



Media Release

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Actelion obtains option to acquire privately-held Trophos

Late-stage Phase III compound in Amyotrophic Lateral Sclerosis to report data in late 2011 – Novel therapeutic approach to be further explored in drug discovery collaboration

ALLSCHWIL, SWITZERLAND and MARSEILLE, FRANCE – 20 July 2010 – Actelion Ltd (SIX: ATLN) and privately-held Trophos SA announced today that they have entered into a binding agreement whereby Actelion has, for EUR 10 million, obtained an exclusive option to acquire privately-held Trophos SA, a clinical stage pharmaceutical company.

Trophos' lead compound olesoxime has completed enrollment into a Phase III study in Amyotrophic Lateral Sclerosis (ALS), an orphan disease also known as Lou Gehrig's disease. This study is expected to report data by the end of 2011; at this time Actelion may exercise the option for an acquisition price between EUR 125 and 195 million in cash, contingent on different regulatory approvals and other clinical progress of Trophos' pipeline.

Simon Buckingham, President, Global Corporate and Business Development: "Trophos has done an excellent job to enroll more than 500 ALS patients into a well-designed pivotal study. Once study results are available, Actelion is ideally positioned to leverage these achievements with our proven global regulatory and marketing expertise in the area of orphan drugs."

Trophos is a clinical stage company with a pipeline of new molecular entities in development for the motor neuron diseases ALS and spinal muscular atrophy (SMA) as well as a novel compound for cardiac ischemia-reperfusion injury.

Damian Marron, Chief Executive Officer at Trophos commented: "Since its inception, Trophos has made significant progress in turning its key expertise in neurodegenerative disorders and orphan diseases into achievements that include advancing our lead compound olesoxime into late stage clinical development. The development of olesoxime has benefited from significant support from patient communities, clinical investigators and the European Union (EU), including Trophos spearheading an EU-funded consortium dedicated to improving the treatment of ALS."

Damian Marron continued: "We are delighted with the option agreement with Actelion, which will bring additional expertise and competencies to enable Trophos' compound to rapidly reach patients following a successful study outcome."

Damian Marron concluded: "I am also pleased with the option agreement as it provides the Trophos' investors an opportunity to realize the value of their investments."

The two companies also agreed on a research collaboration to allow Actelion access to Trophos' proprietary CNS assay technology and compound library. The technology mimics neuronal degeneration processes in the test tube and is used to screen chemical compounds for their ability to block these processes.

Martine Clozel, MD and Chief Scientific Officer at Actelion commented: "Trophos has a pioneering approach and proprietary expertise that has enabled the development of high throughput screens using primary neurons as well as the ability to broadly profile more advanced compounds. This is of great value to Actelion as we have developed a large in-house compound library and significant expertise in the field of neurological disorders."

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Notes to Editor:

About the agreement

On July 19, 2010, Actelion signed an acquisition agreement to purchase Trophos SA, a French clinical stage pharmaceutical company developing drugs for patients with neurodegenerative diseases. The acquisition is contingent on the exercise of an option, whereas Actelion has the right to terminate the agreement at any time during the option period. The option will become effective upon payment of EUR 10 million by Actelion and end two months after Actelion's receipt of the results of an ongoing Phase III study with olesoxime but not later than December 31, 2012. The purchase consideration will be paid in cash and is partially contingent on market approval of olesoxime by the US Food and Drug Administration (FDA) as well as overall pipeline progression of other compounds. Consequently, the purchase price might vary between EUR 125 million and 195 million.

About Olesoxime

Olesoxime is Trophos' lead compound of a proprietary mitochondrial pore modulator series. Preclinical studies have demonstrated that olesoxime promotes the function and survival of neurons and other cell types under disease-relevant stress conditions, through interactions with the mitochondrial permeability transition pore (mPTP). Olesoxime has been shown to be active in the SOD1 model of ALS (Bordet et al., JPET 322:709-720, 2007).

Phase I studies in healthy volunteers and Phase Ib studies in ALS patients demonstrated that olesoxime is well-tolerated. These studies also helped to determine the dose regimen used in the pivotal Phase III study.

About the Phase III study in ALS

The study is an 18-month randomized, parallel group, double-blind, placebo-controlled trial evaluating the efficacy and safety of olesoxime against placebo and has benefited from protocol advice obtained from the European Medical Agency EMA. The study completed enrollment in the first quarter of 2010 with 512 patients diagnosed with ALS between 6 and 36 months before enrollment and receiving standard of care. Olesoxime is dosed at 330 mg once-a-day oral capsules. The study is being undertaken in 15 centers in France, Germany, UK, Belgium and Spain and is part of a 3-year collaborative project named MitoTarget (Grant Agreement No: HEALTH-F2-2008-223388) for which the European Commission has awarded a grant of nearly EUR 6 million.

The primary endpoint of the study is the overall 18-month survival rate. Secondary endpoints include the ALS Functional Rating Scale, time to assisted ventilation, vital capacity (a measure of respiratory function), Manual Muscular Testing and quality of life.

About Amyotrophic Lateral Sclerosis

Amyotrophic Lateral Sclerosis (ALS), often referred to as "Lou Gehrig's Disease", is the most common motor neuron disease with a prevalence of 2-3 per 100,000 (30,000 patients in US; 45,000 in Europe at any given time).

Most people who develop ALS are between the ages of 40 and 70 (average age of 55) and over 80% die three to five years after diagnosis. The most common form of ALS is sporadic, but 5-10% of cases are inherited in a dominant manner (familial ALS).

Early symptoms of ALS include muscle weakness in arms and legs; later difficulties in breathing and swallowing are generally the cause of death. There is no treatment today that halts disease progression in ALS patients.

About Trophos' discovery strategy

The Trophos discovery strategy involves recreating neuronal degeneration processes in the test tube and screening chemical compounds for their ability to block these processes. Disease-relevant assays are developed using the specific neurons affected in each disease, for example: motor neurons for amyotrophic lateral sclerosis (ALS) and spinal muscular atrophy (SMA), striatal neurons for Huntington's disease and cortical neurons for Alzheimer's Disease. In essence, Trophos considers the neuron as a cellular test tube filled with the diverse products of the 30,000 genes expressed by the cell under the conditions that are as close to the physiological environment as possible.

About Trophos SA

Trophos SA is a clinical stage pharmaceutical company developing innovative therapeutics for indications with under-served needs in neurology and cardiology. The company has a novel and proprietary cholesterol-oxime based chemistry platform generating a pipeline of drug candidates with the lead product, olesoxime, fully enrolled in a Phase III study in ALS patients and a second product in the cardiovascular field entering Phase I clinical development. Trophos' mitochondrial pore modulator compounds enhance the function and survival of stressed cells via modulation of dysfunctional mitochondria through interactions at the permeability transition pore (mPTP). Recently published clinical studies support the therapeutic rationale for mitochondria-targeted drugs in neurology (Alzheimer's disease) and cardiology (ischemia-reperfusion injury), which Trophos is uniquely placed to exploit. Trophos was founded in 1999 by Antoine Beret and Michel Delaage, former CEO and CSO respectively of Immunotech, and has been financed by funds represented by Amundi PEF (formerly SGAM AI), Turenne Capital Partenaires, Viveris Management, OTC Asset Management, Sofimac, Sofipaca, CM-CIC Capital Privé, Blue Medical Investment and the Association Française contre les Myopathies (AFM). Trophos has also received non-equity financing from the European Commission, the AFM, the Agence Nationale de Recherche and Oseo Innovation.

About 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 2,300 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[®]).

For further information please contact:

For Actelion Ltd:

Roland Haefeli

Vice President, Head of Investor Relations & Public Affairs

Actelion Pharmaceuticals Ltd, Gewerbestrasse 16, CH-4123 Allschwil

+41 61 565 62 62

+1 650 624 69 36

<http://www.actelion.com>

For Trophos SA:

Andrew Lloyd & Associates

Andrew Lloyd / Neil Hunter

Tel: +44 1273 675100

allo@ala.com / neil@ala.com

<http://www.trophos.com>