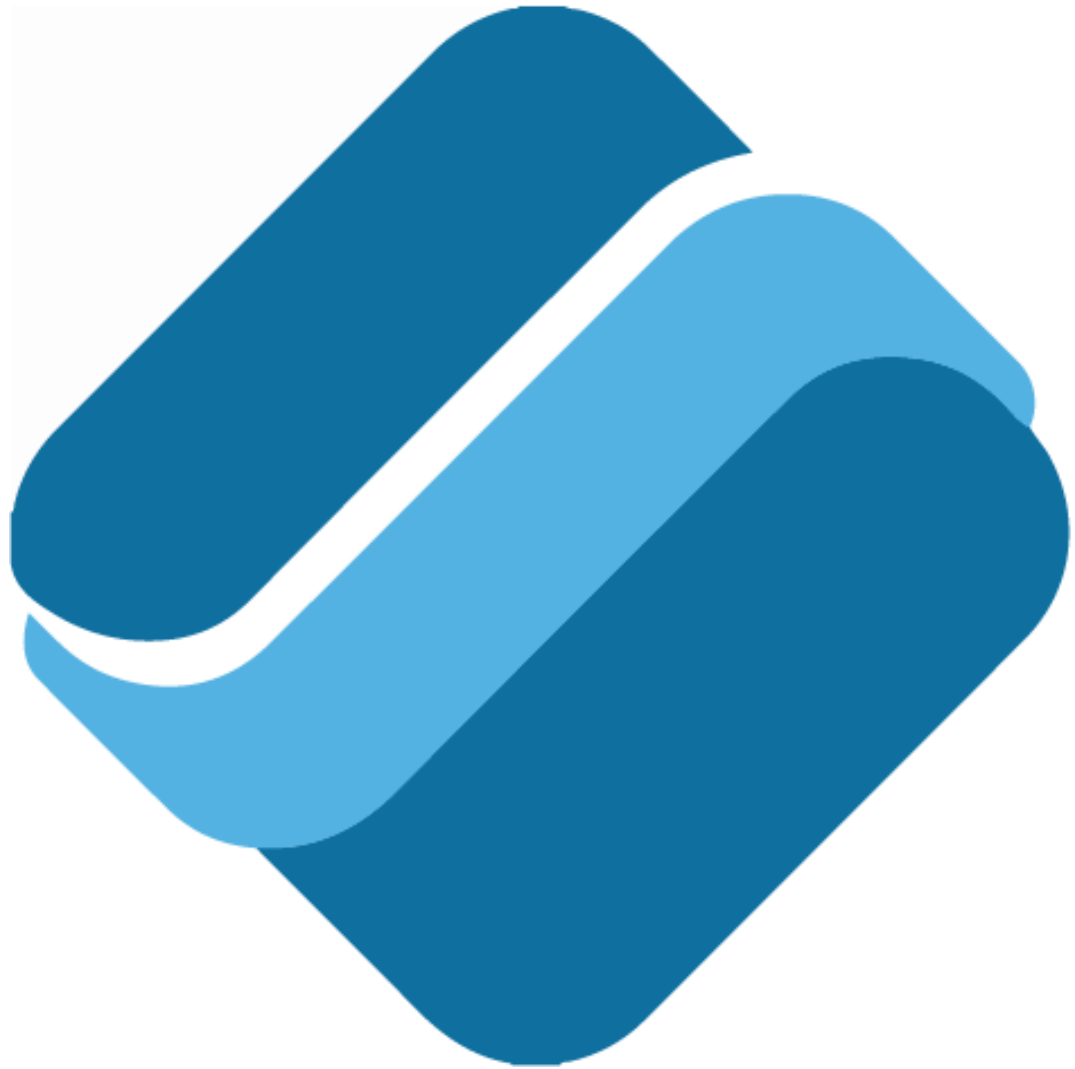
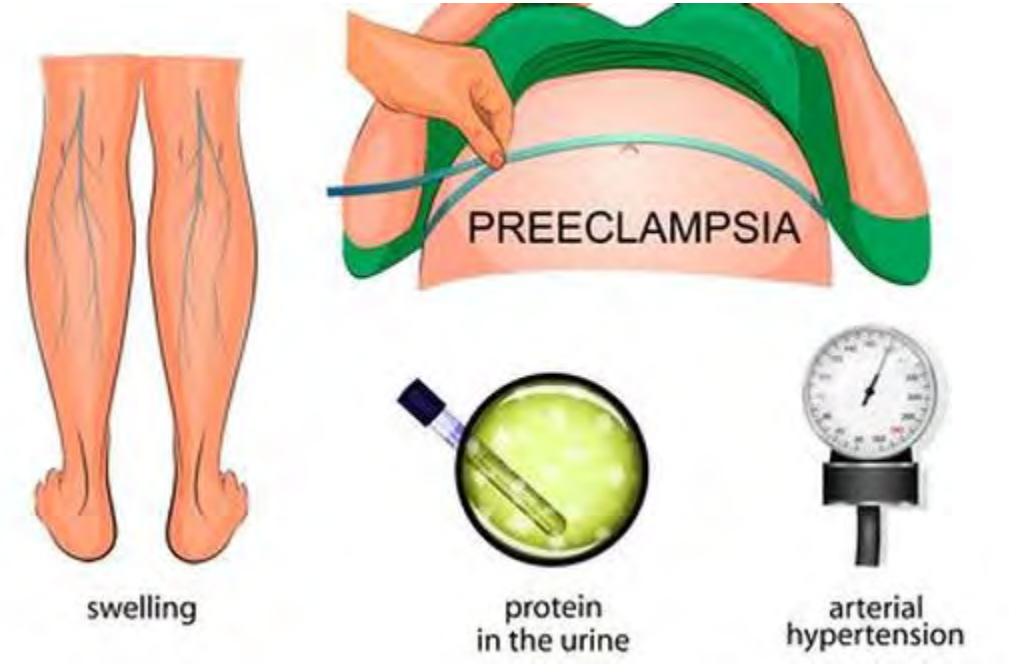


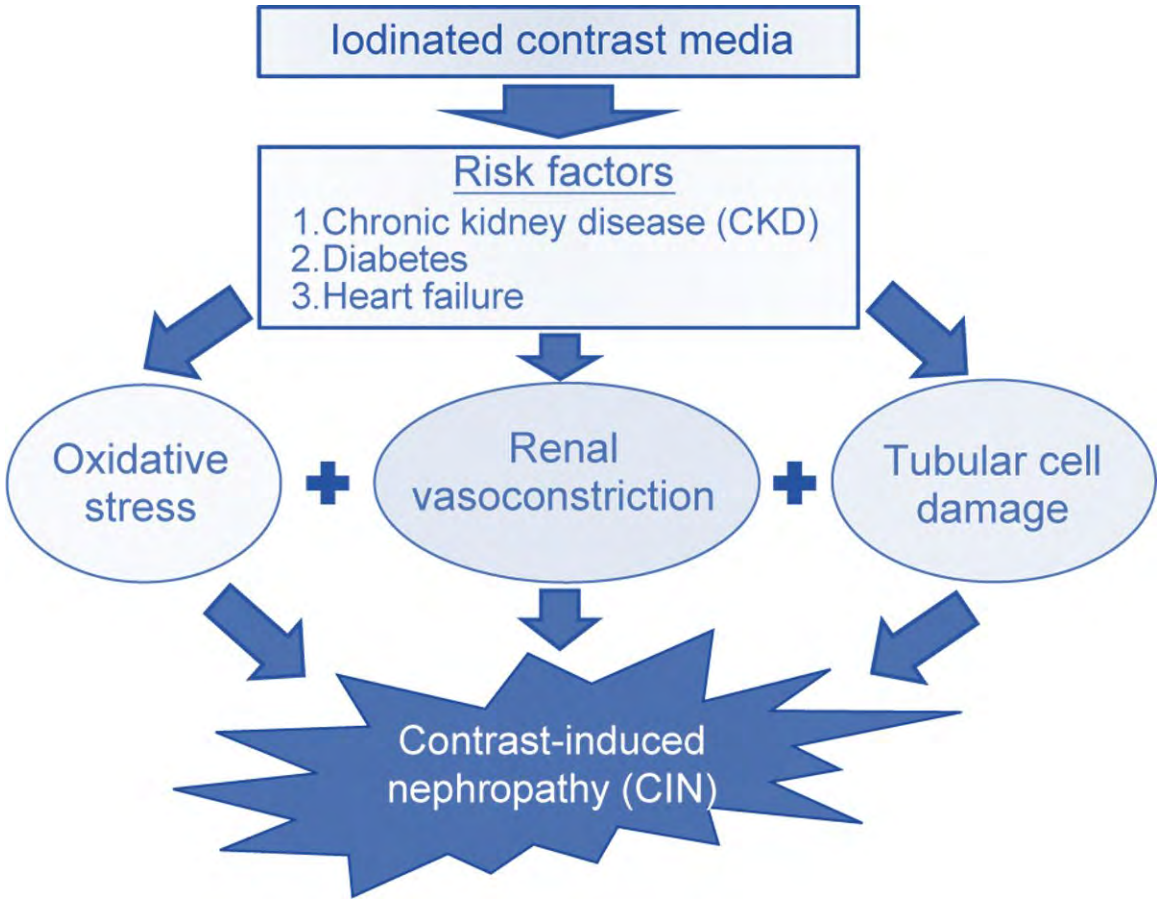
New Activities with rhC1INH: Pre-eclampsia



- Pre-eclampsia (PE) is a disorder of pregnancy characterized by the onset of high blood pressure and often a significant amount of protein in the urine.
- When it arises, the condition begins after 20 weeks of pregnancy. In severe disease there may be red blood cell breakdown, a low blood platelet count, impaired liver function, kidney dysfunction, swelling, shortness of breath due to fluid in the lungs, or visual disturbances.
- Pre-eclampsia increases the risk of poor outcomes for both the mother and the baby. If left untreated, it may result in seizures at which point it is known as eclampsia.

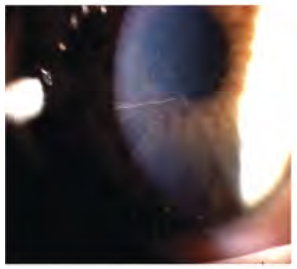
A brief overview of Preeclampsia
J Clin Med Res. 2014 Feb; 6(1): 1–7.
2013 Dec 13. doi: 10.4021/jocmr1682w

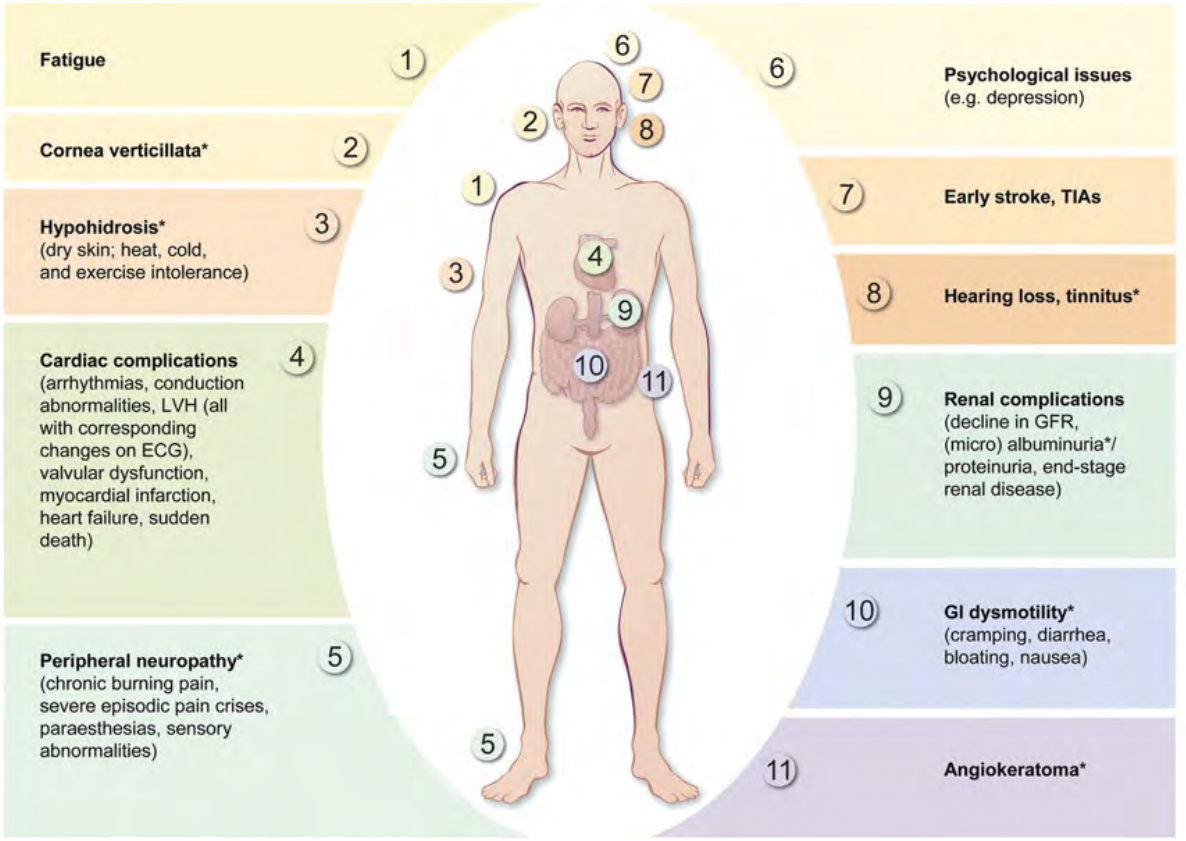
New Activities with rhC1INH: Contrast Induced Nephropathy (CIN)

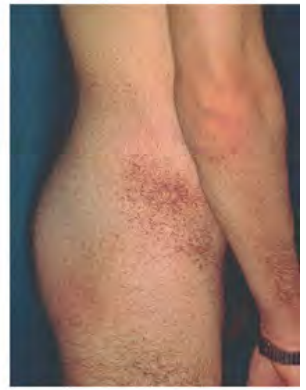


- (CIN) is a form of kidney damage in which there has been recent exposure to medical imaging contrast material without another clear cause for the acute kidney injury.

Fabry's diseases







Ref.: Zaheer Y, et al,
http://dx.doi.org/10.1093/eurheartj/ehs166 802-808
First published online: 26 June 2012

Symptoms: renal dysfunction,cardiac abnormalities,cerebrovascular manifestations,dermatological signs,ocular and auditory symptoms, neurological symptoms