March 2, 2021

Rigel Reports Fourth Quarter and Full Year 2020 Financial Results and Provides Business Update

- Fourth quarter total revenues of $18.5 million; full year total revenues of $108.6 million

- Rigel will receive an upfront cash payment of $125 million from Lilly for the strategic collaboration to develop Rigel's RIP1 inhibitor program

- Launched a Phase 3 clinical trial of fostamatinib in hospitalized COVID-19 patients

- Top line data from NIH/NHLBI-sponsored Phase 2 trial of fostamatinib in COVID-19 patients expected in April 2021

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

SOUTH SAN FRANCISCO, Calif., March 2, 2021 /PRNewswire/ -- Rigel Pharmaceuticals, Inc. (Nasdaq: RIGL) today reported financial results for the fourth quarter and full year ended December 31, 2020, 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.

"Our team has shown its resilience as we continue to execute on our mission to serve patients who have diseases where few or no approved treatment options exist," said Raul Rodriguez, Rigel's president and CEO. "Despite the challenges brought on by 2020, we successfully expanded our global ITP reach and positioned ourselves for potential success in wAIHA, announced a major collaboration with Lilly to develop RIP1 inhibitors, and launched a comprehensive COVID-19 clinical program which has gained the support from the NIH, DOD, and several universities. Importantly, we also continued to explore opportunities in immunology, and more recently heme-onc, with our IRAK 1/4 inhibitor program."

Business Update

In February 2021, Rigel and Eli Lilly (Lilly) announced a global strategic collaboration to co-develop and commercialize R552, Rigel's receptor-interacting serine/threonine-protein kinase 1 (RIP1) inhibitor, for all indications, including autoimmune and inflammatory diseases. In addition, Lilly will lead the development and commercialization of all RIP1 inhibitors in central nervous system (CNS) indications. Under the terms of the agreement, Lilly will pay an upfront cash payment to Rigel of $125 million and is eligible to receive up to $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 launched a Phase 3 clinical trial to evaluate fostamatinib for the treatment of hospitalized COVID-19 patients. The Phase 3 trial is designed to evaluate the safety and efficacy of fostamatinib in hospitalized COVID-19 patients without respiratory failure that have certain high-risk prognostic factors. This multi-center, double-blind, placebo-controlled, adaptive design study is expected to enroll over 300 evaluable patients that will be randomly assigned 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.

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 Rigel's Phase 3 clinical trial of fostamatinib in hospitalized COVID-19 patients.

The Phase 2 clinical trial of fostamatinib in hospitalized COVID-19 patients sponsored by the National Heart, Lung, and Blood Institute (NHLBI), part of the National Institutes of Health (NIH), in collaboration with Inova Health System has currently enrolled 57 patients. This is a double-blind, placebo-controlled Phase 2 clinical trial that is randomly assigning fostamatinib plus standard of care (SOC) or matched placebo plus SOC (1:1) to approximately 60 evaluable patients who are a 5 to 7 on the 8-point ordinal scale (requiring supplemental oxygen via nasal canula or non-invasive ventilation, requiring mechanical ventilation or extracorporeal membrane oxygenation). The primary endpoint of this study is cumulative incidence of serious adverse events (SAE) through day 29. The NHLBI and Rigel expect to report topline data from this clinical trial in April 2021.

Imperial College London's ongoing Phase 2 clinical trial of fostamatinib in hospitalized COVID-19 patients is a two-stage open label, controlled trial with patients randomized (1:1:1) to fostamatinib, ruxolitinib, or standard of care. The study has currently enrolled 106 patients. Treatment will be administered twice daily for 14 days and patients will receive a follow-up assessment at day 14 and day 28 after the first dose. The primary objective will be to determine the efficacy of fostamatinib and the efficacy of ruxolitinib compared to standard of care to reduce the proportion of hospitalized patients progressing from mild or moderate to severe COVID-19 pneumonia.

Enrollment is progressing in Rigel's FORWARD study, a Phase 3 pivotal trial of TAVALISSE in warm autoimmune hemolytic anemia (wAIHA). The study has enrolled 66 of 90 patients targeted for enrollment. Rigel has reached agreement with the U.S. Food and Drug Administration (FDA) on the durable response measure for the primary efficacy endpoint of the study as well as the inclusion of additional secondary endpoints. The company recently announced that the FDA had granted Fast Track designation to TAVALISSE for the

treatment of wAIHA. The FDA previously granted TAVALISSE Orphan Drug designation for this indication. If approved, TAVALISSE may be the first-to-market therapy for patients with wAIHA.

Rigel continues to advance the development of its IRAK1/4 program, which includes R835, an orally available, potent and selective inhibitor that inhibits both IRAK1 and IRAK4. The company is currently identifying therapeutic opportunities in the areas of hematology/oncology and rare immunology diseases.

In November 2020, Rigel and its partner Medison Pharma announced that Health Canada approved the new drug submission (NDS) for TAVALISSE for the treatment of thrombocytopenia in adult patients with chronic ITP who have had an insufficient response to other treatments. Medison is also anticipating a decision on a New Drug Application (NDA) in Israel in Q2 2021. In July 2020, Rigel and its partner Grifols S.A. announced the launch of TAVLESSE in Germany and the United Kingdom following approval by the European Commission in January 2020. Currently, TAVALISSE is in a Phase 3b clinical trial in Japan with Rigel's partner Kissei. This trial is required for approval by the Pharmaceuticals and Medical Devices Agency (PMDA) in Japan.

Financial Update

For the fourth quarter of 2020, Rigel reported a net loss of $19.2 million, or $0.11 per share, compared to net loss of $17.2 million, or $0.10 per share, in the same period of 2019.

In the fourth quarter of 2020, total revenues were $18.5 million, consisting of $17.8 million in TAVALISSE net product sales and $697,000 in contract revenues from collaborations. TAVALISSE net product sales of $17.8 million increased by 28% from $13.8 million in the fourth quarter of 2019. Contract revenues from collaborations of $697,000 for the fourth quarter of 2020 consisted of $500,000 from Grifols related to an option for commercialization in additional territories and $197,000 in revenues earned from the performance of certain research and development services from Rigel's collaboration agreement with Grifols.

Rigel reported total costs and expenses of $37.3 million in the fourth quarter of 2020, compared to $32.7 million for the same period in 2019. The increase in costs and expenses was due to the increase in research and development costs primarily related to the continued work on Rigel's Phase 1 clinical trial in IRAK 1/4 inhibitor program, as well as its recently launched Phase 3 clinical trial for hospitalized COVID-19 patients, partially offset by the decrease in costs on Rigel's ongoing Phase 3 clinical trial in wAIHA.

For the full year ended December 31, 2020, Rigel reported a net loss of $29.7 million, or $0.18 per share, compared to a net loss of $66.9 million, or $0.40 per share, for the same period of 2019.

Rigel reported total revenues of $108.6 million for the year ended December 31, 2020, compared to $59.3 million in the same period of 2019. Total revenues for the year ended December 31, 2020 consisted of $61.7 million in TAVALISSE net product sales and $46.9 million in contract revenues from collaborations. TAVALISSE net product sales of $61.7 million increased by 41% from $43.8 million in 2019. The increase in contract revenues from collaborations related to revenue from the upfront fee previously received in 2019, as well as the milestone payment received from Grifols in the first quarter of 2020 upon EC approval of the MAA for fostamatinib in Europe, and the $2.1 million in contract revenues for the

achievement of a milestone in accordance with the amended license and collaboration agreement with Daiichi-Sankyo, partially offset by the developmental and commercial milestones from its various collaborative partners in 2019.

Total costs and expenses for the year ended December 31, 2020, were $137.6 million, compared to $128.4 million, for the same period of 2019. The increase in total costs and expenses was primarily related to the increases in research and development costs due to the completion of Rigel's Phase 1 clinical trial in RIP1 inhibitor program, ongoing work on recently launched Phase 3 clinical trial for hospitalized COVID-19 patients, continued work on Rigel's Phase 1 clinical trial in IRAK 1/4 inhibitor program and ongoing Phase 3 clinical trials in wAIHA, as well as increases in personnel-related costs, partially offset by decreases in various third-party costs due to the COVID-19 pandemic.

As of December 31, 2020, Rigel had cash, cash equivalents and short-term investments of $57.3 million, compared to $98.1 million as of December 31, 2019.

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.

About R552[6]

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[6]

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).

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

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 ability to achieve development, regulatory and commercial milestone payments, as well as tiered royalties; Rigel's expected operating results for the year ending and as of December 31, 2020, including net sales and cash, cash equivalents and short-term investments; expectations related to the market opportunity for fostamatinib as a COVID-19 therapeutic; Rigel's ability to further develop its clinical stage product candidates; 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 Quarterly Report on Form 10-Q for the period ended September 30, 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 December 31,		Year Ended December 31,	
	2020	2019	2020	2019
	(unaudited)			
Revenues:				
	$	$		$
Product sales, net	17,753	13,829	61,696	43,772
Contract revenues from collaborations	697	1,571	46,925	15,516
Total revenues	18,450	15,400	108,621	59,288
Costs and expenses:				
Cost of product sales	321	178	895	906
Research and development (see Note A)	15,138	14,247	60,101	52,885
Selling, general and administrative (see Note A)	21,818	18,312	76,598	74,588
Total costs and expenses	37,277	32,737	137,594	128,379
Loss from operations	(18,827)	(17,337)	(28,973)	(69,091)
Interest income	19	464	582	2,532
Interest expense	(429)	(327)	(1,353)	(335)
	$	$	$	$
Net loss	(19,237)	(17,200)	(29,744)	(66,894)
	$	$	$	$
Net loss per share, basic and diluted	(0.11)	(0.10)	(0.18)	(0.40)
Weighted average shares used in computing net loss per share, basic and diluted	169,039	167,619	168,754	167,400

Note A

Stock-based compensation expense included in:				
	$	$	$	$
Selling, general and administrative	1,242	934	5,223	6,453
Research and development	388	477	2,072	2,662
	$	$	$	$
	1,630	1,411	7,295	9,115

SUMMARY BALANCE SHEET DATA
(in thousands)

	December 31,	
	2020	2019
	$	$
Cash, cash equivalents and short-term investments	57,327	98,078
Total assets	110,378	147,569
Stockholders' equity	34,026	53,815

View original content to download multimedia:http://www.prnewswire.com/news-releases/rigel-reports-fourth-quarter-and-full-year-2020-financial-results-and-provides-business-update-301238978.html

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