

# Pharming Group NV

Unaudited Full Year Results for 2018

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# Forward Looking Statements

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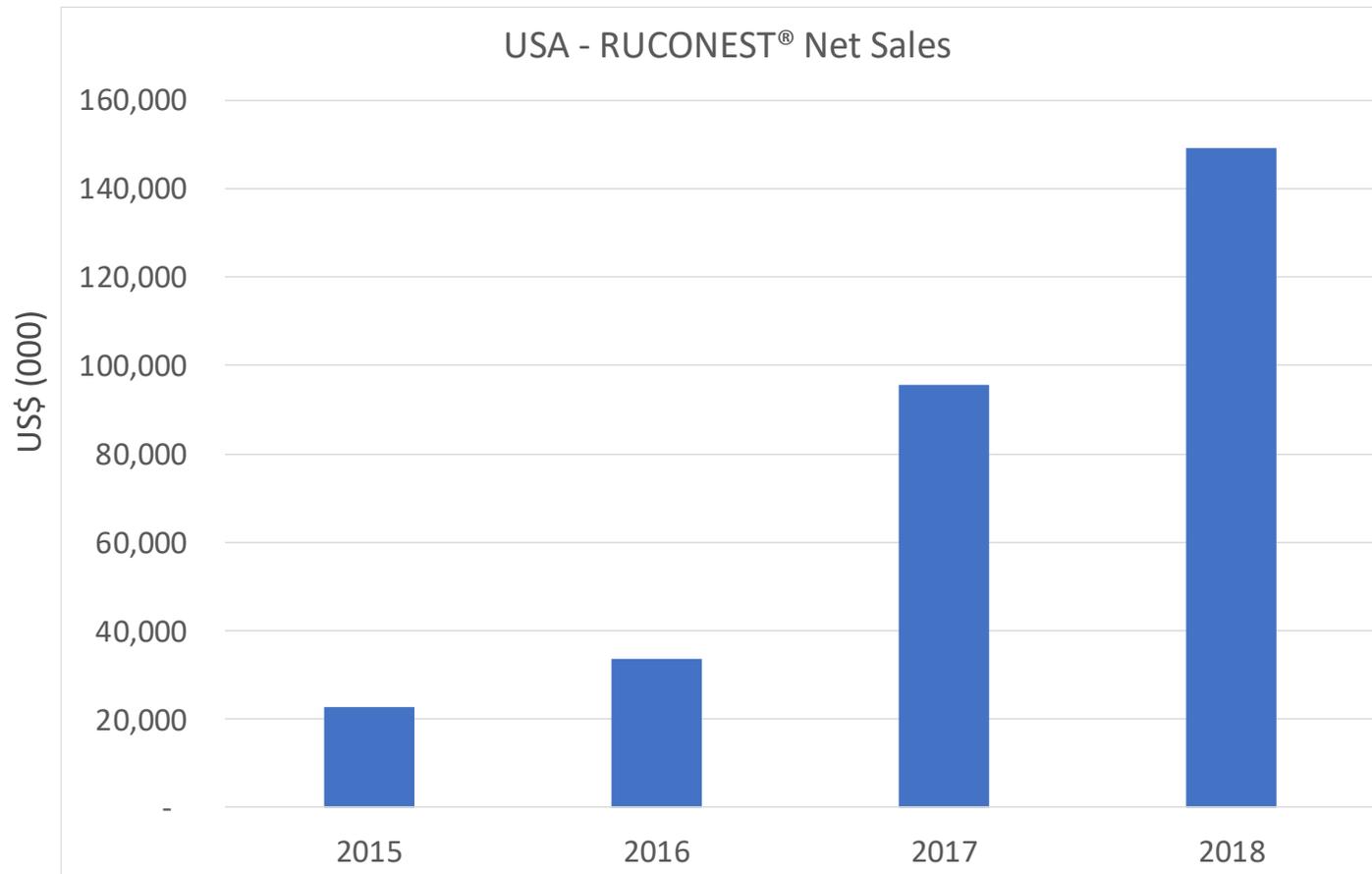
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1. 2018 Operational Highlights
2. Three Pillars of Internal Growth
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# Strong execution of commercial strategy

- Revenues from product sales for the year increased by 51.2% to €134.1 million (FY 2017: €88.7 million), as a result of increasing patient numbers
- The effect of Pharming's approach to US commercialisation can be seen in the chart below:



- Recent launches of subcutaneous injected plasma-derived C1 inhibitor product and subcutaneous long-acting antibody increase future competitive pressure in the prophylaxis segment
- New treatments feature higher responder rates than previous IV plasma-derived C1-inhibitor prophylaxis
- Continued need for effective and reliable treatment for breakthrough attacks, because over half of patients continue to have breakthrough attacks
- The management of HAE typically improves by having multiple types of treatments at hand over time
- Reliable and consistent response when treating attacks of HAE, and the corresponding increasing positive patient experiences, means that RUCONEST continues to find its place as preferred/ultimate treatment
- Opportunity now both for severely-affected patients and for treatment of breakthrough attacks associated with the new prophylaxis products, providing scope for continued growth

- Publications at various scientific congresses throughout the year underpinning reliability and consistency of response to RUCONEST therapy further
- These include the first investigator-initiated (observational) real-world study comparing re-dosing frequency of C1 esterase inhibitor therapies versus icatibant (bradykinin inhibition) therapy in 69 acute HAE attacks
- The data showed that properly dosed, RUCONEST® and other C1 esterase therapies would typically stop HAE attacks on the first treatment and that icatibant (marketed as the world's best-selling HAE drug, Firazyr®) required frequent (and multiple) re-dosing to treat an attack of HAE
- The full results are being written up now by the investigators now for publication shortly

# Three pillars for strong organic growth

- Focusing on investment in our three pillars of organic growth outlined at the Capital Markets Day in June 2018:
  - Convenience of RUCONEST® within the HAE space to meet patients' needs – new intramuscular, subcutaneous and intradermal versions under development
  - Development of RUCONEST®/rhC1INH outside the HAE space to tackle large unmet medical needs for which there are no current approved or effective therapies: initially pre-eclampsia and acute kidney injury
  - New protein replacement products which address significant shortcomings of existing therapies for Pompe and Fabry diseases
- Received a complete response letter for the use of RUCONEST® for the prophylaxis of HAE in September, as result of the FDA being unable to cross the final remaining statistical hurdle in a small sub-group of patients.
  - This issue will be addressed as part of new prophylaxis studies with more convenient forms of RUCONEST as outlined above

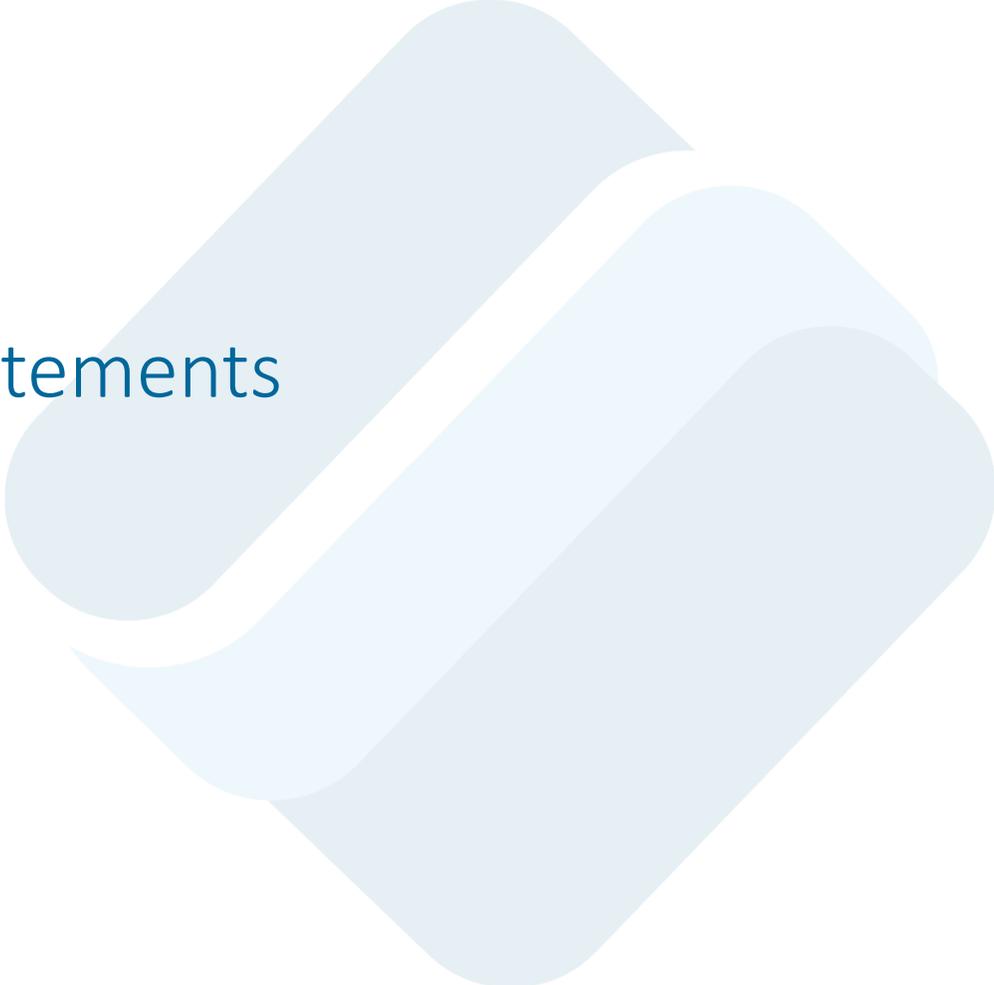
- Positive results delivered in October from a Phase II investigator-initiated study of RUCONEST® in a double-blind, placebo-controlled clinical trial in patients at risk of nephropathy resulting from contrast-enhanced examinations
- The result show clear clinically-significant benefit in patients undergoing percutaneous coronary interventions (“PCI”) such as stent insertions or valve replacements
- The intent-to-treat analysis showed that patients on RUCONEST® had a median percentage change in peak urinary Neutrophil Gelatinase-Associated Lipocalin within 48 hours of 11.3% in the RUCONEST® arm and 205.2% in the placebo arm (p=0.001)
- The overall assessment of the study also showed trends that patients undergoing more invasive interventions and procedures requiring higher volumes of contrast medium experienced a stronger benefit from the RUCONEST® treatment
- As a result, Pharming is now embarking on its own program for acute kidney injury in patients undergoing PCIs accompanied by contrast-enhanced examinations

- An initial study to investigate safety (and efficacy signals) in pregnant women diagnosed with pre-eclampsia has been designed and was submitted to the regulatory authorities and ethics committees in Netherlands and Australia last year
- All study preparations are completed
- Any study in pregnant women, and especially studies in distressed pregnant women, are necessarily very carefully designed, planned and reviewed
- Now awaiting confirmation for final approvals to initiate the study from the relevant ethics committees
- Following ethics committee approval, the study is expected to start soon
- Ethics committee approval and first patient in will be reported by press release

## Pompe development program

- Continuing to develop  $\alpha$ -glucosidase from our platform
- Initiated dialogues with key opinion leaders on future clinical program designs
- Input/ feedback will be sought from regulatory authorities
- Currently manufacturing material for IND-enabling studies and clinical trial material

Fabry program also running - continues to trail Pompe program by approximately one year



# Financial Statements

- Total revenues increased by 51% to €135.1 million (including €0.8 million of license revenue) from €89.6 million (including €0.9 million in license revenue) in 2017
- Operating results improved to a profit of €40.6 million from €21.9 million in 2017, before a one-off non-cash adjustment of €2.6 million for previously-capitalised development costs in respect of a superseded new version of RUCONEST®
  - This improvement was made despite a considerable increase in commercialization and clinical research activities,
- The net result was a profit of €25.0 million (2017: loss of €76.2 million), mainly as a result of the strong operating performance and improved capital structure
- The Cash position increased from €60.0 million at the end of 2017 to €81.5 million at the end of 2018, despite paying down over €14.5 million (\$16.7 million) of the Orbimed loan in the second half of the year
- Shareholders' equity increased from €16.1 million to €61.8 million due mainly to the net result and revenues from option and warrant exercises

- Recently started to repay the \$100m 4 year refinance debt facility with OrbiMed
  - €14.5 million (\$16.7 million) paid down from the principal amount in Q3 and Q4 2018
- All convertible bonds now redeemed or converted and almost all warrants now exercised, so that there will be no further significant fair value adjustments after 2018
- This means that Pharming now has a clean capital structure, with just the employee share option plans making the (6.6%) difference between the issued and fully-diluted capital
- The capital structure does allow for us to consider small acquisitions where these are clearly in shareholders' interests, but larger deals will be brought to shareholders for approval

# Income Statement – Operating Profit

Amounts in € '000	2018	2017 restated *
Product sales	134,326	88,677
License fees	804	943
<b>Revenues</b>	<b>135,130</b>	<b>89,620</b>
Costs of sales	(22,180)	(12,445)
<b>Gross profit</b>	<b>112,950</b>	<b>77,175</b>
Other income	684	790
Research and development	(28,882)	(18,657)
General and administrative	(12,221)	(5,974)
Marketing and sales	(34,539)	(31,422)
<b>Costs</b>	<b>(75,642)</b>	<b>(56,053)</b>
<b>Operating result</b>	<b>37,992</b>	<b>21,912</b>

# Income Statement - Net result

Amounts in € '000	2018	2017 restated *
Fair value gain (loss) on revaluation derivatives *	(495)	(42,063)
Other financial income and expenses *	(36,640)	(65,538)
<b>Financial income and expenses</b>	<b>(37,135)</b>	<b>(107,601)</b>
<b>Result before income tax</b>	<b>857</b>	<b>(85,689)</b>
Income tax credit (expense)	24,136	9,442
<b>Net result for the year</b>	<b>24,993</b>	<b>(76,247)</b>
<b>Attributable to:</b>		
Owners of the parent	24,993	(76,247)
<b>Total net result</b>	<b>24,993</b>	<b>(76,247)</b>
Basic earnings per share (€)	0.041	(0.152)
Fully-diluted earnings per share (€)	0.038	n/a

# Balance Sheet - Assets

Amounts in € '000	2018	2017 restated*
<b>Non-current assets</b>		
Intangible assets	52,435	56,631
Property, plant and equipment	8,402	8,234
Long-term prepayments	2,006	2,296
Deferred tax assets	35,082	9,442
Restricted cash	1,204	1,336
<b>Total non-current assets</b>	<b>99,129</b>	<b>77,939</b>
<b>Current assets</b>		
Inventories	17,315	18,334
Trade and other receivables	17,814	11,260
Cash and cash equivalents	80,311	58,657
<b>Total current assets</b>	<b>115,440</b>	<b>88,251</b>
<b>Total assets</b>	<b>214,569</b>	<b>166,190</b>

# Balance Sheet - Liabilities

Amounts in € '000	2018	2017 restated*
<b>Equity</b>		
Share capital	6,215	5,790
Share premium *	387,525	363,818
Legal reserves	(590)	(938)
Accumulated deficit	(331,399)	(352,560)
<b>Shareholders' equity</b>	<b>61,751</b>	<b>16,110</b>
<b>Non-current liabilities</b>		
Loans and borrowings *	37,267	59,161
Deferred tax liabilities	87	-
Contract liabilities	667	1,467
Finance lease liabilities	164	390
Other financial liabilities	32,034	28,319
<b>Total non-current liabilities</b>	<b>70,219</b>	<b>89,337</b>
<b>Current liabilities</b>		
Loans and borrowings *	35,235	22,398
Contract liabilities	800	804
Derivative financial liabilities *	228	10,080
Trade and other payables	28,589	27,198
Finance lease liabilities	263	263
Other financial liabilities	17,484	-
<b>Total current liabilities</b>	<b>82,599</b>	<b>60,743</b>
<b>Total equity and liabilities</b>	<b>214,569</b>	<b>166,190</b>

# Cash flow – Operating Activities

Amounts in €'000		2018	2017
<b>Operating result</b>		<b>37,992</b>	<b>21,912</b>
<b><i>Non-cash adjustments:</i></b>			
Depreciation, amortisation, impairment		6,559	3,415
Accrued employee benefits		3,270	2,712
Deferred license fees		(804)	(943)
<b>Operating cash flows before changes in working capital</b>		<b>47,017</b>	<b>27,096</b>
<b><i>Changes in working capital:</i></b>			
Inventories		1,019	(393)
Trade and other receivables		(6,554)	(3,345)
Payables and other current liabilities		1,391	14,837
<b>Total changes in working capital</b>		<b>(4,144)</b>	<b>11,099</b>
Changes in non-current assets, liabilities and equity		(1,098)	15
<b>Cash generated from (used in) operations before interest and taxes</b>		<b>41,775</b>	<b>38,210</b>
Interest received		18	3
Income taxes paid		(1,417)	-

# Cash flow - Overall

Amounts in €'000		2018	2017
<b>Net cash flows generated from (used in) operating activities</b>		<b>40,376</b>	<b>38,213</b>
Capital expenditure for property, plant and equipment		(2,496)	(3,248)
Investment intangible assets		(1,273)	(2,797)
<b>Net cash flows used in investing activities</b>		<b>(3,769)</b>	<b>(6,045)</b>
Proceeds of loans and borrowings		-	91,333
Payments of transaction fees and expenses		-	(3,352)
Prepayment on loans and borrowings		(15,137)	(86,258)
Redemption bonds		(2,257)	(3,934)
Interests on loans		(11,063)	(7,877)
Proceeds of equity and warrants		10,496	6,833
<b>Net cash flows generated from (used in) financing activities</b>		<b>(17,961)</b>	<b>(3,255)</b>
<b>Increase (decrease) of cash</b>		<b>18,646</b>	<b>28,913</b>
Exchange rate effects		2,876	(1,057)
Cash and cash equivalents at 1 January		59,993	32,137
<b>Total cash and cash equivalents at 31 December</b>		<b>81,515</b>	<b>59,993</b>

# Outlook 2019



For the remainder of 2019, the Company expects:

- Continued growth in revenues from sales of RUCONEST<sup>®</sup>, mainly driven by the US and Western Europe operations
- Continued achievement of positive net earnings during the year
- Continued investment in the expansion of production of RUCONEST<sup>®</sup> in order to meet the growing demand for RUCONEST<sup>®</sup> internationally
- Investment in further clinical trial programs for RUCONEST<sup>®</sup> with low-volume concentrated liquid intramuscular and subcutaneous versions of RUCONEST<sup>®</sup> for both acute treatment and prophylaxis of HAE, as well as research into other more convenient routes of administration.
- Investment in clinical trials to explore additional indications for RUCONEST<sup>®</sup>
- Investment in development of the new pipeline programs in Pompe disease and Fabry's disease, and other new development opportunities and assets as these occur
- Increasing marketing activity where this can be profitable for Pharming, such as opening new countries for RUCONEST<sup>®</sup>

**No further financial guidance for 2019 is provided.**

## Tickers:

- ENXTAM: PHARM
- Bloomberg: PHAR.AS