Actelion Ltd. is a leading biopharmaceutical company focused on the discovery, development and commercialization of innovative drugs for diseases with significant unmet medical needs. The company has its corporate headquarters in Allschwil/ Basel, Switzerland where it was founded in 1997. Its shares have been listed on the SIX Swiss Exchange (ticker symbol ALTN) since 2000. In September 2008, Actelion shares began trading as part of the blue-chip SMI® (Swiss Market Index).

The company has proven its ability to discover new compounds and to rapidly move them from research through development to commercialization. In particular, Actelion scientists were among the first to work in the field of endothelin receptor antagonists (ERA), leading to Tracleer® and now the tailored ERA Opsumit®.

Actelion has over 30 operative affiliates around the world including the United States, Canada, Brazil, Australia, Japan, Switzerland and a number of EU countries. These subsidiaries provide distribution, sales and marketing services.
FINANCIAL OVERVIEW

<table>
<thead>
<tr>
<th></th>
<th>FY Results 2016</th>
<th>FY Results 2015</th>
<th>% Variance in CHF</th>
<th>% Variance at CER*</th>
</tr>
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<tbody>
<tr>
<td>Product sales</td>
<td>2,412</td>
<td>2,042</td>
<td>18</td>
<td>15</td>
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<tr>
<td>Core operating expenses</td>
<td>1,420</td>
<td>1,288</td>
<td>16</td>
<td>14</td>
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<tr>
<td>Core operating income</td>
<td>992</td>
<td>814</td>
<td>22</td>
<td>17</td>
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<tr>
<td>Diluted Core EPS</td>
<td>8.18</td>
<td>6.16</td>
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<tr>
<td>Diluted US GAAP EPS</td>
<td>6.46</td>
<td>4.91</td>
<td>32</td>
<td>25</td>
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<tr>
<td>No of shares in calculation (million)</td>
<td>107.8</td>
<td>112.5</td>
<td>-</td>
<td>-</td>
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</table>

The full financial statements can be found on www.actelion.com.

COMPANY STRATEGY
Since its founding more than fifteen years ago, Actelion has become a new kind of biopharmaceutical company: one that blends biotech’s innovation, speed and flexibility with big pharma’s operating discipline and excellence in execution. With the intrinsic belief that innovation in all domains is the key to growth, Actelion has built a promising pipeline, seven approved products, and commercial operations in over 30 countries. Actelion’s more than 2,600 employees have a common purpose: to improve patients’ lives by creating innovative medicines that make a real difference.

**Driving growth**
Our strategy is built on four principles:

- **Drive innovation forward.** Pursue top quality science, internally and externally, balanced with medical need and commercial potential.
- **Leverage our global presence.** Expand innovative commercial capabilities to new customers and regions. Manage alliances, putting the product first.
- **Maximize the value of innovation.** Develop projects ourselves and seek partners or out-license when necessary to maximize value.
- **Insist on the highest quality in all we do.** Quality is crucial and needs to be ingrained across all functions.

EMployees

<table>
<thead>
<tr>
<th>Employees Actelion Group (December 2016)</th>
<th>Total: 2,624</th>
</tr>
</thead>
<tbody>
<tr>
<td>Drug Discovery</td>
<td>1,443</td>
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<tr>
<td>Corporate Functions</td>
<td>341</td>
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<tr>
<td>Clinical Development</td>
<td>388</td>
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<tr>
<td>Marketing &amp; Sales</td>
<td>452</td>
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</table>
MARKETED PRODUCTS

Our PAH Franchise
Pulmonary arterial hypertension (PAH) is a chronic, life-threatening disorder characterized by abnormally high blood pressure in the arteries between the heart and lungs of an affected individual.

Actelion’s PAH franchise encompasses oral, inhaled and intravenous formulations of compounds, for patients at various stages in the course of this disease (PAH Functional Classes II–IV), enabling us to deliver treatments across the entire continuum of care.

Opsumit®
Opsumit (macitentan), an orally available endothelin receptor antagonist, resulted from a tailored drug discovery process in Actelion’s laboratories.

Opsumit is commercially available in over 45 markets, including the US (since November 2013), Germany (since January 2014) and Japan (since June 2015). The registration process for other countries is ongoing.

Tracleer®
Tracleer (bosentan), an orally available endothelin receptor antagonist, was the first oral treatment approved for PAH.

Tracleer is commercially available in over 60 markets, including the US [since November 2001], the European Union [since May 2002], and Japan [since April 2005].

In addition to the indication in PAH, Tracleer is approved in the EU for the reduction in the number of new digital ulcers in patients suffering from systemic sclerosis and ongoing digital ulcer disease.

Uptravi®
Uptravi (selexipag), originally discovered and synthesized by Nippon Shinyaku, is the only approved oral, selective IP receptor agonist targeting the prostacyclin pathway in PAH.

Uptravi is commercially available in 9 countries including the US [since January 2016] and Germany [since June 2016]. Market authorization has been received in Australia, Canada, the European Union, Japan, New Zealand, South Korea, Switzerland and the US. The registration process for other countries is ongoing.

Veletri®
Veletri (epoprostenol for injection), an intravenous prostacyclin, is stable at room temperature (77°F/25°C) for up to 24 hours, removing the need for patients to carry ice packs.

Veletri is commercially available in 17 markets, including the US [since 2010], Switzerland and Canada, marketed as Caripul® [since 2012], Japan, marketed as Epoprostenol “ACT”, and some European markets [since 2013]. The registration process for other countries is ongoing.

Ventavis®
Ventavis (iloprost), an inhaled formulation of iloprost, is a synthetic compound structurally similar to prostacyclin (PGI2).


More information on our products can be found in Actelion’s Marketed Products fact sheet.
Our Specialty Products
Actelion is creating specialty franchises alongside PAH – discovering, developing and/or in-licensing/acquiring products in new therapeutic areas.

Valchlor®
Valchlor (mechlorethamine) 0.016% gel is applied topically once daily to affected areas of the skin. Valchlor is currently only available in the US and is approved for the topical treatment of Stage IA and IB mycosis fungoides-type cutaneous T-cell lymphoma in patients who have received prior skin-directed therapy.

Valchlor is commercially available in the US (since November 2013) and Israel (since April 2016).

Zavesca®
Zavesca (miglustat) available as oral capsules, is a glucosylceramide synthase inhibitor indicated as monotherapy for the treatment of adult patients with mild to moderate type I Gaucher disease (GD-1) for whom enzyme replacement therapy is not a therapeutic option.

Zavesca is commercially available for the treatment of GD-1 in 47 countries, including the US and the European Union (since 2003).

In the European Union, Zavesca is also indicated for the treatment of progressive neurological manifestations in adult patients and paediatric patients with Niemann-Pick type C (NP-C) disease, a very rare, invariably progressive and eventually fatal neurodegenerative genetic disorder affecting both children and adults.

Zavesca is commercially available for the treatment of NP-C in 46 countries, including the European Union (since 2009) and Japan, marketed as Brazaves® (since 2012).

More information on our products can be found in Actelion’s Marketed Products fact sheet.
**CLINICAL DEVELOPMENT**
Actelion’s promising development pipeline comprises novel compounds addressing a broad range of diseases, including cardiovascular and immunological disorders as well as central nervous system disorders and infectious disease.

Actelion’s late-stage product candidates include: a novel antibiotic, cadazolid, under investigation for *Clostridium difficile*-associated diarrhea (CDAD) and a S1P1 receptor modulator, ponesimod, investigated in multiple sclerosis.

More information on these and our other development activities can be found in Actelion’s Clinical Development fact sheet.

**DRUG DISCOVERY**
Actelion’s efforts in drug discovery focus on the design, synthesis and optimization of small molecular weight molecules, which are active on molecular target families. This focus allows high productivity in the generation of innovative compounds potentially addressing a wide range of high unmet medical needs.

Initially, the company looked solely at G-protein coupled receptors (GPCRs) and a specific enzyme family known as aspartic proteinases. As the company’s capabilities have expanded, so too have the target platforms, adding anti-infective’s, ion channels and a broad range of soluble enzymes.

More information on our platforms of expertise can be found in Actelion’s Drug Discovery fact sheet.
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<thead>
<tr>
<th>Phase</th>
<th>Compound</th>
<th>Indication</th>
<th>Study</th>
<th>Status</th>
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<tr>
<td>III</td>
<td>Cadazolid</td>
<td><em>Clostridium difficile</em>-associated diarrhea</td>
<td>IMPACT</td>
<td>Ongoing</td>
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<tr>
<td></td>
<td>Macitentan</td>
<td>Pediatric PAH</td>
<td>TOMORROW</td>
<td>Initiating</td>
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<tr>
<td></td>
<td>Macitentan</td>
<td>Portopulmonary hypertension (PoPH)</td>
<td>PORTICO</td>
<td>Ongoing</td>
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<tr>
<td></td>
<td>Macitentan</td>
<td>Fontan-palliated</td>
<td>RUBATO</td>
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<td>Ponesimod</td>
<td>Multiple sclerosis</td>
<td>OPTIMUM</td>
<td>Ongoing</td>
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<td></td>
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<td>Multiple sclerosis</td>
<td>POINT</td>
<td>Ongoing</td>
</tr>
<tr>
<td>II</td>
<td>Cenerimod</td>
<td>Systemic lupus erythematosus</td>
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<td>Ongoing</td>
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<tr>
<td></td>
<td>Clazosentan</td>
<td>Reversal of vasospasm associated with aneurysmal subarachnoid hemorrhage</td>
<td>REVERSE</td>
<td>Ongoing</td>
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<td></td>
<td>Dual Orexin Receptor Antagonist</td>
<td>Insomnia</td>
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<td>Ongoing</td>
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<tr>
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<td>Endothelin Receptor Antagonist</td>
<td>Specialty cardiovascular disorders</td>
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<tr>
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<td>Macitentan</td>
<td>Chronic thromboembolic pulmonary hypertension</td>
<td>MERIT</td>
<td>Complete</td>
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<td>Ib</td>
<td>Lucerastat</td>
<td>Fabry disease</td>
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<td>Complete</td>
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<td>I</td>
<td>New Chemical Entity</td>
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<td>Ongoing</td>
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<td>New Chemical Entity</td>
<td>Inflammatory disorders</td>
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<td>Ongoing</td>
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<td></td>
<td>Selective Orexin 1 Receptor Antagonist</td>
<td>Neurological disorders</td>
<td></td>
<td>Ongoing</td>
</tr>
<tr>
<td></td>
<td>T-type Calcium Channel Blocker</td>
<td>Neurological disorders</td>
<td></td>
<td>Ongoing</td>
</tr>
</tbody>
</table>
ACTELION’S PARTNERSHIPS
Actelion has a dedicated team focused on identifying innovation from external sources that complements our business approach. Once identified, Actelion is rapid, proactive and open in creating benefits for both parties. We commit ourselves to the project we share with our partner, to make the product a global success.

Actelion/ReveraGen
Actelion has obtained an exclusive option to in-license ReveraGen’s lead compound vamorolone for the treatment of Duchenne Muscular Dystrophy at two different stages in its development.

Actelion/Nippon Shinyaku Alliance
Actelion and Nippon Shinyaku entered into an exclusive worldwide alliance in April 2008 to collaborate on selexipag, the first selective oral prostacyclin IP receptor agonist, for patients suffering from pulmonary arterial hypertension (PAH). This compound was originally discovered and synthesized by Nippon Shinyaku.

Actelion/Bayer Schering Pharma AG Alliance
Actelion holds the exclusive US rights for inhaled iloprost, sold under the brand name Ventavis®, the first approved inhaled treatment for pulmonary arterial hypertension (PAH), licensed from Bayer Schering Pharma (through the acquisition of CoTherix Inc.).

COMPANY MILESTONES
2017  Actelion enters into a definitive transaction agreement with Johnson & Johnson
2016  Agreement on in-licensing option for vamorolone from ReveraGen
Initiation of a Phase II program with Actelion’s dual orexin receptor antagonist in insomnia
Initiation of a Phase III program with macitentan in children with PAH
Uptravi is launched in the US and in Germany
Uptravi is approved by the European Medicines Agency for the treatment of PAH
2015  Uptravi is approved by the US FDA for the treatment of PAH
Creation of Vaxxilon - together with the Max Planck Society
Initiation of Phase III program with ponesimod in patients with relapsing multiple sclerosis
2014  Selexipag meets primary endpoint in pivotal Phase III GRIPHON outcome study in patients with PAH
2013  Initiation of Phase III program with cadazolid in patients with Clostridium difficile-associated diarrhea
Valchlor is added to Actelion’s specialty portfolio in the US
Opsumit is approved in the US and the EU and launched in the US for PAH
2012  Macitentan meets primary endpoint in pivotal Phase III SERAPHIN outcome study in patients with pulmonary arterial hypertension
2010  Veletri is launched in the US further strengthening Actelion’s PAH franchise
2009  Tracleer receives EU approval of pediatric formulation for the treatment of PAH
Zavesca receives EU approval for Niemann-Pick type C disease
2008  Tracleer receives EU approval for treatment of patients with mildly symptomatic PAH
Actelion and Nippon Shinyaku enter into a license agreement on novel orally available IP receptor agonist for the treatment of PAH
2007  Tracleer receives EU approval for reduction of number of new digital ulcerations in systemic sclerosis patients
2006  Definitive agreement to acquire US-based CoTherix, Inc. adding Ventavis® to Actelion’s product offerings in the US
2003  First approval of Zavesca® for the treatment of Type 1 Gaucher disease
2001  First approval of Tracleer for the treatment of pulmonary arterial hypertension (PAH)
2000  Initial Public Offering (IPO); Actelion shares are listed on the Swiss New Market Stock Exchange with a record valuation of CHF 1.2 billion
1997  Foundation of Actelion December 17, 1997
Disclaimer: This fact sheet has the sole purpose to provide members of the public with general information about the activities of Actelion Ltd and its associated companies. The forward-looking statements in this fact sheet are based on current expectations and belief of company management, which are subject to numerous risks and uncertainties.

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