Pharming Group reports interim financial results for the first nine months of 2018

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

***Leiden, The Netherlands*, 25 October 2018:** Pharming Group N.V. ("Pharming" or "the Company") (Euronext Amsterdam: PHARM) presents its (unaudited) interim financial report for the first nine months and the third quarter ended 30 September 2018.

Financial highlights

- Net product sales for the first nine months of 2018 increased to €97.7 million (Q3: €38.6 million), an increase of approximately 74% on a like-for-like basis compared to €56.0 million for the first nine months of 2017), as a result of the increasing numbers of patients using RUCONEST® in the USA and in Europe.
- US net product sales for the first nine months of 2018 increased to €92.9 million (Q3: €36.5 million), an increase of 77% compared to €52.5 million for the first nine months of 2017. In the rest of the world, product sales for the first nine months of 2018 increased to €4.8 million (Q3: €2.1 million), an increase of approximately 37% compared to €3.5 million the first nine months of 2017.
- Total revenues for the first nine months of 2018 increased by 73% to €98.3 million (including €0.6 million of license revenue) from €56.7 million in the first nine months of 2017 (including €0.7 million in license revenue).
- Operating profits rose by 144% to €31.0 million in the first nine months of 2018, compared to €12.7 million in the first nine months of 2017. Operating profits also rose by 82% quarter on quarter, to €14.7 million in the third quarter from €8.1 million in the second quarter of 2018. These improvements were made despite a considerable increase in operating costs, mainly relating to improvements in production capacity and development costs for the new indications and formulations of RUCONEST®.
- Net profit for the first nine months was €11.7 million (Q3: €5.4 million), compared with a loss as originally stated of €37.7 million in the same period last year. The 77% improvement quarter on quarter resulted mainly from the improvement in sales in the USA.
- Positive cashflows during the third quarter of 2018 were driven by increasing revenues above the cash required for costs and repayment of the first quarterly instalment of €7.5 million of the principal amount of the Company's outstanding loan including associated fees. This resulted in an increase in the cash position to €72.2 million from €66.9 million at June 30 2018 (€38.6 million at 30 September 2017).
- The equity position improved from €40.7 million at the end of June 2018 to €48.2 million at the end of the third quarter of 2018 (End of third quarter 2017: €6.2 million), mainly due to the net result and the exercise of employee options during the quarter. Other financial liabilities, which

refers to the contingent consideration for the milestones, has been divided into current and non-current elements, reflecting the probability of paying the first milestone in 2019.

- Inventories changed from €23.2 million at the end of the second quarter of 2018 to €21.0 million at the end of the third quarter of 2018 (End of third quarter 2017: €18.0 million), mainly due to slightly larger than anticipated sales demands in the US and Europe.
- Net profits were affected by an adjustment to the policy on effective interest on the $100 million loan to reflect normal fees due on the contracted quarterly repayments that are being accounted for now in the effective interest rate instead of being taken directly to the income statement. This amendment will amount to €4.3 million spread out over the four year lifetime of the loan, and if this policy had been implemented from the start of the loan, there would have been additional non-cash finance costs of €0.9 million in 2017. For the full year, the corrected loss would therefore have been €80.8 million, and this will be shown in the comparables column at the full year results. Additional changes reflected in net profit include an increase of €5.8 million for contingent consideration increases, as the likelihood of milestone payments increases, and this provision listed under other financial liabilities has been split into current and non-current segments. The net result also includes an increase in the deferred taxation asset of €5.7 million reflecting the faster-than-expected growth in profits likely to be taxable.
- Since the last reporting date of 30 June 2018, the company has issued a total of 6,947,881 shares and recorded 282,001 options in connection with a number of exercises of options under the current schemes. The number of issued shares as at 25 October 2018 is 617,358,943. The fully diluted number of shares as at 25 October 2018 is 657,578,717.

Operational highlights during the third quarter

- In September 2018, the Company received a Complete Response Letter (CRL) from the U.S. Food and Drug Administration (FDA) regarding the supplemental Biologics License Application (sBLA) for RUCONEST® to expand the current indication to include prophylaxis in patients with hereditary angioedema (HAE). Based on their review, the FDA has requested an additional clinical trial to evaluate the effectiveness of RUCONEST® further in HAE prophylaxis. The Company is following up with the FDA on the exact path to approval and what study would be suitable. The development of intra-muscular, sub-cutaneous and pain-free intra-dermal delivery of RUCONEST® is not affected by this and continues as planned.
- Also in September, the Company filed a clinical trial application (CTA) with the European Medicines Agency in the Netherlands to initiate the clinical development of RUCONEST to treat and prevent pre-eclampsia. A similar filing has also been made in Australia.

Operational highlights since the reporting date

- Last week, Pharming announced positive results from a Phase II investigator-initiated study of RUCONEST® (recombinant human C1 esterase inhibitor, or "rhC1INH") in a double-blind, placebo-controlled clinical trial in patients at risk of nephropathy resulting from contrast-enhanced examinations.

  The study was led by Dr. Michael Osthoff at the University Hospital Basel, Basel, Switzerland. 75 eligible patients with known moderate to severe renal function impairment were given either 50 units per kg (up to 4200 units) of RUCONEST® (n=37) or placebo (n=38) immediately prior to treatment with standard-of care contrast medium as part of an elective coronary angiography with or without a percutaneous coronary intervention ("PCI"), and then a second identical treatment four hours after the intervention .

In the overall study, RUCONEST® showed a statistically-significant effect (p= 0.038) in reducing the rise in urinary Neutrophil Gelatinase-Associated Lipocalin (NGAL), the primary endpoint for the study and a generally recognized early marker of acute renal injury, in patients with diagnosed renal function impairment undergoing interventions enhanced with standard contrast media such as PCIs. 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).

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. The treatment also showed an excellent safety profile comparable to the placebo group – a particularly significant observation considering the high-risk patient group included in the study (average age approximately 77 years, with multiple comorbidities and impaired kidney function).

This data therefore supports additional clinical investigations for the use of rhC1INH in a new indication where there is significant unmet medical need.

Sijmen de Vries, Chief Executive Officer, commented:

"I am pleased to report very good results indeed today in a period of intense competition. The revenue and profit performance reinforces the success of our in-market strategy for RUCONEST®. The encouraging growth for RUCONEST® despite that competition also gives us confidence in our approach to ensure all patients have the least uncertainty, discomfort and disruption from their condition possible.
In addition, we continue to make good progress in our pipeline, reporting positive data from the first investigator-initiated study of RUCONEST® in contrast-induced nephropathy, and filing clinical trial applications to initiate our clinical development of RUCONEST® to treat and prevent pre-eclampsia. We also look forward to the data from the comparative investigator-initiated study in HAE at the end of the year, and to new filings for development of additional forms of RUCONEST® in the near future."

Financial summary

9 months to 30 September

Amounts in €m except per share data	*2018 3rd Quarter*	*2018 1st 9 months*	*2017 1st 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	

Chief Executive Officer Commentary

- During the third quarter of 2018 we demonstrated strong growth in RUCONEST® sales, building on positive progress reported in our half-year results, and validating our marketing approach to ensure all patients have access to potentially the best treatment for their HAE attacks before those attacks develop into painful symptoms.
- More recently, we delivered on important groundwork for future growth opportunities; we submitted a CTA for the first study in the development of RUCONEST® to treat and prevent pre-eclampsia, which may enable RUCONEST® to be the first real product able to ameliorate this appalling condition for expectant mothers; and we received positive results (detailed below) from the investigator initiated study in the large indication of acute kidney injury resulting from contrast-enhanced vascular interventions and examinations. This strong positive progress will now enable us to plan the next steps in clinical development for this indication..
- Looking forward to the remainder of 2018 and beyond, we expect sales in the last quarter of this year to be in the same range as Q3 despite competitive pressure as more and more patients become familiar with the ease and power of treating their HAE with RUCONEST® and for 2018 to become the first net profitable year for Pharming. We expect to start 2019 on a very strong footing with many new opportunities to enhance shareholder value very significantly in the future.

Dr Sijmen de Vries
Chief Executive Officer

Outlook

For the remainder of 2018, the Company expects:

- 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

About Pharming Group N.V.

Pharming is a specialty pharmaceutical company developing innovative products for the safe, effective treatment of rare diseases and unmet medical needs. Pharming's lead product, RUCONEST® (conestat

alfa) is a recombinant human C1 esterase inhibitor approved for the treatment of acute Hereditary Angioedema ("HAE") attacks in patients in Europe, the US, Israel and South Korea. The product is available on a named-patient basis in other territories where it has not yet obtained marketing authorization.

RUCONEST® is distributed by Pharming in Austria, France, Germany, Luxembourg, the Netherlands, the United Kingdom and the United States of America. Pharming holds commercialisation rights in Algeria, Andorra, Bahrain, Belgium, Ireland, Jordan, Kuwait, Lebanon, Morocco, Oman, Portugal, Qatar, Syria, Spain, Switzerland, Tunisia, United Arab Emirates and Yemen. In some of these countries distribution is made in association with the HAEi Global Access Program (GAP).

RUCONEST® is distributed by Swedish Orphan Biovitrum AB (publ) (SS: SOBI) in the other EU countries, and in Azerbaijan, Belarus, Georgia, Iceland, Kazakhstan, Liechtenstein, Norway, Russia, Serbia and Ukraine.

RUCONEST® is distributed in Argentina, Colombia, Costa Rica, the Dominican Republic, Panama, and Venezuela by Cytobioteck, in South Korea by HyupJin Corporation and in Israel by Kamada.

RUCONEST® is also being examined for approval for the treatment of HAE in young children (2-13 years of age) and evaluated for various additional follow-on indications.

Pharming's technology platform includes a unique, GMP-compliant, validated process for the production of pure recombinant human proteins that has proven capable of producing industrial quantities of high quality recombinant human proteins in a more economical and less immunogenetic way compared with current cell-line based methods. Leads for enzyme replacement therapy ("ERT") for Pompe and Fabry's diseases are being optimized at present, with additional programs not involving ERT also being explored at an early stage at present.

Pharming has a long-term partnership with the China State Institute of Pharmaceutical Industry ("CSIPI"), a Sinopharm company, for joint global development of new products, starting with recombinant human Factor VIII for the treatment of Haemophilia A. Pre-clinical development and manufacturing will take place to global standards at CSIPI and are funded by CSIPI. Clinical development will be shared between the partners with each partner taking the costs for their territories under the partnership.

Additional information is available on the Pharming website: www.pharming.com

Forward-looking Statements

This press release of Pharming Group N.V. and its subsidiaries ("Pharming", the "Company" or the "Group") may contain forward-looking statements including without limitation those regarding Pharming's financial projections, market expectations, developments, partnerships, plans, strategies and capital expenditures.

The Company cautions that such forward-looking statements may involve certain risks and uncertainties, and actual results may differ. Risks and uncertainties include without limitation the effect of competitive, political and economic factors, legal claims, the Company's ability to protect intellectual property, fluctuations in exchange and interest rates, changes in taxation laws or rates, changes in legislation or accountancy practices and the Company's ability to identify, develop and successfully commercialise new products, markets or technologies.

As a result, the Company's actual performance, position and financial results and statements may differ materially from the plans, goals and expectations set forth in such forward-looking statements. The Company assumes no obligation to update any forward-looking statements or information, which should be taken as of their respective dates of issue, unless required by laws or regulations.

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Pharming Group N.V.

Consolidated Interim Financial Statements (Unaudited)
For the first nine months ended 30 September 2018

Appendix: Main Financial Statements reported in US dollars

(This appendix is not part of the Consolidated Interim Financial Statements)

Consolidated Statement of Income
For the first nine months ended 30 September

Amounts in €'000, except per share data	YTD 2018	YTD 2017
Product sales	97,677	55,987
License fees	603	741
Revenues	**98,280**	**56,728**
Costs of sales	**(15,844)**	**(7,919)**
Gross profit	**82,436**	**48,809**
Other income	**473**	**607**
Research and development	(17,568)	(13,068)
General and administrative	(8,321)	(4,308)
Marketing and sales	(26,005)	(19,315)
Costs	**(51,894)**	**(36,691)**
Operating result	**31,015**	**12,725**
Fair value gain (loss) on revaluation derivatives	(725)	(15,186)
Other financial income and expenses	(18,498)	(35,248)
Financial income and expenses	**(19,223)**	**(50,434)**
Result before income tax	**11,792**	**(37,709)**
Income tax expense	(76)	-
Net result for the period	**11,716**	**(37,709)**
Attributable to:		
Owners of the parent	11,716	(37,709)
Total net result	**11,716**	**(37,709)**
Basic earnings per share (€)	**0.019**	**(0.077)**
Fully-diluted earnings per share (€)	**0.017**	**n/a**

Consolidated Statement of Comprehensive Income
For the first nine months ended 30 September

Amounts in €'000	YTD 2018	YTD 2017
Net result for the period	11,716	(37,709)
Currency translation differences	90	(482)
Items that may be subsequently reclassified to profit or loss	90	(482)
Other comprehensive income, net of tax	90	(482)
Total comprehensive income for the period	11,806	(38,191)
Attributable to:		
Owners of the parent	11,806	(38,191)

Consolidated Balance Sheet
As at date shown

Amounts in €'000	30 September 2018	31 December 2017
Intangible assets	56,322	56,631
Property, plant and equipment	8,298	8,234
Long term prepayment	1,968	2,296
Deferred tax asset	9,392	9,442
Restricted cash	1,191	1,336
Non-current assets	**77,171**	**77,939**
Inventories	20,951	18,334
Trade and other receivables	23,570	11,260
Cash and cash equivalents	71,025	58,657
Current assets	**115,546**	**88,251**
Total assets	**192,717**	**166,190**
Share capital	6,172	5,790
Share premium	391,023	370,220
Legal reserves	(848)	(938)
Accumulated deficit	(348,174)	(356,270)
Shareholders' equity	**48,173**	**18,802**
Loans and borrowings	41,432	58,684
Deferred license fee income	867	1,467
Finance lease liabilities	185	390
Other financial liabilities	15,773	28,319
Non-current liabilities	**58,257**	**88,860**
Loans and borrowings	35,924	21,962
Deferred license fee income	800	804
Derivative financial liabilities	889	8,301
Trade and other payables	31,167	27,198
Finance lease liabilities	263	263
Other financial liabilities	17,244	-
Current liabilities	**86,287**	**58,528**
Total equity and liabilities	**192,717**	**166,190**

Consolidated Statement of Cash Flows
For the first nine months ended 30 September

Amounts in €'000	YTD 2018	YTD 2017
Operating result	31,015	12,725
Non-cash adjustments:		
Depreciation, amortization	2,907	2,543
Accrued employee benefits	2,316	1,308
Deferred license fees	(603)	(741)
Operating cash flows before changes in working capital	35,635	15,835
Changes in working capital:		
Inventories	(2,617)	(54)
Trade and other receivables	(12,310)	(9,358)
Payables and other current liabilities	3,969	2,977
Total changes in working capital	(10,958)	(6,435)
Changes in non-current assets, liabilities and equity	1,347	524
Cash generated from (used in) operations before interest and taxes	26,024	9,924
Interest received	-	-
Net cash flows generated from (used in) operating activities	26,024	9,924
Capital expenditure for property, plant and equipment	(2,057)	(2,518)
Investment intangible assets	(1,826)	(2,189)
Net cash flows used in investing activities	(3,883)	(4,707)
Proceeds of loans and borrowings	-	89,181
Payments of transaction fees and expenses	-	(16,051)
Prepayments and interests on loans and borrowings	(17,990)	(76,984)
Proceeds of equity and warrants	7,949	6,110
Net cash flows generated from (used in) financing activities	(10,041)	2,256
Increase (decrease) of cash	12,100	7,473
Exchange rate effects	123	(973)
Cash and cash equivalents at 1 January	59,993	32,137
Total cash and cash equivalents at 30 September	72,216	38,637
Of which restricted cash	1,191	248
Cash and cash equivalents at 30 September	71,025	38,389

Appendix: Main Financial Statements reported in US dollars

The original Financial Statements are reported in Euros. In case of differences of interpretation between the Financial Statements in US Dollars and the Financial Statements in Euros, the Financial Statements in Euros will prevail.

Consolidated Statement of Income in US Dollars
For the first nine months ended 30 September

Amounts in $'000, except per share data	YTD 2018	YTD 2017
Product sales	116,799	62,789
License fees	722	831
Revenues	**117,521**	**63,621**
Costs of sales	**(18,946)**	**(8,881)**
Gross profit	**98,575**	**54,740**
Other income	**566**	**681**
Research and development	(21,007)	(14,656)
General and administrative	(9,950)	(4,831)
Marketing and sales	(31,097)	(21,662)
Costs	**(62,054)**	**(41,149)**
Operating result	**37,087**	**14,271**
Fair value gain (loss) on revaluation derivatives	(867)	(17,031)
Other financial income and expenses	(22,541)	(39,531)
Financial income and expenses	**(23,408)**	**(56,561)**
Result before income tax	**13,679**	**(42,291)**
Income tax expense	(91)	-
Net result for the period	**13,588**	**(42,291)**
Attributable to:		
Owners of the parent	13,588	(42,291)
Total net result	**13,588**	**(42,291)**
Basic earnings per share ($)	**0.022**	**(0.086)**
Fully-diluted earnings per share ($)	**0.020**	**n/a**

Consolidated Balance Sheet in US Dollars

As at date shown

Amounts in $'000	30 September 2018	31 December 2017
Intangible assets	65,323	67,827
Property, plant and equipment	9,624	9,862
Long term prepayment	2,283	2,749
Deferred tax asset	10,892	11,309
Restricted cash	1,381	1,600
Non-current assets	**89,503**	**93,347**
Inventories	24,299	21,958
Trade and other receivables	27,337	13,487
Cash and cash equivalents	82,374	70,254
Current assets	**134,010**	**105,699**
Total assets	**223,513**	**199,046**
Share capital	7,158	6,935
Share premium	453,509	443,412
Legal reserves	(984)	(1,124)
Accumulated deficit	(403,812)	(426,703)
Shareholders' equity	**55,871**	**22,520**
Loans and borrowings	48,054	70,286
Deferred license fee income	1,005	1,757
Finance lease liabilities	215	467
Other financial liabilities	18,292	33,918
Non-current liabilities	**67,566**	**106,428**
Loans and borrowings	41,665	26,304
Deferred license fee income	928	962
Derivative financial liabilities	1,031	9,942
Trade and other payables	36,147	32,575
Finance lease liabilities	305	315
Other financial liabilities	20,000	-
Current liabilities	**100,076**	**70,098**
Total equity and liabilities	**223,513**	**199,046**