International Trial Shows Clinical Efficacy of Recombinant Fusion Protein Linking Coagulation Factor IX with Albumin (rIX-FP) for Once Weekly Prophylaxis in Patients with Severe Hemophilia B

Results of Phase I/II PROLONG-9FP trial program also demonstrate efficacy in treatment of bleeding episodes and improved pharmacokinetics of rIX-FP compared to current treatment options; recombinant albumin fusion platform forms basis of innovation

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Data presented by CSL Behring today showed clinical efficacy of a once-weekly dosing regimen of recombinant fusion protein linking coagulation Factor IX with albumin (rIX-FP). Results of the study were presented during an oral session at the International Society on Thrombosis and Haemostasis (ISTH) congress in Amsterdam.

CSL Behring, in collaboration with its parent company, CSL Limited (ASX: CSL), is developing rIX-FP for the prophylaxis and treatment of bleeding episodes in patients with hemophilia B as part of the PROLONG-9FP clinical study program.

"Hemophilia B is characterized by factor IX deficiency, which prevents normal blood clotting. People with this condition require factor IX infusions two to three times a week to achieve a significant reduction in bleeding," said Professor Uri Martinowitz, Director of the Institute of Thrombosis and Hemostasis and the Israeli National Hemophilia Center at Chaim Sheba Medical Center in Tel Hashomer, Israel. "This trial showed that less frequent infusions were needed with CSL Behring's investigational rIX-FP compared to currently available FIX products used on-demand or prophylactically to prevent or treat bleeding episodes."

The trial enrolled 17 hemophilia B patients, 13 of whom received weekly prophylactic treatment for approximately 11 months, and four of whom received on-demand treatment only when bleeding occurred. All prophylaxis patients maintained weekly treatment for the entire study, with mean and median annualized spontaneous bleeding rates of 1.26 and 1.13, respectively. All bleeding was treated successfully with two or fewer infusions of rIX-FP, with 95.3 percent of events treated with a single infusion.

Following a single infusion of 25 IU/kg rIX-FP, the mean FIX activity level was 3.8 percent and 2.7 percent above baseline at day 7 and 14, respectively, and the half-life (time taken for half of the treatment to be eliminated or metabolized) of rIX-FP was 94 hours.

"We are very encouraged by these results demonstrating efficacy of a once-weekly infusion of our investigational rIX-FP, as we selected albumin as the ideal genetic fusion partner for our recombinant Factor IX due to its inherently long half-life and good tolerability profile," said Dr. Stefan Schulte, Vice President of Research and Development, CSL Behring. "We look forward to advancing rIX-FP in clinical trials as we are committed to developing recombinant factor therapies that have the potential to improve compliance and ease prophylaxis. This may, in turn, improve the quality of life for people with hemophilia B."

About the Phase I/II Study
The Phase I/II study was an open-label, multicenter, clinical study of rIX-FP in previously treated patients with severe hemophilia B (FIX ≤ 2%), and part of CSL Behring's PROLONG - 9FP clinical developmental program.

The objectives of the study were to evaluate the prevention of bleeding episodes during once weekly prophylaxis and to assess the hemostatic efficacy of rIX-FP for the treatment of bleeding, in addition to safety and pharmacokinetic (PK) assessments. The study consisted of a 10 to 14 day PK evaluation period, and a three to 12 month safety and efficacy evaluation period, during which patients received either on-demand or prophylaxis treatment. More information about the study design can be found at www.clinicaltrials.gov.

About rIX-FP
CSL Behring engineered rIX-FP to extend the half-life of factor IX through genetic fusion with recombinant albumin. CSL Behring selected albumin as the ideal recombinant genetic fusion partner for its coagulation factor proteins due to its long physiological half-life. In addition, albumin has been shown to have a good tolerability profile, low potential for immunogenic reactions and a well-known mechanism of clearance compared to some other technologies. The cleavable linker connecting recombinant factor IX and recombinant albumin has been specifically designed to preserve the native function of the coagulation factor in the fusion protein, while benefiting from recombinant albumin's long physiological half-life.

About Hemophilia
Hemophilia is an inherited bleeding disorder characterized by prolonged or spontaneous bleeding, especially into the muscles and joints. In nearly all cases, it affects only males. The disease is caused by deficient or defective blood coagulation proteins known as factor VIII or IX. The most common form of the disease is hemophilia A, or classic hemophilia, in which the clotting factor VIII is either deficient or defective. Hemophilia A affects approximately 1 in 5,000 to 10,000 people. Hemophilia B is characterized by deficient or defective factor IX. Hemophilia B affects approximately 1 in 25,000 to 50,000 people. The recommended treatment for patients who are factor deficient is to treat by replacement factor therapy.

About CSL Behring
CSL Behring is a leader in the plasma protein therapeutics industry. Committed to saving lives and improving the quality of life for people with rare and serious diseases, the company manufactures and markets a range of plasma-derived and recombinant therapies worldwide.

CSL Behring therapies are used around the world to treat coagulation disorders including hemophilia and von Willebrand disease, primary immune deficiencies, hereditary angioedema and inherited respiratory disease, and neurological disorders in certain markets.

The company's products are also used in cardiac surgery, organ transplantation, burn treatment and to prevent hemolytic diseases in the newborn. CSL Behring operates one of the world's largest plasma collection networks, CSL Plasma. CSL Behring is a subsidiary of CSL Limited (ASX: CSL), a biopharmaceutical company headquartered in Melbourne, Australia. For more information, visit www.cslbehring.com.

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