ACTELION'S COMPANY PROFILE

On June 16, 2017, Actelion became a member of the Janssen Group of Pharmaceutical Companies of Johnson & Johnson. Actelion is committed to leverage Johnson & Johnson's global presence and commercial strength to accelerate growth and patient access to our important therapies. With its unparalleled portfolio of outcome-based PAH treatments, Actelion will continue to focus on advancing the science and patient care in PAH as well as advance pipeline products in new therapeutic areas to find additional meaningfully differentiated products to benefit patients.

As one of the Janssen Pharmaceutical Companies of Johnson & Johnson, Actelion will maintain its leadership position in this space. Janssen has established Pulmonary Hypertension (PH) as a sixth therapeutic area (TA) of focus, complementing its current TAs in Cardiovascular & Metabolism, Immunology, Infectious Diseases & Vaccines, Neuroscience and Oncology. Actelion is excited to continue its 20-year legacy of ground-breaking innovation in this important disease area.

Actelion also has treatments for other serious illnesses including type 1 Gaucher disease, Niemann-Pick type C disease, digital ulcers in patients suffering from systemic sclerosis, and mycosis fungoides in patients with cutaneous T-cell lymphoma.

Through Janssen's science and patient focus, and geographic and commercial reach, Actelion is looking forward to bringing its in-market medicines to even more patients who need them around the world and accelerating market entry for its promising late-stage development assets.

CONTENTS

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.

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.

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.

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.

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.

Ventavis®

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

Actelion has marketed Ventavis in the US since 2007. Bayer Healthcare markets Ventavis elsewhere.

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).

Ledaga®

Ledaga (chlormethine) is an alkylating drug indicated for the treatment of mycosis fungoides-type cutaneous T-Cell lymphoma (MF-CTCL) formulated as a topical, once-daily, colorless gel. Ledaga is indicated for the topical treatment of mycosis fungoides-type cutaneous T-cell lymphoma (MF type CTCL) in adult patients.

In March 2017, the European Commission has granted marketing authorization for the use of Ledaga (chlormethine gel) 160 micrograms/g for the treatment of mycosis fungoides-type cutaneous T-cell lymphoma (MF-CTCL). Subject to fulfilling the agreed commitments and achieving market access in various countries, a potential first European launch of Ledaga is not expected before H1 2018.

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 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.

DEVELOPMENT PIPELINE – ACTELION'S FOCUS ON HIGH UNMET MEDICAL NEEDS

	Compound	Indication	Study	Status
Phase III	**Cadazolid**	*Clostridium difficile*-associated diarrhea	IMPACT	Complete
	Macitentan	Pediatric PAH	TOMORROW	Initiating
	Macitentan	Portopulmonary hypertension (PoPH)	PORTICO	Ongoing
	Macitentan	Fontan-palliated	RUBATO	Initiating
	Ponesimod	Multiple sclerosis	OPTIMUM	Ongoing
	Ponesimod	Multiple sclerosis	POINT	Ongoing
Phase II	**Macitentan**	Chronic thromboembolic pulmonary hypertension	MERIT	Complete

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 On 16 June Actelion becomes a Janssen Pharmaceutical Company of 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

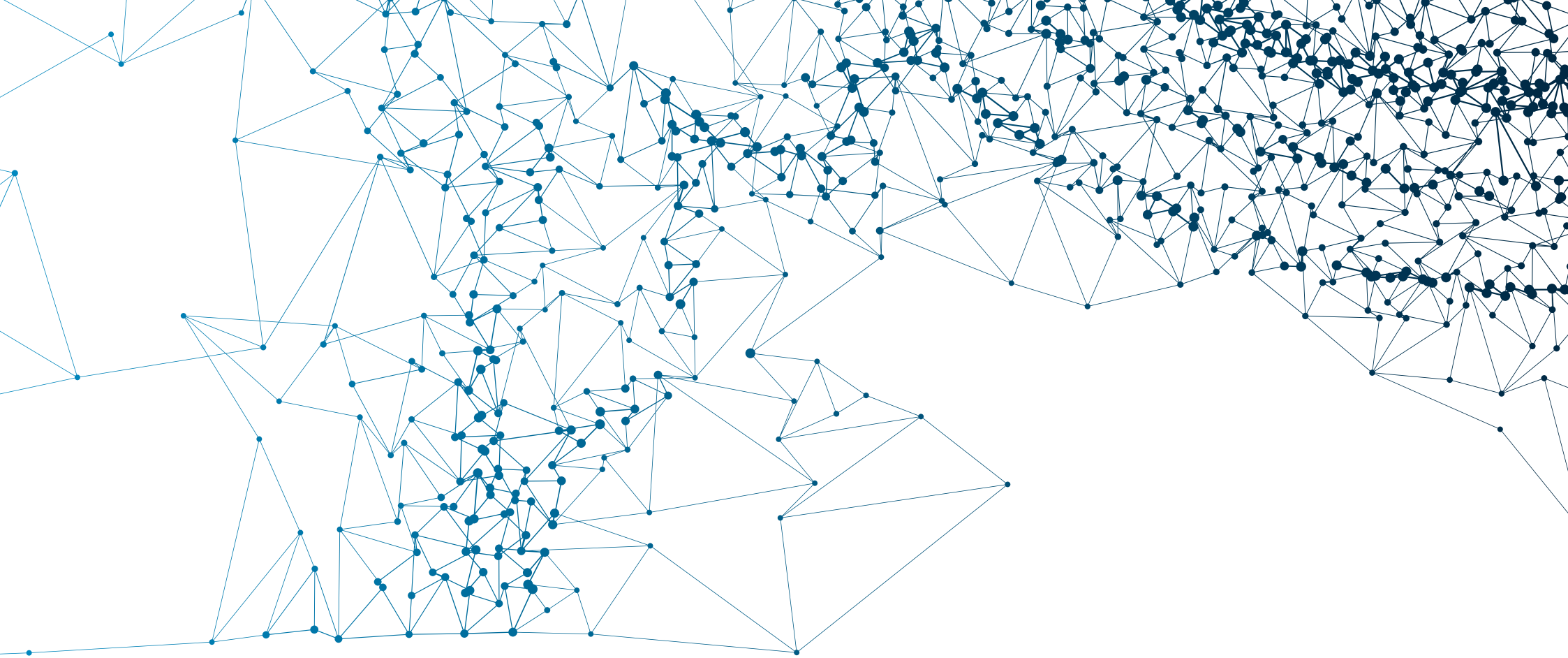
2006 Definitive agreement to acquire US-based CoTherix, Inc. adding Ventavis® to Actelion's product offerings in the US

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



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COMPANY
PROFILE

ACTELION
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OF Johnson&Johnson