Media Release

30 April 2012

Actelion's macitentan meets primary endpoint in pivotal Phase III SERAPHIN outcome study in patients with pulmonary arterial hypertension

ALLSCHWIL/BASEL, SWITZERLAND – 30 April 2012 – Actelion (SIX: ATLN) announced today that initial analysis indicates that the pivotal, long-term, event-driven study SERAPHIN with macitentan, a novel dual endothelin receptor antagonist, in 742 patients suffering from pulmonary arterial hypertension (PAH) and treated for up to three and a half years, has met its primary endpoint.

Macitentan, at both the 3 mg and 10 mg dose, decreased the risk of a morbidity/mortality event over the treatment period versus placebo. This risk was reduced by 45 percent in the 10 mg dose group ($p<0.0001$). At 3 mg, the observed risk reduction was 30 percent ($p=0.0108$). Treatment with macitentan in the SERAPHIN study was well tolerated.

Jean-Paul Clozel, M.D. and Chief Executive Officer of Actelion commented: "I am extremely pleased with the outstanding SERAPHIN results. We are committed to working with the Health Authorities to bring this potentially important advancement in PAH to patients as soon as possible. Submission of the registration dossier to Health Authorities worldwide is expected by the fourth quarter of 2012."

Lewis J. Rubin, M.D., Emeritus Professor, University of California, San Diego and Senior Advisor on SERAPHIN commented: "With this well-designed PAH study, Actelion pursued an ambitious goal to focus on outcome benefits as the primary endpoint. The impressive results of this landmark study are setting a new standard in how to conduct studies in this devastating disease."

Gerald Simonneau M.D., Professor of Pneumology and Head of the Department of Pulmonary Disease and Intensive Care Unit, Hospital Antoine Beclere-Clamart, France and Senior Advisor on SERAPHIN commented: "As a physician with more than 30 years experience in the fight against this terrible disease, I am very excited by the outcome of this study. These results represent an important milestone in the history of clinical trials in PAH and show that macitentan has the potential to offer a new treatment paradigm for these patients."

Secondary efficacy endpoints, including change from baseline to month 6 in six-minute walk-distance, change from baseline to month 6 in WHO functional class and time - over the whole treatment period - to either death due to PAH or hospitalization due to PAH, also showed a dose-dependent effect ($p<0.05$ for either dose). A trend in favor of 10 mg macitentan was observed on all-cause mortality (p=ns).

Guy Braunstein, M.D. and Head of Global Clinical Development commented: “Our thanks go to the investigators and their staff in almost 40 countries, who participated in this compelling study. I truly believe that these results with macitentan will translate into clinical benefits for patients suffering from PAH. The Company will now rapidly analyze this largest ever clinical study in PAH in full detail, in view of regulatory filings later this year.”

Full data from this study will be made available through scientific disclosure at upcoming congresses and publications.

About the safety and tolerability in SERAPHIN

The safety set comprised 741 patients (randomized 1:1:1), who received at least one dose of study treatment. Mean exposure to study treatment was 85.3 weeks for placebo patients (n=249), 99.5 weeks for patients on 3 mg (n=250) and 103.9 weeks for patients on 10 mg (n=242).

Macitentan in this patient population was well tolerated. The number of adverse events reported and patients discontinuing treatment due to adverse events was similar across all groups.

Elevations of liver alanine or aspartate aminotransferases greater than three times the upper limit of normal were observed in 4.5 percent of patients receiving placebo, in 3.6 percent of patients on 3 mg of macitentan and in 3.4 percent of patients on 10 mg of macitentan. In addition, no difference was observed between macitentan and placebo on fluid retention (edema).

A decrease in hemoglobin - reported as an adverse event - was observed more frequently on macitentan than placebo, with no difference in treatment discontinuation between groups.

About the SERAPHIN study

SERAPHIN (Study with an Endothelin Receptor Antagonist in Pulmonary arterial Hypertension to Improve cliNical outcome) was the largest randomized, controlled study in PAH patients with a long-term treatment to include a clearly defined morbidity/mortality primary end-point [1]. The pivotal Phase III study was designed to evaluate the efficacy and safety of macitentan – a novel dual endothelin receptor antagonist that resulted from

a tailored drug discovery process – through the primary endpoint of time to first morbidity and all-cause mortality event in patients with symptomatic PAH.

Global enrollment was completed in December 2009 with a total of 742 patients. Patients were randomized 1:1:1 to receive two different doses of macitentan (3 mg and 10 mg once daily) or placebo. Patients were allowed to receive PAH background therapy throughout the study, either PDE-5 inhibitors or oral/inhaled prostanoids. This event-driven study was conducted in 151 centers from almost 40 countries in North and Latin America, Europe, Asia-Pacific and Africa and was completed in the first half of 2012, with 287 patients having an adjudicated event.

About macitentan

Macitentan is a novel dual endothelin receptor antagonist that resulted from a tailored drug discovery process. Macitentan has a number of potentially key beneficial characteristics. i.e. increased *in vivo* preclinical efficacy vs. existing ERAs resulting from sustained receptor binding and tissue penetration properties. A clinical pharmacology program indicated a low propensity of macitentan for drug-drug interactions [2, 3, 4].

About macitentan in other clinical development programs

Macitentan is currently investigated in a pivotal Phase III program in patients with ischemic digital ulcers associated with systemic sclerosis, initiated in December 2011. Additionally, following excellent preclinical results, a Phase I/Ib open-label study was initiated with macitentan in patients with recurring glioblastoma.

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Notes to the Editor

About Pulmonary Arterial Hypertension

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 with developments of treatment guidelines and new therapies. Drugs targeting the 3 pathways that have been established in the pathogenesis of PAH are endothelin receptor antagonists (ERAs),

prostacyclins 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 clinical trial data have highlighted the need for early intervention, goal-oriented treatment and combination therapy.

Despite these advances in PAH, survival rates are unacceptably low and PAH remains incurable.

References

1. For a general discussion of a clinically meaningful outcome end-point, please see: Proceedings of the 4th world symposium on pulmonary hypertension. J Am Coll Cardiol 2009;54(1 Suppl).
2. Sidharta PN et al. Macitentan: Entry-into-humans study with a new endothelin receptor antagonist. Eur J Clin Pharmacol. 2011;67(10):977-84
3. Bruderer S et al. Effect of cyclosporine A and rifampin on the pharmacokinetics of macitentan, a tissue-targeting dual endothelin receptor antagonist. AAPS J. 2012;14(1):68-78.
4. Bruderer S et al. Absorption, distribution, metabolism, and excretion of macitentan, a dual endothelin receptor antagonist, in humans. Epub Mar 30, 2012

For more information on Actelion's offerings in the area of PAH, please refer to www.actelion.com

Actelion Ltd.

Actelion Ltd is a biopharmaceutical company with its corporate headquarters in Allschwil/Basel, Switzerland. Actelion's first drug Tracleer®, an orally available dual endothelin receptor antagonist, has been approved as a therapy for pulmonary arterial hypertension. Actelion markets Tracleer through its own subsidiaries in key markets worldwide, including the United States (based in South San Francisco), the European Union, Japan, Canada, Australia and Switzerland. Actelion, founded in late 1997, is a leading player in innovative science related to the endothelium - the single layer of cells separating every blood vessel from the blood stream. Actelion's over 2,500 employees focus on the discovery, development and marketing of innovative drugs for significant unmet medical needs. 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 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.

Conference Call / Audiocast

Actelion Ltd will host a Conference Call / Audiocast on on Monday, 30 April 2012, at 14.00 CET / 13.00 BST / 08.00 a.m. EST.

Date/Time:

30 April 2012	14.00 hrs – 15.00 hrs	Basel (CET)
	13.00 hrs – 14.00 hrs	UK (BST)
	08.00 a.m. – 09.00 a.m.	US (EST)

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)44 580 00 74
	UK:	+44 (0)203 367 94 53
	US:	+1 866 907 59 23

Participant's mode:

Listen-Only with possibility to open individual lines during Q&A session. Participants will be asked for their Name and Company.

Audiocast Access:

Audiocast participants should visit the Actelion website www.actelion.com 10-15 minutes before the conference is due to start.

Participant's mode:

Listen only

Audiocast Replay:

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