May 3, 2022

Rigel Reports First Quarter 2022 Financial Results and Provides Business Update

- *First quarter TAVALISSE® net product sales of $16.2 million and total revenues of $16.7 million*
- *Topline data from FORWARD study, a pivotal, Phase 3 clinical trial of fostamatinib in warm autoimmune hemolytic anemia (wAIHA), on track for mid-2022*
- *Management to host a conference call and webcast today at 4:30 p.m. Eastern Time and will be joined by Key Opinion Leader and FORWARD trial investigator, Caroline Piatek, M.D.*

SOUTH SAN FRANCISCO, Calif., May 3, 2022 /PRNewswire/ -- Rigel Pharmaceuticals, Inc. (Nasdaq: RIGL) today reported financial results for the first quarter ended March 31, 2022, 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.

"Rigel delivered on a solid quarter of TAVALISSE sales in ITP, and we believe we are well positioned for increasing sales in upcoming quarters," said Raul Rodriguez, Rigel's president and CEO. "wAIHA is a highly synergistic indication to ITP and we are very excited to be reporting Phase 3 fostamatinib results from our wAIHA trial in mid-2022. If approved, the addition of fostamatinib as a first-to-market therapy in wAIHA is a key next step in building a world-class hem-onc franchise."

Business Update

- In the first quarter of 2022, 1,836 bottles of TAVALISSE were shipped directly to patients and clinics, representing the highest number of bottles shipped to patients and clinics in a quarter since launch and an increase of 15% compared to the first quarter of 2021. Rigel believes these achievements are the result of several factors, including Rigel's salesforce expansion completed in September 2021 and a substantial increase in in-person physician interactions in the quarter, and an expansion of market access for TAVALISSE to now include broad commercial coverage, with preferred status on three key national formularies.

- Rigel is on track to report topline data from its FORWARD trial, a pivotal Phase 3

clinical trial of fostamatinib, an oral SYK inhibitor, in patients with wAIHA in mid-2022. If the data is positive, Rigel expects to proceed with regulatory filings and if approved, fostamatinib has the potential to be the first-to-market therapy for patients with wAIHA in 2023.

- In the first quarter of 2022, Rigel announced the publication of data in the *American Journal of Hematology* from the open label, multicenter, Phase 2 clinical study of fostamatinib in adults with wAIHA who had failed at least one prior treatment.

- Rigel's pivotal Phase 3 clinical trial evaluating fostamatinib in high-risk patients hospitalized with COVID-19 has enrolled 268 of the targeted 308 patients as of May 2, 2022. Due to the recent decline in COVID-19 hospitalizations, Rigel is reviewing strategies to complete enrollment and report topline results before the end of the year, including potentially completing the trial with fewer patients than the initial targeted enrollment of 308 patients.

- New data on fostamatinib in hospitalized patients with COVID-19 will be presented by collaborators from the National Institutes of Health and Inova Fairfax Hospital at the American Thoracic Society 2022 Annual Meeting being held in San Francisco, May 13 – 18, 2022.

- Startup activities are ongoing in Rigel's open-label, Phase 1b clinical trial of R289, a potent and selective IRAK1/4 inhibitor, in patients with lower-risk myeloid dysplastic syndrome (LR-MDS) who are refractory/resistant to prior therapies. The primary endpoint for this trial is safety with key secondary endpoints including preliminary efficacy and evaluation of pharmacokinetic properties. Rigel will also collect key biomarker data to further characterize R289's mechanism of action in LR-MDS.

- Partner Eli Lilly continues to advance R552, a potent and selective RIPK1 inhibitor, with the initial Phase 2 study in an immunologic disease indication now anticipated to begin in the first quarter of 2023. RIPK1 is implicated in a broad range of key inflammatory cellular processes and plays a key role in tumor necrosis factor (TNF) signaling, especially in the induction of pro-inflammatory necroptosis.

- On April 27, 2022, Rigel's partner Kissei Pharmaceutical Co., Ltd. (Kissei) announced that a new drug application (NDA) was submitted to Japan's Pharmaceuticals and Medical Devices Agency (PMDA) for fostamatinib in chronic ITP. Rigel has an exclusive license and supply agreement with Kissei 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 2022, Rigel reported a net loss of $27.4 million, or $0.16 per basic and diluted share, compared to a net income of $39.5 million, or $0.23 per basic share and $0.22 per diluted share for the same period of 2021.

In the first quarter of 2022, total revenues were $16.7 million, consisting of $16.2 million in TAVALISSE net product sales and $0.5 million in contract revenues from collaborations. TAVALISSE net product sales of $16.2 million increased by 31% from $12.4 million in the first quarter of 2021.

Rigel reported total costs and expenses of $43.0 million in the first quarter of 2022, compared to $39.3 million for the same period in 2021. The increase in costs and expenses was primarily due to increased commercial activities related to the sales force expansion in late 2021, increased research and development costs related to the development of Rigel's IRAK 1/4 inhibitor program, and increased personnel related costs and stock-based compensation expense, partially offset by decreased research and development costs related to the Phase 3 clinical trial of fostamatinib for wAIHA and the ongoing Phase 3 clinical trial of fostamatinib in hospitalized patients with COVID-19.

As of March 31, 2022, Rigel had cash, cash equivalents and short-term investments of $107.5 million, compared to $125.0 million as of December 31, 2021.

Conference Call and Webcast Today at 4:30pm Eastern Time, with KOL and FORWARD Trial Investigator, Caroline Piatek, M.D.

Rigel will hold a live conference call and webcast today at 4:30pm Eastern Time (1:30pm Pacific Time) to discuss financial results and provide an update on the business. The conference call will also feature a presentation by Caroline Piatek, M.D., Associate Professor of Clinical Medicine, Jane Anne Nohl Division of Hematology at the Keck School of Medicine of the University of Southern California, Key Opinion Leader, and FORWARD trial investigator. Dr. Piatek will discuss the current treatment landscape, unmet medical need, patient journey and how she may incorporate fostamatinib, if approved, into clinical practice in wAIHA.

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 lead to the destruction of the body's own red blood cells. Warm antibody AIHA (wAIHA), which is the most common form of AIHA, is characterized by the presence of antibodies that react with the red blood cell surface at body temperature. wAIHA affects approximately 36,000 adult patients in the U.S.[1] and can be a severe, debilitating disease. To date, there are no disease-targeted therapies approved for wAIHA, despite the unmet medical need that exists for these patients.

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.[2] 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.[3]

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 pro-inflammatory cytokine release by monocytes and macrophages, production of neutrophil extracellular traps (NETs) by neutrophils, and platelet aggregation.[4,5,6,7] Furthermore, SYK inhibition in neutrophils and platelets may lead to decreased thrombo-inflammation, 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 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.TAVALISSEUSPI.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, the United Kingdom (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)[8]; a Phase 3 clinical trial (NCT04629703) for the treatment of hospitalized high-risk patients with COVID-19[8]; and an NIH/NHLBI-sponsored Phase 3 clinical trial (ACTIV-4 Host Tissue Trial, NCT04924660) for the treatment of COVID-19 in hospitalized patients.

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

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

1. Prevalence: A. Zanella, et al, *haematologica* 2014; 99(10); % Warm AIHA: T. Kalfa; Hematology Am Soc Hematol Educ Program. 2016 Dec 2; 2016(1): 690–697
2. Berlin DA, Gulick RM, and Martinez FJ. *Severe Covid-19*. N Engl J Med 2020. DOI: https://doi.org/10.1056/NEJMcp2009575
3. 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
4. Hoepel W et al. *High titers and low fucosylation of early human anti–SARS-CoV-2 IgG promote inflammation by alveolar macrophages*. Science Translational Medicine 02 Jun 2021. DOI: https://www.doi.org/10.1126/scitranslmed.abf8654
5. Sung P-S and Hsieh S-L. *CLEC2 and CLEC5A: Pathogenic Host Factors in Acute Viral Infections*. Frontiers in Immunology December 6, 2019. DOI: https://doi.org/10.3389/fimmu.2019.02867
6. Strich J et al. *Fostamatinib Inhibits Neutrophils Extracellular Traps Induced by COVID-19 Patient Plasma: A Potential Therapeutic*. Journal of Infectious Disease March 15, 2021. DOI: https://doi.org/10.1093/infdis/jiaa789
7. Bye AP et al. *Aberrant glycosylation of anti-SARS-CoV-2 IgG is a pro-thrombotic stimulus for platelets*. BioRxiv March 26, 2021. DOI: https://doi.org/10.1101/2021.03.26.437014
8. *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 timing of reporting topline data from the FORWARD trial in patients with wAIHA; enrollment and reporting of data from the Company's Phase 3 clinical trial of fostamatinib in hospitalized COVID-19 patients; the commercial success of TAVALISSE in the U.S. and TAVLESSE in Europe, including expectations related to the potential and market opportunity for fostamatinib as therapeutic for, among other things, wAIHA and COVID-19, the commercialization of fostamatinib in international markets, Rigel's ability to further develop its clinical stage and early-stage product candidates and programs including its IRAK1/4 inhibitor program, 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, 2021 and subsequent filings. 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, except as required by law.

Contact for Investors & Media:
Jodi Sievers - Rigel Pharmaceuticals
Phone: 650.624.1232
Email: ir@rigel.com

RIGEL PHARMACEUTICALS, INC.
STATEMENTS OF OPERATIONS
(in thousands, except per share amounts)

	Three Months Ended March 31,	
	2022	**2021**
	(unaudited)	
Revenues:		
Product sales, net	$ 16,197	$ 12,376
Contract revenues from collaborations	538	65,642
Government contract	-	3,000
Total revenues	16,735	81,018
Costs and expenses:		
Cost of product sales	121	316
Research and development (see Note A)	15,474	16,826
Selling, general and administrative (see Note A)	27,401	22,121
Total costs and expenses	42,996	39,263
Income (loss) from operations	(26,261)	41,755
Interest income	21	1
Interest expense	(1,205)	(485)
Income (loss) before income taxes	(27,445)	41,271
Provision for income taxes	-	1,771
Net income (loss)	$ (27,445)	$ 39,500
Net income (loss) per share, basic and diluted		
Basic	$ (0.16)	$ 0.23
Diluted	$ (0.16)	$ 0.22
Weighted average shares used in computing net income (loss) per share, basic and diluted		
Basic	171,774	169,800
Diluted	171,774	176,069

Note A

Stock-based compensation expense included in:		
Selling, general and administrative	$ 2,739	$ 2,053
Research and development	468	586
	$ 3,207	$ 2,639

SUMMARY BALANCE SHEET DATA
(in thousands)

	March 31, 2022	**December 31, 2021 (1)**
	(unaudited)	
Cash, cash equivalents and short-term investments	$ 107,519	$ 124,967
Total assets	149,074	167,328
Stockholders' equity	6,798	30,374

(1) Derived from audited financial statements

View original content to download multimedia:https://www.prnewswire.com/news-releases/rigel-reports-first-quarter-2022-financial-results-and-provides-business-update-301538880.html

SOURCE Rigel Pharmaceuticals, Inc.