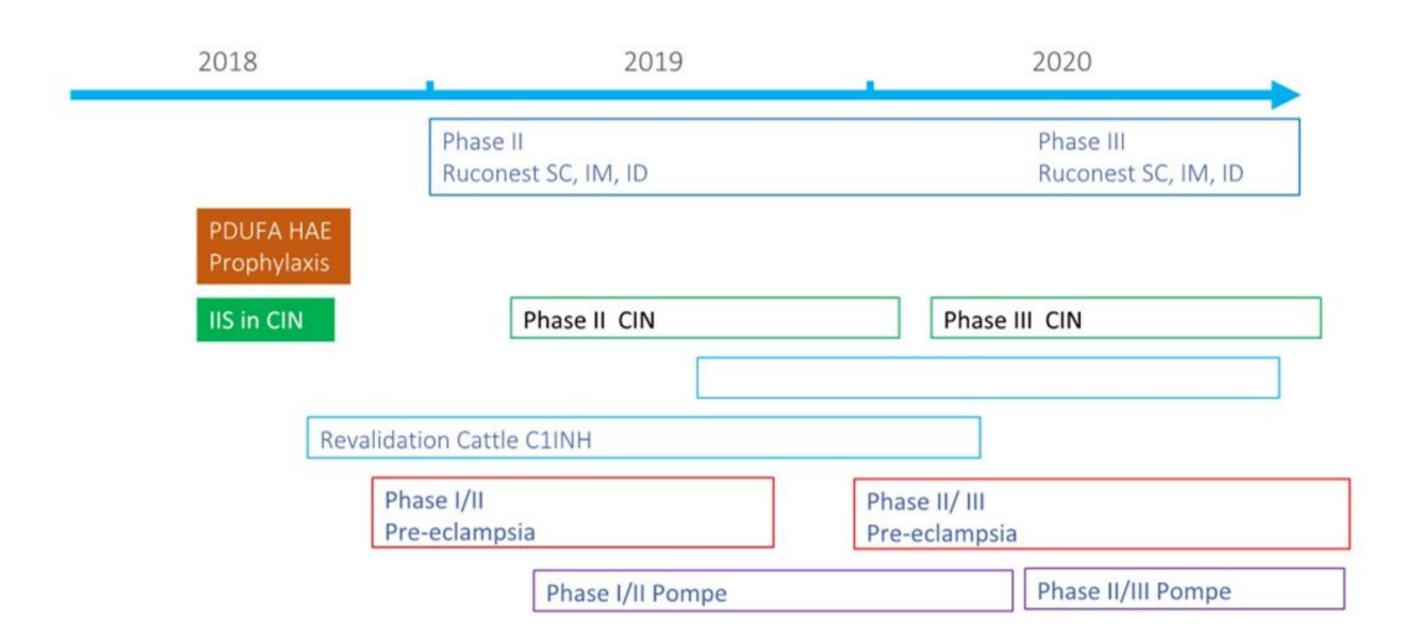
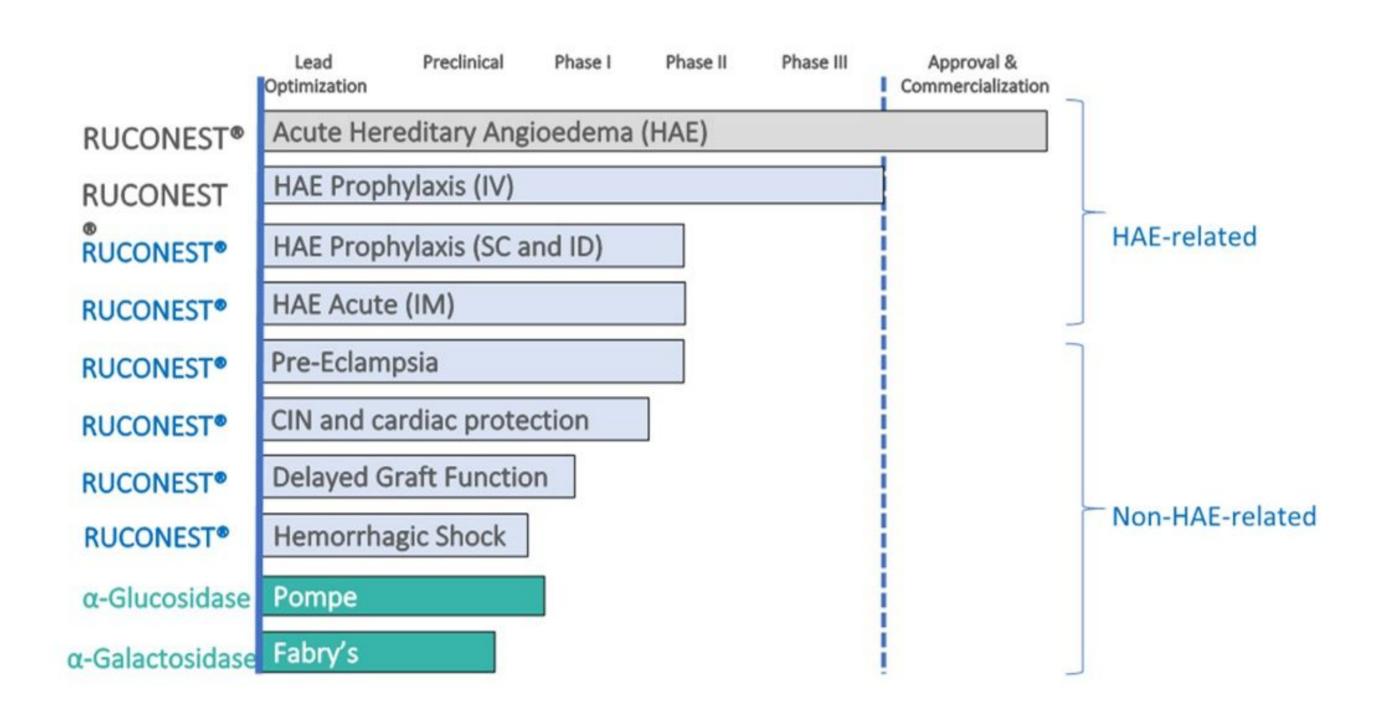
Newsflow



Assumed solely for purpose of diagram: positive results of studies



Expansion of pipeline to multiple products and markets



Update: Contrast-induced Nephropathy (CIN)



Basel Study results:

Positive results from a Phase II investigator-initiated study of RUCONEST®

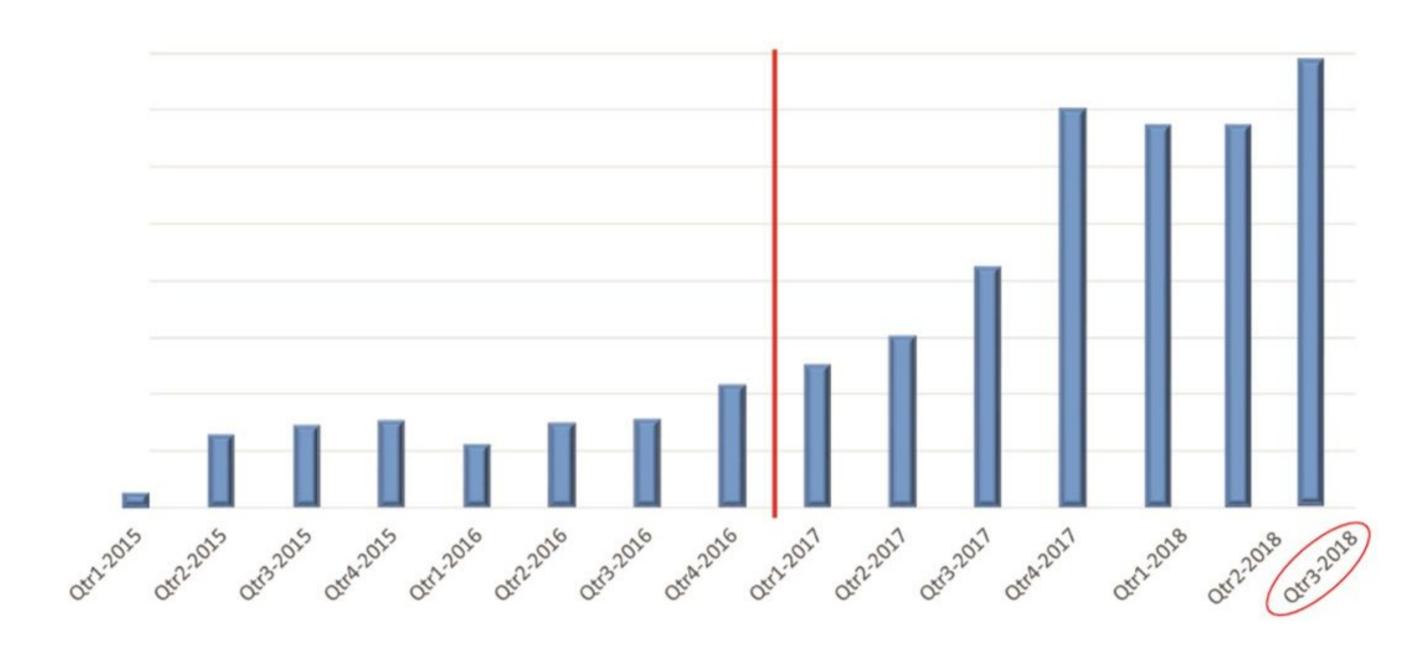
- RUCONEST® showed a statistically significant effect (p=0.038) in reducing Neutrophil Gelatinase-Associated Lipocalin (NGAL).
- The results were especially clear in the sub-group of patients (n=30) undergoing PCI. The intent-to -treat analysis in this group showed that patients on RUCONEST® had a median increase in peak urinary NGAL concentration within 48 hours of 1.8 ng/ml compared with an increase of 26.2 ng/ml in the placebo arm (p=0.04).
- This corresponds to a clear difference in the median percentage change in the peak urinary NGAL level within 48 hours of 11.3% in the RUCONEST® arm and 205.2% in the placebo arm (p=0.001).
- Overall patients undergoing invasive procedures requiring high volumes of contrast media experienced a stronger benefit from the RUCONEST® treatment.

Next steps:

Following positive results, we will continue discussion with Dr. Osthoff and other experts
with the aim to perform further clinical development to establish efficacy and efficiency of
RUCONEST® treatment in patient groups likely to experience the greatest benefit.

US quarterly sales development in volumes





First 9 months 2018: Financial Results



9 months to 30 September

Amounts in €m except per share data	2018 3 rd Quarter	2018 1# 9 months	2017 1# 9 months	% Change
Income Statement				
Revenue from product sales	38.6	97.7	56.0	74%
Other revenue	0.2	0.6	0.7	(14%)
Total revenue	38.8	98.3	56.7	73%
Gross profit	32.4	82.4	48.8	69%
Operating result	14.7	31.0	12.7	144%
Net result	5.4	11.7	(37.7)	131%
Balance Sheet				
Cash & marketable securities	72.2	72.2	38.6	87%
Share Information				
Earnings per share (€): - Undiluted	0.009	0.019	(0.077)	125%
- Fully diluted	0.008	0.017	n/a	

Compared with the first nine months of 2017 (on a like-for-like basis):

 Product revenues up 74% to €97.7 million, operating profit up 144% to €31.0 million, net profit up 131% to 11.7 million

Compared with the last quarter ended 30 June 2018:

- Product revenues up 30% to €38.6 million, operating profit up 82% to €14.7 million, net profit up 77% to €5.4 million
- Cash increased to €72.2 million (after €7.5m repayment of debt) to invest in key growth drivers

Outlook



- FY 2018 revenues from product sales to be in the range of most analysts' forecasts and for the fourth quarter results to be in the same range as the third quarter, driven by continued underlying demand balanced by increasing competition
- Achievement of a continued positive net result, continued operating profit and positive cashflows for the remaining quarter
- Continued investment in the production of RUCONEST® in order to ensure continuity of supply to the growing markets in the US, Europe and the RoW
- Approval for the pre-eclampsia study and commencement of that study
- Continued and enhanced support for patients in all territories, as we continue to believe that RUCONEST® represents a fast, effective, reliable and safe therapy option for all HAE patients no matter their situation
- Continued progress in the new pipeline programs in Pompe disease and Fabry's disease, and additional development opportunities and assets as they occur

Increasing sales and continued positive results

Three horizons of growth



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
- Contrast-induced Nephropathy
- Others such as 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