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Rigel Initiates Phase 3 Studies of Fostamatinib in ITP

SOUTH SAN FRANCISCO, Calif., July 16, 2014 /PRNewswire/ -- Rigel Pharmaceuticals, Inc. (Nasdaq: RIGL) today announced the initiation of a Phase 3 clinical program for its oral SYK inhibitor, fostamatinib, in patients with ITP (immune thrombocytopenic purpura). The focus of these clinical studies is to evaluate the potential of fostamatinib to increase the platelet counts of patients with chronic ITP. Fostamatinib may provide a novel therapeutic for the underlying cause of this autoimmune disease of the blood.

"Based on our extensive clinical experience with this product candidate, which includes more than 4,500 patient-years of data, we hope to demonstrate that fostamatinib can provide a new treatment option for patients with chronic ITP," said James M. Gower, chairman and chief

executive officer of Rigel. Results of Rigel's Phase 2 clinical study, published in *Blood* (volume 113, number 14), showed that fostamatinib significantly increased the platelet counts of certain ITP patients, including those who had failed other currently available agents.

Fostamatinib in ITP Phase 3 Program Design

A total of 150 ITP patients will be randomized into two identical multi-center, double-blind, placebo-controlled clinical studies. The patients will have been diagnosed with persistent or chronic ITP, and have blood platelet counts consistently below 30,000 per microliter of blood. Two thirds of the subjects will receive fostamatinib orally at 100 mg bid (twice daily), the other third will receive placebo on the same schedule. Subjects are expected to remain

on treatment for 24 weeks. At week 4 of treatment, subjects who meet certain platelet count and tolerability thresholds will have their dosage of fostamatinib (or corresponding placebo) increased to 150 mg bid.

The primary efficacy endpoint of this program is a stable platelet response by week 24 with platelet counts at or above 50,000 per microliter of blood for at least 4 of the final 6 qualifying blood draws. Results are expected at year-end 2015.

Immune Thrombocytopenic Purpura

Chronic ITP affects an estimated 60,000 – 125,000 people in the US. In patients with ITP, the immune system attacks and destroys the body's own blood platelets, which play an active role in blood clotting and healing. ITP patients can suffer extraordinary bruising, bleeding and fatigue as a result of low platelet counts. Current therapies for ITP include steroids, blood platelet production boosters (TPOs) and splenectomy. Fostamatinib is the only potential therapy that may address the autoimmune basis of the disease.

Taken in tablet form, fostamatinib blocks the activation of SYK kinase inside immune cells. ITP causes the body to produce antibodies that attach to healthy platelets in the blood stream. Immune cells recognize these antibodies and affix to them, which activates the SYK enzyme inside the immune cell, and triggers the destruction of the antibody and the attached platelet. When SYK is inhibited by fostamatinib, it interrupts this immune cell function and allows the platelets to escape destruction.

About Rigel (www.rigel.com)

Rigel Pharmaceuticals, Inc. is a clinical-stage drug development company that discovers and develops novel, small-molecule drugs for the treatment of inflammatory and autoimmune diseases, as well as muscle disorders. Rigel's pioneering research focuses on intracellular signaling pathways and related targets that are critical to disease mechanisms. Rigel currently has five product candidates in development: fostamatinib, an oral SYK inhibitor in Phase 3 clinical trials for ITP and expected to enter into a Phase 2 clinical trial for IgA nephropathy in the second half of 2014; R348, a topical JAK/SYK inhibitor currently in Phase 2 clinical trials for dry eye; R118, an AMPK activator in Phase 1 development; and two oncology product candidates in Phase 1 development with partners BerGenBio and Daiichi Sankyo.

This press release contains "forward-looking" statements, including, without limitation, statements related to Rigel's clinical development plans, including the timing, design and nature of planned clinical trials and the timing and nature of results of those trials, as well as the potential activity of fostamatinib with respect to ITP and the ability of fostamatinib to provide a new treatment option for patients with chronic ITP. 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 "planned," "will," "may," "expect," 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, the availability of resources to develop Rigel's product candidates, Rigel's need for additional capital in the future to sufficiently fund Rigel's operations and research, the uncertain timing of completion of and the success of clinical trials, market competition, risks associated with and Rigel's dependence on Rigel's corporate

partnerships, 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, 2014. 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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