
Pharming

Pharming Group

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HC Wainwright Conference

New York City

09 September 2019

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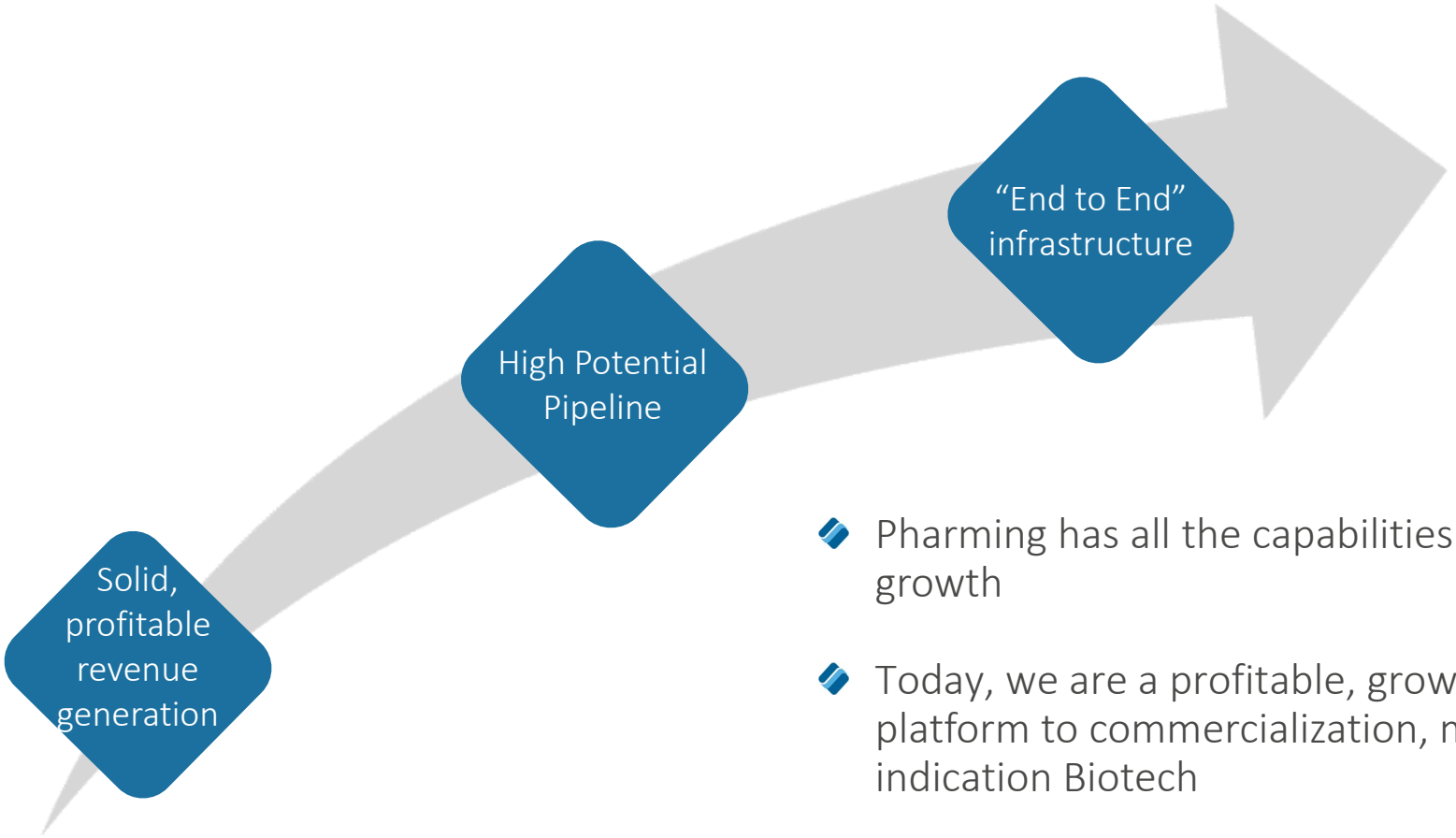
Company Overview

- **Public Company:** Euronext: PHARM: ~€750million (~$820 million)
- **Domiciled:** the Netherlands, ~220 employees globally
- **Focus:** Rare and Ultra-rare disease development and commercialization
  - Marketed product: **RUCONEST®**
  - Recombinant human C1-esterase inhibitor (enzyme replacement therapy)
  - For acute angioedema attacks in patients with hereditary angioedema (HAE)
  - Marketed in USA, EU, LatAm, Korea and Israel with other territories coming
- **Profitable and cash flow positive with 1H2019 net sales of €78M and expecting continued growth in sales**

Pharming today and into the future



- Pharming has all the capabilities needed for sustainable, high growth
- Today, we are a profitable, growing and fully integrated, from platform to commercialization, multiple product, multiple indication Biotech
- From this we can drive lasting additional growth by developing innovative solutions in select rare, ultra-rare and specialty diseases

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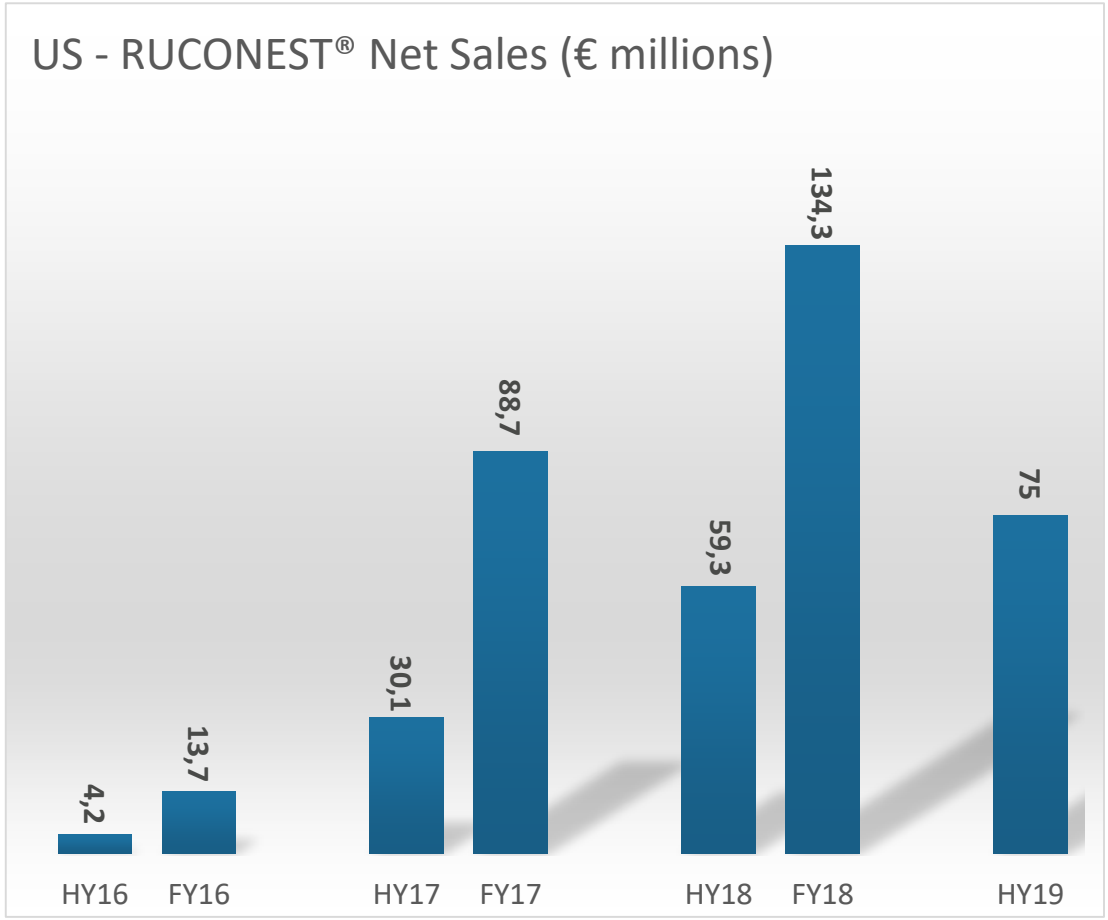
Solid, profitable revenue generation

RUCONEST® : Strong Execution of Commercial Strategy

- HAE market which is a complex, serious disease with many idiosyncrasies. The current approved therapies all address certain segments/ phenotypes of HAE.
- RUCONEST®; as the only recombinant PRT, due to it's dosing and method of administration, serves a segment the other therapies are unable to serve in an adequate way.
- Pharming, as a result of the solid RUCONEST®; business, has a strong balance sheet with growing cash position

Pharming is in a very strong position to execute and grow



RUCONEST®: Patient Segmentation

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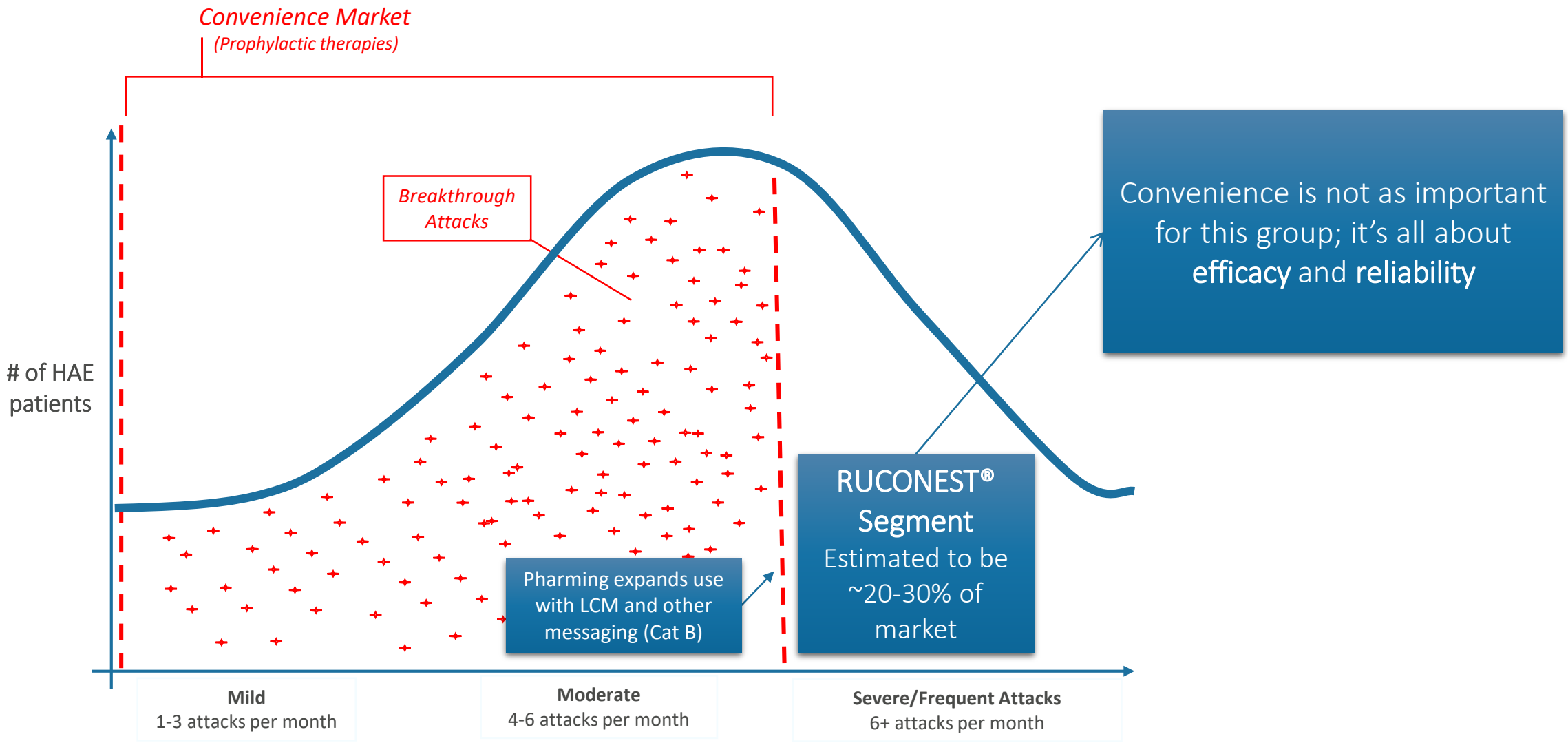
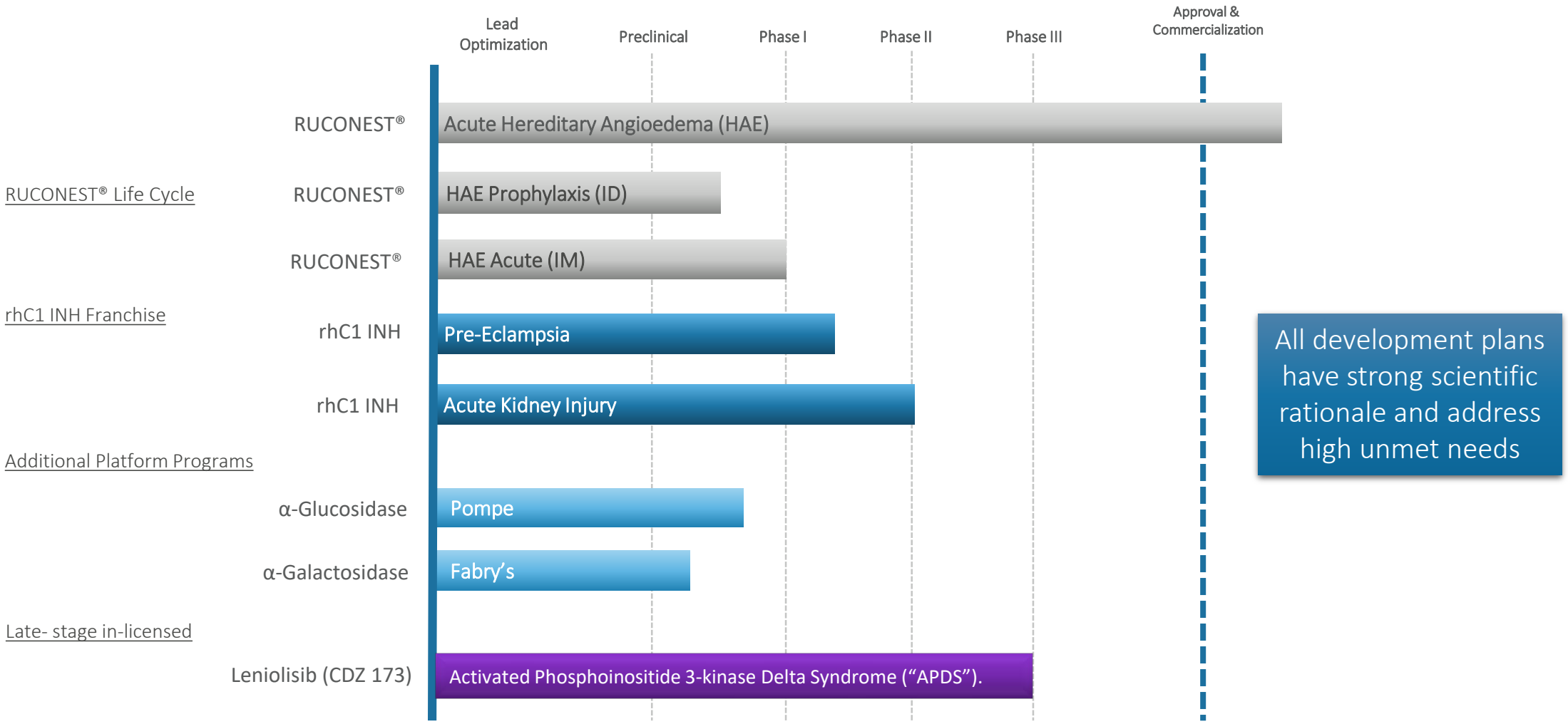


1. Banerji A, et al. Allergy Asthma Proc. 2018;39:212-223

High Potential Pipeline

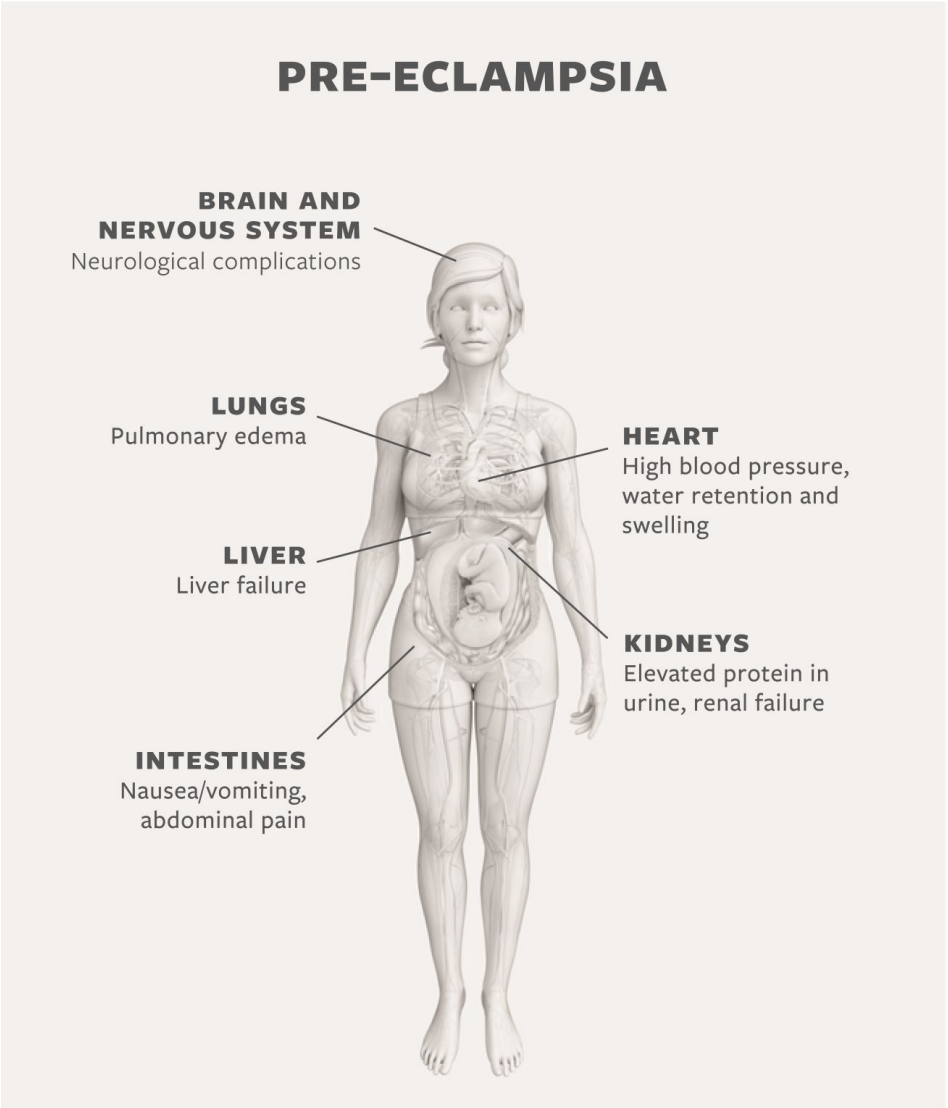
High Potential Pipeline



All development plans have strong scientific rationale and address high unmet needs

Pre-eclampsia- 1 of 3 Serious Conditions of Late Pregnancy

- 19% of all first Pregnancies Affected
  - 8% Spontaneous Preterm Birth
  - 6% Foetal growth restriction
  - 5% Pre Eclampsia



Pre-eclampsia

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Table 1 Analytical data (mean ± 1 SD) in normal pregnancy, preeclampsia and in non-pregnant women

	(A) Normal pregnancy (n = 20)	(B) Mild preeclampsia (n = 17)	(C) Moderate preeclampsia (n = 10)	(D) Non-pregnant women (n = 20)
C1-INH activity (%)	74.3 ± 15.5	64.4 ± 14.0	55.5 ± 15.8	95.1 ± 10.8
C1-INH antigen (%)	68.2 ± 10.4	62.7 ± 13.3	53.1 ± 8.8	86.5 ± 12.2

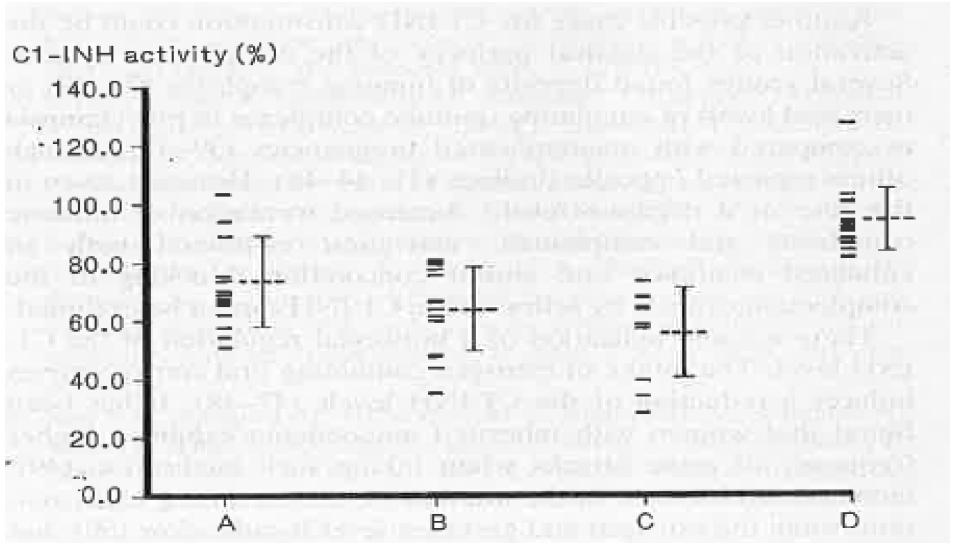

Fig. 2 Scattergram of C1-INH activities. Uncomplicated pregnancies (A), mild preeclampsia (B), moderate preeclampsia (C) and non-pregnant controls (D)

- High unmet need with no current treatment
- Significant cost to healthcare system and families
- Challenging disease to study; demands thoughtful, ethical approach

Acute Kidney Injury (AKI) resulting from Contrast Medium (CM)

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- First described in the 1950's
- Radiographic contrast medium are responsible for 11% of cases of hospital-acquired renal insufficiency, the third most common cause of renal failure after impaired renal perfusion and the use of nephrotoxic medications.
- AKI from CM is responsible for a third of all hospital-acquired acute kidney injury (AKI) and affects between 1% and 2% of the general population and up to 50% of high-risk subgroups following coronary angiography (CA) or percutaneous coronary intervention (PCI).[1]

Table 1 | Risk factors for the development of CIN

Fixed (non-modifiable) risk factors	Modifiable risk factors
Older age	Volume of CM
Diabetes mellitus	Hypotension
Pre-existing renal failure	Anemia and blood loss
Advanced CHF	Dehydration
Low LVEF	Low serum albumin level (<35 g/l)
Acute myocardial infarction	ACE inhibitors
Cardiogenic shock	Diuretics
Renal transplant	Non-steroidal anti-inflammatory drugs
	Nephrotoxic antibiotics
	IABP

Abbreviations: ACE, angiotensin-converting enzyme; CHF, congestive heart failure; CIN, contrast-induced nephropathy; CM, contrast media; IABP, intra-aortic balloon pump; LVEF, left ventricular ejection fraction.

[1] *Mehran R, Nikolsky E. Contrast-induced nephropathy: definition, epidemiology, and patients at risk. Kidney Int Suppl 2006:S11–15*
CIN= Contrast induced nephropathy

Acute Kidney Injury (AKI) Resulting from Contrast Medium (CM)

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- Screening
- Informed consent
- Randomization
- Baseline samples
- Hydration

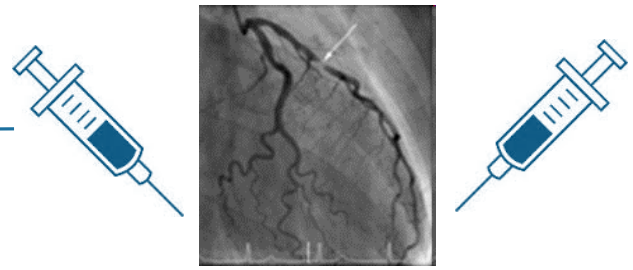
- Blood/urine sampling
- Discharge

Telephone interview

Day -1/0 | Day 0 | Day +1 | Day +2 | Week +4 and +12

Group 1: rhC1INH
<84kg: 50 U/kg
>84kg: 4200 U

Group 2: placebo
NaCl 0.9%

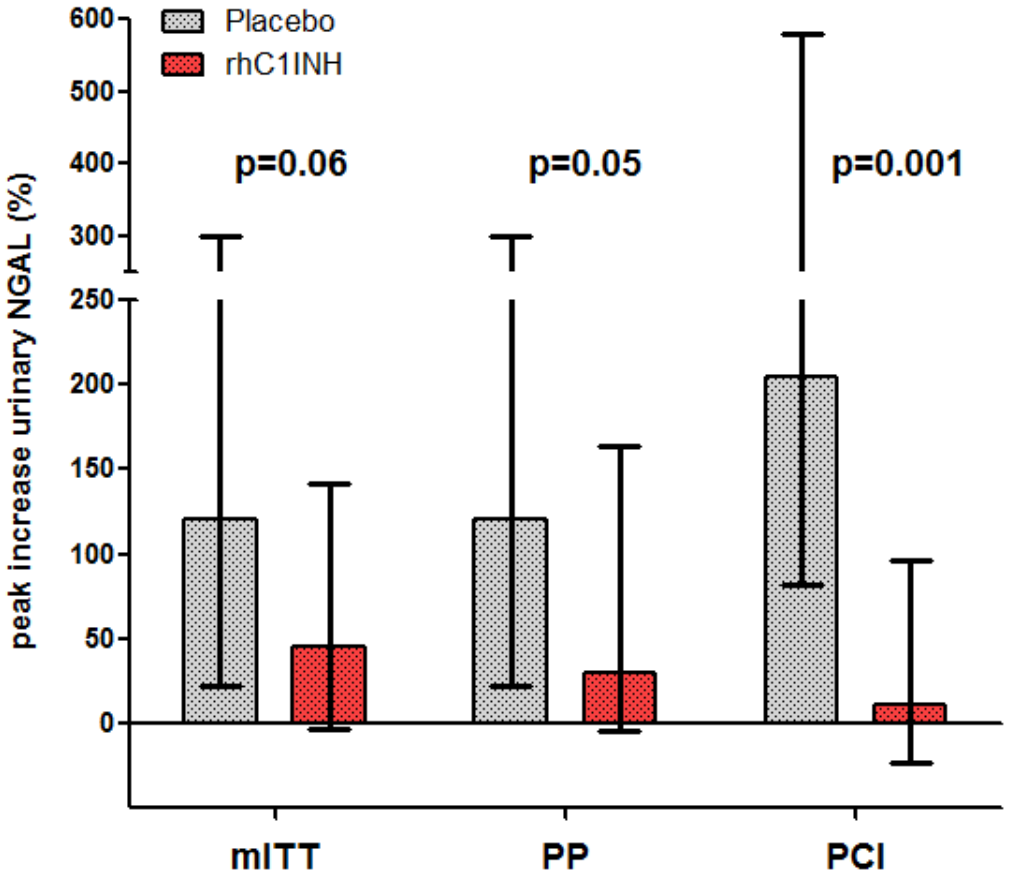


Outpatient visit
- Blood/urine sampling

Study completed M. Osthoff MD et al.,University Hospital Basel, CH

Proof of Concept

Relative urine NGAL peak increase 48 h, (%)



Leveraging our “End to End” Infrastructure

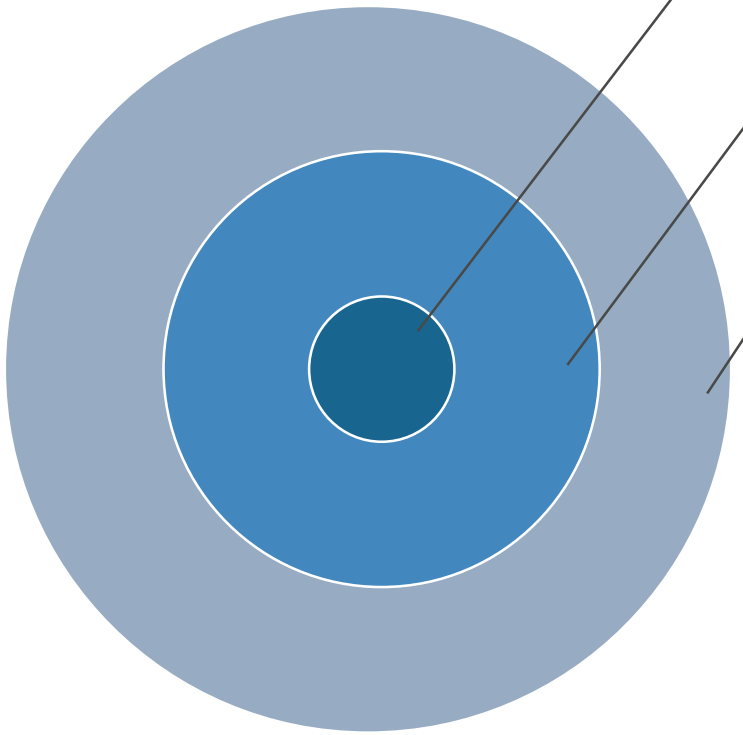
Leniolisib for APDS licensed from Novartis

- Leniolisib is a selective PI3Kinaseδ inhibitor
- Leniolisib is in a registration-enabling study for the treatment of APDS
- APDS is a recently discovered primary immune deficiency caused by a mutation in the PIK3CD gene that increases activity of PI3Kδ, a promoter of activity in the immune system, an ultra-rare, debilitating disease with no approved treatment
- Beginning in childhood, recurrent infections, particularly in the lungs, sinuses, and ears. Over time, recurrent respiratory tract infections can lead to a condition called bronchiectasis, cause breathing problems.

Leniolisib for APDS licensed from Novartis

- People with activated PI3K-delta syndrome may also have chronic active viral infections, commonly Epstein-Barr virus or cytomegalovirus infections. Sufferers also frequently develop lymphomas and other cancers.
- APDS is treated by immunologists; the main physicians treating HAE and therefore already addressed by Pharming commercialization teams
- Upfront payment of $20 million (€17.9 million)
- If approved, the drug is expected to reach the market in 2H 2021 or 1H 2022

Investment Strategy: Focus, Leverage our Strengths



Active Business Development aimed at capital efficient and smart investments that leverage commercial infrastructure ahead of maturation of new indications/ new internal pipeline projects

Financial Performance & 2019 Outlook

H1 2019: Financial Results

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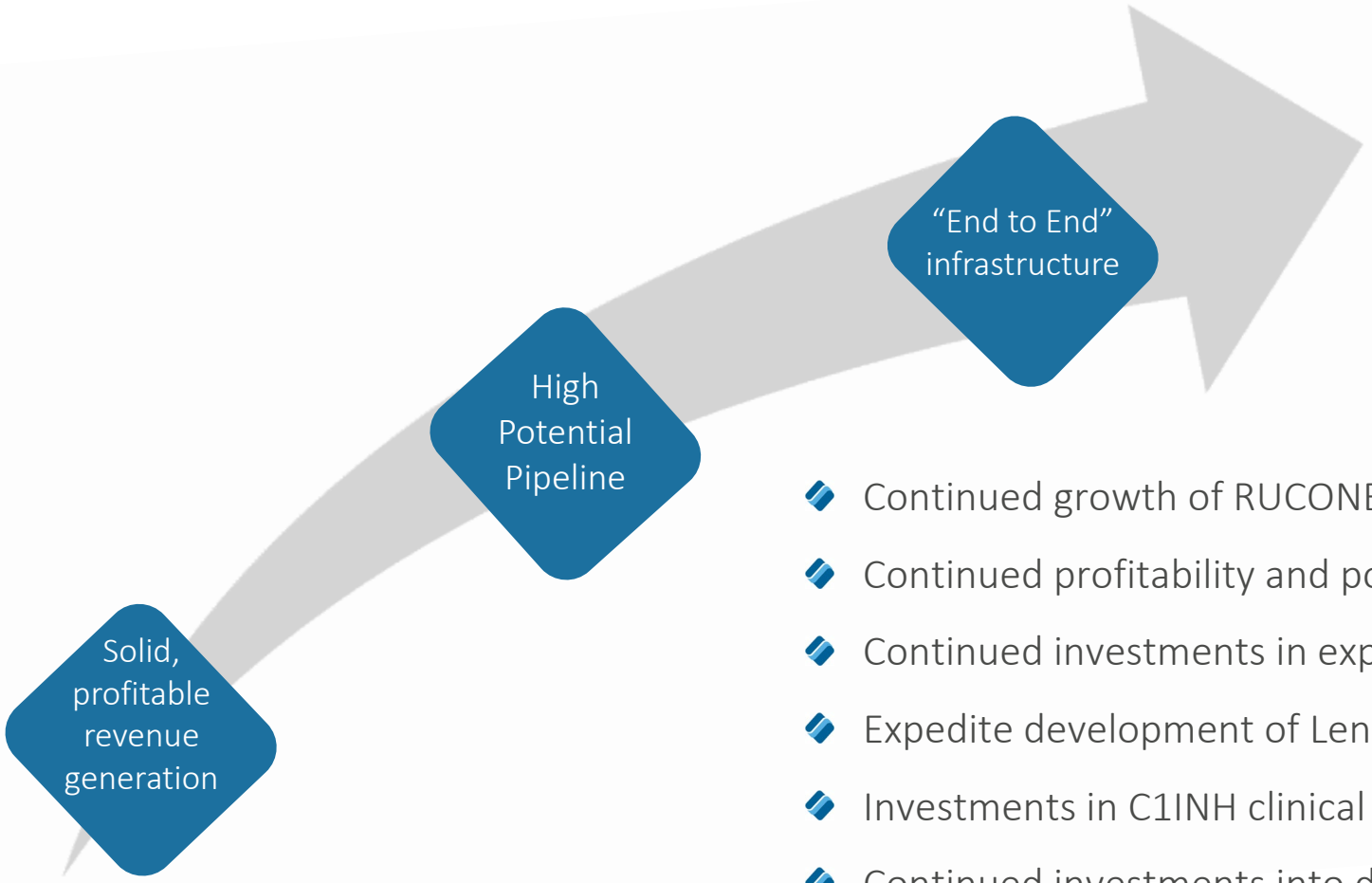
Amounts in €m except per share data	2019 1st Half	2018 1st Half *restated	% Change
Income Statement			
Revenues	77.9	59.5	31%
Gross profit	67.0	50.0	34%
Operating result	24.6	16.3	51%
Net result	13.6	8.5*	60%
Balance Sheet			
Cash & marketable securities	65.3	66.9	(2%)
Share Information			
Earnings per share (€): Undiluted	0.022	0.014*	57%
Fully diluted	0.020	0.013*	54%

** After restatement on the basis set out above and in Note 4 to the Financial Statements in the Annual Report 2018.*

Summary and Outlook 2019 and beyond



- Continued growth of RUCONEST® sales
- Continued profitability and positive cash flows
- Continued investments in expansion of manufacturing
- Expedite development of Leniolisib to FDA and EMA approval
- Investments in C1INH clinical trials for PE and AKI
- Continued investments into development PRT for Pompe and Fabry
- Re- evaluation of most advantageous route of administration for RUCONEST®

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