January 13, 2020

Rigel Pharmaceuticals Provides Business Update

- TAVALISSE® preliminary 4Q net product sales of approximately $13.8 mln, YoY increase of 90%; preliminary 4Q total revenues of approximately $15.4 mln

- Received European approval of fostamatinib disodium hexahydrate for adult patients with chronic ITP

- Phase 3 pivotal trial for warm AIHA on track with 20 patients enrolled, including 15 in last 2 months

SOUTH SAN FRANCISCO, Calif., Jan. 13, 2020 /PRNewswire/ -- Rigel Pharmaceuticals, Inc. (Nasdaq: RIGL) today provided a business update, including preliminary product revenue and total TAVALISSE® (fostamatinib disodium hexahydrate) bottles for the quarter; the European Commision's (EC) approval of fostamatinib disodium hexahydrate (fostamatinib) for adult patients with chronic immune thrombocytopenia (ITP); and enrollment progress in its Phase 3 pivotal trial for warm autoimmune hemolytic anemia (AIHA). The company's president and CEO, Raul Rodriguez, will provide a more detailed company overview during his presentation taking place on Thursday, January 16 at 7:30am PT at the 38th Annual J.P. Morgan Healthcare Conference.

"Rigel has built a solid foundation for continued growth, anchored by increased revenues from our U.S. commercial business. With European approval of fostamatinib, we expect to generate revenue from royalty payments beginning in the second half of the year, and importantly, we will receive a $20 million milestone from our partner, Grifols, S.A.," stated Mr. Rodriguez, Rigel's president and CEO. "Enrollment of our Phase 3 clinical trial in warm AIHA is accelerating with the ongoing activation of trial sites. This is an extremely attractive market for Rigel given the lack of an FDA-approved therapy and the synergies with our ITP commercial business. Additionally, we are making meaningful progress with our novel IRAK 1/4 and RIP1 inhibitor programs and are excited about their potential value."

Preliminary Financial Update

While Rigel is still in the process of determining final results for the fourth quarter of 2019, the company expects to report net product sales of approximately $13.8 million, compared to

$7.3 million in the same period of 2018, an increase of 90%.

Contract revenues from collaborations for the quarter ended December 31, 2019, are expected to be approximately $1.6 million, which consists of a $1.5 million fee earned pursuant to an amendment in October 2019 of the license and collaboration agreement with Aclaris dated August 27, 2015, as well as deferred revenue from its collaboration with Grifols related to the performance of certain research and development services.

For the fourth quarter 2019, Rigel expects to report total revenues of approximately $15.4 million.

The company expects to report cash, cash equivalents and short-term investments as of December 31, 2019 of approximately $98.0 million, compared to $128.5 million as of December 31, 2018.

The above information is preliminary, has not been audited and is subject to change upon completion of the audit of the company's financial statements as of and for the year ended December 31, 2019.

Commercial Update

Growing TAVALISSE in the U.S. Market

During the fourth quarter of 2019, a total of 1,518 bottles of TAVALISSE were sold in the U.S. of which 1,422 were shipped directly to patients and clinics. The refill rate at month 4 increased to approximately 54%, reflecting the benefit patients are experiencing as TAVALISSE is used more frequently in earlier line treatment and physicians become more experienced with its use.

Heading into 2020, Rigel is poised for further growth with plans to expand its salesforce by six people in key markets. In addition, the company intends to continue its ongoing data initiatives and leverage recently presented post-hoc data showing a 78% response rate in ITP patients who received TAVALISSE in second line use.

EC Approval of Fostamatinib and Product Launch

Rigel today announced that it has received EC approval of its marketing authorization application (MAA) for fostamatinib for the treatment of chronic immune thrombocytopenia in adult patients who are refractory to other treatments. With this approval, the company will receive a milestone payment of $20 million based on terms of its collaboration with Grifols, S.A. During the regulatory review process, Grifols began preparing to launch the product in the major European markets and is now able to begin the regulatory processes for marketing in the individual countries. The company expects to generate incremental revenue from European sales of fostamatinib in the second half of this year in the form of royalty payments.

Portfolio Update

Phase 3 Trial in Warm AIHA

Since the launch of FORWARD (Fostamatinib Research in Warm Antibody AIHA Disease), Rigel's Phase 3 pivotal trial in warm AIHA, the company has been working with clinics and regulatory authorities and has established a vast majority of the more than 100 clinical trial sites planned across 22 countries. With the number of opened sites growing rapidly, over 45 in the last three months, enrollment of the trial has accelerated as planned with 15 of 20 total

patients enrolled in the last two months. The trial remains on track to complete enrollment in mid-2020.

Clinical Development Pipeline

In the fourth quarter of 2019, Rigel announced results from its Phase 1 clinical trial of R835[1], its interleukin-1 receptor-associated kinase 1/4 (IRAK1/4) inhibitor. This novel molecule is the only asset in clinical development that is a dual inhibitor of IRAK1 and IRAK4 and has been shown preclinically to block inflammatory cytokine production in response to toll-like receptor (TLR) and the interleukin-1 receptor (IL-1R) family signaling. The Phase 1 trial established proof-of-mechanism by demonstrating the inhibition of inflammatory cytokine production in an LPS (lipopolysaccharide) challenge.

In addition, Rigel recently initiated a Phase 1 trial of its receptor-interacting protein kinase (RIP1) inhibitor, R552[1]. RIP1 is believed to play a critical role in necroptosis, a form of regulated cell death implicated in inflammatory and neurodegenerative diseases. Initial data from Rigel's ongoing Phase 1 suggests R552 has an attractive pharmacokinetic (PK) and safety profile with a half-life of approximately 15 hours. In earlier preclinical studies, the molecule was shown to prevent joint and skin inflammation in a RIP1 kinase-mediated murine model.

Both assets target pathways that are of high interest in the biopharma industry and are widely thought to have significant potential in the treatment of inflammatory and immune-mediated diseases.

38th Annual J.P. Morgan Webcast Presentation Details

Rigel's presentation will be webcast and is scheduled to take place Thursday, January 16 at 7:30am PT. To access the live and subsequently archived webcast, go to the Investor Relations section of the company's website at www.rigel.com. Please connect to the website several minutes prior to the start of the live webcast to ensure adequate time for any software download that may be necessary.

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 45,000 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 R835[1]

The investigational candidate, R835, is an orally available, potent and selective inhibitor of IRAK1 and IRAK4 that has been shown preclinically to block inflammatory cytokine production in response to toll-like receptor (TLR) and the interleukin-1 receptor (IL-1R) family signaling. TLRs and IL-1Rs play a critical role in the innate immune response, and dysregulation of these pathways can lead to a variety of inflammatory pathological conditions. R835 treatment demonstrates amelioration of clinical symptoms in multiple rodent models of inflammatory disease including psoriasis, arthritis, lupus, multiple sclerosis and gout. The safety and efficacy of R835 has not been established by the FDA or any healthcare authority.

About R552[1]

The investigational candidate, R552, is an orally available, potent and selective inhibitor of receptor-interacting protein kinase (RIP1). RIP1 is believed to play a critical role in necroptosis. Necroptosis is a form of regulated cell death where the rupturing of cells leads to the dispersion of their inner contents, which induces immune responses and enhances inflammation. In preclinical studies, R552 prevented joint and skin inflammation in a RIP1-mediated murine model of inflammation and tissue damage. The safety and efficacy of R552 has not been established by the FDA or any healthcare authority.

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, visitwww.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 clinical programs include a Phase 3 study of fostamatinib in warm autoimmune hemolytic anemia (AIHA); a recently completed Phase 1 study of R835[1], a proprietary molecule from its interleukin receptor associated kinase (IRAK) inhibitor program; and an ongoing Phase 1 study of R552[1], a proprietary molecule from its receptor-interacting protein kinase (RIP) 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 commercial success of TAVALISSE in the U.S.; Rigel's ability to broaden its pipeline of assets targeting immune-mediated diseases; Rigel's efforts to expand fostamatinib in Europe and to expand its salesforce in key markets; Rigel's regulatory and collaborative efforts in Europe to make fostamatinib available to ITP patients more globally; the utility of fostamatinib in other indications, including warm autoimmune hemolytic anemia; Rigel's ability to achieve development and commercial milestones; Rigel's expected operating results for the quarter ending and as of December 31, 2019, including net sales and cash, cash equivalents and short-term investments; expectations related to the market opportunity for ITP in the European market, and the design, timing, enrollment and results of Rigel's 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 "goals", "potential", "preliminary", "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 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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