PRESS RELEASE

Stockholm, Sweden and Dublin, Ireland 7 December 2016

Sobi™ and Horizon Pharma enter five-year distribution agreement for Ravicti® and Ammonaps® outside the United States

Swedish Orphan Biovitrum AB (publ) (Sobi™) and Horizon Pharma plc (NASDAQ: HZNP) ("Horizon") today announced that the companies have entered into a five-year distribution agreement for Ravicti® (glycerol phenylbutyrate) in European countries, including United Kingdom, Germany, France, Italy and Spain and for Ammonaps® (sodium phenylbutryate) in the same European countries and certain Middle Eastern countries. Under the agreement, Sobi will have exclusive marketing, sales and distribution rights for the two medicines in the territory until 31 December 2021. Horizon has the ability to terminate the agreement after two years subject to certain pre-defined termination fees. Sobi currently distributes Ravicti in certain Middle Eastern countries and Ammonaps in certain European and Middle Eastern countries.

Ravicti and Ammonaps are authorised by the European Commission and are indicated for the treatment of Urea Cycle Disorders (UCD).

"We are happy to build on our successful relationship with Horizon Pharma to support providing treatment to people living with UCDs", says Alan Raffensperger, Chief Operating Officer of Sobi and continues: "This distribution agreement will allow us to leverage our existing expertise and extensive experience within the area of UCDs. Our main focus now will turn to implementing patient access activities with the objective to launch Ravicti in Europe during 2017."

"Sobi has been a trusted partner in Europe and the Middle East for the last three years," says Francoise de Craecker, group vice president and general manager, EMEA, orphan business unit, Horizon Pharma plc. "We believe that Sobi's current distribution of Ammonaps uniquely qualifies it to effectively provide Ravicti to people living with UCDs in European markets."

_____

About Ravicti®

Ravicti is indicated for use in all 28 Member States of the European Union and 3 Member States of the European Economic Area as a nitrogen-binding agent for chronic management of adult and pediatric patients two months of age and older with UCDs who cannot be managed by dietary protein restriction and/or amino acid supplementation alone.

Swedish Orphan Biovitrum AB (publ)
Postal address: SE-112 76 Stockholm, Sweden
Phone: +46 8 697 20 00 | www.sobi.com

Important Safety Information

LIMITATIONS OF USE:

- Ravicti is not indicated for the treatment of acute hyperammonemia in patients with UCDs because more rapidly acting interventions are essential to reduce plasma ammonia levels.
- The safety and efficacy of Ravicti for the treatment of patients with N-acetylglutamate synthase (NAGS) and CITRIN (citrullinaemia type 2) deficiency has not been established.
- The use of RAVICTI in patients <2 months of age is not recommended as the safety and efficacy of Ravicti in this age group has not been established.

CONTRAINDICATIONS:

- Hypersensitivity to the active substance.
- Treatment of acute hyperammonaemia.

WARNINGS AND PRECAUTIONS:

- Phenylacetate acid (PAA), the major metabolite of Ravicti, may be toxic at levels ≥500 µg/mL. Reduce Ravicti dosage if symptoms of neurotoxicity, including vomiting, nausea, headache, somnolence, confusion, or sleepiness are present in the absence of high ammonia or other intercurrent illnesses.
- Low or absent pancreatic enzymes or intestinal disease resulting in fat malabsorption may result in reduced or absent digestion of Ravicti and/or absorption of phenylbutyrate and reduced control of plasma ammonia. Monitor ammonia levels closely.
- Studies in animals have shown reproductive toxicity (see section 5.3). There are limited data regarding the use of glycerol phenylbutyrate in pregnant women. Glycerol phenylbutyrate is not recommended during pregnancy and in women of childbearing potential not using contraception.
- It is unknown whether glycerol phenylbutyrate or its metabolites are excreted in human milk. A risk to the newborns/infants cannot be excluded. A decision must be made whether to discontinue breastfeeding or to discontinue/abstain from glycerol phenylbutyrate therapy taking into account the benefit of breast-feeding for the child and the benefit of therapy for the woman.

ADVERSE REACTIONS:

- Assessment of adverse reactions was based on exposure in 114 UCD patients (65 adults and 49 children between the ages of 2 months and 17 years) with deficiencies in CPS, OTC, ASS, ASL, ARG, or HHH across 4 short term and 3 long term clinical studies, in which 90 patients completed 12 months duration (median exposure = 51 weeks).
- At the beginning of the treatment, abdominal pain, nausea, diarrhoea, and/or headache may occur; these reactions usually disappear within a few days even if treatment is continued. The most frequently reported adverse reactions (>5%) during glycerol phenylbutyrate treatment were diarrhoea, flatulence, and headache (8.8% each); decreased appetite (7.0%), vomiting (6.1%); and fatigue, nausea and, skin odour abnormal (5.3% each).

DRUG INTERACTIONS:

- Concomitant use of medicinal products known to inhibit lipase should be given with caution as glycerol phenylbutyrate is hydrolysed by digestive lipase into phenylbutyrate acid and glycerol. This may be associated with increased risk of medicinal product interactions with lipase inhibitors and with lipase contained in pancreatic enzyme replacement therapies.
- A potential effect on CYP2D6 isoenzyme cannot be excluded and caution is advised for patients who receive medicinal products that are CYP2D6 substrates.
- Glycerol phenylbutyrate and/or its metabolites, PAA and PBA, have been shown to be weak inducers of CYP3A4 enzyme in vivo. In vivo exposure to glycerol phenylbutyrate has resulted in decreased systemic exposure to midazolam of approximately 32% and increased exposure to the 1-hydroxy metabolite of midazolam, suggesting that steady-state dosing of glycerol phenylbutyrate results in CYP3A4 induction. The potential for interaction of glycerol phenylbutyrate as a CYP3A4 inducer and those products predominantly metabolised by the CYP3A4 pathway is possible. Therefore, therapeutic effects and/or metabolite levels of medicinal products, including some oral contraceptives that are substrates for this enzyme may be reduced and their full effects cannot be guaranteed, following coadministration with glycerol phenylbutyrate.
- Corticosteroids, valproic acid, haloperidol and probenecid may have the potential to affect ammonia levels.

About Ammonaps®
Ammonaps (sodium phenylbutyrate) is used to treat Urea Cycle Disorders (UCD). UCD is a group of serious conditions in which patients suffer from deficiencies in the enzymes required to remove ammonia from the blood stream.

Ammonaps is used to reduce levels of ammonia and glutamine in the blood. Ammonaps is used with other treatments and a special diet for the long-term management of patients with urea cycle disorders where there is lack of one or more of the following enzymes: carbamyl phosphate synthetase, ornithine transcarbamylase, or argininosuccinate synthetase. Ammonaps is indicated in all patients with neonatal-onset presentation (complete enzyme deficiencies, presenting within the first 28 days of life). It is also indicated in patients with late-onset disease (partial enzyme deficiencies, presenting after the first month of life) who have a history of hyperammonaemic encephalopathy.

Important Safety Information

CONTRAINDICATIONS:
- Pregnancy.
- Breast-feeding.
- Hypersensitivity to the active substance or to any of the excipients

WARNINGS AND PRECAUTIONS:
- Ammonaps tablets should not be used in patients with dysphagia due to the potential risk of oesophageal ulceration if tablets are not promptly delivered to the stomach.
- Each Ammonaps tablet contains 62 mg (2.7 mmol) of sodium, corresponding to 2.5 g (108 mmol) of sodium per 20 g of sodium phenylbutyrate, which is the maximum daily dose. Ammonaps should therefore be used with caution in patients with congestive heart failure or severe renal insufficiency, and in clinical conditions where there is sodium retention with oedema.
- Ammonaps granules contain 124 mg (5.4 mmol) of sodium per gram of sodium phenylbutyrate, corresponding to 2.5 g (108 mmol) of sodium per 20 g of sodium phenylbutyrate, which is the maximum daily dose. Ammonaps should therefore be used with caution in patients with congestive heart failure or severe renal insufficiency, and in clinical conditions where there is sodium retention with oedema.
- Since the metabolism and excretion of sodium phenylbutyrate involves the liver and kidneys, Ammonaps should be used with caution in patients with hepatic or renal insufficiency.
- Serum potassium should be monitored during therapy since renal excretion of phenylacetylglutamine may induce a urinary loss of potassium.
- Even on therapy, acute hyperammonaemic encephalopathy may occur in a number of patients.
- Ammonaps is not recommended for the management of acute hyperammonaemia, which is a medical emergency.
- In children unable to swallow tablets, it is recommended to use Ammonaps granules instead.

ADVERSE REACTIONS:
- In clinical trials with Ammonaps, 56 % of the patients experienced at least one adverse event and 78 % of these adverse events were considered as not related to Ammonaps.
- Adverse reactions mainly involved the reproductive and gastrointestinal system.

DRUG INTERACTIONS:
- Concurrent administration of probenecid may affect renal excretion of the conjugation product of sodium phenylbutyrate.
- There have been published reports of hyperammonaemia being induced by haloperidol and by valproate. Corticosteroids may cause the breakdown of body protein and thus increase plasma ammonia levels. More frequent monitoring of plasma ammonia levels is advised when these medications have to be used.

About Sobi™
Sobi is an international specialty healthcare company dedicated to rare diseases. Sobi's mission is to develop and deliver innovative therapies and services to improve the lives of patients. The product portfolio is primarily focused on Haemophilia, Inflammation and Genetic diseases. Sobi also markets a portfolio of specialty and rare disease products across Europe, the Middle East, North Africa and Russia for partner companies. Sobi is a pioneer in biotechnology with world-class capabilities in protein biochemistry and biologics manufacturing. In 2015, Sobi had total revenues of SEK 3.2 billion (USD 385 M) and about 700 employees. The share (STO: SOBI) is listed on Nasdaq Stockholm. More information is available at www.sobi.com.

About Horizon Pharma plc
Horizon Pharma plc is a biopharmaceutical company focused on improving patients' lives by identifying, developing, acquiring and commercializing differentiated and accessible medicines that address unmet medical needs. The Company markets 11 medicines through its orphan, rheumatology and primary care business units. For more information, please visit www.horizonpharma.com. Follow @HZNPplc on Twitter or view careers on our LinkedIn page.

Forward Looking Statements
This press release contains forward-looking statements, including statements regarding the potential benefits that may be derived from the distribution agreement between Horizon Pharma and Sobi, and plans and expected timing with respect to launching Ravicti in Europe. These forward-looking statements are based on management expectations and assumptions as of the date of this press release, and actual results may differ materially from those in these forward-looking statements as a result of various factors. These factors include risks that the distribution agreement is terminated early, that either party does not comply with its obligations under the distribution agreement, that adequate pricing and reimbursement for Ravicti in Europe is not available, and those factors described in Horizon Pharma's filings with the United States Securities and Exchange Commission, including those discussed under the caption "Risk Factors" in those filings. Forward-looking statements speak only as of the date of this press release and Horizon does not undertake any obligation to update or revise these statements, except as may be required by law.

For more information please contact:

Media relations
Linda Holmström, Senior Communications Manager
T: + 46 708 73 40 95, + 46 8 697 31 74
linda.holmstrom@sobi.com

Investor relations
Jörgen Winroth, Vice President, Head of Investor Relations
T: +1 347-224-0819, +1 212-579-0506, +46 8 697 2135
jorgen.winroth@sobi.com

Horizon Pharma contacts:

Media
Geoff Curtis
Senior Vice President, Corporate Communications
media@horizonpharma.com

Investor relations
Tina Ventura
Senior Vice President, Investor Relations
Investor-relations@horizonpharma.com