



Horizon Pharma plc Reports Strong First-Quarter 2018 Orphan and Rheumatology Net Sales Growth; Increases Full-Year 2018 Guidance and Announces New Company Operating Structure to Enhance Focus on Rare Diseases

-- First-Quarter 2018 Orphan and Rheumatology Net Sales of \$172.2 Million Increased 11 Percent; Represented 77 Percent of Total Company Net Sales --

-- First-Quarter 2018 KRYSTEXXA Net Sales Growth of 48 Percent; Increasing Full-Year 2018 Net Sales Growth Guidance to More Than 65 Percent --

-- 50 Percent of Patients Now Enrolled in Teprotumumab Phase 3 Clinical Trial --

-- Establishing New Operating Structure Effective in Second-Quarter 2018, With Two Operating Segments: Orphan and Rheumatology; Primary Care --

-- First-Quarter 2018 Net Sales of \$223.9 Million; First-Quarter 2018 GAAP Net Loss of \$157.3 Million; Adjusted EBITDA of \$33.6 Million --

-- Increasing Full-Year 2018 Net Sales Guidance Range to \$1.170 Billion to \$1.200 Billion and Adjusted EBITDA Guidance Range to \$390 Million to \$415 Million --

DUBLIN, IRELAND – May 9, 2018 – Horizon Pharma plc (NASDAQ: HZNP) announced its first-quarter 2018 financial results today and increased its full-year 2018 net sales and adjusted EBITDA guidance. The Company also announced that, effective in the second-quarter 2018, the Company is realigning its operating structure and will report financial results as two separate operating segments: its strategic growth business, orphan and rheumatology; and primary care. The new operating structure reflects the evolution of the Company’s strategy and vision of transitioning Horizon Pharma to a biopharmaceutical company focused on rare disease medicines.

“Our orphan and rheumatology medicines represented approximately 77 percent of the Company’s first-quarter net sales and generated double-digit growth driven by 48 percent growth of KRYSTEXXA,” said Timothy P. Walbert, chairman, president and chief executive officer, Horizon Pharma plc. “Our decision to operate orphan and rheumatology separately from primary care marks a pivotal next step in our ongoing strategic transformation to a company focused on rare disease medicines. We made significant advancements during the first quarter toward our goal, including progress ahead of our expectations in the Phase 3 clinical trial for our key rare disease medicine in development, teprotumumab, which is now 50 percent enrolled.”



Financial Highlights

(in millions except for per share amounts and percentages)	<u>Q1 18</u>	<u>Q1 17</u>	<u>% Change</u>
Net sales	\$ 223.9	\$ 220.9	1
Net loss	(157.3)	(90.6)	(74)
Non-GAAP net income	4.8	35.0	(86)
Adjusted EBITDA	33.6	51.9	(35)
Net loss per share - diluted	(0.96)	(0.56)	(71)
Non-GAAP earnings per share - diluted	0.03	0.21	(86)

First-Quarter and Recent Company Highlights

- New Head of R&D and Chief Scientific Officer:** Shao-Lee Lin, M.D., Ph.D., joined the Company in January 2018 as executive vice president, head of research and development (R&D) and chief scientific officer. Dr. Lin is an accomplished pharmaceutical executive, physician and scientist with more than 20 years of academic and clinical research experience and will accelerate the development of a robust pipeline to drive the Company's next phase of growth.

“We are committed to establishing Horizon Pharma as a leader in the rare disease space, and one of our goals to support that objective is to enhance the capabilities of our R&D organization,” said Lin. “We are well on our way, having made several important additions to the organization that expand our development capabilities, support our business development team in evaluating and identifying development-stage opportunities and lead our therapeutic areas from a clinical development strategy and portfolio management perspective. Enhancing our R&D organization will enable us to maximize our on-market medicines and develop new medicines for patients with unmet needs – and in the case of rare diseases, some of the most significantly underserved patients.”

- Intellectual Property Update:** The Company recently received two Notices of Allowance from the U.S. Patent and Trademark Office for U.S. patent application numbers 15/457,643 and 15/687,132, both entitled "Methods of Therapeutic Monitoring of Nitrogen Scavenging Drugs" that cover RAVICTI. The U.S. patents scheduled to issue from these applications will expire on Sept. 22, 2030. After issuance, the Company plans to list the U.S. patents in the FDA's Approved Drug Products with Therapeutic Equivalence Evaluations, or Orange Book.



Research and Development

Orphan Candidates and Programs:

- **Teprotumumab:** The Phase 3 clinical trial for teprotumumab, the Company's fully human monoclonal antibody IGF-1R-inhibitor in development for the treatment of thyroid eye disease (TED), a rare eye disease, is 50 percent enrolled and is on track for enrollment completion by year end, or earlier. The pivotal confirmatory study is evaluating teprotumumab for the treatment of moderate-to-severe active TED, which has no FDA-approved treatments. The Company estimates peak annual U.S. net sales of more than \$750 million for teprotumumab, assuming U.S. FDA approval.

Rheumatology Pipeline Candidates and Programs:

- **Immunomodulation Studies:** The evaluation of the use of immunomodulation therapies to enhance the response rate to KRYSTEXXA is being studied in two investigator-initiated trials, using two different immunomodulators, both of which are commonly used by rheumatologists. **REduCing Immunogenicity to Pegloticase (RECIPE)** is a double-blind, placebo controlled trial to evaluate the impact of a 12-week course of immunomodulating therapy with daily doses of mycophenolate mofetil (MMF). **Tolerization Reduces Intolerance to Pegloticase and Prolongs the Urate Lowering Effect (TRIPLE)** is an exploratory, open-label adaptive trial with multiple patient cohorts, including a cohort to evaluate the impact of adding daily doses of the immunomodulator azathioprine for a two-week run-in period, followed by KRYSTEXXA every two weeks for a total of 13 doses along with daily doses of azathioprine.
- **New Rheumatology Programs:** In January 2018, the Company announced two development programs for next-generation biologics for uncontrolled gout (chronic gout that is refractory to conventional therapies) to support and sustain the Company's market leadership in uncontrolled gout: **HZN-003** and **PASylated uricase technology**. HZN-003 is a pre-clinical, genetically engineered uricase derivative with optimized uricase and optimized PEGylation technology. PASylated uricase technology may improve the half-life of uricase, and the Company is collaborating with a third party to identify a lead candidate that could use the technology to construct a next-generation gout biologic. The Company also announced the addition of **HZN-002**, a pre-clinical, novel dexamethasone conjugate with the potential to address inflammatory diseases through its targeted delivery technology.

New Operating Structure Aligned with Long-term Strategy

Given the Company's focus on rare disease medicines, effective in the second quarter of 2018, the Company is realigning its structure to operate its strategic growth business, orphan and rheumatology, separate from its primary care business. The new structure allows the Company to more efficiently allocate its resources to address unmet treatment needs for patients with rare diseases.



As part of the new operating structure, the Company has realigned its commercial operations under a new leadership position, executive vice president and chief commercial officer, and recently promoted Vikram Karnani to that role. Karnani was most recently senior vice president, rheumatology business unit, leading the successful growth to date of KRYSTEXXA. In addition, aligned with the new operating structure, the Company is adding critical R&D leadership roles to support the orphan and rheumatology segment, including recently hired clinical development heads for both of these therapeutic areas.

As a result of these changes, in the second-quarter of 2018, the Company will begin reporting its financial results as two separate operating segments: the orphan and rheumatology segment, the Company's strategic rare disease-focused business and the primary care segment, reporting net sales and operating income for each segment.

First-Quarter 2018 Total Company Financial Results

Note: For additional detail and reconciliation of non-GAAP financial measures to the most directly comparable GAAP financial measures, please refer to the tables at the end of this release.

- **Total Net Sales:** First-quarter net sales were \$223.9 million, an increase of 1.4 percent, driven by continued strong growth of the Company's orphan and rheumatology medicines. Net sales of \$220.9 million in the first quarter of 2017 included PROCYSBI and QUINSAIR net sales of \$4.9 million in Europe, the Middle East and Africa (EMEA) regions. The EMEA marketing rights to PROCYSBI and QUINSAIR were divested in June 2017. Excluding the 2017 EMEA net sales of PROCYSBI and QUINSAIR, year-over-year growth would have been 3.7 percent.
- **Gross Profit:** Under U.S. GAAP in the first quarter of 2018, the gross profit ratio was 48.1 percent compared to 37.0 percent in the first quarter of 2017. The non-GAAP gross profit ratio in the first quarter of 2018 was 87.0 percent compared to 88.5 percent in the first quarter of 2017.
- **Operating Expenses:** R&D expenses were 7.9 percent of net sales; and selling, general and administrative (SG&A) expenses were 80.2 percent of net sales. Non-GAAP R&D expenses were 6.8 percent of net sales, and non-GAAP SG&A expenses were 65.2 percent of net sales.
- **Income Tax Rate:** The income tax rate in the first quarter of 2018 on a GAAP basis was 0.2 percent and on a non-GAAP basis was 44.5 percent.
- **Net (Loss) Income:** On a GAAP basis in the first quarter of 2018, net loss was \$157.3 million. First-quarter 2018 non-GAAP net income was \$4.8 million.
- **Adjusted EBITDA:** In the first quarter of 2018, adjusted EBITDA was \$33.6 million.
- **Earnings (Loss) per Share:** On a GAAP basis in the first quarter of 2018, diluted loss per share was \$0.96; in the first quarter of 2017, diluted loss per share was \$0.56. Non-GAAP diluted earnings per share in the first quarter of 2018 and 2017 were \$0.03 and \$0.21, respectively. Weighted average shares outstanding used for calculating GAAP diluted loss per share and non-GAAP diluted earnings per share in the first quarter of 2018 were 164.5 million and 167.8 million, respectively.



First-Quarter Orphan and Rheumatology Net Sales

(in millions except for percentages)	Q1 18	Q1 17	% Change
Orphan	\$ 114.7	\$ 112.5	2
RAVICTI [®]	49.1	43.9	12
PROCYSBI ^{®(1)}	34.9	34.3	2
ACTIMMUNE [®]	24.9	26.2	(5)
BUPHENYL [®]	5.7	6.3	(9)
QUINSAIR ^{™(1)}	0.1	1.8	(93)
Rheumatology	57.5	42.8	35
KRYSTEXXA [®]	46.7	31.6	48
RAYOS [®]	10.7	10.3	4
LODOTRA [®]	0.1	0.9	(87)
Orphan and Rheumatology Net Sales	\$ 172.2	\$ 155.3	11

(1) On June 23, 2017, Horizon Pharma completed the divestiture of a European subsidiary that owned the marketing rights to PROCYSBI and QUINSAIR in Europe, the Middle East and Africa to Chiesi Farmaceutici S.p.A. Horizon Pharma retains marketing rights for the two medicines in the United States, Canada, Latin America and Asia.

- Combined first-quarter 2018 net sales of orphan and rheumatology medicines of \$172.2 million increased 11 percent, driven by continued strong KRYSTEXXA vial growth, as well as growth of RAVICTI and PROCYSBI. Combined first-quarter 2017 net sales of orphan and rheumatology medicines of \$155.3 million included EMEA net sales of PROCYSBI and QUINSAIR, which were divested in June 2017, of \$4.9 million. Excluding the 2017 EMEA net sales of PROCYSBI and QUINSAIR from combined orphan and rheumatology net sales, year-over-year growth would have been 15 percent.

First-Quarter Primary Care Net Sales

(in millions except for percentages)	Q1 18	Q1 17	% Change
Primary Care			
PENNSAID [®] 2%	26.8	41.6	(36)
DUEXIS [®]	15.7	17.7	(12)
VIMOVO [®]	8.4	4.9	72
MIGERGOT [®]	0.8	1.4	(47)
Primary Care Net Sales	51.7	65.6	(21)

- First-quarter 2018 net sales of primary care medicines were \$51.7 million, negatively impacted by seasonality, to a somewhat greater degree than originally anticipated. First-quarter net sales also included a negative \$14 million impact from an additional accrual for medicines in the wholesale and retail channel following the Company's price action. Excluding the additional accrual, which did not occur in first-quarter 2017, first-quarter 2018 primary care net sales on a pro-forma basis were similar to first-quarter 2017, reflecting the stability of this business.



Cash Flow Statement and Balance Sheet Highlights

- On a GAAP basis in the first quarter of 2018, operating cash flow was negative \$60.8 million. Non-GAAP operating cash flow was negative \$52.7 million in the first quarter of 2018, as expected. GAAP and non-GAAP operating cash flow in the first quarter of 2017 included a significant one-time working capital benefit associated with the implementation of managed care contracts for certain primary care medicines.
- The Company had cash and cash equivalents of \$674.3 million as of March 31, 2018.
- As of March 31, 2018, the total principal amount of debt outstanding was \$2.019 billion, which comprised \$844 million in senior secured term loans due 2024; \$300 million senior notes due 2024; \$475 million senior notes due 2023; and \$400 million exchangeable senior notes due 2022. As of March 31, 2018, net debt was \$1.344 billion.

Full-Year 2018 Guidance

The Company now expects full-year 2018 net sales guidance of \$1.170 billion to \$1.200 billion, an increase from the previous range of \$1.150 billion to \$1.180 billion. Full-year 2018 adjusted EBITDA is now expected to be \$390 million to \$415 million, an increase from the previous range of \$370 million to \$395 million. Company guidance now assumes a delay in the implementation of a U.S. government rule related to 340B entity drug pricing to July 1, 2019, following the U.S. Department of Health and Human Services' proposal to delay the effective date to that date. As a result, the Company now expects full-year 2018 net sales growth for KRYSTEXXA of more than 65 percent.

Webcast

At 8 a.m. EST / 1 p.m. IST today, the Company will host a live webcast to review its financial and operating results and provide a general business update. The live webcast and a replay may be accessed at <http://ir.horizon-pharma.com>. Please connect to the Company's website at least 15 minutes prior to the live webcast to ensure adequate time for any software download that may be needed to access the webcast. A replay of the webcast will be available approximately two hours after the live webcast.

About Horizon Pharma plc

Horizon Pharma plc is focused on researching, developing and commercializing innovative medicines that address unmet treatment needs for rare and rheumatic diseases. By fostering a growing pipeline of medicines in development and exploring all potential uses for currently marketed medicines, we strive to make a powerful difference for patients, their caregivers and physicians. For us, it's personal: by living up to our own potential, we are helping others live up to theirs. For more information, please visit www.horizonpharma.com. Follow @HZNPplc on Twitter, like us on Facebook or explore career opportunities on LinkedIn.



Note Regarding Use of Non-GAAP Financial Measures

EBITDA, or earnings before interest, taxes, depreciation and amortization, and adjusted EBITDA are used and provided by Horizon Pharma as non-GAAP financial measures. Horizon Pharma provides certain other financial measures such as non-GAAP net income, non-GAAP diluted earnings per share, non-GAAP gross profit and gross profit ratio, non-GAAP operating expenses, non-GAAP tax rate and non-GAAP operating cash flow, each of which include adjustments to GAAP figures. These non-GAAP measures are intended to provide additional information on Horizon Pharma's performance, operations, expenses, profitability and cash flows. Adjustments to Horizon Pharma's GAAP figures as well as EBITDA exclude acquisition and/or divestiture-related expenses, charges related to the discontinuation of ACTIMMUNE development for Friedreich's ataxia, gain from divestiture, an upfront fee for a license of a patent, a litigation settlement, loss on debt extinguishment, costs of debt refinancing, drug manufacturing harmonization costs, restructuring and realignment costs, as well as non-cash items such as share-based compensation, depreciation and amortization, royalty accretion, non-cash interest expense, long-lived asset impairment charges, impacts of contingent royalty liability remeasurements and other non-cash adjustments. Certain other special items or substantive events may also be included in the non-GAAP adjustments periodically when their magnitude is significant within the periods incurred. Horizon maintains an established non-GAAP cost policy that guides the determination of what costs will be excluded in non-GAAP measures. Horizon Pharma believes that these non-GAAP financial measures, when considered together with the GAAP figures, can enhance an overall understanding of Horizon Pharma's financial and operating performance. The non-GAAP financial measures are included with the intent of providing investors with a more complete understanding of the Company's historical and expected 2018 financial results and trends and to facilitate comparisons between periods and with respect to projected information. In addition, these non-GAAP financial measures are among the indicators Horizon Pharma's management uses for planning and forecasting purposes and measuring the Company's performance. For example, adjusted EBITDA is used by Horizon Pharma as one measure of management performance under certain incentive compensation arrangements. These non-GAAP financial measures should be considered in addition to, and not as a substitute for, or superior to, financial measures calculated in accordance with GAAP. The non-GAAP financial measures used by the Company may be calculated differently from, and therefore may not be comparable to, non-GAAP financial measures used by other companies. Horizon Pharma has not provided a reconciliation of its full-year 2018 adjusted EBITDA outlook to an expected net income (loss) outlook because certain items such as acquisition/divestiture-related expenses and share-based compensation that are a component of net income (loss) cannot be reasonably projected due to the significant impact of changes in Horizon Pharma's stock price, the variability associated with the size or timing of acquisitions/divestitures and other factors. These components of net income (loss) could significantly impact Horizon Pharma's actual net income (loss).



Forward-Looking Statements

This press release contains forward-looking statements, including, but not limited to, statements related to Horizon Pharma's full-year 2018 net sales and adjusted EBITDA guidance, expected growth of certain medicines, estimated peak annual net sales of certain of Horizon Pharma's medicines and pipeline candidates, expected financial performance in future periods, expected timing and scope of clinical trials, including the Phase 3 clinical trial of teprotumumab, expected increases in investment in Horizon Pharma's rare disease medicine pipeline and the impact thereof, expected enhancements to Horizon Pharma's R&D function, potential market opportunity for Horizon Pharma's medicines in approved and potential additional indications, and business and other statements that are not historical facts. These forward-looking statements are based on Horizon Pharma's current expectations and inherently involve significant risks and uncertainties. Actual results and the timing of events could differ materially from those anticipated in such forward-looking statements as a result of these risks and uncertainties, which include, without limitation, risks that Horizon Pharma's actual future financial and operating results may differ from its expectations or goals; Horizon Pharma's ability to grow net sales from existing products; the availability of coverage and adequate reimbursement and pricing from government and third-party payers and risks relating to Horizon Pharma's ability to successfully implement its business strategies; risks inherent in developing novel medicine candidates, such as teprotumumab, and existing medicines for new indications; risks related to acquisition integration and achieving projected benefits; risks associated with regulatory approvals; risks in the ability to recruit, train and retain qualified personnel; competition, including potential generic competition; the ability to protect intellectual property and defend patents; regulatory obligations and oversight, including any changes in the legal and regulatory environment in which Horizon Pharma operates and those risks detailed from time-to-time under the caption "Risk Factors" and elsewhere in Horizon Pharma's filings and reports with the SEC. Horizon Pharma undertakes no duty or obligation to update any forward-looking statements contained in this press release as a result of new information.

Contacts:

Investors:

Tina Ventura
Senior Vice President,
Investor Relations
investor-relations@horizonpharma.com

Ruth Venning
Executive Director,
Investor Relations
investor-relations@horizonpharma.com

U.S. Media:

Geoff Curtis
Senior Vice President,
Corporate Affairs & Chief Communications Officer
media@horizonpharma.com

Ireland Media:

Ray Gordon
Gordon MRM
ray@gordonmrm.ie



Horizon Pharma plc
Condensed Consolidated Statements of Operations (Unaudited)
(in thousands, except share and per share data)

	Three Months Ended March 31,	
	2018	2017
Net sales	\$ 223,881	\$ 220,859
Cost of goods sold	116,092	139,116
Gross profit	<u>107,789</u>	<u>81,743</u>
OPERATING EXPENSES:		
Research and development	17,645	13,061
Selling, general and administrative	179,599	174,065
Impairment of long-lived asset	37,853	-
Total operating expenses	<u>235,097</u>	<u>187,126</u>
Operating loss	<u>(127,308)</u>	<u>(105,383)</u>
OTHER EXPENSE, NET:		
Interest expense, net	(30,454)	(31,983)
Foreign exchange loss	(110)	(259)
Loss on debt extinguishment	-	(533)
Other income, net	178	35
Total other expense, net	<u>(30,386)</u>	<u>(32,740)</u>
Loss before benefit for income taxes	(157,694)	(138,123)
Benefit for income taxes	<u>(367)</u>	<u>(47,553)</u>
Net loss	<u>\$ (157,327)</u>	<u>\$ (90,570)</u>
Net loss per ordinary share - basic and diluted	<u>\$ (0.96)</u>	<u>\$ (0.56)</u>
Weighted average ordinary shares outstanding - basic and diluted	<u>164,549,502</u>	<u>161,972,052</u>



Horizon Pharma plc
Condensed Consolidated Balance Sheets (Unaudited)
(in thousands, except share data)

	As of	
	March 31, 2018	December 31, 2017
ASSETS		
CURRENT ASSETS:		
Cash and cash equivalents	\$ 674,330	\$ 751,368
Restricted cash	6,390	6,529
Accounts receivable, net	404,208	405,214
Inventories, net	47,365	61,655
Prepaid expenses and other current assets	52,805	43,402
Total current assets	1,185,098	1,268,168
Property and equipment, net	19,488	20,405
Developed technology, net	2,338,942	2,443,949
Other intangible assets, net	5,241	5,441
Goodwill	426,441	426,441
Deferred tax assets, net	859	3,470
Other assets	26,776	36,081
Total assets	\$ 4,002,845	\$ 4,203,955
LIABILITIES AND SHAREHOLDERS' EQUITY		
CURRENT LIABILITIES:		
Long-term debt—current portion	\$ 38,446	\$ 10,625
Accounts payable	41,271	34,681
Accrued expenses	180,448	175,697
Accrued trade discounts and rebates	429,701	501,753
Accrued royalties—current portion	65,534	65,328
Deferred revenues—current portion	3,812	6,885
Total current liabilities	759,212	794,969
LONG-TERM LIABILITIES:		
Exchangeable notes, net	318,669	314,384
Long-term debt, net, net of current	1,547,912	1,576,646
Accrued royalties, net of current	291,456	291,185
Deferred revenues, net of current	-	9,713
Deferred tax liabilities, net	157,472	157,945
Other long-term liabilities	67,029	68,015
Total long-term liabilities	2,382,538	2,417,888
COMMITMENTS AND CONTINGENCIES		
SHAREHOLDERS' EQUITY:		
Ordinary shares, \$0.0001 nominal value; 300,000,000 shares authorized; 165,359,893 and 164,785,083 shares issued at March 31, 2018 and December 31, 2017, respectively, and 164,975,527 and 164,400,717 shares outstanding at March 31, 2018 and December 31, 2017, respectively	17	16
Treasury stock, 384,366 ordinary shares at March 31, 2018 and December 31, 2017	(4,585)	(4,585)
Additional paid-in capital	2,274,254	2,248,979
Accumulated other comprehensive loss	(520)	(983)
Accumulated deficit	(1,408,071)	(1,252,329)
Total shareholders' equity	861,095	991,098
Total liabilities and shareholders' equity	\$ 4,002,845	\$ 4,203,955



Horizon Pharma plc
Condensed Consolidated Statements of Cash Flows (Unaudited)
(in thousands)

	Three Months Ended March 31,	
	2018	2017
CASH FLOWS FROM OPERATING ACTIVITIES:		
Net loss	\$ (157,327)	\$ (90,570)
Adjustments to reconcile net loss to net cash (used in) provided by operating activities		
Depreciation and amortization expense	68,907	71,483
Equity-settled share-based compensation	27,833	28,837
Royalty accretion	14,718	12,959
Royalty liability remeasurement	(2,151)	(2,944)
Impairment of long-lived asset	37,853	-
Amortization of debt discount and deferred financing costs	5,496	5,423
Deferred income taxes	1,680	(47,695)
Loss on debt extinguishment	-	533
Foreign exchange and other adjustments	(120)	787
Changes in operating assets and liabilities:		
Accounts receivable	1,064	(94,377)
Inventories	14,290	37,050
Prepaid expenses and other current assets	(9,805)	(2,445)
Accounts payable	6,528	36,078
Accrued trade discounts and rebates	(72,120)	116,079
Accrued expenses and accrued royalties	4,454	(46,040)
Deferred revenues	(1,484)	(618)
Other non-current assets and liabilities	(627)	266
Net cash (used in) provided by operating activities	<u>(60,811)</u>	<u>24,806</u>
CASH FLOWS FROM INVESTING ACTIVITIES:		
Payment related to license agreement	(12,000)	-
Purchases of property and equipment	(665)	(1,423)
Net cash used in investing activities	<u>(12,665)</u>	<u>(1,423)</u>
CASH FLOWS FROM FINANCING ACTIVITIES:		
Repayment of term loans	(2,125)	(772,750)
Net proceeds from term loans	-	847,768
Proceeds (refunds) related to the ESPP plan	14	(173)
Proceeds from the issuance of ordinary shares in connection with stock option exercises	945	544
Payment of employee withholding taxes relating to share-based awards	(3,517)	(4,277)
Net cash (used in) provided by financing activities	<u>(4,683)</u>	<u>71,112</u>
Effect of foreign exchange rate changes on cash, cash equivalents and restricted cash	<u>982</u>	<u>(298)</u>
Net (decrease) increase in cash, cash equivalents and restricted cash	(77,177)	94,197
Cash, cash equivalents and restricted cash, beginning of the period ⁽¹⁾	757,897	516,150
Cash, cash equivalents and restricted cash, end of the period ⁽¹⁾	<u>\$ 680,720</u>	<u>\$ 610,347</u>

(1) Amounts include restricted cash balance in accordance with ASU No. 2016-18.



Horizon Pharma plc
GAAP to Non-GAAP Reconciliations
Net Income and Earnings Per Share (Unaudited)
(in thousands, except share and per share data)

	Three Months Ended March 31,	
	2018	2017
GAAP net loss	\$ (157,327)	\$ (90,570)
Non-GAAP adjustments:		
Impairment of long-lived asset	37,853	-
Remeasurement of royalties for medicines acquired through business combinations	(2,151)	(2,944)
Acquisition/divestiture-related costs	3,911	10,039
Restructuring and realignment costs	3,342	-
Amortization, accretion and step-up:		
Intangible amortization expense	67,355	69,677
Accretion of royalty liabilities	14,719	12,959
Amortization of debt discount and deferred financing costs	5,496	5,423
Inventory step-up expense	17,076	40,595
Share-based compensation	27,833	28,469
Depreciation expense	1,552	1,806
Charges relating to discontinuation of Friedreich's ataxia program	950	-
Drug substance harmonization costs	804	4,299
Upfront and milestone payments related to license agreements	90	-
Fees related to term loan refinancings	27	4,143
Loss on debt extinguishment	-	533
Royalties for medicines acquired through business combinations	(12,521)	(11,317)
Total of pre-tax non-GAAP adjustments	166,336	163,682
Income tax effect of pre-tax non-GAAP adjustments	31,683	(38,103)
Other non-GAAP income tax adjustments	(35,893)	-
Total of non-GAAP adjustments	162,126	125,579
Non-GAAP Net Income	\$ 4,799	\$ 35,009
Non-GAAP Earnings Per Share:		
Weighted average shares - Basic	164,549,502	161,972,052
Non-GAAP Earnings Per Share - Basic:		
GAAP loss per share - Basic	(0.96)	(0.56)
Non-GAAP adjustments	0.99	0.78
Non-GAAP earnings per share - Basic	0.03	0.22
Weighted average shares - Diluted		
Weighted average shares - Basic	164,549,502	161,972,052
Ordinary share equivalents	3,201,430	2,895,016
Weighted average shares - Diluted	167,750,932	164,867,068
Non-GAAP Earnings Per Share - Diluted		
GAAP loss per share - Diluted	(0.96)	(0.56)
Non-GAAP adjustments	0.99	0.78
Diluted earnings per share effect of ordinary share equivalents	-	(0.01)
Non-GAAP earnings per share - Diluted	0.03	0.21



Horizon Pharma plc
GAAP to Non-GAAP Reconciliations
EBITDA, Gross Profit and Operating Cash Flow (Unaudited)
(in thousands, except percentages)

	Three Months Ended March 31,	
	2018	2017
EBITDA and Adjusted EBITDA:		
GAAP net loss	\$ (157,327)	\$ (90,570)
Depreciation	1,552	1,806
Amortization, accretion and step-up:		
Intangible amortization expense	67,355	69,677
Accretion of royalty liabilities	14,719	12,959
Amortization of deferred revenue	-	(204)
Inventory step-up expense	17,076	40,595
Interest expense, net (including amortization of debt discount and deferred financing costs)	30,454	31,983
Benefit for income taxes	(367)	(47,553)
EBITDA	\$ (26,538)	\$ 18,693
Other non-GAAP adjustments:		
Impairment of long-lived asset	37,853	-
Remeasurement of royalties for medicines acquired through business combinations	(2,151)	(2,944)
Acquisition/divestiture-related costs	3,911	10,039
Restructuring and realignment costs	3,342	-
Share-based compensation	27,833	28,469
Charges relating to discontinuation of Friedreich's ataxia program	950	-
Drug substance harmonization costs	804	4,299
Upfront and milestone payments related to license agreements	90	-
Fees related to term loan refinancing	27	4,143
Loss on debt extinguishment	-	533
Royalties for medicines acquired through business combinations	(12,521)	(11,317)
Total of other non-GAAP adjustments	60,138	33,222
Adjusted EBITDA	\$ 33,600	\$ 51,915
GAAP gross profit	\$ 107,789	\$ 81,743
Non-GAAP gross profit adjustments:		
Acquisition/divestiture-related costs	19	80
Share-based compensation	783	428
Remeasurement of royalties for medicines acquired through business combinations	(2,151)	(2,944)
Intangible amortization expense	67,155	69,474
Accretion of royalty liabilities	14,719	12,959
Inventory step-up expense	17,076	40,595
Depreciation	176	183
Charges relating to discontinuation of Friedreich's ataxia program	950	-
Drug substance harmonization costs	804	4,299
Royalties for medicines acquired through business combinations	(12,521)	(11,317)
Total of Non-GAAP adjustments	87,010	113,757
Non-GAAP gross profit	\$ 194,799	\$ 195,500
GAAP gross profit %	48.1%	37.0%
Non-GAAP gross profit %	87.0%	88.5%
Non-GAAP operating cash flow:		
GAAP cash provided by operating activities	\$ (60,811)	\$ 24,806
Cash payments for acquisition/divestiture-related costs	3,958	20,392
Cash payment for litigation settlement	-	16,250
Cash payments for upfront and milestone payments related to licence agreement	275	-
Cash payments for charges relating to clinical trial wind-down costs	3,399	482
Cash payments relating to term loan refinancing	18	3,312
Cash payments for restructuring and realignment costs	447	-
Non-GAAP operating cash flow	\$ (52,714)	\$ 65,242



Horizon Pharma plc
GAAP to Non-GAAP Tax Rate Reconciliation (Unaudited)
(in millions, except percentages)

Q1 2018					
	Pre-tax Net (Loss) Income	Income Tax (Benefit) Expense	Tax Rate	Net (Loss) Income	Diluted (Loss) Earnings Per Share
As reported - GAAP	\$ (157.7)	\$ (0.4)	0.2%	\$ (157.2)	\$ (0.96)
Non-GAAP adjustments	166.3	4.2		162.1	
Non-GAAP	\$ 8.6	\$ 3.8	44.5%	\$ 4.9	\$ 0.03

Q1 2017					
	Pre-tax Net (Loss) Income	Income Tax (Benefit) Expense	Tax Rate	Net (Loss) Income	Diluted (Loss) Earnings Per Share
As reported - GAAP	\$ (138.1)	\$ (47.6)	34.4%	\$ (90.6)	\$ (0.56)
Non-GAAP adjustments	163.7	38.1		125.7	
Non-GAAP	\$ 25.6	\$ (9.4)	(37.0)%	\$ 35.0	\$ 0.21



Horizon Pharma plc
Certain Income Statement Line Items - Non-GAAP Adjusted
For the Three Months Ended March 31, 2018 and March 31, 2017
(in thousands) (Unaudited)

Horizon Pharma plc
Certain Income Statement Line Items - Non-GAAP Adjusted
For the Three Months Ended March 31, 2018
(Unaudited)

	COGS	Research & Development	Selling, General & Administrative	Impairment of long-lived asset	Interest Expense	Income Tax Benefit (Expense)
\$	(116,092)	(17,645)	(179,599)	(37,853)	(30,454)	367
	-	-	-	37,853	-	-
	(2,151)	-	-	-	-	-
	19	(85)	3,977	-	-	-
	-	-	3,342	-	-	-
	67,155	-	200	-	5,496	-
	-	-	-	-	-	-
	14,719	-	-	-	-	-
	17,076	-	-	-	-	-
	783	2,440	24,610	-	-	-
	176	-	1,376	-	-	-
	804	-	-	-	-	-
	-	90	-	-	-	-
	-	-	27	-	-	-
	950	-	-	-	-	-
	(12,521)	-	-	-	-	-
	-	-	-	-	-	31,683
	-	-	-	-	-	(35,893)
	87,010	2,445	33,552	37,853	5,496	(42,110)
\$	(29,082)	(15,200)	(146,067)	-	(24,958)	(3,843)

Horizon Pharma plc
Certain Income Statement Line Items - Non-GAAP Adjusted
For the Three Months Ended March 31, 2017
(Unaudited)

	COGS	Research & Development	Selling, General & Administrative	Interest Expense	Loss on Debt Extinguishment	Income Tax Benefit (Expense)
\$	(139,116)	(13,061)	(174,065)	(31,983)	(533)	47,553
	80	177	9,782	-	-	-
	69,474	-	203	-	-	-
	-	-	-	5,423	-	-
	12,959	-	-	-	-	-
	40,595	-	-	-	-	-
	428	2,049	25,992	-	-	-
	183	-	1,623	-	-	-
	4,299	-	-	-	-	-
	(2,944)	-	-	-	-	-
	-	-	4,143	-	-	-
	-	-	-	-	533	-
	(11,317)	-	-	-	-	-
	-	-	-	-	-	(38,103)
	113,757	2,226	41,743	5,423	533	(38,103)
\$	(25,359)	(10,835)	(132,322)	(26,560)	-	9,450

GAAP as reported

Non-GAAP Adjustments (in thousands):

- Impairment of long-lived asset⁽¹⁾
- Re measurement of royalties for medicines acquired through business combinations⁽²⁾
- Acquisition/divestiture-related costs⁽³⁾
- Restructuring and realignment costs⁽⁴⁾
- Amortization, accretion and step-up:
 - Intangible amortization expense⁽⁵⁾
 - Amortization of debt discount and deferred financing costs⁽⁶⁾
 - Accretion of royalty liability⁽⁷⁾
 - Inventory step-up expense⁽⁸⁾
- Share-based compensation⁽⁹⁾
- Depreciation expense⁽¹⁰⁾
- Drug substance harmonization costs⁽¹¹⁾
- Upfront and milestone payments related to license agreements⁽¹²⁾
- Fees related to term loan refinancing⁽¹³⁾
- Charges relating to discontinuation of Friedrich's ataxia program⁽¹⁴⁾
- Royalties for medicines acquired through business combinations⁽¹⁵⁾
- Income tax effect on pre-tax non-GAAP adjustments⁽¹⁶⁾
- Other non-GAAP income adjustments⁽¹⁷⁾
- Total of non-GAAP adjustments

Non-GAAP

GAAP as reported

Non-GAAP Adjustments (in thousands):

- Acquisition/divestiture-related costs⁽³⁾
- Amortization, accretion and step-up:
 - Intangible amortization expense⁽⁵⁾
 - Amortization of debt discount and deferred financing costs⁽⁶⁾
 - Accretion of royalty liability⁽⁷⁾
 - Inventory step-up expense⁽⁸⁾
- Share-based compensation⁽⁹⁾
- Depreciation expense⁽¹⁰⁾
- Drug substance harmonization costs⁽¹¹⁾
- Re measurement of royalties for medicines acquired through business combinations⁽²⁾
- Fees related to term loan refinancing⁽¹³⁾
- Loss on debt extinguishment⁽¹⁸⁾
- Royalties for medicines acquired through business combinations⁽¹⁵⁾
- Income tax effect on pre-tax non-GAAP adjustments⁽¹⁶⁾
- Total of non-GAAP adjustments

Non-GAAP



NOTES FOR CERTAIN INCOME STATEMENT LINE ITEMS - NON-GAAP

- (1) During the three months ended March 31, 2018, the Company recorded an impairment of \$37.9 million to write off the book value of developed technology related to PROCYSBI in Canada and Latin America due to lower than anticipated future net sales.
- (2) At the time of the Company's acquisition of the rights to ACTIMMUNE, BUPHENYL, KRSTEXXA, MIGERGOT, PROCYSBI, RAVICTI and VIMOVO, the Company estimated the fair value of contingent royalties payable to third parties using an income approach under the discounted cash flow method, which included revenue projections and other assumptions the Company made to determine the fair value. If the Company significantly overperforms or underperforms against its original revenue projections or it becomes necessary to make changes to assumptions as a result of a triggering event, the Company is required to reassess the fair value of the contingent royalties payable. Any subsequent adjustment to fair value is recorded in the period such adjustment is made as either an increase or decrease to royalties payable, with a corresponding increase or decrease in cost of goods sold, in accordance with established accounting policies. The Company recorded net decreases of \$2.2 million and \$2.9 million to cost of goods sold to adjust the amount of the contingent royalty liabilities relating to PROCYSBI during the first quarter of 2018, and to KRSTEXXA and VIMOVO during the first quarter of 2017, respectively.
- (3) Expenses, including legal and consulting fees, incurred in connection with the Company's acquisitions and divestitures have been excluded.
- (4) Represents expenses, including severance costs and consulting fees, related to the restructuring and realignment activities.
- (5) Intangible amortization expenses are associated with the Company's intellectual property rights, developed technology and customer relationships of ACTIMMUNE, BUPHENYL, KRSTEXXA, LODOTRA, MIGERGOT, PENNSAID 2%, PROCYSBI, RAVICTI, RAYOS and VIMOVO.
- (6) Represents amortization of debt discount and deferred financing costs associated with the Company's debt.
- (7) Represents accretion expense associated with the ACTIMMUNE, BUPHENYL, KRSTEXXA, MIGERGOT, PROCYSBI, QUINSAIR, RAVICTI and VIMOVO contingent royalty liabilities.
- (8) During the three months ended March 31, 2018, the Company recognized in cost of goods sold, \$17.1 million for inventory step-up expense, primarily related to KRSTEXXA inventory sold.

During the three months ended March 31, 2017, the Company recognized in cost of goods sold, \$14.4 million for inventory step-up expense related to KRSTEXXA inventory sold and \$26.1 million for inventory step-up expense related to PROCYSBI and QUINSAIR inventory sold.
- (9) Represents share-based compensation expense associated with the Company's stock option, restricted stock unit and performance stock unit grants to its employees and non-employees, its cash-settled long-term incentive plan and its employee stock purchase plan.
- (10) Represents depreciation expense related to the Company's property, equipment, software and leasehold improvements.
- (11) During the year ended December 31, 2016, the Company committed to spend \$14.9 million related to the harmonization of the manufacturing processes for ACTIMMUNE and IMUKIN drug substance. During the three months ended March 31, 2018, the Company incurred costs of \$0.8 million related to these activities that qualify for exclusion in the Company's non-GAAP financial measures under its non-GAAP cost policy.
- (12) Represents upfront and milestone payments related to license agreements.
- (13) Represents arrangement and other fees relating to the refinancing of the Company's term loans during the first quarter 2018.



- (14) During the three months ended March 31, 2018, charges relating to discontinuation of the Friedreich's ataxia program include a \$1.0 million increase in cost of goods sold relating to the purchase of additional units of ACTIMMUNE.
- (15) Royalties of \$12.5 million were incurred during the three months ended March 31, 2018, based on the periods' net sales for ACTIMMUNE, BUPHENYL, KRYSTEXXA, MIGERGOT, PROCYSBI, QUINSAIR, RAVICTI and VIMOVO.
- (16) Income tax adjustments on pre-tax non-GAAP adjustments represent the estimated income tax impact of each pre-tax non-GAAP adjustment based on the statutory income tax rate of the applicable jurisdictions for each non-GAAP adjustment.
- (17) Other non-GAAP income tax adjustments during the three months ended March 31, 2018 reflect a measurement period adjustment relating to Notice 2018-28 that was issued by the U.S. Treasury Department and the U.S. Internal Revenue Service in April 2018 ("the notice"). In accordance with the measurement period provisions under SAB 118 and the guidance in the notice the Company reinstated the deferred tax asset related to its U.S. interest expense carryforwards under Section 163(j) based on the new U.S. federal tax rate of 21 percent. The impact of the deferred tax asset reinstatement in accordance with SAB 118 was a \$35.9 million increase to the Company's benefit for income taxes and a corresponding decrease to the U.S. group net deferred tax liability position.
- (18) During the three months ended March 31, 2017, the Company recorded a loss on debt extinguishment of \$0.5 million, comprised the write-off of \$0.4 million in debt discount and deferred financing costs, and an early redemption payment of \$0.1 million.