July 14, 2021

New TAVALISSE® Data Analyses To Be Presented at International Society on Thrombosis and Haemostasis (ISTH) 2021 Congress

--In a Post-hoc Analysis of Patients With at Least One Year of Treatment, 91% of ITP Patients Achieved Platelet Counts ≥ 50,000/µL and Incidence of Bleeding Events and Use of Rescue Therapy Decreased With Continuous Treatment--

SOUTH SAN FRANCISCO, Calif., July 14, 2021 /PRNewswire/ -- Rigel Pharmaceuticals, Inc. (Nasdaq: RIGL), today announced that data from its clinical development program for TAVALISSE® (fostamatinib disodium hexahydrate) tablets will be highlighted in two presentations at the upcoming International Society on Thrombosis and Haemostasis (ISTH) Virtual Congress, taking place online from July 17 – 21, 2021.

"For patients who remain on TAVALISSE for a year or more, we found that their initial response was not the final response and platelet counts continued to increase over time," said Wolfgang Dummer, MD, PhD, chief medical officer for Rigel. "This is due to TAVALISSE's mechanism of action, SYK inhibition, which blocks the autoimmune response that leads to platelet destruction, allowing the patient's platelet count to build in a natural and gradual way."

"The post-hoc analyses that will be presented at ISTH show that treatment with TAVALISSE results in continuously increasing platelet counts over the first year of treatment, highlighting the potential benefit for ITP patients of longer-term treatment," said Raul Rodriguez, Rigel's president and CEO. "By evaluating these patient groups, we are able to provide clinicians with further insight into how duration of therapy and treatment in earlier lines of therapy can potentially impact clinical outcomes."

Fostamatinib is an oral drug designed to inhibit spleen tyrosine kinase (SYK), a key signaling component of the autoimmune process that leads to platelet destruction in immune thrombocytopenia (ITP). 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.

All ePosters will be made available online during the Poster Networking Session on the conference website on Saturday, July 17, 2021, at 4:00 p.m. Eastern Time.

Abstract Presentation Number: PB0815
Title: *Time Course of Response to Fostamatinib, an Oral Spleen Tyrosine Kinase (SYK) Inhibitor for the Treatment of Immune Thrombocytopenia (ITP)*
Presenting Author: Michelle Sholzberg, MDCM, Division Head of Hematology-Oncology and the Medical Director of the Coagulation Laboratory at St. Michael's Hospital, Toronto

In a post-hoc analysis of the Rigel FIT Phase 3 program, for the 58 patients treated for ≥1 year, clinical endpoints were assessed in 3-month increments over the first year:

- Patients treated for ≥1 year, had a response rate (achieved a platelet count ≥50,000/μL) of 91% (53/58), 71% (41/58) within 12 weeks, with median platelet counts continuing to increase in each 3-month period of the first year of treatment.
- Incidence of bleeding events and use of rescue therapy continually decreased in these 58 patients over the first year of treatment with fostamatinib.

Abstract Presentation Number: PB0833
Title: *Treatment of Immune Thrombocytopenia (ITP) in the COVID-19 Era: Fostamatinib, an Oral Spleen Tyrosine Kinase (SYK) Inhibitor*
Presenting Authors: Nichola Cooper, Asad Charania, Alice Hart, Christine Ademokun, and Robert Numerof

A review of safety and efficacy data from the fostamatinib Phase 3 program evaluated dosing and titration, incidence of infections, thrombocytosis and thrombosis, and the immune response to pathogens to assess the potential benefits of TAVALISSE for the unique challenges of treating ITP during the pandemic. Findings include:

- A reduced need for office visits due to oral administration, easy titration, and low incidence of thrombocytosis.
- Fostamatinib's unique mechanism of action provide increased hemostasis and may reduce the risk of thrombosis.
- Fostamatinib is not an immunosuppressant drug.

In addition to the poster presentations, Rigel is participating in a virtual symposium:
Title: *Role of Spleen Tyrosine Kinase (SYK) and SYK Inhibition in Disease Pathways*
Date and Time: Sunday, July 18, 2021, 12:30 - 1:30 p.m. ET
Presenters:

- Dr. Vadim Markovtsov – SYK in Human Biology and Disease Pathways
- Dr. Nichola Cooper – Clinical Trials with SYK Inhibitors
- Dr. Craig Kessler, Moderator

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 include fatigue, 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. In addition to fostamatinib, 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 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 about 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, visitwww.fda.gov/medwatch **or call 1-800-FDA-1088 (800-332-1088).**

TAVALISSE and TAVLESSE are registered trademarks of Rigel Pharmaceuticals, Inc.

About Rigel
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 hematologic disorders, cancer and rare immune 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) tablets, 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. The product is also commercially available in Europe (TAVLESSE) and Canada (TAVALISSE) for the treatment of chronic immune thrombocytopenia in adult patients.

Fostamatinib is currently being studied in a Phase 3 clinical trial (NCT03764618) for the treatment of warm autoimmune hemolytic anemia (wAIHA)[1]; a Phase 3 clinical trial (NCT04629703) for the treatment of hospitalized high-risk patients with mild-to-moderate COVID-19[1]; an NIH/NHLBI-sponsored Phase 3 clinical trial for the treatment of COVID-19 in hospitalized patients on oxygen therapy, and a Phase 2 clinical trial for the treatment of COVID-19 being conducted by Imperial College London. An NIH/NHLBI-sponsored Phase 2 clinical trial for the treatment of hospitalized patients with COVID-19, in collaboration with Inova Health System, was recently completed.

Rigel's other clinical programs include its interleukin receptor-associated kinase (IRAK) inhibitor program, and a receptor-interacting serine/threonine-protein kinase (RIP1) inhibitor program in clinical development with partner Eli Lilly and Company. In addition, Rigel has product candidates in development with partners AstraZeneca, BerGenBio ASA, and Daiichi Sankyo.

For further information, visit www.rigel.com or follow us on Twitter or LinkedIn.

[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 potential benefit for ITP patients of longer-term treatment with TAVALISSE. 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 "potential", "may", "expects", 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 quarter ended March 31, 2021. In addition, the COVID-19 pandemic may result in further delays in Rigel's studies, trials and sales, or impact Rigel's ability to obtain supply of TAVALISSE. 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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