November 7, 2019

Rigel Announces Upcoming Data Presentations at the 61st ASH Annual Meeting & Exposition

78% response rate in ITP patients who received fostamatinib as second-line therapy in post-hoc analysis

SOUTH SAN FRANCISCO, Calif., Nov. 7, 2019 /PRNewswire/ -- Rigel Pharmaceuticals, Inc. (Nasdaq: RIGL), today announced that data related to TAVALISSE® (fostamatinib disodium hexahydrate) tablets has been accepted for two poster presentations at the 61st American Society of Hematology (ASH) Annual Meeting & Exposition to be held December 7-10, 2019, in Orlando, FL.

Rigel conducted a post-hoc data analysis from a Phase 3 clinical program of TAVALISSE in adult patients with immune thrombocytopenia (ITP). In this analysis, 32 patients received fostamatinib as a second-line therapy, and 78% (25/32) achieved ≥1 platelet count of ≥50,000/μL (without rescue therapy). Adverse events were manageable and consistent with those previously reported with fostamatinib.

"These data show a high response rate in the early-line treatment of adult ITP," said Raul Rodriguez, Rigel's President and CEO. "We believe that these data, coupled with TAVALISSE's differentiated and targeted mechanism of action, provide an attractive treatment approach for early-line patients. These patients represent the vast majority of the adult ITP population."

Additionally, in a Phase 2 open-label study of fostamatinib in patients with warm antibody autoimmune hemolytic anemia (wAIHA), data showed that 44% (11/25) of evaluable patients met the primary efficacy endpoint of a hemoglobin (Hgb) level >10 g/dL with an increase of ≥2 g/dL from baseline by week 24. Including one late responder at week 30, the overall response rate was 48% (12/25). Adverse events were manageable and consistent with those previously reported with fostamatinib.

Fostamatinib disodium hexahydrate is an oral drug designed to inhibit spleen tyrosine kinase (SYK), a key signaling component of the body's immune process that leads to platelet destruction in ITP and proposed red blood cell destruction in AIHA. Fostamatinib is commercially available in the U.S. under the brand name TAVALISSE (fostamatinib

disodium hexahydrate) tablets and is the first and only SYK inhibitor indicated for the treatment of thrombocytopenia in adult patients with chronic ITP who have had an insufficient response to a previous treatment. TAVALISSE is currently being investigated in a Phase 3 trial for wAIHA, a rare, serious blood disorder for which there are no approved therapies.

Poster Presentations

Abstract #1069

Enhanced Responses to Fostamatinib as Second-Line Therapy and in Persistent Immune Thrombocytopenia (ITP) Patients

Session Name: 311. Disorders of Platelet Number or Function: Poster I
Presenter: Ralph Boccia
Date: Saturday, December 7, 2019
Presentation Time: 5:30 PM-7:30 PM EST
Location: Hall B (Orange County Convention Center)

Abstract #3518

Fostamatinib[1], a Spleen Tyrosine Kinase (SYK) Inhibitor, for the Treatment of Warm Antibody Autoimmune Hemolytic Anemia (wAIHA): Final Results of the Phase 2, Multicenter, Open-Label Study

Session Name: 101. Red Cells and Erythropoiesis, Structure and Function, Metabolism, and Survival, Excluding Iron: Poster III
Presenter: Kerry Rogers
Date: Monday, December 9, 2019
Presentation Time: 6:00 PM - 8:00 PM EST
Location: Hall B (Orange County Convention Center)

The conference abstracts can be accessed here.

To learn more about Rigel Pharmaceuticals and TAVALISSE® visit booth #415 during ASH 2019.

About ITP

In patients with ITP (immune thrombocytopenia), the immune system attacks and destroys the body's own blood platelets, which play an active role in blood clotting and healing. Common symptoms of ITP are excessive bruising and bleeding. People suffering with chronic ITP may live with an increased risk of severe bleeding events that can result in serious medical complications or even death. Current therapies for ITP include steroids, blood platelet production boosters (TPO-RAs) and splenectomy. However, not all patients respond to existing therapies. As a result, there remains a significant medical need for additional treatment options for patients with ITP.

About AIHA

Autoimmune hemolytic anemia (AIHA) is a rare, serious blood disorder in which the immune system produces antibodies that result in the destruction of the body's own red blood cells. AIHA affects approximately 40,000 adult patients in the U.S. and can be a severe, debilitating disease. To date, there are no disease-targeted therapies approved for AIHA, despite the unmet medical need that exists for these patients. Warm antibody AIHA (wAIHA), the most common form of AIHA, is characterized by the presence of antibodies that react with the red blood cell surface at body temperature.

About TAVALISSE

Indication

TAVALISSE® (fostamatinib disodium hexahydrate) tablets is indicated for the treatment of thrombocytopenia in adult patients with chronic immune thrombocytopenia (ITP) who have had an insufficient response to a previous treatment.

Important Safety Information

Warnings and Precautions

- Hypertension can occur with TAVALISSE treatment. Patients with pre-existing hypertension may be more susceptible to the hypertensive effects. Monitor blood pressure every 2 weeks until stable, then monthly, and adjust or initiate antihypertensive therapy for blood pressure control maintenance during therapy. If increased blood pressure persists, TAVALISSE interruption, reduction, or discontinuation may be required.
- Elevated liver function tests (LFTs), mainly ALT and AST, can occur with TAVALISSE. Monitor LFTs monthly during treatment. If ALT or AST increase to >3 x upper limit of normal, manage hepatotoxicity using TAVALISSE interruption, reduction, or discontinuation.
- Diarrhea occurred in 31% of patients and severe diarrhea occurred in 1% of patients treated with TAVALISSE. Monitor patients for the development of diarrhea and manage using supportive care measures early after the onset of symptoms. If diarrhea becomes severe (≥Grade 3), interrupt, reduce dose or discontinue TAVALISSE.
- Neutropenia occurred in 6% of patients treated with TAVALISSE; febrile neutropenia occurred in 1% of patients. Monitor the ANC monthly and for infection during treatment. Manage toxicity with TAVALISSE interruption, reduction, or discontinuation.
- TAVALISSE can cause fetal harm when administered to pregnant women. Advise pregnant women the potential risk to a fetus. Advise females of reproductive potential to use effective contraception during treatment and for at least 1 month after the last dose. Verify pregnancy status prior to initiating TAVALISSE. It is unknown if TAVALISSE or its metabolite is present in human milk. Because of the potential for serious adverse reactions in a breastfed child, advise a lactating woman not to breastfeed during TAVALISSE treatment and for at least 1 month after the last dose.

Drug Interactions

- Concomitant use of TAVALISSE with strong CYP3A4 inhibitors increases exposure to the major active metabolite of TAVALISSE (R406), which may increase the risk of adverse reactions. Monitor for toxicities that may require a reduction in TAVALISSE dose.
- It is not recommended to use TAVALISSE with strong CYP3A4 inducers, as concomitant use reduces exposure to R406.
- Concomitant use of TAVALISSE may increase concentrations of some CYP3A4 substrate drugs and may require a dose reduction of the CYP3A4 substrate drug.
- Concomitant use of TAVALISSE may increase concentrations of BCRP substrate drugs (eg, rosuvastatin) and P-Glycoprotein (P-gp) substrate drugs (eg, digoxin), which may require a dose reduction of the BCRP and P-gp substrate drug.

Adverse Reactions

- Serious adverse drug reactions in the ITP double-blind studies were febrile neutropenia, diarrhea, pneumonia, and hypertensive crisis, which occurred in 1% of TAVALISSE patients. In addition, severe adverse reactions occurred including dyspnea and hypertension (both 2%), neutropenia, arthralgia, chest pain, diarrhea, dizziness, nephrolithiasis, pain in extremity, toothache, syncope, and hypoxia (all 1%).
- Common adverse reactions (≥5% and more common than placebo) from FIT-1 and FIT-2 included: diarrhea, hypertension, nausea, dizziness, ALT and AST increased, respiratory infection, rash, abdominal pain, fatigue, chest pain, and neutropenia.

Please see www.TAVALISSE.com for full Prescribing Information.

**To report side effects of prescription drugs to the FDA,
visit www.fda.gov/medwatch or call 1-800-FDA-1088 (800-332-1088).**

TAVALISSE is a trademark of Rigel Pharmaceuticals, Inc.

About Rigel (www.rigel.com)
Rigel Pharmaceuticals, Inc., is a biotechnology company dedicated to discovering, developing and providing novel small molecule drugs that significantly improve the lives of patients with immune and hematologic disorders, cancer and rare diseases. Rigel's pioneering research focuses on signaling pathways that are critical to disease mechanisms. The company's first FDA approved product is TAVALISSE® (fostamatinib disodium hexahydrate), the only oral spleen tyrosine kinase (SYK) inhibitor, for the treatment of adult patients with chronic immune thrombocytopenia who have had an insufficient response to a previous treatment. Rigel's current clinical programs include a Phase 3 study of fostamatinib in autoimmune hemolytic anemia (AIHA); a recently completed Phase 1 study of R835, a proprietary molecule from its interleukin receptor associated kinase (IRAK) program; and an ongoing Phase 1 study of R552, a proprietary molecule from its receptor-interacting protein kinase (RIP1) inhibitor program. In addition, Rigel has product candidates in clinical development with partners Aclaris Therapeutics, AstraZeneca, BerGenBio ASA, and Daiichi Sankyo.

[1] The product for this use or indication is investigational and has not been proven safe or effective by any regulatory authority.

Forward Looking Statements
This release contains forward-looking statements relating to, among other things, the safety, tolerability, design, timing and results of Rigel's products, product candidates and clinical trials. Any statements contained in this press release that are not statements of historical fact may be deemed to be forward-looking statements. Words such as "planned", "will", "may", "expects", "anticipates", "estimates", "hopes", "believes" and similar expressions are intended to identify these forward-looking statements. These forward-looking statements are based on Rigel's current expectations and inherently involve significant risks and uncertainties. Actual results and the timing of events could differ materially from those anticipated in such forward looking statements as a result of these risks and uncertainties, which include, without limitation, risks and uncertainties associated with the commercialization and marketing of TAVALISSE; risks that the FDA, EMA or other regulatory authorities may make adverse decisions regarding fostamatinib; risks that TAVALISSE clinical trials may not be predictive of real-world results or of results in subsequent clinical trials; risks that TAVALISSE may have unintended side effects, adverse

reactions or incidents of misuses; the availability of resources to develop Rigel's product candidates; market competition; as well as other risks detailed from time to time in Rigel's reports filed with the Securities and Exchange Commission, including its Quarterly Report on Form 10-Q for the period ended September 30, 2019. Rigel does not undertake any obligation to update forward-looking statements and expressly disclaims any obligation or undertaking to release publicly any updates or revisions to any forward-looking statements contained herein.

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View original content to download multimedia:http://www.prnewswire.com/news-releases/rigel-announces-upcoming-data-presentations-at-the-61st-ash-annual-meeting--exposition-300953564.html

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