May 5, 2021

Rigel Reports First Quarter 2021 Financial Results and Provides Business Update

- Reported positive topline results of fostamatinib in Phase 2 clinical trial in hospitalized patients with COVID-19

- Net product sales of $12.4 million and total revenues of $81.0 million

- Received upfront cash payment of $125 million from Eli Lilly for the exclusive license agreement for Rigel's RIP1 inhibitor program

- Conference call and webcast today at 4:30PM Eastern Time

SOUTH SAN FRANCISCO, Calif., May 5, 2021 /PRNewswire/ -- Rigel Pharmaceuticals, Inc. (Nasdaq: RIGL) today reported financial results for the first quarter ended March 31, 2021, including sales of TAVALISSE® (fostamatinib disodium hexahydrate) tablets, for the treatment of adults with chronic immune thrombocytopenia (ITP) who have had an insufficient response to a previous treatment.

"We continued to make significant progress in achieving important corporate and clinical milestones in the first quarter, which is a true testament to the dedication of our team to bring meaningful treatment options to those patients who need them the most," said Raul Rodriguez, Rigel's president and CEO. "Following our recent announcement of positive topline data from the Phase 2 trial of fostamatinib in hospitalized patients with COVID-19 and our partnership with Eli Lilly for our RIP1 inhibitor program, we are excited by the prospects that the rest of 2021 holds for Rigel."

Business Update

In April 2021, Rigel reported positive topline data from the multi-center, Phase 2 clinical trial evaluating the safety of fostamatinib, Rigel's oral spleen tyrosine kinase (SYK) inhibitor, for the treatment of hospitalized patients with COVID-19. The trial met its primary endpoint of safety, and showed broad and consistent improvement in numerous efficacy endpoints including mortality, ordinal scale assessment, and number of days in the ICU. The trial was conducted in collaboration with the National Heart, Lung, and Blood Institute (NHLBI), part of the National Institutes of Health (NIH), and Inova Health System. The NHLBI is expected to publish a full analysis of the trial data in a peer-reviewed journal. Rigel is discussing these

results with health authorities, including the U.S. Food and Drug Administration (FDA), and intends to apply for Emergency Use Authorization (EUA) for fostamatinib for the treatment of hospitalized COVID-19 patients.

Rigel's Phase 3 clinical trial to further evaluate fostamatinib in hospitalized patients with COVID-19 is currently enrolling. Rigel was awarded $16.5 million from the U.S. Department of Defense's (DOD) Joint Program Executive Office for Chemical, Biological, Radiological and Nuclear Defense (JPEO-CBRND) to support this Phase 3 clinical trial. The study is designed to evaluate fostamatinib for prevention of progression to severe disease in hospitalized patients with COVID-19 without respiratory failure that have certain high-risk prognostic factors. This multi-center, double-blind, placebo-controlled study will randomly assign patients to either fostamatinib plus standard of care (SOC) or matched placebo plus SOC (1:1). Treatment will be administered orally twice daily for 14 days. There will be a follow-up period to day 60. The primary endpoint of this study is the proportion of subjects who progress to severe/critical disease within 29 days. In addition, Rigel's COVID-19 program includes an investigator-sponsored trial currently being conducted by Imperial College London.

Rigel's FORWARD study, a Phase 3 pivotal trial of TAVALISSE in patients with warm autoimmune hemolytic anemia (wAIHA), has enrolled 72 of 90 patients as of May 5, 2021. If approved, TAVALISSE has the potential to be the first to market therapy for patients with wAIHA. The FDA has granted Fast Track designation as well as Orphan Drug designation to TAVALISSE for the treatment of wAIHA.

In the first quarter of 2021, 1,599 bottles of TAVALISSE were shipped to patients and clinics, an increase of 14% year over year. Net product sales for the first quarter decreased 2% year over year to $12.4 million. During the quarter, the company experienced typical first quarter reimbursement issues such as the resetting of co-pays and the Medicare donut hole, along with physician and patient access issues created by the COVID-19 pandemic. Incrementally, the company's net product sales were negatively impacted by a significant 235 bottle decrease in bottles remaining in its distribution channels compared to Q4 2020.

In April 2021, Rigel received a $125 million upfront cash payment from its collaboration with Eli Lilly and Company (Lilly), following the expiration of the waiting period under the Hart-Scott Rodino Antitrust Improvements Act of 1976. In February 2021, Rigel and Lilly entered into a global exclusive license agreement and strategic collaboration to develop and commercialize Rigel's R552, a receptor-interacting serine/threonine-protein kinase 1 (RIP1) inhibitor, for all indications including autoimmune and inflammatory diseases. Rigel and Lilly are currently focused on the planning for the initiation of a Phase 2 clinical trial. Pursuant to the collaboration, Lilly will also lead all clinical development of central nervous system (CNS) penetrating RIP1 inhibitors. Rigel is eligible to receive up to an additional $835 million in potential development, regulatory and commercial milestone payments, as well as tiered royalties that will vary depending upon Rigel's clinical development investment.

Rigel is also advancing the development of its IRAK1/4 program, where it intends to pursue both hematology/oncology and rare immune disease opportunities. Rigel has begun discussions with the FDA regarding initiating a Phase 2 clinical trial in low-risk myelodysplastic syndrome (MDS) and is also in discussions regarding academic medical collaborations in this indication. In rare immune diseases, the company is exploring opportunities including palmoplantar pustulosis (PPP), hidradenitis suppurativa (HS), and

others.

Rigel's partner Kissei Pharmaceutical Co., Ltd. (Kissei) has completed enrollment of its Phase 3 clinical trial of fostamatinib in adult Japanese patients with chronic ITP. In October 2018, Rigel and Kissei entered into an exclusive agreement to develop and commercialize fostamatinib in all current and potential indications in Japan, China, Taiwan, and the Republic of Korea.

Financial Update

For the first quarter of 2021, Rigel reported net income of $39.5 million, or $0.23 per basic share and $0.22 per diluted share, compared to net income of $21.2 million, or $0.13 per basic and diluted share, for the same period of 2020.

In the first quarter of 2021, total revenues were $81.0 million, consisting of $12.4 million in TAVALISSE net product sales, $65.6 million in contract revenues from collaborations and $3.0 million in government contract revenue.

Contract revenues from collaborations of $65.6 million for the first quarter of 2021 consisted of $60.6 million in revenue related to Rigel's license agreement with Lilly, $4.0 million in revenue related to the grant of non-exclusive license of a certain patent to an unrelated third-party company, and $1.0 million in revenue for the delivery of drug supply under its collaboration agreement with Grifols. Government contract revenue was related to the income that Rigel recognized pursuant to the agreement it entered into in January 2021 with the DOD to support Rigel's ongoing Phase 3 clinical trial of fostamatinib in hospitalized patients with COVID-19.

Rigel reported total costs and expenses of $39.3 million in the first quarter of 2021, compared to $34.7 million for the same period in 2020. The increase in costs and expenses was primarily due to increases in personnel-related costs, stock-based compensation expense, and research and development costs related to its ongoing Phase 3 clinical trial in hospitalized patients with COVID-19 and development of its IRAK 1/4 inhibitor program, partially offset by the decrease in research and development costs due to the completion of a Phase 1 clinical trial for its RIP1 inhibitor program.

As of March 31, 2021, Rigel had cash, cash equivalents and short-term investments of $39.3 million, compared to $57.3 million as of December 31, 2020. Cash, cash equivalents and short-term investments as of March 31, 2021, does not include the $125.0 million upfront payment received from Lilly in April 2021.

Conference Call and Webcast with Slides Today at 4:30pm Eastern Time

Rigel will hold a live conference call and webcast today at 4:30pm Eastern Time (1:30pm Pacific Time).

Participants can access the live conference call by dialing (877) 407-3088 (domestic) or (201) 389-0927 (international). The conference call and accompanying slides will also be webcast live and can be accessed from the Investor Relations section of the company's website at www.rigel.com. The webcast will be archived and available for replay after the call via the Rigel website.

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 COVID-19 & SYK Inhibition

COVID-19 is the infectious disease caused by Severe Acute Respiratory Syndrome Coronavirus-2 (SARS-CoV-2). SARS-CoV-2 primarily infects the upper and lower respiratory tract and can lead to acute respiratory distress syndrome (ARDS). Additionally, some patients develop other organ dysfunction including myocardial injury, acute kidney injury, shock resulting in endothelial dysfunction and subsequently micro and macrovascular thrombosis.[1] Much of the underlying pathology of SARS-CoV-2 is thought to be secondary to a hyperinflammatory immune response associated with increased risk of thrombosis.[2]

SYK is involved in the intracellular signaling pathways of many different immune cells. Therefore, SYK inhibition may improve outcomes in patients with COVID-19 via inhibition of key Fc gamma receptor (FcγR) and c-type lectin receptor (CLR) mediated drivers of pathology, such as inflammatory cytokine release by monocytes and macrophages, production of neutrophil extracellular traps (NETs) by neutrophils, and platelet aggregation.[3,4,5] Furthermore, SYK inhibition in neutrophils and platelets may lead to decreased thromboinflammation, alleviating organ dysfunction in critically ill patients with COVID-19.

For more information on Rigel's comprehensive clinical program in COVID-19, go to: https://www.rigel.com/pipeline/proprietary-programs/covid-19

About R552

The investigational candidate, R552, is an orally available, potent and selective inhibitor of receptor-interacting serine/threonine-protein kinase 1 (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 R835

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 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 and TAVLESSE are registered trademarks 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 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 for the treatment of warm autoimmune hemolytic anemia (wAIHA)[6]; a Phase 3 clinical trial for the treatment of hospitalized patients with COVID-19[6]; an NIH/NHLBI-sponsored Phase 2 clinical trial for the treatment of hospitalized patients with COVID-19, in collaboration with Inova Health System; and a Phase 2 clinical trial for the treatment of COVID-19 being conducted by Imperial College London.

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.

1. Berlin DA, Gulick RM, Martinez FJ. Severe Covid-19. N Engl J Med 2020
2. Becker RC. COVID-19 Update: COVID-19 associated coagulopathy. Journal of Thrombosis and Thrombolysis May 15, 2020. DOI: https://doi.org/10.1007/s11239-020-02134-3
3. Hoepel W. et al. Anti-SARS-CoV-2 IgG from severely ill COVID-19 patients promotes macrophage hyper-inflammatory responses. bioRxiv July 13, 2020. DOI: https://doi.org/10.1101/2020.07.13.190140
4. Sung P-S and Hsieh S-L (2019) CLEC2 and CLEC5A: Pathogenic Host Factors in Acute Viral Infections. Front. Immunol. 10:2867. DOI: https://doi.org/10.3389/fimmu.2019.02867
5. Behnen M. Immobilized Immune Complexes Induce Neutrophil Extracellular Trap Release by Human Neutrophil Granulocytes via Fcγ RIIIB and Mac-1. The Journal of Immunology July 2014. DOI: https://doi.org/10.4049/jimmunol.1400478
6. *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. and TAVLESSE in Europe; Rigel's intention to apply for EUA for fostamatinib for the treatment of hospitalized COVID-19 patients; Rigel's ability to achieve development, regulatory and commercial milestone payments, as well as tiered royalties; expectations related to the market opportunity for fostamatinib as a COVID-19 therapeutic; Rigel's ability to further develop its clinical stage and early stage product candidates and programs; and Rigel's partnering efforts. 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 Annual Report on Form 10-K for the year ended December 31, 2020. 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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RIGEL PHARMACEUTICALS, INC.
STATEMENTS OF OPERATIONS
(in thousands, except per share amounts)

	Three Months Ended March 31,	
	2021	**2020**
	(unaudited)	
Revenues:		
Product sales, net	$ 12,376	$ 12,680
Contract revenues from collaborations	65,642	43,081
Government contract	3,000	—
Total revenues	81,018	55,761
Costs and expenses:		
Cost of product sales	316	155
Research and development (see Note A)	16,826	16,149
Selling, general and administrative (see Note A)	22,121	18,430
Total costs and expenses	39,263	34,734
Income from operations	41,755	21,027
Interest income	1	358
Interest expense	(485)	(142)
Income before income taxes	41,271	21,243
Provision for income taxes	1,771	-
Net income	$ 39,500	$ 21,243
Net loss per share, basic and diluted		
Basic	$ 0.23	$ 0.13
Diluted	$ 0.22	$ 0.13
Weighted average shares used in computing net loss per share, basic and diluted		
Basic	169,800	168,469
Diluted	176,069	168,568

Note A
Stock-based compensation expense included in:

Selling, general and administrative	$ 2,053	$ 1,330
Research and development	586	694
	$ 2,639	$ 2,024

SUMMARY BALANCE SHEET DATA
(in thousands)

	March 31, 2021	**December 31, 2020 (1)**
Cash, cash equivalents and short-term investments	$ 39,345	$ 57,327
Total assets	215,993	110,378
Stockholders' equity	78,298	34,026

(1) Derived from audited financial statements

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