Media Release

14 February 2017

Actelion announces excellent financial results for 2016

ALLSCHWIL/BASEL, SWITZERLAND – 14 February 2017 – Actelion Ltd (SIX: ATLN) today announced its results for the full year 2016.

FINANCIAL HIGHLIGHTS

- Sales growing to CHF 2,412 million (+15% at CER)
- Opsumit sales continue strong trajectory and grow to CHF 831 million (+57% at CER)
- Uptravi sales reach CHF 245 million in first year of launch – driven by the US
- US GAAP operating income grows to CHF 789 million (+14% at CER)
- Core operating income grows to CHF 992 million (+17% at CER)

USD 30 billion proposal by Johnson & Johnson to acquire Actelion

PROPOSED TRANSACTION HIGHLIGHTS

- Actelion to be acquired by Johnson & Johnson for $ 30 billion with spin-out of new R&D company, listed on Swiss stock exchange
- Actelion shareholders to receive 280 US dollars per Actelion share in all-cash tender offer and one share of new R&D company for each Actelion share as stock dividend
- Johnson & Johnson to acquire Actelion's marketed products in particular its leading PAH franchise
- Johnson & Johnson to also acquire global rights to Actelion's promising advanced late-stage therapies, ponesimod and cadazolid
- New R&D company launching with cash of CHF 1 billion to continue the culture of innovation with early stage R&D pipeline
- Johnson & Johnson will also receive an option on an endothelin receptor antagonist (ACT-132577) currently being developed for resistant hypertension

FINANCIAL OVERVIEW

in CHF million (except for per share data)	FY 2016	FY 2015	% variance in CHF	% variance at CER(1)
US GAAP results				
Net revenue	**2,418**	2,045	18	15
Operating income	**789**	656	20	14
Net income	**696**	552	26	19
Diluted EPS	**6.46**	4.91	32	25
Core performance(2)				
Product sales	**2,412**	2,042	18	15
Core operating income	**992**	814	22	17
Core net income	**881**	693	27	22
Core diluted EPS	**8.18**	6.16	33	27

Cash flow	FY 2016	FY 2015
Operating cash flow	920	658
Capital expenditure	(57)	(44)
Free cash flow	90	(800)
Net cash position as of 31 December	495	405

(1) CER percentage changes are calculated by reconsolidating both the 2015 and 2016 results at constant currencies (the average monthly exchange rates for 2015).

(2) Actelion continues to measure and report core operating performance, which management believes more accurately reflects the underlying business performance. The Group believes that these non-GAAP financial measurements provide useful supplementary information to investors. These non-GAAP measures are reported in addition to, not as a substitute for, US GAAP financial performance.

Jean-Paul Clozel, MD, Chief Executive Officer, commented: "With Johnson & Johnson's proposed acquisition of Actelion and the spin-out of a new R&D company, we have created unprecedented value for all of our stakeholders. Our current PAH portfolio and our late-stage pipeline will have expanded potential as part of Johnson & Johnson. With the creation of a new R&D company we also have the opportunity to realize the value potential we have created with our discovery engine and early-stage pipeline. I am very proud of what we've achieved, and I am very excited about the challenges and opportunities ahead."

Otto Schwarz, Chief Operating Officer, commented: "The significant clinical utility of Opsumit resulted in continued strong patient uptake with more than 21,000 patients currently receiving therapy. Moreover, after just one year on the US market, we can say that the Uptravi launch has been very successful by any standards, proving the high unmet medical need for oral prostacyclin therapy and validating our commercial strategy. I strongly believe that, as a part of the Johnson & Johnson family of companies, we will be

able to serve even more patients by opening new markets and creating additional opportunities for our products."

André C. Muller, Chief Financial Officer, commented: "Actelion's 2016 performance has been impressive with the company delivering record sales and earnings. The proposed transaction with Johnson & Johnson announced on 26 January will enable Actelion shareholders to not only monetize their holdings at 280 US dollars per share but also retain future upside potential with the distribution of 1 share in the newly created R&D company for every Actelion share. Both companies are now working to finalize the operational and financial details of the split and prepare the listing of the new company."

SALES UPDATE

Actelion's excellent commercial performance during 2016 was driven by the outstanding Uptravi launch in the US and Opsumit's sustained strong growth trajectory. During the fourth quarter of 2016, combined sales of the company's outcome-based PAH portfolio – Opsumit, Uptravi and Veletri – reached 55% of total sales, demonstrating the significant progress made in the fundamental transformation of the PAH business.

In the US, sales increased by 25% at CER, driven by the strong Uptravi launch, the continued Opsumit momentum due to share gains in an expanding ERA market. European sales were 1% higher compared to 2015. A strong Opsumit performance and solid Tracleer use in the digital ulcer indication were impacted by continued pricing pressure and market erosion from bosentan generics, particularly in Spain. Sales in Japan increased by 19% at CER, driven by very strong sales of Opsumit (launched in June 2015), Tracleer momentum in the digital ulcer indication and Zavesca (Japanese trade name Brazaves).

Comparing average exchange rates for 2016 to 2015, the Swiss franc weakened, mostly against the US dollar, euro and Japanese yen, resulting in a positive currency variance of 63 million Swiss francs.

Sales by product – FY2016

			% variance	
in CHF millions	**FY 2016**	**FY 2015**	**in CHF**	**at CER**
Opsumit®	831	516	61	57
Tracleer®	1,020	1,212	-16	-18
Uptravi®	245	-	nm	nm
Veletri®	97	83	17	12
Ventavis®	73	105	-30	-32
Valchlor®	35	27	30	27
Zavesca®	104	92	13	12
Others	8	7	7	8
Total product sales	**2,412**	**2,042**	**18**	**15**

*nm = not meaningful

Sales by product – Q4 2016

			% variance	
in CHF millions	**Q4 2016**	**Q4 2015**	**in CHF**	**at CER**
Opsumit®	235	162	45	43
Tracleer®	229	278	-17	-19
Uptravi®	85	-	nm	nm
Veletri®	26	23	12	9
Ventavis®	15	24	-37	-38
Valchlor®	10	8	20	19
Zavesca®	26	24	10	10
Others	2	2	-6	-7
Total product sales	**627**	**519**	**21**	**19**

Sales by region – FY 2016

			% variance	
in CHF millions	**FY 2016**	**FY 2015**	**in CHF**	**at CER**
United States	1,306	1,026	27	25
Europe*	646	634	2	1
Japan	258	190	36	19
Rest of the world	201	192	5	6
Total product sales	**2,412**	**2,042**	**18**	**15**

*Europe = EU28 and Switzerland

Sales by region – Q4 2016

			% variance	
in CHF millions	**Q4 2016**	**Q4 2015**	**in CHF**	**at CER**
United States	342	259	32	30
Europe*	162	159	2	4
Japan	77	58	31	17
Rest of the world	47	42	10	9
Total product sales	**627**	**519**	**21**	**19**

*Europe = EU28 and Switzerland

PAH FRANCHISE

Opsumit®

Sales of Opsumit (macitentan) amounted to 831 million Swiss francs for 2016, an increase of 57% at CER compared to 2015. The strong growth across all regions and all relevant markets (Opsumit is now available in almost 40 markets) was driven by solid quarterly increases in the number of patients treated in an expanding ERA market due to increased use in combination with PDE-5 inhibitors, and some upgrades from Tracleer, notably in Japan.

Uptravi®

Sales of Uptravi (selexipag) amounted to 245 million Swiss francs for 2016. Since the US launch at the beginning of January 2016, patient demand has continued to increase with sales of 232 million Swiss francs (which includes 30 million Swiss francs for the build-up of inventory in the US). For the fourth quarter, US

sales amounted to 77 million Swiss francs, compared to 66 million Swiss francs in the third quarter, 45 million Swiss francs in the second quarter and 15 million Swiss francs in the first quarter of 2016. In other geographies, Uptravi sales were driven by the particularly successful launch in Germany. Uptravi is also available in several other markets; it was most recently launched with full reimbursement in the Netherlands and Switzerland.

At the end of 2016, just over 2,400 patients were being treated with Uptravi globally, with more than 1,900 patients coming from the US.

Tracleer®

Sales of Tracleer (bosentan) amounted to 1,020 million Swiss francs for 2016, a decrease of 18% at CER compared to 2015. This was driven to a large extent by volume erosion resulting from the significant impact of Opsumit uptake on the Tracleer patient base and by increased generic competition, notably in Spain, where generic bosentan entered the market in January 2016. Tracleer sales were supported by the digital ulcer indication in Europe and Japan.

Following the Pediatric Investigation Plan (PIP) compliance statement from the European Committee for Medicinal Products for Human Use (CHMP), applications for extension of the Supplementary Protection Certificate (SPC) were granted in all possible 19 EU countries until the end of August 2017.

Veletri®

Sales of Veletri (epoprostenol for injection) amounted to 97 million Swiss francs for 2016, an increase of 12% at CER compared to 2015. This increase was mostly driven by France, Italy, Spain and the UK. Demand in Japan, where it is marketed as Epoprostenol ACT, remained strong, however sales growth was mitigated by a 12% price cut effective March 1, 2016. In February 2017, Actelion Pharmaceuticals Japan proudly announced that Epoprostenol ACT received a label extension for dosage and administration in pediatric patients with PAH.

Ventavis®

Sales of Ventavis (iloprost) amounted to 73 million Swiss francs for 2016, a decrease of 32% at CER compared to 2015 due to competitive pressures, including the availability of Uptravi. Underlying units decreased by 37%.

SPECIALTY PRODUCTS

Valchlor®

Sales of Valchlor (mechlorethamine) amounted to 35 million Swiss francs for 2016, an increase of 27% at CER compared to 2015. In the US, the company has made good progress in establishing Valchlor as a valuable option in the treatment algorithm for early-stage mycosis fungoides, a type of Cutaneous T-Cell Lymphoma (MF-CTCL).

In December 2016, the Committee for Medicinal Products for Human Use (CHMP), the scientific committee of the European Medicines Agency (EMA), issued a positive opinion for the use of chlormethine gel 160 micrograms/g (Ledaga®) for the treatment of mycosis fungoides-type cutaneous T-cell lymphoma (MF-CTCL)

in adult patients and recommended that the European Commission approves the product. The European Commission is expected to issue a final decision by the end of February 2017.

Zavesca®

Sales of Zavesca (miglustat) amounted to 104 million Swiss francs for 2016, an increase of 12% at CER compared to 2015.

Sales in the US were strong, due to a relatively low prior year base as a consequence of an inventory adjustment. In Europe, sales were flat due to the launch of generic miglustat (for the type 1 Gaucher disease indication only), which mitigated the continued strong, double-digit growth in the Niemann-Pick type C (NP-C) indication. Globally, the number of patients receiving Zavesca grew by 6%, compared to 2015, which was driven by a 13% increase in the treatment of patients with NP-C.

CORE R&D EXPENDITURE

The excellent commercial performance enabled Actelion to advance both the late and earlier stage pipeline, resulting in increased R&D expenditure which translates into a ratio of R&D core operating expenses to sales of 21%, slightly higher than in 2015. Core R&D expenses amounted to 509 million Swiss francs, an increase of 25% at CER. This increase was driven by higher clinical trial expenses, mainly driven by the strong recruitment in the Phase III OPTIMUM study (ponesimod in multiple sclerosis; announced in April 2015) and the Phase III IMPACT study (Cadazolid in *Clostridium difficile* associated diarrhea), as well as costs related to the preparation and initiation of Phase II studies for Actelion's new ERA in specialty cardiovascular disorders and DORA in insomnia.

CORE OPERATING INCOME

Core operating income amounted to 992 million Swiss francs, an increase of 17% at CER.

CORE EPS

Diluted core earnings per share were CHF 8.18 for the full year 2016, an increase of 27% at CER compared to the same period of 2015.

DELIVERING VALUE TO SHAREHOLDERS

In-keeping with its commitment to maximizing shareholder value, Actelion returned 428 million Swiss francs to shareholders through the second-line share buyback as well as the increased dividend of CHF 1.50 per share, paid in May 2016. Actelion's shares performed strongly throughout 2016 regardless of the extraordinary volatility created by the strategic discussions initiated in late November. The unaffected share price performance up until the strategic discussions became public was an increase of approximately 15%. At the end of the year, Actelion's stock traded at 220.5 Swiss francs per share, an increase of 58% for the calendar year. The resulting total shareholder return (TSR) for 2016 amounted to 59%.

On 26 January 2017, Actelion and Johnson & Johnson jointly announced that they have entered into a definitive transaction agreement under which Johnson & Johnson will launch an all-cash tender offer in Switzerland to acquire all of the outstanding shares of Actelion for 280 US dollars per share. Additionally, Actelion shareholders will receive one share of a newly created R&D company that will be spun out concurrently with the closing of the proposed transaction.

CLINICAL DEVELOPMENT PIPELINE

The pipeline continued to strengthen with substantial progress made with several compounds:

- The ongoing Phase III program IMPACT investigating cadazolid treatment in patients suffering from *Clostridium difficile*-associated diarrhea is progressing well; results are expected in the first half of 2017.
- In the third quarter of 2016, Actelion announced the initiation of the Phase III POINT study, which investigates the use of combination therapy with ponesimod, an orally active, selective sphingosine-1-phosphate receptor 1 ($S1P_1$) immunomodulator, and dimethyl fumarate (Tecfidera®) for patients with relapsing multiple sclerosis (RMS). The POINT study - which will be conducted under a Special Protocol Assessment (SPA) agreement with the FDA - is the first to assess the concurrent administration of two oral therapies in MS with the objective to improve disease control in this progressive, debilitating neurological disorder. Ponesimod is also being studied in the Phase III OPTIMUM study to compare the efficacy and safety of ponesimod with teriflunomide (Aubagio®) in patients with RMS. The study is making good progress, with enrollment expected to be complete in Q1 2017.
- Also in the third quarter of 2016, the company advanced its new dual orexin receptor antagonist (DORA) into Phase II development in patients with insomnia. The Phase II program consists of two studies, one in adult and one in elderly patients, and is designed to evaluate the effect of Actelion's DORA versus placebo on sleep maintenance and sleep initiation, as well as next-day residual effect and next-day performance. The study in adults also includes a zolpidem reference arm. The decision to move into a Phase II program was based on excellent data collected from the preclinical and Phase I clinical program, as well as a thorough understanding of the potential of dual orexin receptor antagonism on sleep efficacy and architecture.
- With macitentan (Opsumit), the company is conducting a pediatric study, TOMORROW, to evaluate the effect of macitentan on delaying disease progression in children with PAH using a pediatric formulation of macitentan. Recruitment is expected to start in Q1 2017.
- Actelion will assess the efficacy and safety of macitentan in stable Fontan-palliated adolescents and adults in the Phase III study RUBATO. The primary objective of this study is to assess the effect of macitentan as compared to placebo on exercise capacity through cardiopulmonary exercise testing (peak VO_2). The duration of the study is expected to be approximately 28 months; the start is planned for mid-2017.
- A Phase II study with macitentan, MERIT, assessed the efficacy, safety and tolerability of macitentan in patients with inoperable chronic thromboembolic pulmonary hypertension (CTEPH). The study was completed in September 2016 and delivered very positive results, meeting its primary endpoint of a significant reduction in pulmonary vascular resistance (PVR) with macitentan compared with placebo, and also showing a significant positive effect on exercise capacity for macitentan over placebo.
- A Phase III study with macitentan, MAESTRO, assessed the effects of macitentan on exercise capacity in patients with Eisenmenger Syndrome. The study was completed in January 2017, but did not meet the primary endpoint of significantly improving exercise capacity with macitentan compared with placebo.
- Lucerastat is being evaluated for the treatment of Fabry disease. In an initial Phase Ib study, patients receiving enzyme replacement therapy who were treated with lucerastat demonstrated a marked decrease in the accumulation of metabolic substrates thought to be responsible for the lesions characteristic of this disease. Actelion is currently in discussions with health authorities to move directly to Phase III.

	Compound	Indication	Study	Status
Phase III	**Cadazolid**[1]	*Clostridium difficile*-associated diarrhea	IMPACT	Ongoing
	Macitentan[1]	Pediatric PAH	TOMORROW	Initiating
	Macitentan[1]	Portopulmonary hypertension (PoPH)	PORTICO	Ongoing
	Macitentan[1]	Fontan-palliated patients	RUBATO	Initiating
	Ponesimod[1]	Multiple sclerosis	OPTIMUM	Ongoing
	Ponesimod[1]	Multiple sclerosis	POINT	Ongoing
Phase II	**Cenerimod**[2]	Systemic lupus erythematosus	-	Ongoing
	Clazosentan[2]	Reversal of vasospasm associated with aneurysmal subarachnoid hemorrhage	REVERSE	Ongoing
	Dual Orexin Receptor Antagonist[2]	Insomnia	-	Ongoing
	Endothelin Receptor Antagonist (ACT-132577)[2]	Specialty cardiovascular disorders	-	Ongoing
	Macitentan[1]	Chronic thromboembolic pulmonary hypertension	MERIT	Complete
Phase Ib	**Lucerastat**[2]	Fabry disease	-	Complete
Phase I	**New Chemical Entity**[2]	Cardiovascular disorders	-	Ongoing
	New Chemical Entity[2]	Inflammatory disorders	-	Ongoing
	Selective Orexin 1 Receptor Antagonist[2]	Neurological disorders	-	Ongoing
	T-type Calcium Channel Blocker[2]	Neurological disorders	-	Ongoing

[1] Upon completion of the proposed transaction these assets would be developed by Johnson & Johnson
[2] Upon completion of the proposed transaction these assets would be developed by the new R&D company

HUMAN RESOURCES

At the end of 2016, Actelion employed 2,624 permanent employees worldwide, an increase of 3% (or 77 permanent positions) compared to the end of 2015.

ANNUAL REPORT

Full details on the progress made in 2016 are available in Actelion's 2016 Annual Report, at www.actelion.com/annual-report.

NOTES TO SHAREHOLDERS:

The next General Meeting of Shareholders will take place on or around 05 April, 2017, the date will be confirmed with the publication of the offer prospectus by Johnson & Johnson on or around 16 February 2017.

In light of the expected completion of the proposed transaction with Johnson & Johnson, the Board of Directors will propose to carry forward the 2016 accumulated profit and therefore not distribute any cash dividend.

RESULTS DAY CENTER

Investor community: To make your job easier, we provide links to all relevant documentation, such as a full financial review, reconciliation US-GAAP to Core results and geographical breakdown by product, from the Results Day Center on our corporate website: www.actelion.com/results-day-center.

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NOTES TO THE EDITOR

ABOUT ACTELION LTD.

Actelion Ltd. is a leading biopharmaceutical company focused on the discovery, development and commercialization of innovative drugs for diseases with significant unmet medical needs.

Actelion is a leader in the field of pulmonary arterial hypertension (PAH). Our portfolio of PAH treatments covers the spectrum of disease, from WHO Functional Class (FC) II through to FC IV, with oral, inhaled and intravenous medications. Although not available in all countries, Actelion has treatments approved by health authorities for a number of specialist diseases including Type 1 Gaucher disease, Niemann-Pick type C disease, Digital Ulcers in patients suffering from systemic sclerosis, and mycosis fungoides type cutaneous T-cell lymphoma.

Founded in late 1997, with now over 2,600 dedicated professionals covering all key markets around the world including Europe, the US, Japan, China, Russia and Mexico, Actelion has its corporate headquarters in Allschwil / Basel, Switzerland.

Actelion shares are traded on the SIX Swiss Exchange (ticker symbol: ATLN) as part of the Swiss blue-chip index SMI (Swiss Market Index SMI®).

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The above information contains certain "forward-looking statements", relating to the company's business, which can be identified by the use of forward-looking terminology such as "estimates", "believes", "expects", "may", "are expected to", "will", "will continue", "should", "would be", "seeks", "pending" or "anticipates" or similar expressions, or by discussions of strategy, plans or intentions. Such statements include descriptions of the company's investment and research and development programs and anticipated expenditures in connection therewith, descriptions of new products expected to be introduced by the company and anticipated customer demand for such products and products in the company's existing portfolio. Such statements reflect the current views of the company with respect to future events and are subject to certain risks, uncertainties and assumptions. Many factors could cause the actual results, performance or achievements of the company to be materially different from any future results, performances or achievements that may be expressed or implied by such forward-looking statements. Should one or more of these risks or uncertainties materialize, or should underlying assumptions prove incorrect, actual results may vary materially from those described herein as anticipated, believed, estimated or expected.