September 25, 2017

Intercept Statement Regarding Ocaliva® (obeticholic acid) Safety and Dosing in Primary Biliary Cholangitis (PBC) Patients

Company to Host Conference Call at 8:30 a.m. ET

NEW YORK, Sept. 25, 2017 (GLOBE NEWSWIRE) -- Intercept Pharmaceuticals, Inc. (Nasdaq:ICPT), a biopharmaceutical company focused on the development and commercialization of novel therapeutics to treat progressive non-viral liver diseases, today provided comment on the Ocaliva (obeticholic acid or OCA) Dear Healthcare Provider (DHCP) letter issued on September 12, 2017, and the subsequent drug safety communication issued by the U.S. Food and Drug Administration (FDA) on September 21, 2017.

Ocaliva was approved in the U.S. in May 2016 (and subsequently in the European Union and Canada) for the treatment of patients with primary biliary cholangitis (PBC) with an inadequate response to, or intolerant of the standard of care, UDCA. PBC is a rare and life-threatening progressive liver disease that primarily afflicts women and is a leading cause of liver failure with resulting need for liver transplant in women. Ocaliva therefore represents an important treatment option for patients with PBC and since its approval more than 3,000 patients have been treated with Ocaliva in the U.S. alone. More than 150 patients are enrolled in ongoing open label phases of Intercept's Phase 2 and Phase 3 clinical trials and have been on OCA treatment for periods ranging from approximately three to seven years.

Label Recommended Ocaliva Dosing
Recommended dosing in the label for Ocaliva in earlier stage PBC patients with no or mild hepatic impairment (non-cirrhotic or Child-Pugh A cirrhosis) starts at 5 mg once daily, increasing after three months to 10 mg once daily based on tolerability and treatment response. However, in late stage patients with moderate or severe hepatic impairment (Child Pugh B or C cirrhosis), recommended dosing starts at 5 mg once weekly, with the possibility to gradually increase to a maximum of 10 mg twice weekly. The reason for this less frequent dosing is that systemic and hepatic concentrations of Ocaliva are predicted to significantly increase in such patients and dose-related liver adverse reactions have previously been documented in PBC patients participating in clinical trials.

Recently Issued Dear Healthcare Provider Letter and FDA Safety Communication
In the course of Intercept's post-marketing pharmacovigilance activities, deaths have been reported in PBC patients with moderate or severe hepatic impairment (Child Pugh B or C cirrhosis). In an analysis performed by Intercept and in consultation with the FDA, Intercept concluded that these patients were prescribed once daily doses of Ocaliva, which is seven times higher than the recommended weekly dose in such patients. As a result, Intercept issued the DHCP letter and the FDA subsequently issued their own safety communication to reinforce recommended label dosing. Both communications remind healthcare providers of the importance of the recommended reduced dosing of Ocaliva in PBC patients with moderate or severe hepatic impairment (Child Pugh B or C cirrhosis), while reiterating the importance of close monitoring of PBC patients for progression of their disease and the occurrence of liver-related adverse reactions.

Actions to Enhance Education About Appropriate Use of Ocaliva
Patient safety is Intercept's highest priority and it is imperative that Ocaliva is dosed according to its approved label. In addition to the DHCP letter, Intercept has taken actions to enhance education about appropriate use of Ocaliva. These initiatives include:

- reeducating physicians on the label, with a focus on ensuring appropriate dosing for patients with moderate or severe hepatic impairment (Child Pugh B or C cirrhosis);
- enhancing monitoring of patients for liver-related adverse reactions; and
- completing adjudication of all reported cases of serious liver injury, including in patients with no or mild hepatic impairment.

Pursuant to the FDA's safety communication, Intercept has begun working with the FDA on updates to the label to better ensure appropriate and safe use of Ocaliva.

Conference Call at 8:30 a.m. ET

Intercept will discuss the FDA statement on a conference call and webcast today at 8:30 a.m. ET. The live event will be available on the investor page of Intercept's website at http://ir.interceptpharma.com or by calling (855) 232-3919 (toll-free

domestic) or (315) 625-6894 (international) five minutes prior to the start time (no passcode is required). A replay of the call will be available on Intercept's website approximately two hours after the completion of the call and will be archived for two weeks.

About Ocaliva® (obeticholic acid)

Ocaliva is indicated in the United States for the treatment of primary biliary cholangitis (PBC) in combination with ursodeoxycholic acid (UDCA) in adults with an inadequate response to UDCA, or as monotherapy in adults unable to tolerate UDCA.

This indication is approved under accelerated approval based on a reduction in alkaline phosphatase (ALP), as a surrogate endpoint which is reasonably likely to predict clinical benefit, including an improvement in liver transplant free-survival. An improvement in survival or disease-related symptoms has not been established. Continued approval for this indication may be contingent upon verification and description of clinical benefit in confirmatory trials. Intercept is currently enrolling COBALT, a Phase 4 clinical outcomes trial of Ocaliva in patients with PBC with the goal of confirming clinical benefit on a post-marketing basis.

In December 2016, Ocaliva received conditional marketing authorization in Europe for the treatment of PBC in combination with UDCA in adults with an inadequate response to UDCA or as monotherapy in adults unable to tolerate UDCA, conditional to the company providing further data post-approval to confirm benefit. For detailed safety information for Ocaliva 5 mg and 10 mg tablets including posology and method of administration, special warnings, drug interactions and adverse drug reactions, please see the European Summary of Product Characteristics that can be found on www.ema.europa.eu.

U.S. IMPORTANT SAFETY INFORMATION
Contraindications
Ocaliva is contraindicated in patients with complete biliary obstruction.

Warnings and Precautions

Liver-Related Adverse Reactions
In two 3-month, placebo-controlled clinical trials a dose-response relationship was observed for the occurrence of liver-related adverse reactions including jaundice, ascites and primary biliary cholangitis flare with dosages of Ocaliva of 10 mg once daily to 50 mg once daily (up to 5-times the highest recommended dosage), as early as one month after starting treatment with Ocaliva.

In a pooled analysis of three placebo-controlled trials in patients with PBC, the exposure-adjusted incidence rates for all serious and otherwise clinically significant liver-related adverse reactions, and isolated elevations in liver biochemical tests, per 100 patient exposure years (PEY) were: 5.2 in the Ocaliva 10 mg group (highest recommended dosage), 19.8 in the Ocaliva 25 mg group (2.5 times the highest recommended dosage) and 54.5 in the Ocaliva 50 mg group (5 times the highest recommended dosage) compared to 2.4 in the placebo group.

Monitor patients during treatment with Ocaliva for elevations in liver biochemical tests and for the development of liver-related adverse reactions. Weigh the potential risks against the benefits of continuing treatment with Ocaliva in patients who have experienced clinically significant liver-related adverse reactions. The maximum recommended dosage of Ocaliva is 10 mg once daily. Adjust the dosage for patients with moderate or severe hepatic impairment.

Discontinue Ocaliva in patients who develop complete biliary obstruction.

Severe Pruritus

Severe pruritus was reported in 23% of patients in the Ocaliva 10 mg arm, 19% of patients in the Ocaliva titration arm and 7% of patients in the placebo arm in the POISE trial, a 12-month double- blind randomized controlled trial of 216 patients. Severe pruritus was defined as intense or widespread itching, interfering with activities of daily living, or causing severe sleep disturbance, or intolerable discomfort, and typically requiring medical interventions. In the subgroup of patients in the Ocaliva titration arm who increased their dosage from 5 mg once daily to 10 mg once daily after 6 months of treatment (n=33), the incidence of severe pruritus was 0% from months 0 to 6 and 15% from months 6 to 12. The median time to onset of severe pruritus was 11, 158 and 75 days for patients in the Ocaliva 10 mg, Ocaliva titration and placebo arms, respectively.

Management strategies include the addition of bile acid resins or antihistamines, Ocaliva dosage reduction and/or temporary interruption of Ocaliva dosing.

Reduction in HDL-C

Patients with PBC generally exhibit hyperlipidemia characterized by a significant elevation in total cholesterol primarily due to increased levels of high density lipoprotein-cholesterol (HDLC). In the POISE trial, dose-dependent reductions from baseline in mean HDL-C levels were observed at 2 weeks in Ocaliva-treated patients, 20% and 9% in the 10 mg and titration arms, respectively, compared to 2% in the placebo arm. At month 12, the reduction from baseline in mean HDL-C level was 19% in the Ocaliva 10 mg arm, 12% in the Ocaliva titration arm and 2% in the placebo arm. Nine patients in the Ocaliva 10 mg arm and six patients in the Ocaliva titration arm, versus three patients in the placebo arm had reductions in HDL-C to less than 40 mg/dL.

Monitor patients for changes in serum lipid levels during treatment. For patients who do not respond to Ocaliva after one year at the highest recommended dosage that can be tolerated (maximum of 10 mg once daily), and who experience a reduction in HDL-C, weigh the potential risks against the benefits of continuing treatment.

Adverse Reactions

The most common adverse reactions from subjects taking Ocaliva (≥5%) were pruritus, fatigue, abdominal pain and discomfort, rash, oropharyngeal pain, dizziness, constipation, arthralgia, thyroid function abnormality and eczema.

Drug Interaction
Bile Acid Binding Resins
Bile acid binding resins such as cholestyramine, colestipol or colesevelam absorb and reduce bile acid absorption and may reduce the absorption, systemic exposure and efficacy of Ocaliva. If taking bile acid binding resins, take Ocaliva at least 4 hours before or 4 hours after (or at as great an interval as possible) taking a bile acid binding resin.

Please see the U.S. Full Prescribing Information for Ocaliva (obeticholic acid) 5 mg and 10 mg tablets.

About Intercept
Intercept is a biopharmaceutical company focused on the development and commercialization of novel therapeutics to treat progressive non-viral liver diseases, including primary biliary cholangitis (PBC), nonalcoholic steatohepatitis (NASH), primary sclerosing cholangitis (PSC) and biliary atresia. Founded in 2002 in New York, Intercept now has operations in the United States, Europe and Canada.

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Safe Harbor Statements

This press release contains "forward-looking statements" within the meaning of the Private Securities Litigation Reform Act of 1995, including, but not limited to, statements on the commercial potential of Ocaliva, Intercept's review and analysis of the cases reported by the FDA and the results thereof, any future events that may be experienced by patients who use Ocaliva and the association of such events with its use, the results of Intercept's educational efforts with healthcare providers and other planned and ongoing initiatives, Intercept's continuing interactions with the FDA, the changes to the label for Ocaliva that may result from Intercept's discussions and negotiations with the FDA and the impact thereof, including in relation to the commercial potential of Ocaliva, Intercept's ongoing and future clinical development programs and any impact thereto, and our strategic directives under the caption "About Intercept." These "forward-looking statements" are based on management's current expectations of future events and are subject to a number of important risks and uncertainties that could cause actual results to differ materially and adversely from those set forth in or implied by such forward-looking statements. These risks and uncertainties include, but are not limited to: the potential benefit and commercial potential of Ocaliva in PBC, and Intercept's ability to maintain its regulatory approval in jurisdictions in which Ocaliva is approved for use in PBC; the initiation, cost, timing, progress and results of Intercept's development activities, preclinical studies and clinical trials; the timing of and Intercept's ability to obtain and maintain regulatory approval of OCA in PBC in countries outside the ones in which it is approved and in indications other than PBC and any other product candidates it may develop such as INT-767; conditions that may be imposed by regulatory authorities on Intercept's

marketing approvals for its products and product candidates such as the need for clinical outcomes data (and not just results based on achievement of a surrogate endpoint), and any related restrictions, limitations, and/or warnings in the label of any approved products and product candidates; Intercept's plans to research, develop and commercialize its product candidates; Intercept's ability to obtain and maintain intellectual property protection for its products and product candidates; Intercept's ability to successfully commercialize its products and product candidates; the size and growth of the markets for Intercept's products and product candidates and its ability to serve those markets; the rate and degree of market acceptance of any of Intercept's products, which may be affected by the reimbursement received from payors; the success of competing drugs that are or become available; regulatory developments in the United States and other countries; the performance of third-party suppliers and manufacturers; the election by Intercept's collaborators to pursue research, development and commercialization activities; Intercept's ability to attract collaborators with development, regulatory and commercialization expertise; Intercept's need for and ability to obtain additional financing; Intercept's estimates regarding expenses, revenues and capital requirements and the accuracy thereof; Intercept's use of cash and short-term investments; Intercept's ability to attract and retain key scientific or management personnel; and other factors discussed under the heading "Risk Factors" contained in our annual report on Form 10-K for the year ended December 31, 2016 filed on March 1, 2017 as well as any updates to these risk factors filed from time to time in our other filings with the Securities and Exchange Commission. All information in this press release is as of the date of the release, and Intercept undertakes no duty to update this information unless required by law.