Jaguar Health Provides Updates Regarding Company's Focus on Rare and Orphan Disease Indications in the U.S. & Europe

January 26, 2023

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Jaguar to take the lead on microvillus inclusion disease in the U.S., with the Company intending to submit an Investigational New Drug (IND) application to the FDA for this ultra-rare disease in 1H 2023

SAN FRANCISCO, CA / ACCESSWIRE / January 26, 2023 / Jaguar Health, Inc. (NASDAQ:JAGX) ("Jaguar" or the "Company") today provided updates regarding the Company's focus on rare and orphan disease indications: short bowel syndrome (SBS), microvillus inclusion disease (MVID), and cholera. Rare disease indications are the sole focus of Napo Therapeutics, the corporation established by Jaguar in Italy in 2021, with an initial focus on SBS. Jaguar is the majority shareholder of Napo Therapeutics.

Microvillus Inclusion Disease (MVID)

The Company intends to submit an Investigational New Drug (IND) application to the U.S. Food and Drug Administration (FDA) in the first half of 2023 for crofelemer, the Company's oral botanical drug, for the indication of MVID, an ultra-rare congenital diarrheal disorder (CDD) condition. MVID is a life-threatening autosomal recessive disease that affects newborns and children and leads to significant morbidity and even death from severe secretory diarrhea. MVID is a subset of various CDDs, which are a group of inherited chronic enteropathies characterized by heterogeneous etiology, and each type of CDD is thus a different disease with a different pathogenetic mechanism. CDDs share a primary common symptom of severe chronic diarrhea, which may produce secondary symptoms, including significant dehydration, metabolic acidosis or alkalosis and malnutrition, which may become life-threatening.

As previously announced, the Company has submitted an Orphan Drug Designation (ODD) application to the FDA for MVID and is awaiting a response on the application. The Orphan Drug Act of 1983, a law passed in the U.S. to facilitate development of drugs for rare diseases, provides for granting special status to a drug or biological product to treat a rare disease or condition upon request of a sponsor. ODD qualifies the sponsor of the drug for various development incentives, including tax credits for qualified clinical testing and relief of filing fees. The European Medicines Agency (EMA) granted ODD for crofelemer in October 2022 for MVID in the European Union.

In May 2022, a clinical collaborator of the Company presented the results of an *in vitro* study evaluating the effectiveness of crofelemer as a potential therapy to mitigate chloride ion secretion in MVID patients at the Digestive Disease Week® (DDW 2022) conference. The results of this study suggested that crofelemer has the clinical potential for symptomatic therapy and/or treatment of MVID.

Short Bowel Syndrome

Crofelemer has received ODD from both the FDA and the EMA for SBS, which is a complex condition characterized by diarrhea and/or severe malabsorption of fluids and nutrients due to surgical resection of bowel segments, congenital anomalies, or disease-associated loss of absorption; any or all of which may require parenteral support (PS) in the form of parenteral nutrition (PN) and/or intravenous (IV) fluids. Investigator-initiated proof-of-concept clinical studies of the safety and effectiveness of crofelemer for reduction of PS in SBS patients with intestinal failure (SBS-IF) are expected to be conducted throughout 2023.

For SBS-IF patients who endure the significant loss of their bowel, the resulting excessive intestinal fluid and electrolyte output may cause lifelong restriction and adjustment of oral intake of food and liquids, leading to the requirement of PN and/or IV fluids to maintain nutrient, electrolyte and fluid balance. The constant need for PS challenges the ability of patients to carry out activities of daily living (ADL), such as attending school or work, and has a significant impact on their daily quality of life (QOL). Furthermore, lifelong PN may lead to potentially life-threatening complications like sepsis and/or organ failures. SBS affects approximately 10,000 to 20,000 people in the United States, according to the Crohn's & Colitis Foundation, and it is estimated that the population of SBS patients in Europe is approximately the same size.

Cholera in the United States

Cholera, an acute diarrheal illness caused by infection of the intestine with the bacterium *Vibrio cholerae*, is an orphan indication in the United States. As announced in September 2022, the FDA has activated the Company's Investigational New Drug (IND) application for NP-300, a novel botanical drug product candidate for the symptomatic relief and treatment of moderate-to-severe diarrhea, with or without concomitant antimicrobial therapy, from bacterial, viral and parasitic infections including *Vibrio cholerae*. Due to the very low incidence and prevalence of cholera in the United States, the

Company plans to request orphan drug designation (ODD) for NP-300 in the U.S.

Upon completion of the requisite development activities to support the New Drug Application (NDA) and subsequent approval of NP-300 by the FDA for the symptomatic relief and treatment of diarrhea from cholera, the Company intends to pursue a Tropical Disease Priority Review Voucher ("TDPRV") under the FDA's financial incentive program to develop NP-300 for this indication. Priority review vouchers are transferable, and in past transactions by other companies have sold for values ranging from $67 million to $350 million, which provides for a potential immediate return on investment upon approval of NP-300 for the cholera-related diarrhea indication.

"Jaguar is focused on two late-stage clinical events in the next approximately 6 to 7 months that we expect to be transformational in terms of value creation and recognition for the Company. Our key near-term clinical activity is our Phase 3 pivotal OnTarget trial of crofelemer for our core follow-on indication of prophylaxis of cancer therapy-related diarrhea (CTD). We expect enrollment in this pivotal trial to complete in the second quarter of 2023," said Lisa Conte, Jaguar's president and CEO. "Our second prioritized clinical program centers around our investigator-initiated proof-of-concept trial of crofelemer for SBS and CDD."

About Crofelemer

Crofelemer is an oral botanical (plant-based) drug extracted and purified from the red bark sap, also referred to as "dragon's blood," of the *Croton lechleri* tree in the Amazon Rainforest. The Company has established a sustainable harvesting program, under fair trade practices, for crofelemer to ensure a high degree of quality, ecological integrity, and support for Indigenous communities. As stated on the FDA's **website**, crofelemer is currently one of only two drugs that have been approved under the FDA's Botanical Guidance definition of a botanical drug product.

About NP-300

NP-300 is a novel oral botanical drug product that is purified and sustainably derived from the red bark sap of the *Croton lechleri* tree, the same source as that for crofelemer, and is planned to be developed under the FDA's Botanical Guidance.

About Jaguar Health, Napo Pharmaceuticals, Napo Therapeutics & Jaguar Animal Health

Jaguar Health, Inc. is a commercial stage pharmaceuticals company focused on developing novel, plant-based, non-opioid, and sustainably derived prescription medicines for people and animals with GI distress, including chronic, debilitating diarrhea. Jaguar Health's wholly owned subsidiary, Napo Pharmaceuticals, Inc., focuses on developing and commercializing proprietary plant-based human pharmaceuticals from plants harvested responsibly from rainforest areas. Our crofelemer drug product candidate is the subject of the OnTarget study, an ongoing pivotal Phase 3 clinical trial for prophylaxis of diarrhea in adult cancer patients receiving targeted therapy. Jaguar Health is the majority shareholder of Napo Therapeutics S.p.A. (f/k/a Napo EU S.p.A.), an Italian corporation established by Jaguar Health in Milan, Italy in 2021 that focuses on expanding crofelemer access in Europe. Jaguar Animal Health is a tradename of Jaguar Health.

For more information about Jaguar Health, please visit https://jaguar.health. For more information about Napo Pharmaceuticals, visit www.napopharma.com. For more information about Napo Therapeutics, visit napotherapeutics.com.

Forward-Looking Statements

Certain statements in this press release constitute "forward-looking statements." These include statements regarding the Company's expectation that it will submit an IND application to the FDA in the first half of 2023 for crofelemer for MVID, the expectation that crofelemer has the clinical potential for symptomatic therapy and/or treatment of MVID, the Company's expectation that investigator-initiated proof-of-concept clinical studies of the safety and effectiveness of crofelemer for reduction of PS in SBS patients with intestinal failure will be conducted throughout 2023, statements regarding the potential for approval of NP-300 by the FDA for the symptomatic relief and treatment of diarrhea from cholera, the Company's expectation that it will pursue a TDPRV for NP-300, and the Company's expectation that two late-stage clinical events in the next approximately 6 to 7 months may be transformational in terms of value creation and recognition for the Company. In some cases, you can identify forward-looking statements by terms such as "may," "will," "should," "expect," "plan," "aim," "anticipate," "could," "intend," "target," "project," "contemplate," "believe," "estimate," "predict," "potential" or "continue" or the negative of these terms or other similar expressions. The forward-looking statements in this release are only predictions. Jaguar has based these forward-looking statements largely on its current expectations and projections about future events. These forward-looking statements speak only as of the date of this release and are subject to a number of risks, uncertainties and assumptions, some of which cannot be predicted or quantified and some of which are beyond Jaguar's control. Except as required by applicable law, Jaguar does not plan to publicly update or revise any forward-looking statements contained herein, whether as a result of any new information, future events, changed circumstances or otherwise.

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