Actelion to provide an update on advancements in its cardiovascular pipeline in PAH and beyond

ALLSCHWIL/BASEL, SWITZERLAND – 07 November 2016 – Actelion Ltd (SIX: ATLN) announced that the company will provide an update on its cardiovascular activities today (Monday, 07 November) in an investor conference call and webcast at 14:00 hrs CET.

Jean-Paul Clozel, M.D. and Chief Executive Officer of Actelion commented: “Actelion has changed the way pulmonary arterial hypertension (PAH) is treated – with therapies across the continuum of care that have improved the long-term outcome for patients suffering from this devastating disease. We are fully committed to PAH and take our leadership role with great responsibility as we optimize the way our drugs are used as well as look for potential new PAH therapeutic targets.”

Jean-Paul Clozel continued: “I am pleased to provide an update on the strong cardiovascular pipeline that we have built beyond PAH. The excellent MERIT results announced today highlight one of the ways patients with different forms of pulmonary hypertension (PH) might benefit from Opsumit. With cardiovascular disease remaining the number one cause of death in many countries, as a cardiologist, I am proud that Actelion is committed to finding new cardiovascular therapies.”

The investor conference call and webcast will be hosted by Actelion’s CEO Jean-Paul Clozel, Head of Global Clinical Development, Guy Braunstein and Chief Scientific Officer, Martine Clozel, who will discuss the following topics:

MACITENTAN (OPSUMIT®)
The company will present its efforts to develop macitentan in new PAH patient populations (WHO Classification Group 1) with label-enabling studies in children (TOMORROW), in patients with Eisenmenger Syndrome (MAESTRO), and in patients with portopulmonary hypertension (PoPH) (PORTICO).

The company will also discuss its commitment to post-launch characterization and safety activities in PAH with an observational registry (OPUS study), studies (SYMPHONY & ORCHESTRA) to validate the patient-reported outcome instrument PAH-SYMPACT®, a study (REPAIR) to evaluate the effect of macitentan on right ventricular remodeling and hemodynamic properties in patients with symptomatic PAH.
Actelion’s efforts to expand the clinical utility of macitentan to new pulmonary hypertension (PH) indications will also be outlined. A study (SOPRANO) to assess the efficacy and safety of macitentan in patients with pulmonary hypertension after Left Ventricular Assist Device Implantation will be presented. In addition, the positive results from the MERIT study in patients with inoperable chronic thromboembolic pulmonary hypertension (CTEPH; WHO Classification Group 4) are now available and will be presented.

As part of investigating macitentan in new cardiovascular indications beyond PH, the company will introduce a Phase III study, RUBATO. This study will assess the efficacy and safety of macitentan in stable Fontan-palliated adolescent and adult patients. In addition, following full evaluation of the pilot MELODY study in patients with combined pre- and post-capillary pulmonary hypertension (CpcPH), the company believes it has identified a heart failure patient population that could most likely benefit from treatment with an endothelin receptor antagonist (ERA). A study is currently being discussed with health authorities.

**SELEXIPAG (UPTRAVI®)**

Following the outstanding launch momentum of Uptravi, the company will present its measures to expand the clinical utility of this important asset.

Actelion is conducting a study (TRANSIT) to assess the tolerability and safety of the transition from inhaled treprostinil to oral selexipag in adult patients with PAH.

To further advance standard of care in PAH, Actelion is conducting a study (TRITON) to compare the efficacy and safety of an initial triple oral treatment regimen of macitentan together with tadalafil and selexipag versus an initial dual oral treatment regimen in newly diagnosed, treatment-naive patients with PAH.

Working closely with health authorities, the company is in the process of developing a strategy for investigating the use of Uptravi in children with PAH.

In addition, an intravenous (i.v.) formulation of selexipag is being developed for the treatment of patients with PAH who are prescribed oral selexipag and who are temporarily unable to take oral medication.

**BOSENTAN IN PEDIATRIC PAH (TRACLEER®)**

As there is no therapy approved for children with PAH in the US, in August of this year, Actelion submitted an NDA for the pediatric appropriate dispersible tablet formulation of Tracleer to the FDA. The company looks forward to further discussions with FDA on this important program and if approved, will make the pediatric formulation available to treat these young patients.
ACTELION’S NEW DUAL ENDOTHELIN RECEPTOR ANTAGONIST: ACT-132577
The company is currently investigating a new potent dual ERA, ACT-132577, the main active metabolite of macitentan, in a dose-finding study in essential hypertension. Once the optimal dose has been identified, the company will investigate this dual ERA in resistant hypertension as a first indication.

NEW CHEMICAL ENTITY
Actelion also has a new chemical entity for cardiovascular indications which is currently in Phase I development.

CLINICAL DEVELOPMENT PIPELINE UPDATE
In addition to the cardiovascular pipeline which Actelion presents today, the company is also pursuing development of in-house innovation in other therapeutic areas. The programs are progressing as communicated with the exception of ponesimod in graft versus host disease, where the company has decided to stop the current study due to difficulty in enrollment.
### Compound Information

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### Notes to the Editor

**ABOUT PULMONARY ARTERIAL HYPERTENSION (PAH)**

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. The symptoms of PAH are non-specific and can range from mild breathlessness and fatigue during normal daily activity to symptoms of right heart failure and severe restrictions on exercise capacity and ultimately reduced life expectancy. PAH is one group within the classification of pulmonary hypertension (PH). This group includes idiopathic PAH.
heritable PAH and PAH caused by factors which include connective tissue disease, HIV infection and congenital heart disease.

The last decade has seen significant advances in the understanding of the pathophysiology of PAH, which has been paralleled by the development of treatment guidelines and new therapies. Drugs targeting the three pathways that have been established in the pathogenesis of PAH are endothelin receptor antagonists (ERAs), prostacyclin receptor agonists, and phosphodiesterase-5 inhibitors. PAH treatments have transformed the prognosis for PAH patients from symptomatic improvements in exercise tolerance 10 years ago to delayed disease progression today. Improved disease awareness and evidence-based guidelines developed from randomized controlled clinical trial data have highlighted the need for early intervention, goal-oriented treatment and combination therapy. Learn more at http://www.pahuman.com/

ABOUT OPSUMIT® (MACITENTAN)

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

In the US, Opsumit is indicated for the treatment of PAH, WHO Group I to delay disease progression. Disease progression included: death, initiation of intravenous (IV) or subcutaneous prostanoids, or clinical worsening of PAH (decreased 6-minute walk distance, worsened PAH symptoms and need for additional PAH treatment). Opsumit also reduced hospitalization for PAH. Effectiveness was established in the long-term study SERAPHIN in PAH patients with predominantly WHO FC II-III symptoms treated for an average of 2 years. Patients were treated with Opsumit monotherapy or in combination with phosphodiesterase-5 inhibitors or inhaled prostanoids. Patients had idiopathic and heritable PAH (57%), PAH caused by connective tissue disorders (31%), and PAH caused by congenital heart disease with repaired shunts (8%).

In Europe, Opsumit is indicated, as monotherapy or in combination, for the long-term treatment of PAH in adult patients of WHO Functional Class (FC) II to III. Efficacy has been shown in a PAH population including idiopathic and heritable PAH, PAH associated with connective tissue disorders, and PAH associated with corrected simple congenital heart disease.

Opsumit is very likely to cause major birth defects. It is contraindicated for use in pregnancy. In the US, Opsumit is distributed under a risk evaluation and mitigation strategy.

ABOUT THE TOMORROW STUDY WITH MACITENTAN

As pediatric PAH physicians have to mostly rely on research data collected in adults when weighing up treatment options, there is a significant medical need for broadening the pediatric knowledge base and showing the benefit of age-appropriate formulations and adequate doses of PAH-specific medications, in order to provide children with PAH with the most appropriate treatment.

In July 2016, Actelion announced that it will be initiating a Phase III study to evaluate the effect of macitentan on delaying disease progression in children with PAH using a pediatric formulation of macitentan (Opsumit). TOMORROW (pediaTric use Of Macitentan tO delay disease pRogressiOn in PAH Worldwide) is a multicenter, controlled, randomized, open-label event-driven study to assess the efficacy, safety and pharmacokinetics of macitentan versus standard of care in children with PAH. The study will enroll children
between the age of 1 month and 18 years in more than 20 countries and is expected to last up to 6 years with patients remaining in the study until the target number of primary efficacy endpoints is met.

The primary efficacy endpoint is defined as time to the first Clinical Event Committee (CEC) confirmed disease progression event, comprising:

- Death (all causes), or
- Atrial septostomy or Pott's anastomosis, or registration on lung transplant list, or
- Hospitalization due to worsening PAH, or
- Clinical worsening of PAH

Due to the open-label nature of the study, the management of investigational centers as well as data management, statistical analysis and coordination of the CEC will be conducted by a CRO. The primary endpoints of the study will be adjudicated by a blinded CEC, in a similar approach to that used in the Phase III SERAPHIN study, where macitentan was studied in adult patients with PAH. An interim analysis for early efficacy or futility is planned when at least 131 CEC-confirmed first disease progression events (70% information fraction) have occurred. If Actelion completes the study as outlined, the company can apply for the extension of the marketing exclusivity for Opsumit both in the US and the European Union.

ABOUT THE MAESTRO STUDY WITH MACITENTAN
Eisenmenger syndrome represents the most advanced form of pulmonary arterial hypertension in conjunction with congenital heart disease (PAH-CHD). It is characterized by eventual reversal of the initial systemic-to-pulmonary shunt due to the increase in pulmonary vascular resistance coinciding with the development of chronic cyanosis and limited exercise capacity.

MAESTRO (MAcitantan in Eisenmenger Syndrome To RestOre exercise capacity) is a Phase III multi-center, double-blind, randomized, placebo-controlled, parallel-group study to evaluate the effects of macitentan on exercise capacity in patients with Eisenmenger syndrome. This study is fully recruited and results are expected early in 2017.

ABOUT THE PORTICO STUDY WITH MACITENTAN
Portopulmonary hypertension (PoPH) is a life-threatening complication of portal hypertension. It is most commonly observed in patients with portal hypertension due to cirrhosis of the liver. The outcome of liver transplants in the presence of PoPH is poor, creating a medical need for PAH-specific therapies in PoPH with the aim of improving pulmonary hemodynamics in order to allow liver transplants to be successfully performed.

PORTICO (PORtopulmonary Hypertension Treatment with maCitentan — a randOmized Clinical Trial) is a randomized, double-blind, placebo-controlled, prospective, multicenter, parallel group Phase IV study to assess the safety and efficacy of macitentan in patients with portopulmonary hypertension (PoPH). The primary objective of the study is to evaluate the effect of 10 mg macitentan on pulmonary vascular resistance (PVR) as compared to placebo. Secondary objectives include the evaluation of the effect of macitentan as compared to placebo on exercise capacity and WHO functional class, as well as the evaluation of the safety and tolerability of macitentan in patients with PoPH.
ABOUT THE OPUS STUDY WITH MACITENTAN

The ongoing OPUS study (OPsumit Users Registry®) is the largest systematic collection of data from patients newly treated with macitentan. OPUS is being conducted at 150 sites across the US, with over 1,000 patients already enrolled (the first patients were enrolled on April 30, 2014). The objective of the study is to characterize the safety profile of macitentan and to describe clinical characteristics and outcomes of patients newly treated with macitentan in the real-world post-marketing setting. The results to date show that the safety profile of macitentan observed in the real-world setting is consistent with that observed in the clinical study setting, with very low incidence of hepatic and hepatobiliary disorders and no unexpected safety findings.

ABOUT THE PAH-SYMPACT® VALIDATION IN THE SYMPHONY & ORCHESTRA STUDIES WITH MACITENTAN

The Pulmonary Arterial Hypertension Symptoms and Impact (PAH-SYMPACT®) questionnaire is a new patient-reported outcome instrument for PAH that captures symptoms and impacts relevant to the patient population. It is the first tool for PAH patients developed following the process outlined in the FDA’s guidance. The psychometric validation of PAH-SYMPACT has been conducted in two prospective, open label studies in patients with PAH: SYMPHONY (in the US), and ORCHESTRA (in the EU). The objective of the studies is to demonstrate the psychometric characteristics of reliability and construct validity of PAH-SYMPACT.

ABOUT THE REPAIR STUDY WITH MACITENTAN

PAH is characterized by a progressive increase in pulmonary arterial pressure (PAP) and in pulmonary vascular resistance (PVR) potentially leading to right heart failure and death.

The primary objective of REPAIR (Right vEntricular Remodeling in Pulmonary Arterial hypertension), a Phase IV study, is to evaluate the effect of 10 mg macitentan on Right Ventricular Stroke Volume assessed by magnetic resonance imaging (MRI) and on PVR assessed with right heart catheterization (RHC) in patients with symptomatic PAH. Secondary objectives include the evaluation of the safety and tolerability of macitentan in symptomatic PAH patients.

ABOUT THE SOPRANO STUDY WITH MACITENTAN

No medical therapy has been established yet to ameliorate pulmonary hypertension (PH) due to left heart disease. Heart transplantation is the only therapy for stage D heart failure, but concomitant PH is one of the major risk factors after heart transplantation. The implantation of a left ventricular assist device (LVAD) is commonly used as a bridge to transplant. The administration of an endothelin receptor antagonist can significantly decrease pulmonary vascular resistance (PVR) and therefore be useful for patients with LVAD implant and severe PH.

SOPRANO (Macitentan in pulmonary hypertenSiOn Post-left ventRiculAr assist device implantation) is a prospective, multicenter, double-blind, randomized, placebo-controlled, parallel group Phase II study to assess the efficacy and safety of macitentan in patients with pulmonary hypertension after Left Ventricular Assist Device Implantation. The primary objective of the study is to evaluate the effect of 10 mg macitentan on PVR as compared to placebo in patients with PH after LVAD implantation. Secondary objectives include the evaluation of the effect of macitentan as compared to placebo on cardio-pulmonary hemodynamics and disease severity in patients after LVAD implantation.

– Actelion to provide an update on advancements in its cardiovascular pipeline in PAH and beyond –
ABOUT THE MERIT STUDY WITH MACITENTAN
CTEPH is the only potentially curable form of PH (WHO Group 4) with pulmonary thromboendarterectomy (PTE) being the primary treatment. However, PTE is not feasible or curative in all CTEPH patients. Operability is determined by CTEPH type and co-morbidities. It is estimated that a third of CTEPH patients are inoperable.

MERIT (Macitentan in the Treatment of Inoperable chronic Thromboembolic pulmonary hypertension) was a Phase II prospective, randomized, placebo-controlled, double-blind, multi-center, parallel-group study to assess the efficacy, safety and tolerability of 10 mg macitentan in patients with inoperable chronic thromboembolic pulmonary hypertension (CTEPH).

In MERIT, 80 inoperable patients were randomized in a 1:1 ratio into 2 treatment groups (macitentan 10 mg or placebo) over a 24 week treatment period. The study started in August 2014 and was completed in September 2016. Patients with symptomatic PH in WHO Functional Class (FC) III or IV at baseline were allowed to receive PH background therapy throughout the study, including PDE-5 inhibitors or oral/inhaled prostanoids. All patients included into the study underwent independent operability assessment based on local or central adjudication committees.

After 16 weeks the treatment effect was a significant 16% reduction in pulmonary vascular resistance (PVR) with macitentan compared with placebo (95% CL: −30%, −1%; p=0.04 intention-to-treat (ITT)). The efficacy observed was consistent across all sub-groups, including patients receiving background PH specific therapy at baseline (61%), including PDE-5 inhibitors (59%). Mean PVR decreased from baseline in both macitentan and placebo groups (geometric mean percent ratios of Week 16/baseline 73.0% and 87.2%, respectively).

The study also showed a significant positive effect of macitentan compared to placebo on exercise capacity. After 24 weeks of treatment, the mean change in 6-minute walk distance (6-MWD) from baseline was an increase of 35 meters (m) in macitentan and 1 m in placebo. The 6-MWD least-squares mean difference at Week 24 was 34.0 meters between macitentan and placebo (95% CL: 2.9, 65.2 m; p=0.03).

Macitentan was well tolerated in this patient population and safety was in general consistent with the known safety profile for macitentan from previous clinical studies. The most frequently reported adverse events that occurred with higher frequency on macitentan vs. placebo were peripheral edema (22.5% vs. 10.0%) and events related to anemia (17.5% vs. 2.5%). Hemoglobin decreases were observed in both macitentan and placebo groups and in only one subject in each group hemoglobin values decreased below 100 g/L during the study.

ABOUT THE RUBATO STUDY WITH MACITENTAN
The Fontan procedure is a life-saving surgical procedure to treat children born with complex heart defects. By connecting the venae cavae directly to the pulmonary artery, it diverts venous blood to the lungs without passing through the absent right ventricle. The Fontan procedure stops cyanosis and improves effort tolerance and survival – however, when patients reach adolescence, there is a decline in exercise capacity that accelerates, with the risk of poor functional outcome in the long-term.
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 prospective, multi-center, double-blind, placebo-controlled parallel group study is to assess the effect of macitentan 10 mg as compared to placebo on exercise capacity through cardiopulmonary exercise testing (peak VO₂). Secondary objectives include the evaluation of the safety and tolerability of macitentan as compared to placebo in stable Fontan-palliated patients. The duration of the study is expected to be approximately 28 months; the start is planned for mid-2017.

ABOUT ACT-132577 IN RESISTANT HYPERTENSION
Actelion's latest dual ERA ACT-132577 is being evaluated in a Phase II prospective, multi-center, double-blind, double-dummy, randomized, placebo- and active reference, parallel group, dose-finding study in patients with essential hypertension (grade 1 and 2) to establish a dose-effect relationship. The results from this study will form the basis for development decisions in specialty cardiovascular disorders. Patients will be randomized in the 6 groups in a 1:1:1:1:1:1 ratio: placebo; dose 1, dose 2, dose 3, dose 4 of Actelion's ERA; and lisinopril 20 mg.

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

In the US, Uptravi is indicated for the treatment of PAH (WHO Group I) to delay disease progression and reduce the risk of hospitalization for PAH. Effectiveness was established in a long-term study in PAH patients with WHO Functional Class II-III symptoms. Patients had idiopathic and heritable PAH (58%), PAH associated with connective tissue disease (29%), and PAH associated with congenital heart disease with repaired shunts (10%).

In Europe, Uptravi is indicated for the long-term treatment of PAH in adult patients with WHO functional class II-III, either as combination therapy in patients insufficiently controlled with an endothelin receptor antagonist (ERA) and/or a phosphodiesterase type 5 (PDE-5) inhibitor, or as monotherapy in patients who are not candidates for these therapies. Efficacy has been shown in a PAH population including idiopathic and heritable PAH, PAH associated with connective tissue disorders, and PAH associated with corrected simple congenital heart disease.

As with other therapies targeting the prostacyclin pathway, hyperthyroidism has been observed with Uptravi. If there are any signs of pulmonary edema, the possibility of pulmonary veno-occlusive disease should be considered and, if confirmed, Uptravi should be discontinued. Other adverse events observed with Uptravi usage were similar in nature to those expected with prostacyclin receptor agonists.

ABOUT THE TRANSIT STUDY WITH SELEXIPAG
Due to the burden of inhaled therapies targeting the prostacyclin pathway, patients and physicians may consider a transition from inhaled therapy to oral therapy. With the Phase IIIb study TRANSIT (TRANSITion from inhaled treprostinil to oral selexipag in adult patients with pulmonary arterial hypertension), Actelion is aiming at providing guidance on this transition. TRANSIT assesses the tolerability and the safety of the
transition from inhaled treprostinil to oral selexipag in adult patients with PAH. During the study, the treatment with inhaled treprostinil is tapered off and simultaneously replaced with selexipag. 34 patients at 12 US sites have been enrolled in the study, which is expected to be completed before the end of 2016.

ABOUT THE TRITON STUDY WITH MACITENTAN & SELEXIPAG

The objective of the Phase III TRITON study is to compare the efficacy and safety of an initial triple oral treatment regimen versus an initial dual oral treatment regimen in newly diagnosed, treatment-naïve patients with PAH. The triple oral combination treatment arm comprises macitentan, tadalafil and selexipag; the dual oral combination treatment arm macitentan, tadalafil and placebo. The primary objective of the study is to evaluate the effect of the triple oral combination treatment on pulmonary vascular resistance (PVR) as compared to the dual oral treatment regimen. Secondary objectives include the evaluation of the triple oral combination treatment as compared to dual oral treatment regimen on exercise capacity (6MWD), disease severity and progression, and safety and tolerability.

ABOUT I.V. SELEXIPAG

Intravenous (i.v.) selexipag is being developed for the treatment of patients with PAH who are currently prescribed oral selexipag and who are temporarily unable to take oral medication. In a single pivotal study that is expected to enroll approximately 20 patients, the tolerability and safety of i.v. selexipag will be assessed in a study design of 1 week oral selexipag – 3 doses i.v. selexipag (administered in hospital) – 1 week oral selexipag. The study is expected to start in the first half of 2017.

References

For a full list of references please see our key literature list available here.

INVESTOR CONFERENCE CALL / WEBCAST

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Conference Call Connect #: Dial-in participants should start calling the number below 10-15 minutes before the conference is due to start.

Dial: Europe: +41 (0)44 583 18 01

UK: +44 (0)203 009 24 60

US: +1 855 228 38 74

Participant's mode: Listen-Only with possibility to open individual lines during Q&A session. Participants will be asked for their name and company.

Webcast Access: Webcast participants should go to the Actelion website http://www.actelion.com 10-15 minutes before the conference is due to start.

Webcast Replay: The archived Investor Webcast will be available for replay through http://www.actelion.com/approximately 60 minutes after the call has ended.

– Actelion to provide an update on advancements in its cardiovascular pipeline in PAH and beyond –
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 2,500 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®). All trademarks are legally protected.

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