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29 June: CSL Behring Receives Positive CHMP Opinion for Respreeza® as Maintenance Treatment for Severe Alpha-1 Antitrypsin Deficiency Patients in Europe

28 July: Swissmedic Accepts for Review CSL Behring’s MAA for rIX-FP for Patients with Hemophilia B

28 July: U.S. FDA Accepts for Review CSL Behring’s Biologics License Application for Its Novel rVIII-SingleChain Therapy for Patients with Hemophilia A

25 August: CSL Behring Enrolls First Patient in Global Phase II/III Study of rVIIa-FP for On-Demand Treatment of Patients with Hemophilia A or B with Inhibitors

25 August: CSL Behring Receives Marketing Authorization for Respreeza® in Europe

8 September: Centers for Medicare and Medicaid Services Extends New Technology Add-On Payment for CSL Behring’s Kcentra® for Third Year

28 September: New Clinical Data on Treatment to Slow the Progression of Emphysema in Patients with Alpha-1 Antitrypsin Deficiency Presented at European Respiratory Society International Congress in Amsterdam

29 October: CSL Behring Announces Winners of its 14th Annual Gettin’ in the Game™ Junior National Championship Program

2 December: CSL Behring to Present New Data for rIX-FP and rVIII-SingleChain at the 57th ASH Annual Meeting & Exposition
7 December: CSL Behring Presents Phase III Data for Its Long-Acting Recombinant Factor IX Albumin Fusion Protein for Hemophilia B at the 57th ASH Annual Meeting & Exposition

8 December: CSL Behring Expands Operations to Russia

18 December: CSL CEO Says Corporate Responsibility and Sustainability are all about Continuous Improvement

21 December: CSL Behring Submits New Drug Application to Japan’s Pharmaceuticals and Medical Devices Agency for rIX-FP for Hemophilia B Patients

22 December: CSL Behring Submits Marketing Authorization Application to the European Medicines Agency for rVIII-SingleChain for Patients with Hemophilia A
FDA Approves New Dosing Option for CSL Behring’s Hizentra®

New Label Supports Individualized Therapy Through Greater Dosing Flexibility. From Daily to Once Every Two Weeks (Biweekly); Underscores CSL Behring’s Commitment to Improving Patients’ Lives

KING OF PRUSSIA, Pa. — 02 February 2015

CSL Behring announced today that the U.S. Food and Drug Administration (FDA) has expanded the administration options for Hizentra®, Immune Globulin Subcutaneous (Human), 20% Liquid, to include the ability to individualize therapy with flexible dosing – treatment at regular intervals from daily to once every two weeks (biweekly) – for people with primary immunodeficiency (PI). Self-administered subcutaneously, Hizentra delivers consistent levels of immunoglobulin G (IgG) regardless of dosing schedule. Hizentra, the first and only 20 percent subcutaneous immunoglobulin, received FDA approval in March 2010 as a once-weekly IgG replacement therapy to help protect people with PI against infections and was approved for biweekly (once every two weeks) dosing in September 2013.

PI is a group of serious diseases that compromise the immune system, leaving patients particularly vulnerable to infection. Approximately 250,000 Americans (or one person per 1,200) have been diagnosed with PI.

"Patient preferences on infusion frequency, time and volume can differ for many reasons, so having a treatment option like Hizentra that can be customized to fit individual lifestyles is important to both patients and the physicians who treat them," said Ralph S. Shapiro, M.D., Director of the Midwest Immunology Clinic. "Most important, flexible dosing options with Hizentra give PI patients the freedom to manage their condition based on their specific needs, while still providing a consistent level of protection against infections."

FDA approval of flexible dosing for Hizentra is based on pharmacometrics (modeling and simulation). Clinical trials using these alternative Hizentra dosing regimens were not conducted.

"CSL Behring understands that managing a life-long disorder can be challenging as patients' lifestyles and treatment requirements may change over time," said Bill Campbell, Senior Vice President, North America Commercial Operations, CSL Behring. "Offering PI patients the option of dosing Hizentra at regular intervals from daily to once every two weeks further underscores our commitment to providing treatment options that improve patients' lives."

For more information about Hizentra individualized therapy, please visit Hizentra.com/Individualize.

About Primary Immunodeficiencies

More than 200 types of PIs exist. For individuals with PI, many of them children, infections may not improve as expected with usual treatments and may even keep returning. As a result, patients may face repeated rounds of antibiotics or hospitalization for treatment. Repeated infections can lead to organ damage, which over time can become life-threatening. Some infections, such as meningitis, can even result in death.

For more information on PI, please visit www.Hizentra.com or contact the leading PI patient advocate groups in the U.S., the Immune Deficiency Foundation and the Jeffrey Modell Foundation.

Important Safety Information

Immune Globulin Subcutaneous (Human), Hizentra®, treats various forms of primary immunodeficiency (PI) in patients age 2 and over.
WARNING: Thrombosis (blood clotting) can occur with immune globulin products, including Hizentra. Risk factors can include: advanced age, prolonged immobilization, a history of blood cloting or hyperviscosity (blood thickness), use of estrogens, installed vascular catheters, and cardiovascular risk factors.

If you are at high risk of thrombosis, your doctor will prescribe Hizentra at the minimum dose and infusion rate practicable and will monitor you for signs of thrombosis and hyperviscosity. Always drink sufficient fluids before administration.

Tell your doctor if you have had a serious reaction to other immune globulin medicines or have been told you also have a deficiency of the immunoglobulin called IgA, as you might not be able to take Hizentra. You should not take Hizentra if you know you have hyperprolinemia (too much proline in your blood).

Infuse Hizentra under your skin only; do not inject into a blood vessel.

Allergic reactions can occur with Hizentra. If your doctor suspects you are having a bad allergic reaction or are going into shock, treatment will be discontinued. Immediately tell your doctor or go to the emergency room if you have signs of such a reaction, including hives, trouble breathing, wheezing, dizziness, or fainting.

Tell your doctor about any side effects that concern you. Immediately report symptoms that could indicate a blood clot, including pain and/or swelling of an arm or leg, with warmth over affected area; discoloration in arm or leg; unexplained shortness of breath; chest pain or discomfort that worsens with deep breathing; unexplained rapid pulse; and numbness or weakness on one side of the body. Your doctor will also monitor symptoms that could indicate hemolysis (depletion of blood red cells), and other potentially serious reactions that have been seen with Ig treatment, including aseptic meningitis syndrome (brain swelling); kidney problems; and transfusion-related acute lung injury.

The most common drug-related adverse reactions in the clinical trial for Hizentra were swelling, pain, redness, heat or itching at the site of injection; headache; back pain; diarrhea; tiredness; cough; rash; itching; nausea and vomiting.

Hizentra is made from components of human blood. The risk of transmission of infectious agents, including viruses and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent, cannot be completely eliminated.

Before being treated with Hizentra, inform your doctor if you are pregnant, nursing or plan to become pregnant. Vaccines (such as measles, mumps and rubella) might not work well if you are using Hizentra. Before receiving any vaccine, tell the healthcare professional you are being treated with Hizentra.

For full prescribing information for Hizentra, including the boxed warning and the patient product information, visit http://www.hizentra.com/consumer/prescribing-information.aspx.

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit http://www.fda.gov/medwatch, or call 1-800-FDA-1088.

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 disease of the newborn.

CSL Behring operates one of the world’s largest plasma collection networks, CSL Plasma. CSL Behring is a global biopharmaceutical company and a member of the CSL Group of companies. The parent company, CSL Limited (ASX:CSL), is headquartered in Melbourne, Australia. For more information, visit http://www.cslbehring.com/.

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U.S. FDA Accepts for Review CSL Behring’s Biologics License Application for rIX-FP for Hemophilia B Patients

CSL Behring Achieves Another Key Milestone in PROLONG-9FP, Its Recombinant Factor IX Fusion Protein Development Program; Continues to Advance CSL Behring’s Legacy of Improving Patient Well-Being

KING OF PRUSSIA, Pa. — 04 February 2015

CSL Behring announced today that the U.S. Food and Drug Administration (FDA) has accepted for review its Biologics License Application (BLA) for the marketing authorization of its long-acting fusion protein linking recombinant coagulation factor IX with recombinant albumin (rIX-FP). Upon FDA approval, rIX-FP will provide hemophilia B patients with a long-acting treatment option with dosing intervals up to 14 days.

“FDA’s decision to accept for review the rIX-FP BLA brings CSL Behring one step closer to providing this innovative therapy to hemophilia B patients in the U.S.,” said Dr. Andrew Cuthbertson, Chief Scientific Officer and R&D Director, CSL Limited. “The development of rIX-FP underscores CSL Behring's protein science capabilities, thorough understanding of the hemophilia community, and commitment to improving the well-being of patients with hemophilia B.”

About PROLONG-9FP Clinical Development Program

CSL Behring's BLA is based on the results from the Phase II/III study (patients ages 12 to 61 years) in the PROLONG-9FP program. The Phase II/III pivotal study was an open-label, multicenter, safety, pharmacokinetic (PK) and efficacy study of rIX-FP in previously treated patients with hemophilia B (FIX ≤ 2%).

This study was designed to compare the change in frequency of spontaneous bleeding events between on-demand treatment and a weekly prophylaxis regimen in patients previously receiving only on-demand treatment; and the number of patients developing inhibitors against factor IX as primary outcome measures. The study evaluated multiple prophylaxis regimens, including 7-day and 14-day intervals. A sub-study evaluated the prevention and control of bleeding in patients with hemophilia B undergoing a surgical procedure.

Study design details for rIX-FP (CSL654) are available at clinicaltrials.gov.

About rIX-FP

CSL Behring engineered rIX-FP to extend the half-life of recombinant factor IX through genetic fusion with recombinant albumin. CSL Behring selected recombinant albumin as its recombinant genetic fusion partner for its coagulation factor proteins due to its long physiological half-life. In addition, recombinant albumin has been shown to have a good tolerability profile, low potential for immunogenic reactions and a well-known mechanism of clearance. 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.

In 2012, the FDA granted Orphan Drug Designation for rIX-FP for the treatment and prophylaxis of bleeding episodes in patients with hemophilia B. The designation includes routine prophylaxis treatment, control and prevention of bleeding episodes, and prevention and control of bleeding in perioperative settings. The FDA’s Orphan Drug Designation program provides orphan status to drugs and biologics defined as those intended for the safe and effective treatment or prevention of rare diseases that affect fewer than 200,000 people in the U.S. Orphan designation qualifies the sponsor
of the product for important tax credits, elimination of FDA license application fees and certain marketing incentives.

**About Hemophilia B**

Hemophilia B (congenital factor IX deficiency) is characterized by deficient or defective factor IX and affects approximately 1 in 25,000 to 50,000 people. Hemophilia B is a congenital bleeding disorder characterized by prolonged or spontaneous bleeding, especially into the muscles, joints, or internal organs. Nearly all hemophilia B patients are male.

**About CSL Behring’s Recombinant Factor Development Program**

rIX-FP for the treatment of hemophilia B is a part of CSL Behring’s Recombinant Factor Development program. The AFFINITY clinical trial program is studying CSL Behring’s recombinant Factor VIII SingleChain (rVIII-SingleChain) to treat hemophilia A. CSL Behring also continues to advance its long-acting recombinant fusion protein linking recombinant coagulation factor VIIa with recombinant albumin (rVIIa-FP) to control bleeding episodes in hemophilia patients who have inhibitors.

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CSL Behring Continues Treatment Access Leadership by Donating Bleeding Disorder Protein Therapies IUs to World Federation of Hemophilia

CSL Behring Pioneered Donations to WFH Program; Further Underscores Commitment to Providing Global Bleeding Disorders Community with Access to Treatment

KING OF PRUSSIA, Pa. — 27 February 2015

In recognition of Rare Disease Day and as part of its ongoing commitment to the global bleeding disorders community, CSL Behring announced today that it is donating 2 million international units (IUs) of protein therapies to the World Federation of Hemophilia (WFH). WFH is an international not-for-profit organization which has worked to improve the lives of people with hemophilia and other inherited bleeding disorders. The donation supports the WFH's Global Alliance for Progress (GAP) Program aimed at improving the diagnosis and treatment of bleeding disorders in developing countries. CSL was the first biotherapies company in the world to make a multiyear commitment to WFH to aid the GAP Program with coagulation factor donations over an extended period of time, starting in 2009. Rare Disease Day, February 28, is coordinated by the European Organization for Rare Diseases (EURORDIS) and by several national alliances and patient organizations around the globe.

"We take great pride in the progress of the GAP Program and are grateful for the generous donations made by CSL Behring," said WFH President Alain Weill. "We look forward to our continued partnership with CSL Behring as we strive to help fulfill our commitment to introduce clotting factor concentrates in developing countries where people who are living with a bleeding disorder may not be able to access appropriate treatment."

"CSL Behring is committed to improving the well-being of people who are living with rare or serious diseases," said Paul Perreault, CEO and Managing Director of CSL Limited. "The GAP Program has made excellent progress over the years and continues to align with our goal to make meaningful differences in the lives of people who are living with serious diseases, such as hemophilia or von Willebrand disease."

In 2012, CSL Behring renewed its product donation contract with WFH, for the period 2013 through 2015. Since 2012, CSL Behring has donated close to 7.7 million IUs of its bleeding disorders therapies to treat people with hemophilia or von Willebrand disease (VWD) in developing countries.

About the Global Alliance for Progress

The Global Alliance for Progress (GAP) Program is a healthcare development project launched in 2003, now entering the Second Decade phase. GAP's goal is to greatly increase the diagnosis and treatment of people with hemophilia and other bleeding disorders in developing countries.

The program aims to close the gap between the number of people born with hemophilia and those who reach adulthood, the gap between the estimated and actual number of people diagnosed with bleeding disorders, and the gap between the volume of treatment product needed versus what is available. CSL Behring supports the WFH GAP Program as one of the program sponsors.

About the World Federation of Hemophilia

For over 50 years, the World Federation of Hemophilia (WFH), an international not-for-profit organization, has worked to improve the lives of people with hemophilia and other inherited bleeding disorders. Established in 1963, it is a global network of patient organizations in 127 countries and has official recognition from the World Health Organization. Visit WFH online at www.wfh.org.
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CSL Behring to Provide $80,000 in Scholarship Funding

Company and Employees Cultivate a Culture of Giving

KING OF PRUSSIA, Pa. — 09 March 2015

CSL Behring announced that it is partnering with Children’s Scholarship Fund Philadelphia (CSFP) to support scholarships for low-income students. CSL Behring will contribute $80,000 to fund scholarships in 2015 and 2016, enabling low-income students in grades K-8 to attend private and parochial schools that partner with CSFP.

Scholarship Fund boasts impressive success record

"Since its founding in 2001, Children's Scholarship Fund Philadelphia has awarded more than 16,500 four-year scholarships to K-8th grade students," said Ina B. Lipman, CSFP Executive Director. "Scholarship recipients' on-time high school graduation rates are greater than 96 percent, well above their peers in public schools who graduate on time at a rate of about 57 percent, with a large majority unprepared to enter the workforce or college."

Developing next generation of scientists and business leaders

Karen Etchberger, CSL Behring’s Executive Vice President, Quality & Business Services, said the company and its employees are committed to supporting key programs in communities in which it operates. CSL Behring’s operational headquarters is located in King of Prussia, Pa.

"Partnering with education-based organizations, like the Children's Scholarship Fund Philadelphia, sets a strong foundation for our future as a global biopharmaceutical company, helping prepare the next generation of scientists, researchers and leaders," said Etchberger.

The partnership is the latest in a host of philanthropic and outreach programs that are the foundation of CSL Behring’s culture of supporting key community programs year-round.

Other 2014-2015 philanthropic and outreach highlights

The partnership with CSFP follows another record-breaking United Way campaign, which raised $787,000 across CSL Behring sites throughout the U.S.:

- $243,000 was raised in CSL Behring’s King of Prussia location for United Way of Greater Philadelphia and Southern New Jersey
- More than $411,000 was raised by CSL Plasma, a subsidiary of CSL Behring, and its employees for United Way chapters where the company’s plasma collection centers are located;
- $133,000 was raised by CSL Behring’s manufacturing site in Kankakee, Ill.;
- CSL Plasma raised $80,000 in a separate campaign for the Immune Deficiency Foundation.

Numerous other charitable works by CSL Behring employees in the past year contribute to the culture of giving.

Culture of giving is year-round

Etchberger emphasized that while CSL Behring and CSL Plasma generated the majority of funds for the United Way, employees gave many hours of their time, as well as monetary donations, supporting a host of other charities such as Big Brothers Big Sisters, Arthritis Foundation, Toys for Tots, Intrepid
Fallen Heroes Fund, United2Feed, American Cancer Society and Ronald McDonald House, to contribute to the culture of giving.

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The Lancet Publishes CSL Behring’s Kcentra® Phase III Data

Study Finds Kcentra® is Superior to Plasma in Patients Needing Warfarin Reversal for Urgent Surgical or Invasive Procedures

KING OF PRUSSIA, Pa. — 17 March 2015

CSL Behring today announced that The Lancet has published results from a Phase III clinical study showing Kcentra® (Prothrombin Complex Concentrate [Human]) to be superior to plasma for the urgent reversal of acquired coagulation factor deficiency induced by vitamin K antagonist (VKA, e.g., warfarin) therapy in adult patients needing an urgent surgery or invasive procedure. Kcentra is the first and only non-activated 4-factor prothrombin complex concentrate (4F-PCC) approved by the U.S. Food and Drug Administration (FDA) for this use and for the urgent reversal of warfarin therapy in adult patients with acute major bleeding.

Each year, approximately three to four million people in the U.S. use warfarin to prevent blood clots from forming following a stroke, heart attack, heart valve surgery, deep vein thrombosis/pulmonary embolism, or certain types of irregular heartbeat, such as atrial fibrillation. Due to a deficiency in blood clotting factors induced by warfarin treatment, patients are at increased risk of bleeding, especially if undergoing an urgent surgery or invasive procedure.

“The results of the Phase III study published in The Lancet show that Kcentra is more effective than plasma for INR [international normalized ratio] reduction and periprocedural hemostasis in adults who are taking warfarin and require an urgent procedure,” said lead study author Joshua N. Goldstein, M.D., Ph.D., attending physician in the Department of Emergency Medicine at Massachusetts General Hospital.

Results of the multicenter, open-label, phase IIib randomized trial showed that in 168 evaluable patients, 90 percent of patients treated with Kcentra achieved effective hemostasis, compared to 75 percent of patients treated with plasma. Additionally, INR reduction to ≤1.3 at 30 minutes after the end of infusion was achieved in 55 percent of patients treated with Kcentra versus 10 percent of patients treated with plasma. Incidences of adverse events, serious adverse events, thromboembolic events, and deaths were similar between the Kcentra and plasma groups. In post-hoc analysis, the median time from start of infusion to start of urgent surgical procedure was shorter in the Kcentra group (3.6 hours [IQR 1.9–10.8]) than in the plasma group (8.5 hours [IQR 2.8–18.7]); (p=0.0098).

“CSL Behring has long been at the forefront of developing innovative protein therapies to help healthcare professionals treat patients with serious medical conditions,” said Bill Campbell, Senior Vice President, North America Commercial Operations, CSL Behring. “Kcentra further underscores our commitment by providing physicians with a treatment option to prevent and manage the risks of excessive bleeding in patients on warfarin who need to undergo an urgent surgery or invasive procedure.”

Kcentra was first approved for use in the U.S. in April 2013 and received a new technology add-on payment (NTAP) designation from the Centers for Medicare and Medicaid Services through September 2015. To be eligible for an NTAP, the product must be new and inadequately paid for under existing MS-DRGs, and must provide a significant clinical improvement over existing therapies.

About Kcentra®

In more than 25 countries, CSL Behring markets Kcentra as Beriplex® or Confidex®. In December 2012, the FDA granted Orphan Drug Designation to Kcentra for the treatment of patients needing urgent reversal of Vitamin K antagonist therapy due to major bleeding and/or surgical procedures. The FDA’s Orphan Drug Designation program provides orphan status to drugs and biologics defined as those intended for the safe and effective treatment or prevention of rare diseases that affect fewer
than 200,000 people in the U.S. Orphan designation qualifies the sponsor of the product for important tax credits, elimination of FDA license application fees and certain marketing incentives.

Important Safety Information

Kcentra®, Prothrombin Complex Concentrate (Human), is a blood coagulation factor replacement product indicated for the urgent reversal of acquired coagulation factor deficiency induced by Vitamin K antagonist (VKA—e.g., warfarin) therapy in adult patients with acute major bleeding or the need for urgent surgery or other invasive procedure. Kcentra is for intravenous use only.

WARNING: ARTERIAL AND VENOUS THROMBOEMBOLIC COMPLICATIONS

Patients being treated with Vitamin K antagonist therapy have underlying disease states that predispose them to thromboembolic events. Potential benefits of reversing VKA should be weighed against the risk of thromboembolic events, especially in patients with history of such events. Resumption of anticoagulation therapy should be carefully considered once the risk of thromboembolic events outweighs the risk of acute bleeding. Both fatal and nonfatal arterial and venous thromboembolic complications have been reported in clinical trials and postmarketing surveillance. Monitor patients receiving Kcentra, and inform them of signs and symptoms of thromboembolic events. Kcentra was not studied in subjects who had a thromboembolic event, myocardial infarction, disseminated intravascular coagulation, cerebral vascular accident, transient ischemic attack, unstable angina pectoris, or severe peripheral vascular disease within the prior three months. Kcentra might not be suitable for patients with thromboembolic events in the prior three months.

Kcentra is contraindicated in patients with known anaphylactic or severe systemic reactions to Kcentra or any of its components (including heparin, Factors II, VII, IX, X, Proteins C and S, Antithrombin III and human albumin). Kcentra is also contraindicated in patients with disseminated intravascular coagulation. Because Kcentra contains heparin, it is contraindicated in patients with heparin-induced thrombocytopenia (HIT).

Hypersensitivity reactions to Kcentra may occur. If patient experiences severe allergic or anaphylactic type reactions, discontinue administration and institute appropriate treatment.

In clinical trials, the most frequent (≥2.8%) adverse reactions observed in subjects receiving Kcentra were headache, nausea/vomiting, hypotension, and anemia. The most serious adverse reactions were thromboembolic events, including stroke, pulmonary embolism and deep vein thrombosis.

Kcentra is derived from human plasma. The risk of transmission of infectious agents, including viruses and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent, cannot be completely eliminated.

The safety and efficacy of Kcentra in pediatric use have not been studied, and Kcentra should be used in women who are pregnant or nursing only if clearly needed.

For more information about Kcentra, please visit www.kcentra.com or call toll-free 1-855-4KCENTRA. For full prescribing information, please visit www.kcentra.com/prescribing-information.aspx.

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European Medicines Agency Commences Review of CSL Behring’s Regulatory Submission for rIX-FP for Hemophilia B Patients

rIX-FP Regulatory Submissions Now Made in EU and U.S.; Underscores CSL Behring’s Legacy of Improving the Care for Patients with Bleeding Disorders

KING OF PRUSSIA, Pa. — 30 March 2015

CSL Behring announced today that the European Medicines Agency (EMA) has started the Centralized Procedure for reviewing the company’s Marketing Authorization Application (MAA) for its long-acting fusion protein linking recombinant coagulation factor IX with recombinant albumin (rIX-FP). Upon European Commission approval, rIX-FP will provide hemophilia B patients in the European Union (EU), as well as the European Economic Area (EEA) countries, with a long-acting treatment option with dosing intervals up to 14 days.

"The EMA beginning its Centralized Procedure for rIX-FP is a significant milestone for CSL Behring’s recombinant factor IX clinical development program and moves us one step closer to bringing this innovative therapy to hemophilia B patients in the EU and EEA," said Dr. Andrew Cuthbertson, Chief Scientific Officer and R&D Director, CSL Limited. "CSL Behring’s rIX-FP for hemophilia B patients further illustrates the company’s protein science capabilities and strong partnership with the hemophilia community."

In February 2015, the U.S. Food and Drug Administration accepted for review CSL Behring's Biologics License Application for rIX-FP. Pivotal data for rIX-FP will be presented during the International Society on Thrombosis and Haemostasis (ISTH) Congress in Toronto in June 2015.

About rIX-FP

CSL Behring engineered rIX-FP to extend the half-life of recombinant factor IX through genetic fusion with recombinant albumin. CSL Behring selected recombinant albumin as its recombinant genetic fusion partner for its coagulation factor proteins due to its long physiological half-life. In addition, recombinant albumin has been shown to have a good tolerability profile, low potential for immunogenic reactions and a well-known mechanism of clearance. 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 PROLONG-9FP Clinical Development Program

CSL Behring’s MAA is based on data from the PROLONG-9FP clinical development program, covering patients from the age of 1 to 61 years. Studies in the program were conducted as open-label, multicenter, safety and efficacy studies of rIX-FP in previously treated patients with hemophilia B (FIX ≤ 2%).

The Phase II/III pivotal study (patients ages 12 to 61 years) was designed to compare the change in frequency of spontaneous bleeding events between on-demand treatment and a weekly prophylaxis regimen in patients previously receiving only on-demand treatment; and the number of patients developing inhibitors against factor IX as primary outcome measures. The study evaluated multiple prophylaxis regimens, including 7-day and 14-day intervals. A sub-study evaluated the prevention and control of bleeding in patients with hemophilia B undergoing a surgical procedure.

The primary outcome measures of the Phase III children study (patients ages 1 to 11 years) are PK parameters of rIX-FP and the number of subjects developing inhibitors against factor IX. All patients received a weekly prophylaxis regimen.
Study design details for rIX-FP (CSL654) are available at clinicaltrials.gov.

**About CSL Behring’s Recombinant Factor Development Program**

rIX-FP for the treatment of hemophilia B is a part of CSL Behring’s Recombinant Factor Development program. The AFFINITY clinical development program is studying CSL Behring’s recombinant Factor VIII SingleChain (rVIII-SingleChain) to treat hemophilia A. CSL Behring also continues to advance its long-acting recombinant fusion protein linking recombinant coagulation factor VIIa with recombinant albumin (rVIIa-FP) to control bleeding episodes in hemophilia patients who have inhibitors.

**About Hemophilia B**

Hemophilia B (congenital factor IX deficiency) is characterized by deficient or defective factor IX and affects approximately 1 in 25,000 to 50,000 people. Hemophilia B is a congenital bleeding disorder characterized by prolonged or spontaneous bleeding, especially into the muscles, joints, or internal organs. Nearly all hemophilia B patients are male.

**About CSL Behring**

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CSL Behring Marks World Hemophilia Day by Renewing Pledge to World Federation of Hemophilia

CSL Behring to Donate 10 Million IUs of its Bleeding Disorder Protein Therapies and to Contribute More than $1.1 Million in Financial Support to WFH

MONTREAL and KING OF PRUSSIA, Pa. — 14 April 2015

In recognition of World Hemophilia Day April 17, the World Federation of Hemophilia (WFH) and CSL Behring announced today that CSL Behring has once again committed to donating bleeding disorder protein therapy international units (IUs) to the WFH Global Alliance for Progress (GAP) Program and other WFH programs, as well as making significant financial contributions to WFH.

As part of the new agreement, CSL Behring will provide 10 million IUs of one or more of its products from its broad portfolio of bleeding disorder protein therapies to the WFH over the period of three years, beginning in 2016. The GAP Program strives to improve the diagnosis and treatment of bleeding disorders in developing countries. In 2009, CSL was the first biotherapies company in the world to make a multiyear commitment to WFH to aid the GAP Program with humanitarian aid coagulation factor donations over an extended period of time. CSL Behring will also provide more than $1.1 million in financial support to WFH’s Corporate Partner and GAP programs over the three year period. World Hemophilia Day is promoted by WFH to increase global awareness of hemophilia and other inherited bleeding disorders.

"WFH established the GAP Program to advance our vision of Treatment for All, working to improve diagnosis and access to care for the millions of people who have a rare bleeding disorder yet remain without treatment," said WFH President Alain Weill. "CSL Behring shares WFH’s vision in improving diagnosis and treatment access. We are once again grateful for the generous pledge made by CSL Behring as we strive to help fulfill our commitment to introduce clotting factor concentrates in developing countries where people who are living with a bleeding disorder may not be able to access appropriate treatment."

Since its first donation in 2009, CSL Behring has provided nearly 14 million IUs of its bleeding disorder protein therapies to treat people with hemophilia or von Willebrand disease (VWD) in developing countries.

"CSL Behring has long been at the forefront of enabling people with serious conditions to lead normal and healthy lives," said Paul Perreault, CEO and Managing Director of CSL Limited. "The GAP Program aligns with CSL Behring’s goal to make meaningful differences in the lives of people who are living with serious diseases, such as hemophilia or von Willebrand disease. We look forward to our continued partnership with WFH as we work together to improve treatment access in developing countries around the world."

About the Global Alliance for Progress Program

The Global Alliance for Progress (GAP) Program is a healthcare development project launched in 2003, now in its Second Decade phase (2013-2022). The overarching goals for the GAP Program will be to increase by 50,000 the worldwide number of people identified/diagnosed with a bleeding disorder, as well as to ensure that 50 percent of those newly diagnosed are from the world’s most
impoverished countries. CSL Behring supports the WFH GAP Program as one of the program sponsors.

About the World Federation of Hemophilia

For over 50 years, the World Federation of Hemophilia (WFH), an international not-for-profit organization, has worked to improve the lives of people with hemophilia and other inherited bleeding disorders. Established in 1963, it is a global network of patient organizations in 127 countries and has official recognition from the World Health Organization. Visit WFH online at www.wfh.org.

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CSL Behring Awards Young Researchers Around the World to Advance Coagulation Science

CSL Behring Professor Heimburger Award Provides 100,000 Euros in Start-Up Grants to Emerging Coagulation Investigators; Underscores CSL Behring’s Longstanding Commitment to Pioneering Science to Improve the Care for Patients with Bleeding Disorders

MARBURG, Germany — 23 April 2015

CSL Behring announced today that the company has named five recipients of the 2015 CSL Behring Professor Heimburger Award for coagulation research. Now in its eighth year, the global awards program helps fund the work of young, emerging researchers by providing start-up grants. The goal of the award is to encourage the next generation of coagulation specialists to establish themselves professionally and to continue innovative research. The total value of the five grants is 100,000 euros.

Recipients of the 2015 CSL Behring Professor Heimburger Award were selected from more than 60 applicants, from 30 countries, by an independent committee of world-renowned clinicians. This year’s recipients, receiving 20,000 euros each, are:

- Ashwini Bennett, M.D.
  Hematology Department, Level 4, Monash Health (Australia)
  Neutrophil extracellular traps: Their role in acute venous thromboembolism, sickle cell crisis and myeloproliferative neoplasms

- Jenny Klintman, M.D.
  Skane University Hospital, Clinical Collaboration Research Unit Lund University (Sweden)
  Clinical evaluation and characterization of suspected atypical hemolytic uremic syndrome (aHUS)

- Tesse Leunissen, M.D.
  University Medical Centre Utrecht (Netherlands)
  High accuracy platelet function testing to predict perioperative micro embolic signals (MES) during carotid endarterectomy (CEA)

- Michelle Sonneveld, M.D.
  Erasmus University Medical Center (Netherlands)
  Complement factor H and the relation with von Willebrand Factor and ADAMTS13

- Bryce Andrew Kerlin, M.D.
  Associate Professor of Pediatrics, The Ohio State University College of Medicine (U.S.)
  Thrombin signaling mechanisms in nephrotic syndrome

"Coagulation disorders can be debilitating for patients and have a negative impact on their quality of life," said Jens Oltrogge, Ph.D., Senior Director, Commercial Development Coagulation, CSL Behring. "Professor Heimburger was a pioneer in improving the care for patients with coagulation disorders. Supporting these young investigators dedicated to advancing coagulation therapy, in his honor, underscores CSL Behring’s longstanding commitment to improving patient outcomes through science and innovation. We would like to congratulate all of the recipients of the 2015 CSL Behring Professor Heimburger Award and wish them success with their research."

More information about the CSL Behring Professor Heimburger Award, including the names of previous winners, can be found at: http://www.cslbehring.com/products/bleeding-disorders/professor-heimburger-award.htm
About Professor Heimburger

Professor Norbert Heimburger was an employee of CSL Behring for more than 30 years and devoted his work to blood coagulation. One of his major contributions in this area was the development of virus-safe plasma products based on pasteurization; a development that led CSL Behring to produce the world’s first virus-inactivated factor VIII concentrate to treat hemophilia patients.

About CSL Behring’s Commitment to Advancing Coagulation Science

CSL Behring has a commitment to innovation and a thorough understanding of the bleeding disorders community. Through this commitment and understanding, CSL Behring continues to advance its recombinant factor development program.

The AFFINITY clinical development program is studying CSL Behring’s recombinant Factor VIII SingleChain (rVIII-SingleChain) to treat hemophilia A and the PROLONG-9FP clinical trial program is studying the company’s recombinant fusion protein linking coagulation factor IX with recombinant albumin (rIX-FP) to treat hemophilia B. Regulatory submissions for rIX-FP are currently being reviewed by the U.S. Food and Drug Administration and European Medicines Agency. CSL Behring also continues to advance its long-acting recombinant fusion protein linking recombinant coagulation factor VIIa with recombinant albumin (rVIIa-FP) to control bleeding episodes in hemophilia patients who have inhibitors.

Pivotal data from CSL Behring’s recombinant factor development program will be presented during the International Society on Thrombosis and Haemostasis (ISTH) Congress in Toronto in June 2015.

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SCIENCE, INFORMATION, AND TECHNOLOGY DRIVING EVOLUTION OF GLOBAL HEALTHCARE

Industry serves critical role in advancing healthcare and improving the well-being of patients, says CSL CEO Paul Perreault

SARDINIA, Italy — 14 May 2015

Scientific advancements, more readily available medical information and emerging technologies are driving greater patient engagement, improving diagnosis and treatment, and enabling people with life-threatening diseases to live longer and healthier lives, said CSL Limited CEO and Managing Director Paul Perreault. CSL is the parent company of CSL Behring.

"It's all about diagnosing sooner, improving our ability to get the right treatment to the right patient at the right time, or even finding a cure," Perreault told global healthcare leaders at the 2015 International Plasma Product Biotechnology meeting this week. "Industry plays a critical role in this evolution. The way to succeed in the new environment is to innovate and adapt. In the end, we are about improving the well-being of patients."

As CEO of a leading global specialty biotherapeutics company, Perreault noted patients and doctors are more informed and engaged today than ever before. The vast amount of medical information available and new advances in technologies have empowered patients and helped doctors to better diagnose and treat those with life-threatening conditions. Advanced diagnostics and genomics are two of the technologies that are making a difference, Perreault said.

Advanced diagnostic technologies

"Technologies such as hand-held devices, advanced diagnostic tests and the Internet are revolutionizing the practice of medicine and the diagnosis of rare diseases, which often went undiagnosed for years," said Perreault. "This is important for physicians who now have information at their fingertips that can mean the difference between successfully diagnosing and treating a patient with a rare medical disorder or not being able to make a diagnosis, which can be frustrating and disheartening for both patient and doctor."

Genomics and personalized medicine

Genomic or precision medicine enables physicians to tailor treatment to the patient, or to better understand how a patient may respond to a particular therapy in advance. While this is well-established with certain cancers, it is less so in the rare disease space.

Gene therapy trials for patients with rare diseases are in progress to correct the gene mutations causing rare or serious conditions. Additionally, while personalized medicine is still in its infancy, advances such as the approval of new flexible dosing regimens for products used to treat immune deficiencies has improved administration convenience and the quality of life for many patients, Perreault said.

Inadequate market access

Perreault said inadequate market access affects many patient groups, but is especially critical for those with rare diseases. "We spend a great deal of time and resources understanding payer needs, the evolution of health technology assessments (HTA) and what these mean for patients, and support constructive engagement with payers," he said. "It's important that payers do not just look at cost alone. The value of therapies needs to be considered, too. Standard HTA assessments should be modified to reflect the special nature of rare diseases, the impact of these therapies on such serious conditions, the societal benefit and the small patient population size involved."
New healthcare delivery and business models

As a result of cost pressures and trying to improve efficiency and quality of care, Perreault said, "We are seeing an evolution of healthcare delivery and business models such as outpatient, home care and new dosage forms, which lead to improved quality of life, change in facility use, and broader and different make-up of healthcare teams." Today, more patients are being treated in outpatient settings, through home care, or Accountable Care Organizations, and often with a broader and different make-up of healthcare teams.

Cost of medicines

Although medicine spending is often portrayed as driving up healthcare costs, medicines are not a main driver of increased healthcare costs in the United States, Perreault noted. In fact, medicines account for a small and declining share of health spending growth. Every dollar spent on healthcare is broken down as follows, according to the Pharmaceutical Research and Manufacturers of America:

- $.07 – government administration and net cost of private health insurance
- $.08 – home health and nursing home care
- $.09 – prescription drugs
- $.21 – physician and clinical services
- $.23 – other
- $.32 – hospital care

Stakeholder Partnerships

In such a complex and evolving global healthcare environment, the engagement of stakeholders as partners in solutions is paramount, Perreault said. For instance, rare disease patient groups are increasing their global presence and capabilities in information sharing, community building and advocacy. Physicians work in tandem with those groups and governments are increasingly engaging them for input, including helping design health programs and policies. Industry has a role in supporting patient care, information sharing, stakeholder capabilities and engagement with all stakeholders. Working together provides the best opportunity for successful evolution.

"Our healthcare systems worldwide have evolved tremendously over the past decade, and they will continue to do so at exponential rates," said Perreault. "Change is constant, but our future will remain bright if we embrace the opportunities that result in better patient care, and if we work together to shape the healthcare environment to increase access and improve outcomes."

About CSL Behring

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National Hemophilia Foundation Honors CSL Behring with Corporate Leadership Award

CSL Behring Recognized for its Longstanding Commitment to Delivering Innovative Products and Programs that Advance the Care of the Bleeding Disorders Community

NEW YORK and KING OF PRUSSIA, Pa. — 22 May 2015

The National Hemophilia Foundation (NHF) has awarded CSL Behring its 2015 Corporate Leadership Award as recognition for the company’s longstanding and unwavering commitment to advancing science and improving the care of the bleeding disorders community. The award was accepted by Paul Perreault, CEO and Managing Director, CSL Limited, during the NHF Annual Spring Soiree in New York City on May 21.

“The National Hemophilia Foundation is dedicated to finding better treatments and cures for bleeding disorders,” said Val Bias, Chief Executive Officer of NHF. “CSL Behring shares NHF’s vision and commitment to providing education, advocacy and research opportunities that strive to improve the well-being of patients and their caregivers. We applaud CSL Behring’s leadership and dedication to delivering innovative products and programs that make a meaningful difference in the lives of the bleeding disorders community.”

World-class research and development, high quality manufacturing, and patient-centered management are the longstanding hallmarks of CSL Behring in serving the bleeding disorders community. Driven by its commitments to the community, CSL Behring develops and delivers innovative protein-based therapies, and leading educational and emotional support programs that improve the lives of people living with hemophilia, von Willebrand disease and other serious bleeding disorders. By closely partnering with the hemophilia community, CSL Behring has achieved significant advancements in its Recombinant Factor Development program which, upon regulatory approvals, will provide hemophilia A and B patients with new treatment options.

The company also continues to advance patient, caregiver and researcher support through highly impactful programs, including:

- **My Source℠** program -- one-stop location for CSL Behring's patient-support resources for the U.S. bleeding disorders community
- **Common Factors** series of educational events
- **My Access℠** cost share program
Gettin’ in the Game™ events and the Gettin’ in the Game™ Junior National Championship program

CSL Behring Professor Heimburger Award for Coagulation Research

Pledge to the World Federation of Hemophilia to donate bleeding disorder therapies and provide financial contributions.

To learn about CSL Behring’s programs for the U.S. bleeding disorders community, please visit: www.MySourceCSL.com.

“CSL Behring has long been at the forefront of protecting the health of people living with a range of serious and chronic medical conditions,” said Paul Perreault, CEO and Managing Director of CSL Limited. “NHF’s goals align with our focus to develop and deliver innovative products and programs that save people’s lives and improve the quality of their lives. On behalf of CSL’s 14,000 employees around the world who are driven by our deep passion and commitment to the global bleeding disorders community, I’d like to thank NHF for recognizing us with this year’s Corporate Leadership Award.”

About the Spring Soiree

The Annual Spring Soiree publically acknowledges outstanding leaders whose exemplary contributions have significantly advanced NHF’s mission to find better treatment and cures for bleeding disorders and to prevent the complications of these disorders.

About NHF

The National Hemophilia Foundation (NHF) is dedicated to finding better treatments and cures for inheritable bleeding disorders and to preventing the complications of these disorders through education, advocacy and research. There are over 400,000 people worldwide who are affected by hemophilia, an inheritable bleeding disorder in which the blood does not clot normally. Seventy percent of these individuals do not have access to treatment and care, resulting in severe joint damage, extreme pain, and possible life-threatening bleeds. NHF is the premier organization in the U.S. serving the bleeding disorders community. For more than 65 years, NHF continues to be a vital resource for prevention education and provides a network of support for the estimated 20,000 Americans living with hemophilia and their families.

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The Lancet Publishes CSL Behring’s Alpha-1 Antitrypsin Deficiency Study Data

Important New Findings Published in The Lancet Support the Use of Zemaira® [Alpha1-Proteinase Inhibitor (Human)] in Patients with Alpha-1 Antitrypsin Deficiency (AATD)

Marburg, Germany — 01 June 2015

Findings from CSL Behring’s RAPID study, the largest placebo-controlled trial ever conducted in patients with alpha-1 antitrypsin deficiency (AATD), demonstrate that the use of Alpha1-Proteinase Inhibitor therapy may slow the progressive loss of lung tissue experienced by these critically ill patients. According to findings of RAPID (Randomized, Placebo-controlled Trial of Augmentation Therapy in Alpha-1 Proteinase Inhibitor Deficiency), published by The Lancet, patients with AATD treated with Alpha1-Proteinase Inhibitor therapy exhibited a lower annual rate of lung density decline compared to placebo, when measured using chest computed tomography, at full inspiration.

“RAPID is regarded as a landmark study validating almost two decades of focus on the lung-density endpoint as the most sensitive way to track lung tissue decline and the seven-year collaboration of an international team of investigators,” said Kenneth R. Chapman, MD, Director of the Asthma & Airway Centre at the University Health Network in Toronto, and lead author of the paper. “Our findings provide additional evidence that treatment with an Alpha1-Proteinase Inhibitor may slow the accelerated loss of lung tissue that is a characteristic of this potentially debilitating disease.”

AATD is a hereditary condition that can severely affect a patient’s lung function. The condition is marked by a low level or absence of alpha-1-proteinase inhibitor (A1-PI), a natural protein that inhibits neutrophil elastase, thereby preventing destruction of lung tissue. Severe deficiency of A1-PI is associated with a strong tendency for the development of emphysema, a form of chronic obstructive pulmonary disease (COPD), and can significantly impact everyday life and life expectancy. According to a recent registry, emphysema affects 54 percent of diagnosed Alpha-1-deficient patients.

On average, it takes over seven years from the time a patient’s lung symptoms first appear until a proper diagnosis is made. And nearly half (43 percent) of patients see at least three physicians before being diagnosed. Approximately 90 percent of individuals with AATD are not diagnosed or are thought to have other conditions, such as asthma or smoking-related COPD.

“We are excited that the results of this important study in Alpha-1 have been published in the highly-respected journal The Lancet,” said John Walsh, co-founder, President and CEO of the Alpha-1 Foundation. “We commend CSL Behring for their outstanding commitment to the Alpha-1 community and advancing the understanding and treatment of the disease. These results further support the use of augmentation therapy in the treatment of Alpha-1, and we hope they bolster efforts of Alpha-1 communities around the world to win access to therapy.”

Study Design and Findings

According to study protocol, the effect of CSL Behring’s Alpha1-Proteinase Inhibitor therapy on the progression of emphysema, the primary endpoint of the study, was measured by computed tomography scan over 24 months and was assessed by the annual rate of lung density loss at total lung capacity (TLC) and functional residual capacity (FRC) combined. The annual rate of lung density loss at TLC and FRC combined were not statically different in treatment group compared to placebo group. However, results showed a 34 percent reduction in the annual rate lung density decline compared with placebo when measured at TLC (or full inspiration).

Serious adverse events were not statistically different between groups with one death in the A1-PI group and three deaths in the placebo group.
RAPID was a multicenter, double-blind, randomized, parallel group, placebo-controlled study comparing the efficacy and safety of CSL Behring’s Alpha1-Proteinase Inhibitor therapy with placebo in patients with emphysema due to AATD. The participants, 180 non-smokers aged 18-65, were randomly assigned to receive A1-PI intravenously 60 mg/kg weekly or placebo.

About Zemaira

Zemaira is a highly-purified form of Alpha1 Proteinase Inhibitor (human) currently approved in Brazil, New Zealand, and the US, where it is indicated for chronic augmentation and maintenance therapy in adults with Alpha1 deficiency and clinical evidence of emphysema.

CSL Behring is seeking additional approvals around the world for this A1-PI therapy, which is currently under review with the European Medicines Agency.

Zemaira is contraindicated in patients with a history of severe systemic reaction to the product or to A1-PI protein, including anaphylaxis. Due to the risk of severe hypersensitivity, Zemaira is also contraindicated in immunoglobulin A-deficient patients with antibodies against IgA.

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CSL Behring to Present Pivotal Data for rVIII-SingleChain and rIX-FP at 2015 ISTH Congress

More than 20 abstracts focusing on investigational and branded products showcase CSL Behring’s expertise and commitment to advancing the care of patients with bleeding disorders

KING OF PRUSSIA, Pa. — 16 June 2015

CSL Behring announced today it will present more than 20 abstracts, including five oral presentations, from across its hematology portfolio of investigational and branded products at the 2015 International Society on Thrombosis and Haemostasis (ISTH) Congress, being held in Toronto June 20-25. The presentations will include pivotal trial data for two of its late-stage recombinant products – its novel recombinant factor VIII SingleChain (rVIII-SingleChain) compound for hemophilia A and its long-acting recombinant factor IX albumin fusion protein (rIX-FP) for hemophilia B.

Hemophilia is an inherited bleeding disorder, primarily affecting males, caused by a shortage of or defect in proteins that prevent the blood from clotting normally. The condition affects more than 175,000 people worldwide, the majority of whom have hemophilia A.1

“CSL is proud to be sharing a significant amount of new scientific and clinical research at ISTH’s 2015 conference,” said Dr. Andrew Cuthbertson, Chief Scientific Officer and Director of R&D, CSL Limited. “I am particularly excited that data from our phase III pivotal studies, for both rVIII-SingleChain and rIX-FP, will be presented publicly for the first time. These product candidates hold great promise and potential, and could offer patients strong and sustained efficacy and improved convenience with less frequent dosing, two key areas of unmet need. These data, along with abstracts for other R&D candidates and products in our coagulation franchise, reinforce the depth and breadth of CSL’s knowledge and commitment to advancing the care of patients with serious medical conditions.”

In addition to presenting new clinical data, CSL Behring will host programs at ISTH 2015 focusing on the unmet needs of people living with hemophilia A and B, as well as other bleeding disorders. Of note is a satellite symposium, “Pioneering Therapeutic Proteins in Hemophilia Care Through Innovative Technologies,” being held on Monday, June 22 from 12:15 – 1:45 p.m. EDT (Room 714, Level 700). Representatives of the CSL Behring coagulation franchise will be available at booth number 1522 throughout ISTH.

rVIII-SingleChain

Oral Presentation

Wednesday, June 24, 8:30–8:45 a.m. EDT

- Late-breaking abstract # LB008: rVIII-SingleChain, results of the pivotal phase I/III PK, efficacy and safety clinical trial in adults and adolescents with severe hemophilia A

Posters

Tuesday, June 23, 6:00-7:30 p.m. EDT

- The FVIII plasma activity of rVIII-SingleChain can be measured in both the one-stage and chromogenic substrate assays. E-Poster # PO089
Physicochemical characterization of recombinant single-chain factor VIII (rVIII-SingleChain). E-Poster # PO146
Population pharmacokinetic model of recombinant single-chain factor VIII (rVIII-SingleChain) in patients with hemophilia A. E-Poster # PO241
Efficacy and safety of rVIII-SingleChain in surgical prophylaxis. E-Poster # PO258
rVIII-SingleChain pharmacokinetics in adults, adolescents and children. E-Poster # PO262

**rIX-FP**

**Oral Presentations**

*Wednesday, June 24, 2:00-3:15 p.m. EDT*

- Efficacy, pharmacokinetics (PK) and safety results of a phase 3 clinical study of recombinant fusion protein linking coagulation factor IX with albumin (rIX-FP) in previously treated children with hemophilia B. Oral Communication # OR346
- Efficacy and safety results of a phase 3 pivotal clinical study of recombinant fusion protein linking coagulation factor IX with albumin (rIX-FP) in previously treated patients with hemophilia B. Oral Communication # OR347
- Population pharmacokinetics (PK) of recombinant fusion protein linking coagulation factor IX with recombinant albumin (rIX-FP) in adult and pediatric patients with severe hemophilia B. Oral Communication # OR350

**Posters**

*Monday, June 22, 5:15–6:30 p.m. EDT*

- Tissue distribution of rIX-FP after intravenous application to rodents. E-Poster # PO152

*Tuesday, June 23, 6:00–7:30 p.m. EDT*

- Structural characterization of recombinant factor IX fusion protein linked with human albumin (rIX-FP). E-Poster # PO144

*Wednesday, June 24, 5:15–6:30 p.m. EDT*

- Efficacy and safety of recombinant fusion protein linking coagulation factor IX with albumin (rIX-FP) in previously treated patients with hemophilia B undergoing a surgical procedure. E-Poster # PO253

**rVIIa-FP**

**Oral Presentation**

*Monday, June 22, 4:15–4:30 p.m. EDT*

- A recombinant fusion protein linking activated coagulation factor VIIa with albumin (rVIIa-FP) binds to neonatal Fc receptor and tissue factor in vitro. Oral Communication # AS016

**Posters**

*Tuesday, June 23, 6:00–7:30 p.m. EDT*
Dosing of rVIIa-FP in clinical studies in hemophilia with inhibitors and factor VII deficiency. E-Poster # PO257

Wednesday, June 24, 5:15–6:30 p.m. EDT

The recombinant fusion protein linking activated factor VIIa to human albumin (rVIIa-FP) provides superior bleeding protection compared to recombinant FVIIa (rFVIIa) in a novel monkey model of acquired factor VIII inhibitors. E-Poster # PO596

Plasma-Derived Factor VIII/VWF

Posters

Monday, June 22, 5:15–6:30 p.m. EDT

- An open-label, multi-center extension study to assess the efficacy and safety of a plasma-derived von Willebrand factor/factor VIII (VWF/FVIII) concentrate in pediatric, adolescent, and adult subjects with von Willebrand disease. E-Poster # PO643
- Plasma-derived, purified, pasteurized von Willebrand factor/factor VIII concentrate in the treatment of patients with von Willebrand disease and haemophilia A: update of a long-term observational study. E-Poster # PO647

Tuesday, June 23, 6:00–7:30 p.m. EDT

- A phase III, open-label, multicenter study to evaluate pharmacokinetics of a plasma-derived von Willebrand factor/factor VIII (VWF/FVIII) concentrate in pediatric subjects with hemophilia A (SWIFTLY-HA study). E-Poster # PO206
- A phase III open-label, multi-center study with a plasma-derived von Willebrand factor/factor VIII concentrate to assess the pharmacokinetics, efficacy, and safety in pediatric subjects with von Willebrand disease (SWIFTLY-VWD study). E-Poster # PO629

Wednesday, June 24, 5:15–6:30 p.m. EDT

- Molar specific activity of Factor VIII concentrates. E-Poster # PO194
- High-purity, plasma-derived, pasteurized factor VIII concentrate in the treatment of patients with hemophilia A: update of a long-term observational study. E-Poster # PO195
- A phase III, open-label, multicentre study to evaluate efficacy and safety of a plasma-derived von Willebrand factor/factor VIII concentrate in pediatric subjects with hemophilia A (SWIFTLY-HA study). E-Poster # PO249

rVWF-FP

Poster

Tuesday, June 23, 6:00–7:30 p.m. EDT

- Improved resolution of high molecular weight multimers of recombinant von Willebrand factor–albumin fusion product by agarose electrophoresis/western blotting. E-Poster # PO673

Pasteurization

Poster
**Wednesday, June 24, 5:15–6:30 p.m. EDT**

- Inactivation of emerging viruses by pasteurization in plasma-derived medicinal products. E-Poster # PO605

**About rVIII-SingleChain**

rVIII-SingleChain is a novel recombinant single-chain factor VIII (FVIII) construct specifically designed for greater molecular stability. It uses a covalent bond that forms one structural entity, a single chain, to improve the stability and half-life of FVIII.

The Phase III trial, a part of the AFFINITY clinical development program, is an open-label, non-randomized, multicenter study evaluating the efficacy, safety and pharmacokinetics of rVIII-SingleChain. Study design details for rVIII-SingleChain (CSL627) are available at clinicaltrials.gov.

**About rIX-FP**

CSL Behring engineered rIX-FP to extend the half-life of recombinant factor IX through genetic fusion with recombinant albumin. CSL Behring selected albumin as its 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. 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.

In **February 2015**, the U.S. Food and Drug Administration accepted for review CSL Behring’s Biologics License Application (BLA) for rIX-FP. In **March 2015**, the European Medicines Agency (EMA) started the Centralized Procedure for reviewing CSL Behring’s Marketing Authorization Application (MAA) for rIX-FP.

The PROLONG-9FP clinical development program for rIX-FP covers patients from the age of 1 to 61 years. Studies in the program were conducted as open-label, multicenter, safety and efficacy studies of rIX-FP in previously treated patients with hemophilia B (FIX ≤ 2%).

Study design details for rIX-FP (CSL654) are available at clinicaltrials.gov.

**About CSL Behring**

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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 disease of the newborn.

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Defining and Differentiating the Value of Therapies Used to Treat Rare Disease Patients is a Challenge for Payers

Panel of experts examines how the value of biopharmaceutical technologies is assessed and how it affects drug development at BIO International Conference

PHILADELPHIA, Pa. — 17 June 2015

The cost of medicines in general and biopharmaceutical therapies used to treat rare diseases in particular has faced increased attention from payers and other stakeholders in recent years.

Determining how the value of biopharmaceutical technologies is assessed presents a special and critically important challenge for stakeholders. It can also have an impact on drug development and patient access to care in the rare disease space, where 400 million people around the world have a rare disease. Approximately 7,000 rare disorders have been identified, but only a relatively small number of therapies are available to treat them.

This afternoon, a panel of experts moderated by Dennis Jackman, CSL Behring’s Senior Vice President of Global Healthcare Policy and External Affairs, will address this question and identify examples of effective mechanisms of defining and differentiating the value to patients and healthcare of rare disease therapies at 2015 BIO International Conference. Panelists include: Amanda Bartelme, Director Avalere Health; Pamela Gavin, COO National Organization for Rare Disorders, and Jim Geraghty, Principal Third Rock Ventures.

“It’s important that healthcare systems worldwide recognize the special nature of rare disease therapies,” Jackman said. “In determining the value of these therapies, consideration must be given to factors such as societal value, severity of the condition, lack of alternative treatments, data generation challenges due to small populations, lack of comparators, and endpoints and other factors. It’s important that payers look at the holistic value of the therapy and not just its cost in order to help ensure access to these lifesaving therapies. Rare disease therapies are still a small portion of the total drug spend, but make a huge difference in the lives of patients and their families. Manufacturers also understand the need to produce assessments that help to demonstrate this value and continue to progress achievable ways to do that.” Jackman noted that, “there are particular challenges in researching and developing rare disease therapies. The way value is assessed and the way access to treatment is granted will have a significant impact on innovation and the sustainability of those efforts, and, ultimately patient care.”

About CSL Behring

CSL Behring is a leader in the biotherapeutics 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 disease of the newborn.

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CSL Behring today presented data from a Phase I/III study on the efficacy and safety of its novel investigational recombinant factor VIII single chain (rVIII-SingleChain) in adolescents and adults with hemophilia A during a late breaking abstract session at the 2015 International Society on Thrombosis and Haemostasis (ISTH) Congress. Overall, patients using rVIII-SingleChain to prevent bleeding (prophylaxis) were well controlled with two to three infusions per week and developed no inhibitors.

Patients using rVIII-SingleChain to prevent bleeding had low annualized bleeding rates (median ABR of 1.14), and an annualized spontaneous bleeding rate (ASBR) of 0.00. rVIII-SingleChain had improved pharmacokinetic parameters compared with octocog alfa, the comparator. Of 848 bleeds treated in the study, 94 percent were successfully controlled with no more than two infusions of rVIII-SingleChain, with 81 percent controlled by only one infusion. The majority of bleeding events treated with rVIII-SingleChain and assessed by investigators (94 percent of 835 assessed bleeding events) were rated as excellent or good. The data are part of the AFFINITY Phase I/III study, an open-label, multi-center trial examining the safety, efficacy and pharmacokinetics of rVIII-SingleChain compared with recombinant human antihemophilic factor VIII (octocog alfa). Study design details for rVIII-SingleChain (CSL627) are available at clinicaltrials.gov.

"In this large-scale study, we observed relatively low annualized bleeding rates and a median of zero spontaneous bleeding events with rVIII-SingleChain for routine prophylaxis for patients with hemophilia A," said Professor Ingrid Pabinger-Fasching, M.D., of the Medical University of Vienna, Austria and lead investigator of the pivotal trial. "As the first and only single chain recombinant factor product, rVIII-SingleChain has the potential to offer improved protection from bleeding with less frequent dosing, and an excellent safety profile thus far."

Results presented in the late breaking session included data on more than 14,000 exposure days in 146 patients on prophylaxis and 27 patients treated on demand for a bleeding event. In total, 120 patients were treated for more than 50 days of exposure; 52 had more than 100 days of exposure. Among patients in the prophylaxis group, 32 percent were dosed twice weekly and 54 percent received treatment three times per week. The most common adverse events were naso-pharyngitis, arthralgia, and headache. Overall, rVIII-SingleChain was well tolerated and no inhibitors have been reported.

"Our novel rVIII-SingleChain was specifically designed to improve the stability and provide longer-lasting hemostatic efficacy of factor VIII, thereby addressing the need to provide hemophilia A patients with a treatment that may require fewer infusions while maintaining its therapeutic effect," said Dr. Andrew Cuthbertson, Chief Scientific Officer and Director of R&D, CSL Limited. "These pivotal data are promising and are supportive of CSL Behring’s commitment to bringing this therapy to the market, and to helping improve the care of people living with hemophilia A."

About rVIII-SingleChain
rVIII-SingleChain is a novel recombinant single-chain factor VIII (FVIII) construct specifically designed for greater molecular stability. It uses a covalent bond that forms one structural entity, a single chain, to improve the stability of FVIII and provide longer-lasting FVIII activity.

About Hemophilia A

Hemophilia A (congenital factor VIII deficiency) is caused by deficient or defective factor VIII. The condition is characterized by prolonged or spontaneous bleeding, especially into the muscles, joints, or internal organs. Affecting approximately 1 in 5,000 to 10,000 people, hemophilia A is the most common form of hemophilia. Nearly all hemophilia A patients are male.

About CSL Behring

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###
TORONTO — 24 June 2015

- Data supports prolonged dosing intervals up to 14 days for routine prophylaxis in hemophilia B patients
- Majority of adult and pediatric patients using rIX-FP for routine prophylaxis had an annualized spontaneous bleeding rate (AsBR) of 0.00
- Surgical sub study results show single dose of rIX-FP sufficient to maintain hemostasis during surgery

CSL Behring today presented data from Phase III studies evaluating the efficacy and long-term safety of its investigational long-acting fusion protein linking recombinant coagulation factor IX with recombinant albumin (rIX-FP). The data, shared in three separate oral presentations at the 2015 International Society on Thrombosis and Haemostasis (ISTH) Congress in Toronto, support the use of rIX-FP for routine prophylaxis, dosed once up to every 14 days, and for on-demand treatment of bleeding episodes in previously-treated adults and children with hemophilia B. The findings also include efficacy and safety results supporting the use of rIX-FP in patients undergoing surgical procedures.

“The pivotal data for rIX-FP are exciting because they suggest the potential for prolonged dosing intervals of up to two weeks for routine prophylaxis,” said Elena Santagostino, M.D., Ph.D., Professor in the Medical School of Clinical and Experimental Hematology at the University of Milan/IRCCS Maggiore Hospital, and lead investigator. “The trials also showed that this less frequent dosing was possible without compromising therapeutic benefit. This suggests rIX-FP, if approved, could be an important new treatment option, especially for patients who lead active lifestyles and require a prophylactic regimen.”

Key Study Findings

- **rIX-FP in previously-treated adults and adolescents** (abstract #OR347): The first Phase III study was a global safety and efficacy trial assessing rIX-FP for prophylaxis treatment once every 7, 10 and 14 days and on-demand treatment of bleeding episodes in 63 previously-treated patients (12-61 years of age) with hemophilia B (FIX activity ≤2% of normal). In one arm, 23 previously on-demand patients received only on-demand treatment for 6 months and then switched to 7-day prophylaxis treatment. In the other arm, 40 patients received 7-day prophylaxis treatment for 6 months and then, if eligible, switched to a 10- or 14-day prophylaxis treatment interval for 12 to 18 months.

In patients who started on-demand treatment then switched to prophylaxis treatment, annualized spontaneous bleeding rates (AsBR) decreased by 100 percent from a median 15.43 during the on-demand treatment phase to 0.00 in the prophylaxis treatment phase (p<0.0001). Among patients who started on 7-day prophylaxis treatment then switched to 10- or 14-day prophylaxis treatment, the median AsBR rate was 0.00. In total, 99 percent of bleeding events were successfully managed with one or two infusions, with 94 percent of bleeds controlled with only one infusion regardless of the cause or location. Patients on 14-day prophylaxis treatment used a median dose of 75 IU/kg, which was 50 percent less therapy compared with their previously used FIX products. None of the patients developed inhibitors to factor IX or antibodies to rIX-FP. Related adverse events were reported in five patients (7.9 percent). The most common adverse reaction in clinical trials was headache. Overall, rIX-FP was well tolerated and no safety concerns were identified.

“These data provide additional evidence of the benefits of CSL Behring’s recombinant albumin fusion technology, which was designed to significantly reduce clearance and provide longer-lasting hemostatic efficacy of factor IX to allow for less frequent dosing,” said Dr. Andrew Cuthbertson, Chief Scientific Officer and Director of R&D, CSL Limited. “CSL Behring has long been committed to protecting the health of people living with a range of serious medical conditions and rIX-FP
exemplifies our promise to developing and delivering innovative products that have the potential to improve the care of patients around the world.”

- **rIX-FP in previously-treated children** (abstract #OR346): A second Phase III global study evaluated the safety and efficacy of rIX-FP for prophylaxis treatment once every 7 days and treatment of bleeding episodes in 27 children, age 1-11 years, with hemophilia B (FIX activity ≤2% of normal). Patients were treated for 12 months and/or 50 exposure days. The median AsBR was 0.00, and similar for patients under age 6 and those between 6 and 11 years. In total, 97 percent of bleeding episodes were successfully managed by one or two infusions, with 89 percent treated with only one infusion. Overall, the pharmacokinetic profile demonstrated a greater than 5-fold longer half-life, reduced clearance for rIX-FP versus other FIX products, supporting prophylaxis treatment intervals every 7 to 14 days. Investigators rated 96 percent of the treatments as effective (excellent or good). A total of 25 patients achieved 50 or more exposure days. None of the patients developed inhibitors to factor IX or antibodies to rIX-FP, and there were no related adverse events or withdrawals from the study.

- **rIX-FP in previously-treated patients undergoing surgery** (abstract #PO253): The third study was a surgical sub-group analysis included in the phase III studies as part of the global PROLONG-9FP clinical program. This abstract evaluated the use of rIX-FP to prevent bleeding during and post-surgery in 10 patients with hemophilia B (ages 8 to 51 years). The results showed that a single dose of rIX-FP was sufficient to maintain hemostasis during surgery. Over the 14-day post-surgical study period, patients needed anywhere from two to seven infusions. Both the consumption and dosing frequency of rIX-FP were considered remarkably low based on the type of surgery performed. The response was rated by investigators as excellent or good during all procedures. None of the patients developed inhibitors to factor FIX or antibodies to rIX-FP, and there were no related adverse events or withdrawals from the study.

**About rIX-FP**

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In February 2015, the U.S. Food and Drug Administration accepted for review CSL Behring’s Biologics License Application (BLA) for rIX-FP. In March 2015, the European Medicines Agency (EMA) started the Centralized Procedure for reviewing CSL Behring’s Marketing Authorization Application (MAA) for rIX-FP.

The PROLONG-9FP clinical development program for rIX-FP covers patients from the age of 1 to 61 years. Studies in the program were conducted as open-label, multicenter, safety and efficacy studies of rIX-FP in previously treated patients with hemophilia B (FIX ≤ 2%). Study design details for rIX-FP (CSL654) are available at clinicaltrials.gov.

**About Hemophilia B**

Hemophilia B (congenital factor IX deficiency) is characterized by deficient or defective factor IX and affects approximately 1 in 25,000 to 50,000 people. Hemophilia B is a congenital bleeding disorder characterized by prolonged or spontaneous bleeding, especially into the muscles, joints, or internal organs. Nearly all hemophilia B patients are male.

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CSL Behring Announces the 2015 Interlaken Leadership Awards Recipient

Awards Program Supports Novel Research of Immunoglobulin Therapy for Neurological Disorders

KING OF PRUSSIA, Pa. — 29 June 2015

CSL Behring announced today that Mohammad Alsharabati, MD, University of Alabama at Birmingham, United States, is the recipient of the 2015 www.interlakenleadershipawards.com for original research in the field of neuroimmunology. This annual global awards program provides monetary grants and/or product supply for investigational use to support research focusing on the potential role of immunoglobulin (Ig) therapy in the treatment of neurological disorders. The global review committee seeks proposals likely to advance innovative medical research and knowledge about the potential role of Ig therapy to improve the lives of patients who have disabling neurological conditions.

Dr. Alsharabati’s proposal to investigate subcutaneous immunoglobulin (SClg) versus intravenous immunoglobulin (IVlg) in the management of neuromuscular junction (NMJ) disorders was chosen for its focus on myasthenia gravis and Lambert Eaton Myasthenic Syndrome. The study will determine if the same constant therapeutic effect seen when treating primary immunodeficiency syndrome patients subcutaneously can be reproduced in NMJ disorder patients.

“While immunoglobulin treatment for NMJ disorders is practiced, there is much to learn about the differences between IVlg and SClg treatment. Specifically, we’ll assess whether SClg treatment can maintain a higher trough level for patients so they do not go through fluctuations in symptoms, thereby improving quality of life and management of their symptoms.” said Dr. Alsharabati. “I’m enthusiastic about conducting this research and am honored to have been selected to receive the Interlaken Leadership Award.”

“CSL Behring is proud to support Dr. Alsharabati in his endeavors advancing NMJ disorders research.” said Karen MacPhail, Senior Director, Immunology, CSL Behring. “Studies like this are focused on improving patients’ lives, which is at the heart of our company values. CSL Behring is committed to supporting innovative research that might lead to the discovery of new treatment options for our patients.”

Since 2010, the Interlaken Leadership Awards has awarded over $5 million in grants and/or study drug for research studying Ig therapy in areas such as neuromyelitis optica (NMO), Duchenne muscular dystrophy (DMD), complex regional pain syndrome (CRPS), paraneoplastic syndromes, autoimmune peripheral neuropathies, and Guillain-Barré syndrome (GBS). All proposals received for the Interlaken Leadership Awards program are evaluated based on scientific merit, strength of hypothesis, relevance to neuroimmunology and feasibility.

About CSL Behring

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CSL Behring Receives Positive CHMP Opinion for Respreeza® as Maintenance Treatment for Severe Alpha-1 Antitrypsin Deficiency Patients in Europe

Respreeza® has been shown to slow the progression of emphysema

MARBURG, Germany — 29 June 2015

CSL Behring announced today that the European Medicines Agency (EMA) Committee for Medicinal Products for Human Use (CHMP) has recommended granting marketing authorization for Respreeza®, a highly purified alpha-1 protein derived from human plasma, indicated to treat patients with alpha-1 antitrypsin deficiency (AATD). AATD is a hereditary condition marked by a lack of the alpha-1 antitrypsin protein, whose main function is to protect the lungs from inflammation. Respreeza® replaces the protein that these patients are missing and raises the alpha-1 antitrypsin levels in their blood, which can help to protect the lungs from damage due to inflammation.

“CSL Behring continues to make strides towards fulfilling our promise to improve the lives of our patients. This positive opinion from CHMP brings us closer to providing Respreeza® as a new treatment option to the AATD community in Europe,” said Lutz Bonacker, Senior Vice President & General Manager Commercial Operations, Europe, CSL Behring.

The CHMP positive opinion will be transmitted to the European Commission (EC) to start the EC decision-making process. The EC may then grant a marketing authorization for Respreeza® as a maintenance treatment to slow the progression of emphysema in adults with documented severe alpha-1 proteinase inhibitor deficiency. CSL Behring’s RAPID (randomized, placebo-controlled trial of augmentation therapy in alpha-1 proteinase inhibitor deficiency) study results were considered as part of this submission. According to findings of the study, patients with AATD treated with alpha-1 proteinase inhibitor therapy exhibited a lower annual rate of lung density decline compared to placebo, when measured using chest computed tomography, at full inspiration. This demonstrated that Respreeza® significantly slows the progression of emphysema in these critically ill patients.

About CSL Behring’s Pulmonary Products

Respreeza® will be indicated for maintenance treatment, and to slow the progression of emphysema in adults with documented severe alpha-1 proteinase inhibitor deficiency (e.g. genotypes PiZZ, PiZ(null), Pi(null,null), PiSZ). Patients are to be under optimal pharmacologic and non-pharmacologic treatment and show evidence of progressive lung disease (e.g. lower forced expiratory volume per second (FEV1) predicted, impaired walking capacity or increased number of exacerbations) as evaluated by a healthcare professional experienced in the treatment of alpha-1 proteinase inhibitor deficiency. Respreeza® is contraindicated in patients with hypersensitivity to the active substance or to any of the excipients and IgA deficient patients with known antibodies against IgA, due to the risk of severe hypersensitivity and anaphylactic reactions.

Zemaira® is a highly-purified form of alpha-1 proteinase inhibitor (human) currently approved in Brazil, New Zealand, and the US, where it is indicated for chronic augmentation and maintenance therapy in adults with AATD and clinical evidence of emphysema. Zemaira® is contraindicated in patients with a history of severe systemic reaction to the product or to A1-PI protein, including anaphylaxis. Due to the risk of severe hypersensitivity, Zemaira® is also contraindicated in immunoglobulin A-deficient patients with antibodies against IgA.

About Alpha-1 Antitrypsin Deficiency

Alpha-1 Antitrypsin Deficiency is an inherited condition that can result in severe lung disease in adults and liver disease at any age, as well as other less known manifestations such as paniculitis, a skin disease. AATD is the most commonly known genetic risk factor for emphysema and is commonly
referred to as genetic COPD. Low levels or absence of the protective protein alpha-1 antitrypsin, which is produced by the liver, characterize AATD.

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Swissmedic Accepts for Review CSL Behring’s MAA for rIX-FP for Patients with Hemophilia B

BERN, Switzerland — 28 July 2015

- Upon approval, rIX-FP will offer hemophilia B patients in Switzerland prolonged dosing intervals of up to 14 days for routine prophylaxis
- rIX-FP regulatory submissions now made in the U.S., European Economic Area and Switzerland
- Latest milestone underscores CSL Behring’s deep commitment to developing and delivering innovative specialty biotherapies that improve the well-being of patients with serious diseases

CSL Behring announced today that Swissmedic has accepted for review a Marketing Authorization Application (MAA) for the company’s investigational long-acting fusion protein linking recombinant coagulation factor IX with recombinant albumin (rIX-FP) for the prophylaxis and treatment of bleeding episodes in people with Hemophilia B. Upon Swissmedic approval, rIX-FP will provide hemophilia B patients with a long-acting treatment option with dosing intervals up to 14 days.

Hemophilia B is a congenital bleeding disorder characterized by deficient or defective factor IX. It affects approximately 1 in 25,000 to 50,000 people. Nearly all patients are male.

"rIX-FP is built on CSL’s strength in protein research and development and scientific leadership in bleeding disorders, coupled with a long-standing relationship with the hemophilia community," said Dr. Andrew Cuthbertson, Chief Scientific Officer and R&D Director, CSL Limited. "Swissmedic initiating its review of the MAA for rIX-FP moves us one step closer to bringing this innovative specialty biotherapy to patients with hemophilia B in Switzerland and further underscores our deep commitment to improving the well-being of people with serious medical conditions."

The submission is based on the PROLONG-9FP clinical development program. PROLONG-9FP includes Phase I through Phase III open-label, multicenter studies evaluating the safety and efficacy of rIX-FP in adults and children (ages 1 to 61 years) with hemophilia B (FIX ≤ 2%) who were previously treated with other factor IX products.

Results from the Phase III studies were recently presented at the 2015 International Society on Thrombosis and Haemostasis (ISTH) Congress in Toronto. The data showed median annualized spontaneous bleeding rates (AsBR) of 0.00 in patients using rIX-FP prophylactically once every 7 days (age 1-11 years) and once up to every 14 days (age 12-61 years). The data also showed that 97 percent (age 1-11 years) and 99 percent (age 12-61 years) of bleeding events were treated with one or two infusions, with the majority of events (89 percent in patients 1-11 years, and 94 percent in patients 12-61 years) treated with only one infusion. Across the Phase III studies, none of the patients developed inhibitors to factor IX or antibodies to rIX-FP. The most common adverse reaction in clinical trials was headache.

About rIX-FP

CSL Behring engineered rIX-FP to extend the half-life of recombinant factor IX through genetic fusion with recombinant albumin. CSL Behring selected recombinant albumin as its recombinant genetic fusion partner for its coagulation factor proteins due to its long physiological half-life.

Earlier this year, CSL Behring announced that the U.S. Food and Drug Administration accepted for review the company’s Biologics License Application (BLA) for rIX-FP, and the European Medicines Agency (EMA) started the Centralized Procedure for reviewing its MAA for rIX-FP.

CSL Behring’s Operations in Switzerland
In June, CSL Behring broke ground on its state-of-the-art coagulation factor plant (CSL Behring Recombinant Facility AG) in Lengnau. CSL Behring expects the plant to be fully operational by 2019. The project is expected to create at least 300 new jobs and stimulate an investment of more than 400 million Swiss Francs in the local economy over the next five years. Once operational, CSL Behring’s recombinant factor products will be manufactured there. This includes rIX-FP as well as CSL Behring’s other recombinant factor candidates in development: rVIII-SingleChain for the treatment of hemophilia A and rVIIa-FP for the treatment of hemophilia A or B with inhibitors as well congenital factor VII deficiency.

For more than 60 years, CSL Behring has been operating a research and production plant in Bern for its immunoglobulins and albumin, employing over 1,300 people.

For more information about CSL Behring’s recombinant products in development to treat hemophilia, visit http://www.cslbehring.com/products/bleeding-disorders/novel-recombinant-hemophilia-treatments.

The information in this news release is exclusively intended to inform the press. CSL Behring explicitly refers to the advertising provisions of the Swiss Pharmaceutical Advertising Law (Art. 31 and 32 of the Federal Act on Medicinal Products and Medical Devices [TPA, SR 812.21] as well as the respective regulation regarding advertising of medicinal products [AWV, SR 812.212.5]), CSL Behring especially refers to the prohibition of advertising prescription-only medicinal products directed at the general public.

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U.S. FDA Accepts for Review CSL Behring’s Biologics License Application for Its Novel rVIII-SingleChain Therapy for Patients with Hemophilia A

rVIII-SingleChain underscores CSL Behring’s deep commitment to developing and delivering specialty biotherapies that improve the well-being of patients with serious diseases


CSL Behring announced today that the U.S. Food and Drug Administration has accepted for review the company’s Biologics License Application (BLA) for its novel investigational recombinant factor VIII single-chain (rVIII-SingleChain) for the treatment of hemophilia A. In the pivotal clinical trial, rVIII-SingleChain met all primary endpoints.

Hemophilia A is a congenital bleeding disorder characterized by deficient or defective factor VIII; nearly all affected patients are male. People with hemophilia A may experience prolonged or spontaneous bleeding, especially into the muscles, joints, or internal organs. The condition affects approximately 1 in 6,000 male births.

“CSL Behring has one of the industry’s largest portfolios of biotherapies that improve the care and well-being of patients with a bleeding disorder,” said Dr. Andrew Cuthbertson, Chief Scientific Officer and Director of R&D, CSL Limited. “Our scientific expertise and relationship with the bleeding disorders community led us to seek further advancements in the care and treatment of hemophilia. Today, we have the only recombinant single-chain factor VIII product in late-stage development for the management of hemophilia A, and we are excited to be one step closer to providing this innovative treatment to patients in the U.S.”

The BLA submission is based on the AFFINITY clinical development program, which includes a phase I/III open-label, multi-center trial examining safety and efficacy. The pharmacokinetics of rVIII-SingleChain compared with recombinant human antihemophilic factor VIII (octocog alfa) was also studied. Study design details for rVII SingleChain are available at clinicaltrials.gov.

Results from the phase I/III study were recently presented at the International Society on Thrombosis and Haemostasis (ISTH) congress in Toronto. Patients treating prophylactically had a median annualized bleeding rate (ABR) of 1.14 and a median annualized spontaneous bleeding rate (AsBR) of 0.00. The data also showed that, of 848 bleeds treated in the study, 94 percent were controlled with no more than two infusions of rVIII-SingleChain, with 81 percent controlled by one infusion. Moreover, hemostatic control of a bleeding event treated with rVIII-SingleChain was assessed by the investigator as excellent or good 94 percent of the time (835 assessed bleeding events).

The results presented included data on more than 14,000 exposure days in 146 patients on prophylaxis and 27 patients treating on demand for a bleeding event. In total, 120 patients were treated for more than 50 days of exposure; 52 had more than 100 days of exposure. In the prophylaxis group, 32 percent of patients were dosed twice weekly and 54 percent received treatment three times per week; the regimen was determined by the investigator. The most common adverse events were naso-pharyngitis, arthralgia, and headache. No inhibitors were reported.

About rVIII-SingleChain

Specifically designed for greater molecular stability, rVIII-SingleChain is the first and only single-chain factor VIII (FVIII) product in late-stage development for the treatment of hemophilia A. rVIII-SingleChain (also known as CSL627) has a strong affinity for von Willebrand factor, leading to greater stability and integrity of FVIII in circulation. For more information about CSL Behring’s recombinant products in development to treat hemophilia, visit http://www.cslbehring.com/products/bleeding-disorders/novel-recombinant-hemophilia-treatments.
About CSL Behring

The people and science of CSL Behring save lives around the world. We develop and deliver innovative specialty biotherapies, driven by our 100-year promise to help people with life-threatening conditions live full lives. With 14,000 employees and operations in 30 countries, CSL applies world-class R&D, high-quality manufacturing and patient-centered management.

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 disease of the newborn.

CSL Behring operates one of the world’s largest plasma collection networks, CSL Plasma. CSL Behring is a global biopharmaceutical company and a member of the CSL Group of companies. The parent company, CSL Limited (ASX:CSL), is headquartered in Melbourne, Australia. For more information, visit www.cslbehring.com.

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CSL Behring Enrolls First Patient in Global Phase II/III Study of rVIIa-FP for On-Demand Treatment of Patients with Hemophilia A or B with Inhibitors

KING OF PRUSSIA, Pa. — 25 August 2015

- Key milestone achieved in CSL Behring’s PROLONG-7FP clinical development program
- PROLONG-7FP is studying CSL Behring’s recombinant fusion protein linking coagulation factor VIIa with albumin (rVIIa-FP)
- Milestone underscores CSL Behring’s focused scientific expertise and deep commitment to developing and delivering innovative specialty biotherapies for people with serious diseases

CSL Behring announced today that the first patient has been enrolled in its Phase II/III clinical study evaluating the pharmacokinetics (PK), efficacy, and safety of the company’s recombinant fusion protein linking coagulation factor VIIa with albumin (rVIIa-FP) for on-demand treatment in patients with congenital hemophilia A or B who have developed an inhibitor to factor VIII or factor IX replacement therapy. The study will enroll approximately 54 male patients, the first of whom was enrolled in Malaysia.

“CSL Behring has a thorough understanding of the bleeding disorders community, focused scientific expertise and a strong commitment to developing and delivering innovative specialty biotherapies that treat serious medical conditions,” said Dr. Andrew Cuthbertson, Chief Scientific Officer and Director of R&D, CSL Limited. “Our commitment, expertise and understanding helped CSL to develop rVIIa-FP, based on the innovative recombinant albumin fusion technology platform, to treat patients with hemophilia A or B with inhibitors as well as congenital factor VII deficiency.”

About Hemophilia A or B with Inhibitors

Hemophilia is an inherited bleeding disorder characterized by prolonged or spontaneous bleeding, especially into the muscles and joints. The disorder is caused by deficient or defective blood coagulation proteins known as factor VIII (hemophilia A) or IX (hemophilia B). The recommended treatment for these patients is factor replacement therapy. Development of inhibitory antibodies against factor VIII or factor IX is a major complication associated with replacement therapy and may partially or completely prevent the efficacy of factor replacement therapy. The incidence of inhibitors in individuals with hemophilia A is estimated to be up to 33 percent and between one and 6 percent in patients with hemophilia B. The most recent World Federation of Hemophilia (WFH) survey identified more than 4,753 patients with hemophilia A and 248 with hemophilia B who have developed an inhibitor worldwide.¹

About rVIIa-FP

Preclinical studies have confirmed that CSL Behring’s rVIIa-FP (also known as CSL689) has favorable pharmacokinetic properties compared with the existing recombinant FVIIa product.² In a phase I study in healthy volunteers, rVIIa-FP showed a good tolerance, and a 3- to 4-fold increase in half-life compared with the commercially available rFVIIa-product (median 8.5h).³

The European Commission granted Orphan Drug Designations for rVIIa-FP for the treatment of patients with hemophilia A or B who have developed an inhibitor as well as the treatment of congenital factor VII deficiency. rVIIa-FP also received Orphan Drug Designation from the U.S. Food and Drug Administration (FDA) for the treatment and prophylaxis of bleeding episodes in patients with congenital hemophilia and inhibitors to coagulation factor VIII or IX. Orphan designation qualifies the sponsor of the product for important benefits such as tax credits for qualified clinical testing and exemption from certain application fees.

For more information about CSL Behring’s recombinant products in development to treat hemophilia, visit http://www.cslbehring.com/products/bleeding-disorders/novel-recombinant-hemophilia-treatments.
About PROLONG-7FP Clinical Development Program

This Phase II/III study in patients with hemophilia A or B who have developed an inhibitor is a part of the PROLONG-7FP clinical development program. This program aims to demonstrate the therapeutic advantages of rVIIa-FP in patients with hemophilia A or hemophilia B who have developed inhibitors as well as in patients with congenital FVII deficiency. A Phase I PK study in patients with congenital FVII deficiency is ongoing.

For more information about the Phase II/III study, please visit clinicaltrials.gov.

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CSL Behring Receives Marketing Authorization for Respreeza® in Europe

*Respreeza® is a maintenance treatment for severe Alpha-1 Antitrypsin Deficiency patients and has been shown to slow the progression of emphysema*

MARBURG, Germany — 25 August 2015

CSL Behring announced today that the European Commission (EC) has granted marketing authorization in all European Union (EU) member states for Respreeza® to treat patients with Alpha-1 antitrypsin deficiency (AATD). Respreeza®, a highly purified Alpha-1 protein derived from human plasma, is the only Alpha-1 proteinase inhibitor that has been proven in a prospective double blind, placebo controlled trial (the RAPID study) to significantly reduce the loss of lung tissue, slowing the progression of emphysema due to AATD. AATD is a hereditary condition marked by a lack of the Alpha-1 antitrypsin protein, whose main function is to protect the lungs from inflammation.

“AATD is a potentially debilitating disease and many affected individuals suffer from serious lung disease,” said Professor Helmut Teschler, MD, Director of the West German Lung Centre at the University of Essen. “With the approval of Respreeza®, healthcare professionals can now provide patients with severe AATD in Europe with a next generation Alpha1-proteinase inhibitor (A1-PI) that provided additional evidence that this augmentation therapy can slow the accelerated loss of lung tissue.”

Frank Willersinn, M.D., Alpha-1 Global Steering Committee Chair and patient representative in Europe added, “We are so glad that Respreeza® has been approved by the European Medicines Agency. We commend CSL Behring for their long-lasting commitment to the Alpha-1 community, now bringing their established Alpha 1-antitrypsin product to Europe, allowing it to be a cornerstone for treatment of AATD patients in the near future.”

CSL Behring has marketed A1-PI for 12 years as Zemaira® in the United States, where it is indicated for chronic augmentation and maintenance therapy in adults with A1-PI deficiency and clinical evidence of emphysema. In US, Zemaira® is contraindicated in patients with a history of severe systemic reaction to the product or to A1-PI protein, including anaphylaxis. Due to the risk of severe hypersensitivity, Zemaira® is also contraindicated in immunoglobulin A (IgA)-deficient patients with antibodies against IgA.

**About Respreeza®**

In EU, Respreeza® is indicated for maintenance treatment, and to slow the progression of emphysema in adults with documented severe A1-PI deficiency (e.g. genotypes PiZZ, PiZ(null), Pi(null,null), PiSZ). Patients are to be under optimal pharmacologic and non-pharmacologic treatment and show evidence of progressive lung disease (e.g. lower forced expiratory volume per second (FEV1) predicted, impaired walking capacity or increased number of exacerbations) as evaluated by a healthcare professional experienced in the treatment of A1-PI deficiency. Respreeza® is contraindicated in patients with hypersensitivity to the active substance or to any of the excipients and IgA deficient patients with known antibodies against IgA, due to the risk of severe hypersensitivity and anaphylactic reactions.

**About the RAPID Study**

The RAPID study (Randomized, Placebo-controlled Trial of Augmentation Therapy in Alpha-1 Proteinase Inhibitor Deficiency) enrolled 180 severely deficient alpha-1 patients in the U.S. and Europe who were treated for 2 years with Respreeza® or Placebo. The study, published in Lancet in June 2015, showed a significant reduction in the rate of lung tissue loss as measured by CT scan lung density. In those patients who continued into the extension study, where all subjects received Respreeza®, the benefit was maintained for another 2 years.
About Alpha-1 Antitrypsin Deficiency

Alpha-1 Antitrypsin Deficiency is an inherited condition that can result in severe lung disease in adults and liver disease at any age, as well as other less known manifestations such as panarititis, a skin disease. AATD is the most commonly known genetic risk factor for emphysema and is commonly referred to as genetic COPD. Low levels or absence of the protective protein alpha-1 antitrypsin, which is produced by the liver, characterize AATD.

About CSL Behring

CSL Behring is a global 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 disease of the newborn.

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Centers for Medicare and Medicaid Services Extends New Technology Add-On Payment for CSL Behring’s Kcentra® for Third Year

Designation Reinforces the Benefit of Kcentra for Warfarin Reversal in the Inpatient Hospital Setting; Kcentra Delivers on CSL Behring’s Promise to Save Lives Using the Latest Technologies

KING OF PRUSSIA, Pa. — 08 September 2015

CSL Behring today announced that the Centers for Medicare and Medicaid Services (CMS) again extended the new technology add-on payment (NTAP) for Kcentra® (Prothrombin Complex Concentrate [Human]). The NTAP for Kcentra is available through September 2016 for eligible Medicare beneficiaries treated in the inpatient hospital setting. Kcentra is the first and only non-activated 4-factor prothrombin complex concentrate (4F-PCC) approved by the U.S. Food and Drug Administration (FDA) for the urgent reversal of acquired coagulation factor deficiency induced by Vitamin K antagonist (VKA, e.g., warfarin) therapy in adult patients with acute major bleeding or in need of an urgent surgery or invasive procedure. Kcentra, first approved for use in the U.S. in April 2013, received its NTAP designation effective October 1, 2013.

"With the extension of the NTAP designation for Kcentra, hospitals will continue to have broad access to this specialty biotherapy for warfarin reversal," said Bill Campbell, Senior Vice President and General Manager, North America, CSL Behring. "Warfarin is an important treatment for millions of people living with cardiovascular disease, but it can lead to severe, potentially life-threatening bleeding. Kcentra restores depleted clotting proteins to reverse the effects of warfarin and demonstrates CSL Behring’s promise to develop and deliver innovative treatments that save lives."

Unlike fresh frozen plasma (FFP), the most widely used agent for warfarin reversal, Kcentra does not require thawing or blood-type matching and can be administered more quickly and with less volume than FFP.

About the New Technology Add-On Payment (NTAP) Policy

The CMS NTAP policy was implemented in 2001 to support timely access to innovative therapies for Medicare beneficiaries in the inpatient hospital setting that are not adequately paid for under the Medicare Severity Diagnosis-Related Groups (MS-DRGs). To be eligible for an NTAP, the product must be new and inadequately paid for under existing MS-DRGs and provide a significant clinical improvement over existing therapies. CMS will continue to reimburse hospitals an additional amount, up to $1,587.50, for cases involving Kcentra that exceed the MS-DRG payment amount.


Prevalence of Warfarin Therapy

Each year, approximately three to four million people in the U.S. are treated with the oral anticoagulant warfarin to prevent blood clots from forming following a stroke, heart attack, heart valve surgery, deep vein thrombosis/pulmonary embolism, or certain types of irregular heartbeat, such as atrial fibrillation. However, because of the deficiency in blood clotting factors induced by warfarin treatment, patients may experience severe bleeding. It is estimated that emergency departments across the U.S. see approximately 29,000 cases annually for warfarin-associated bleeding.

About Kcentra®

CSL Behring markets Kcentra as Beriplex® or Conﬁdex® in more than 25 countries. In December 2012, the FDA granted Orphan Drug Designation to Kcentra for the treatment of patients needing...
urgent reversal of Vitamin K antagonist therapy due to major bleeding and/or surgical procedures. The FDA's Orphan Drug Designation program provides orphan status to unique drugs and biologics defined as those intended for the safe and effective treatment or prevention of rare diseases that affect fewer than 200,000 people in the U.S. Orphan designation qualifies the sponsor of the product for important benefits such as tax credits for qualified clinical testing and exemption from certain application fees.

Important Safety Information

Kcentra®, Prothrombin Complex Concentrate (Human), is a blood coagulation factor replacement product indicated for the urgent reversal of acquired coagulation factor deficiency induced by Vitamin K antagonist (VKA; e.g., warfarin) therapy in adult patients with acute major bleeding or the need for urgent surgery or other invasive procedure. Kcentra is for intravenous use only.

WARNING: ARTERIAL AND VENOUS THROMBOEMBOLIC COMPLICATIONS

Patients being treated with Vitamin K antagonist therapy have underlying disease states that predispose them to thromboembolic events. Potential benefits of reversing VKA should be weighed against the risk of thromboembolic events, especially in patients with history of such events. Resumption of anticoagulation therapy should be carefully considered once the risk of thromboembolic events outweighs the risk of acute bleeding. Both fatal and nonfatal arterial and venous thromboembolic complications have been reported in clinical trials and postmarketing surveillance. Monitor patients receiving Kcentra, and inform them of signs and symptoms of thromboembolic events. Kcentra was not studied in subjects who had a thromboembolic event, myocardial infarction, disseminated intravascular coagulation, cerebral vascular accident, transient ischemic attack, unstable angina pectoris, or severe peripheral vascular disease within the prior three months. Kcentra might not be suitable for patients with thromboembolic events in the prior three months.

Kcentra is contraindicated in patients with known anaphylactic or severe systemic reactions to Kcentra or any of its components (including heparin, Factors II, VII, IX, X, Proteins C and S, Antithrombin III and human albumin). Kcentra is also contraindicated in patients with disseminated intravascular coagulation. Because Kcentra contains heparin, it is contraindicated in patients with heparin-induced thrombocytopenia (HIT).

Hypersensitivity reactions to Kcentra may occur. If patient experiences severe allergic or anaphylactic type reactions, discontinue administration and institute appropriate treatment.

In clinical trials, the most frequent (≥2.8%) adverse reactions observed in subjects receiving Kcentra were headache, nausea/vomiting, hypotension, and anemia. The most serious adverse reactions were thromboembolic events, including stroke, pulmonary embolism and deep vein thrombosis.

Kcentra is derived from human plasma. The risk of transmission of infectious agents, including viruses and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent, cannot be completely eliminated.

The safety and efficacy of Kcentra in pediatric use have not been studied, and Kcentra should be used in women who are pregnant or nursing only if clearly needed.

For more information about Kcentra, please visit www.kcentra.com or call toll-free 1-855-4KCENTRA. For full prescribing information, please visit www.kcentra.com/prescribing-information.aspx.

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New Clinical Data on Treatment to Slow the Progression of Emphysema in Patients with Alpha-1 Antitrypsin Deficiency Presented at European Respiratory Society International Congress in Amsterdam

MARBURG, Germany — 28 September 2015

CSL Behring today hosted a symposium highlighting an option to slow the progression of emphysema in adults with documented severe alpha-1 antitrypsin deficiency (AATD), during the European Respiratory Society (ERS) International Congress, which is being held from 26-30 September in Amsterdam, Netherlands. AATD is a hereditary condition marked by a lack of the alpha-1 antitrypsin protein, whose main function is to protect the lungs from inflammation.

The symposium was chaired by Professor Gerry McElvaney, MD, Royal College of Surgeons in Ireland, and included presentations by Professor Kenneth R. Chapman, MD, (Canada), Dr. David Parr, MD (United Kingdom), Dr. Emer Reeves, MD (Ireland) and Dr. Niels Seersholm, MD (Denmark).

During the symposium, Dr. Chapman highlighted evidence from the recently published RAPID trial of alpha-1 proteinase inhibitor (A1-PI) therapy. Chapman notes, “RAPID is regarded as a landmark study validating almost two decades of focus on the lung-density endpoint as the most sensitive way to track lung tissue decline in this potentially debilitating disease. Our findings, for the first time in a single randomized-controlled trial, provided additional evidence that treatment with an alpha-1 proteinase inhibitor provides a clinical benefit by slowing the accelerated loss of lung tissue.”

Professor McElvaney also presented data on the RAPID Extension trial, where eligible subjects received A1-PI for another 2 years, in an oral presentation in a separate late-breaker session on 27 September. “The extended trial showed that, over 48 months, A1-PI reduced the rate of lung density decline. The treatment effect was not only maintained by patients treated for all 4 years, but also showed significant reduction in terms of absolute lung density loss in patients that were on placebo for the first 2 years and then switched to active therapy in the last 2 years.” said, Professor McElvaney. “These results further confirm the findings of the RAPID trial and support the use of augmentation therapy in the treatment of severe AATD.”

Data from the RAPID and RAPID Extension trials were the basis for the marketing application in the EU for Respreeza®, a highly purified alpha-1 protein derived from human plasma, in the treatment of severe AATD. The European Commission granted approval of Respreeza® on 20 August, 2015. It is now marketed by CSL Behring in Europe and the first national launches of the product are imminent. Respreeza® is the only alpha-1 proteinase inhibitor that has been proven in a prospective double blind, placebo controlled trial (the RAPID study) to significantly reduce the loss of lung tissue, slowing the progression of emphysema due to AATD.

About the RAPID Program

RAPID Trial - The RAPID trial was a multicenter, double-blind, randomized, parallel group, placebo-controlled study comparing the efficacy and safety of Respreeza®, CSL Behring’s A1-PI therapy, with placebo in patients with emphysema due to AATD. The participants, 180 non-smokers aged 18-65, were randomly assigned to receive A1-PI intravenously 60 mg/kg weekly or placebo.

RAPID Trial Study Design and Findings - According to study protocol, the effect of Respreeza® on the progression of emphysema, the primary endpoint of the study, was measured by computed tomography scan over 24 months and was assessed by the annual rate of lung density loss at total lung capacity (TLC) and functional residual capacity (FRC). The annual rate of lung density loss at TLC (i.e., full aspiration) showed a statistically significant 34 percent reduction in the annual rate lung density decline compared with placebo. There was 24 percent reduction when measured at FRC (i.e., resting exhalation), which was not significant. Serious adverse events were not statistically different between groups with one death in the A1-PI group and three deaths in the placebo group.
RAPID Trial Extension - Eligible subjects completing the 2-year RAPID trial were enrolled into a 2-year, open label RAPID Extension trial where they either continued to receive weekly infusions of Respreeza® for another 2 years (Early Start group) or were switched from placebo to receive 2 years of Respreeza® therapy (Delayed Start group).

The Early Start group maintained an advantage over placebo across the 48 month period. The Delayed Start group showed a statistically significant reduction in the absolute loss of lung density in response to the administration of Respreeza® in the last 2 years compared to the loss in the first 2 years while exposed to placebo. The number and proportion of fast decliners (annual rate of lung density loss > 2g/L/y) was reduced in association with treatment with Respreeza® across both studies. The 4-year data also demonstrated moderate, statistically significant correlations between changes in lung density decline and changes in FEV1, FEV1% predicted and FVC.

These effects across both studies demonstrate a consistent disease modifying effect of A1-PI therapy, and suggest that early treatment may be more appropriate to effectively slow the progression of emphysema.

About Respreeza®

In EU, Respreeza® is indicated for maintenance treatment, and to slow the progression of emphysema in adults with documented severe A1-PI deficiency (e.g. genotypes PIZZ, PIZ(null), Pi(null,null), PiSZ). Patients are to be under optimal pharmacologic and non-pharmacologic treatment and show evidence of progressive lung disease (e.g. lower forced expiratory volume per second (FEV1) predicted, impaired walking capacity or increased number of exacerbations) as evaluated by a healthcare professional experienced in the treatment of A1-PI deficiency. Respreeza® is contraindicated in patients with hypersensitivity to the active substance or to any of the excipients and IgA deficient patients with known antibodies against IgA, due to the risk of severe hypersensitivity and anaphylactic reactions.

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CSL Behring Announces Winners of its 14th Annual Gettin’ in the Game℠ Junior National Championship Program

JNC Demonstrates CSL Behring’s Promise to Develop and Deliver Support Programs for Kids with Hemophilia and/or von Willebrand disease (VWD) and Their Families

PHOENIX — 29 October 2015

CSL Behring announced that Tyler Reitano from the Cascade Foundation of Southern Arizona and Maklain Briggs from the Utah Hemophilia Foundation are the national winners of the 2015 Gettin’ in the Game℠ Junior National Championship (JNC) program in golf and baseball, respectively. The JNC, launched in 2002, is CSL Behring’s annual baseball and golf competition that encourages kids to remain active despite the challenges a bleeding disorder can pose, while allowing them to develop life-long connections with other members of the community.

“Through innovative products and programs, CSL Behring helps people with life threatening medical conditions live full lives,” said Bill Campbell, Senior Vice President and General Manager, North America, CSL Behring. "The JNC is one program that delivers on our promise of making a real and lasting difference in the lives of people in the bleeding disorders community. Congratulations to Tyler and Maklain and all of the competitors who participated in this year’s event.”

CSL Behring’s JNC program is the first and only national golf and baseball competition designed specifically for the bleeding disorders community. In addition to the competitive activities, this event provides children and their caregivers with educational information and opportunities to interact with others in the community. The JNC features accomplished athletes, who themselves have been diagnosed with a bleeding disorder. These athletes hold clinics to help educate children and their families about the fundamentals of golf and baseball and share their stories with participants to encourage them to remain active as a part of managing their condition.

For more information about the JNC program or the wide variety of resources for the bleeding disorders community offered by CSL Behring, please visit www.MySourceCSL.com. CSL Behring’s My Source program provides one-stop access to educational information, financial support and community connections to individuals living with hemophilia and/or VWD.

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CSL Behring to Present New Data for rIX-FP and rVIII-SingleChain at the 57th ASH Annual Meeting & Exposition

KING OF PRUSSIA, Pa — 02 December 2015

Abstracts include pivotal efficacy and safety data for CSL Behring’s long-acting recombinant albumin fusion protein for hemophilia B; CSL Behring’s recombinant coagulation factor development program reinforces the company’s promise to develop specialty biotherapies that help people with serious medical conditions live full lives.

CSL Behring announced today that it will present several abstracts from its recombinant coagulation factor development programs at the American Society of Hematology’s (ASH) 57th ASH Annual Meeting in Orlando, December 5-8. An oral presentation will focus on the pivotal trial for CSL Behring’s novel, long-acting albumin fusion protein linking recombinant coagulation factor IX with recombinant albumin (rIX-FP) for the treatment of hemophilia B.

Hemophilia is an inherited bleeding disorder caused by deficient or defective proteins that prevent the blood from clotting properly. According to the National Hemophilia Foundation, the condition affects approximately 400,000 people worldwide, the majority of whom have hemophilia A.

“The clinical findings that CSL Behring is presenting at ASH reinforce our promise to advance hemophilia care by developing innovative specialty biotherapies that have the potential to improve patients’ lives,” said Dr. Andrew Cuthbertson, Chief Scientific Officer and Director of R&D, CSL Limited. “For nearly a century, CSL has developed and delivered innovations that have helped people with serious medical conditions live full lives. Our recombinant coagulation factor development pipeline is another example of our R&D focus to deliver treatment advancements that patients and providers want.”

CSL Behring medical representatives will be available at booth number 228 throughout the ASH Annual Meeting. Key abstracts from CSL Behring at ASH include:

rIX-FP

**Oral Presentation**

*Monday, December 7, 10:30 a.m. – 12:00 p.m. ET*

- Efficacy and safety results of PROLONG-9FP clinical program of recombinant fusion protein linking coagulation factor IX with albumin (rIX-FP) in previously treated patients with hemophilia B. Session Name: 322. Disorders of Coagulation or Fibrinolysis: Novel Treatment Strategies in Hemophilia. [Oral Presentation Abstract #548](#)

**Posters**

*Saturday, December 5, 5:30 p.m. – 7:30 p.m. ET*

- Long-Term Use of coagulation factor IX (Recombinant) albumin fusion protein (rIX-FP) in previously treated patients with hemophilia B in the PROLONG-9FP program. [Poster Presentation Abstract # 1096](#)

*Sunday, December 6, 6:00 p.m. – 8:00 p.m. ET*
• Efficacy and safety of recombinant fusion protein linking coagulation factor IX with albumin (rIX-FP) in previously treated patients with hemophilia B undergoing major orthopedic surgeries. 

Poster Presentation Abstract # 2298

rVIII-SingleChain

Posters

Saturday, December 5, 5:30 p.m. - 7:30 p.m. ET

• Pharmacokinetic properties of different rFVIII products in rabbits, rats and FVIII ko mice. Poster Presentation Abstract #1078

Monday, December 7, 6:00-9:00 p.m. ET

• rVIII-SingleChain in surgical prophylaxis: Efficacy and safety of continuous and bolus infusion. Poster Presentation Abstract # 3525

Monday, December 7, 6:00-8:00 p.m. ET

• rVIII-SingleChain pharmacokinetics and safety in children less than 12 years of age. Poster Presentation Abstract #3524

About rIX-FP

CSL Behring engineered rIX-FP to extend the half-life of recombinant factor IX through genetic fusion with recombinant albumin. CSL Behring selected albumin as its recombinant genetic fusion partner due to its long physiological half-life. In addition, albumin has been shown to have a favorable tolerability profile, low potential for immunogenic reactions and a well-known mechanism of clearance. 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.

The U.S. Food and Drug Administration, European Medicines Agency and Swissmedic are currently reviewing CSL Behring’s license applications for rIX-FP. Upon regulatory approvals, rIX-FP will provide hemophilia B patients with a long-acting treatment option with dosing intervals up to 14 days.

The PROLONG-9FP clinical development program for rIX-FP covers patients from the age of 1 to 61 years. Studies in the program were conducted as open-label, multicenter, safety and efficacy studies of rIX-FP in previously treated patients with hemophilia B (factor IX level ≤ 2%). Study design details for rIX-FP (CSL654) are available at clinicaltrials.gov (NCT01496274).

About rVIII-SingleChain

Specifically designed for greater molecular stability, rVIII-SingleChain is the first and only recombinant single-chain factor VIII (FVIII) product in late-stage development for the treatment of hemophilia A. rVIII-SingleChain has a strong affinity for von Willebrand factor, leading to greater stability and integrity of factor VIII in circulation. The stability of rVIII-SingleChain leads to a longer-lasting therapeutic effect with reduced dosing frequency.

In July, the U.S. Food and Drug Administration accepted for review CSL Behring’s BLA for rVIII-SingleChain. The BLA submission is based on the AFFINITY clinical development program, which includes a phase I/III open-label, multi-center trial examining safety and efficacy. The pharmacokinetics of rVIII-SingleChain compared with recombinant human antihemophilic factor VIII
(octocog alfa) were also studied. Study design details for rVIII-SingleChain (CSL627) are available at clinicaltrials.gov (NCT01486927).

For more information about CSL Behring’s recombinant coagulation factor products in development to treat hemophilia, visit http://www.cslbehring.com/products/bleeding-disorders/novel-recombinant-hemophilia-treatments.

About CSL

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###
CSL Behring Presents Phase III Data for Its Long-Acting Recombinant Factor IX Albumin Fusion Protein for Hemophilia B at the 57th ASH Annual Meeting & Exposition

ORLANDO, Fla. — 07 December 2015

- Data from ongoing extension study evaluate efficacy and long-term safety of rIX-FP when dosed up to once every 14 days for routine prophylaxis
- The median annualized spontaneous bleeding rate (AsBR) was zero for all treatment intervals
- rIX-FP demonstrates CSL Behring’s promise to develop innovative specialty biotherapies that help people with serious medical conditions live full lives

Global biotherapeutics leader CSL Behring today presented data from its Phase III PROLONG-9FP clinical program evaluating the efficacy and long-term safety of its investigational long-acting fusion protein linking recombinant coagulation factor IX with recombinant albumin (rIX-FP). The data, from an ongoing extension study and two pivotal Phase III studies, assessed rIX-FP for routine prophylaxis in previously-treated adults with hemophilia B, at dosing intervals of up to 14 days. The findings were presented during an oral presentation at the American Society of Hematology’s (ASH) 57th ASH Annual Meeting and Exposition in Orlando, along with a second abstract reporting efficacy and safety results of rIX-FP in patients undergoing surgical procedures.

“The findings from the ongoing extension study are promising and confirm the data we saw in our pivotal studies, with prolonged dosing intervals of up to 14 days with rIX-FP for routine prophylaxis,” said Elena Santagostino, M.D., Ph.D., Professor in the Medical School of Clinical and Experimental Hematology at the University of Milan/IRCCS Maggiore Hospital, and lead investigator. “rIX-FP has been developed with a pharmacokinetic profile that allows for less frequent dosing. Through this ongoing study, we are continuing to evaluate longer treatment intervals and the potential impact on preventing bleeding in select patients.”

Key Study Findings

- **Efficacy and safety of rIX-FP in previously-treated children, adolescents, and adults (Oral Presentation Abstract #548):** The first abstract reported on two completed Phase III studies (CSL654-3001 and CSL654-3002) assessing the safety and efficacy of rIX-FP for prophylaxis treatment in previously-treated patients (ages 1 to 61) with hemophilia B (factor IX activity ≤2%). The first study (CSL654-3001) included 63 patients, ages 12 to 61. These patients either received prophylaxis treatment (once every seven days for six months and then, if eligible, a 10- or 14-day prophylaxis treatment interval for 12 to 18 months) or only on-demand treatment for six months and then switched to seven-day prophylaxis treatment. The second study (CSL654-3002) included 27 children (ages 1 to 11) who received seven-day prophylaxis treatment for approximately 12 months. Overall, the median annualized spontaneous bleeding rate (AsBR) was 0.00 for all treatment intervals.

  During the extension study, 76 patients from the phase III studies continued their prophylaxis regimen but additional patients switched to longer treatment intervals, including the 10- and 14-day intervals in patients younger than 12 and more prolonged treatment intervals in patients older than 18. At least 50 patients achieved 100 exposure days of treatment without developing an inhibitor to factor IX or antibodies to rIX-FP. Overall, no serious adverse reactions were seen during the studies and favorable long-term tolerability was demonstrated.

“At CSL, we focus our world-class research and development to help people with serious medical conditions, such as hemophilia B, live full lives,” said Dr. Andrew Cuthbertson, Chief Scientific Officer and Director of R&D, CSL Limited. “These latest results from our pivotal research program demonstrate the potential for rIX-FP to help patients maintain factor IX levels over a long period of time with greater freedom from frequent infusions. This is an important attribute for patients who require a prophylactic regimen but don’t want treatment to disrupt their active lives.”
rIX-FP in previously-treated patients undergoing surgery (poster presentation abstract #2298): The second abstract reported on a surgical sub-study included in the Phase III studies, including the ongoing extension study, as part of the global PROLONG-9FP clinical program. This abstract evaluated the use of rIX-FP in five major orthopedic surgeries to prevent bleeding during and post-surgery in four patients with hemophilia B. For all procedures, the investigators rated the response as “excellent” or “good,” and a single pre-operative dose of rIX-FP maintained hemostasis during surgery. Over the 14-day perioperative period, patients needed six or seven infusions. The median rIX-FP consumption prior to surgery and post-surgery was 340 IU/kg, which was lower than with conventional therapies. None of the patients developed inhibitors to factor IX or antibodies to rIX-FP.

About Hemophilia B

Hemophilia B is a congenital bleeding disorder characterized by deficient or defective factor IX; nearly all affected patients are male. People with hemophilia B may experience prolonged or spontaneous bleeding, especially into the muscles, joints, or internal organs. According to U.S. Centers for Disease Control and Prevention, the condition affects approximately one in 25,000 male births.

About rIX-FP

CSL Behring engineered rIX-FP to extend the half-life of recombinant factor IX through genetic fusion with recombinant albumin. CSL Behring selected albumin as its recombinant genetic fusion partner due to its long physiological half-life. In addition, albumin has been shown to have a favorable tolerability profile, low potential for immunogenic reactions and a well-known mechanism of clearance. 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.

The U.S. Food and Drug Administration, European Medicines Agency and Swissmedic are currently reviewing CSL Behring’s license applications for rIX-FP. For more information about CSL Behring’s recombinant coagulation factor products in development to treat hemophilia, visit http://www.cslbehring.com/products/bleeding-disorders/novel-recombinant-hemophilia-treatments.

About CSL

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###
CSL Behring Expands Operations to Russia

Moscow opening reinforces CSL Behring’s promise to deliver its world-class therapies to patients with rare and serious diseases around the world.

KING OF PRUSSIA, Pa. — 08 December 2015

Global biotherapeutics leader CSL Behring announced today its latest geographic expansion to provide more patients with greater access to treatment by opening operations in Russia. This is particularly significant in Russia where the healthcare system has some unmet needs for state-of-the-art biotherapies and blood plasma products.

The annual usage of certain classes of these medicines in Russia is much lower than it should be. For example, consumption of immunoglobulins in Russia per capita is 10 to 20 times lower than in the USA and some European countries.

With operations in over 30 countries, CSL Behring focuses on developing novel, protein-based therapies for the treatment of bleeding disorders, immune deficiencies, inherited respiratory disease and hereditary angioedema, and for neurological disorders in certain markets. CSL Behring is a subsidiary of CSL Limited.

CSL CEO and Managing Director Paul Perreault, who joined Russian healthcare leaders and patient advocates at the office opening today, said the new office enables CSL Behring to partner more closely with the Russian Federation, healthcare providers, patient groups and the scientific community. He noted CSL Behring will be closer to patients, listen to them more carefully, and better understand their medical needs. The company currently has seven products registered in Russia.

"It is this process of listening and engaging that enhances our ability to deliver new and innovative medicines that make such a huge difference in people’s lives," Perreault said. He added that it will now be easier to launch products in Russia and offer new therapy options to doctors and their patients.

In addition, CSL Behring is investigating opportunities to contribute to the development of the Russian pharmaceutical industry, and identify the best ways to partner with the Russian government. As an example, because the amount of human plasma that is currently collected in Russia is insufficient to meet the growing demand for protein-based medicines, Perreault said it may be possible to transfer CSL Behring’s plasma collection technology to Russia, and initiate toll manufacturing in that country.

Over the last five years, CSL has invested more than $2 billion in R&D, employing more than 1,100 R&D experts.

"We are fortunate to have a robust R&D pipeline, with many projects within each stage of development," Perreault said at the opening. “Our world-class commercial operation, combined with our large and focused R&D team and operational excellence, enable us to quickly identify, develop and deliver innovations that patients and healthcare providers want. We are excited to expand the availability of our lifesaving therapies in Russia."

According to Perreault, CSL has made significant advancements in its recombinant factor development program for the treatment of hemophilia. “Once approved, our recombinant factor IX fusion protein for hemophilia B and our recombinant single-chain factor VIII for hemophilia A will provide patients with novel treatment options that have the potential to give patients extended dosing intervals,” Perreault said.
About CSL

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###
CSL CEO Says Corporate Responsibility and Sustainability are all about Continuous Improvement

*The global biotherapeutics company issued its 2014-2015 Corporate Responsibility Report, which details company performance in key social, environmental and economic priority areas.*

KING OF PRUSSIA, PA — 18 December 2015

CSL Limited (ASX:CSL) has recorded another strong performance in corporate responsibility, delivering on its promise to responsibly achieve growth through the disciplined execution of its business strategy, investing in research and development to advance therapies for unmet medical needs, growing its core portfolio of products by expanding indications and markets, and through its commitment to productivity and efficiency.

CSL Limited CEO and Managing Director Paul Perreault emphasizes that corporate responsibility and sustainability are all about continuous improvement. “At CSL, we are motivated by the challenges and opportunities this brings. Tackling these with our stakeholders wherever we can makes the pursuit of sustainability even more rewarding.”

CSL’s economic, social and environmental achievements and challenges include:

- Economic contribution direct to local economies of US$5 billion, including global community investment of US$28.4 million to patient, biomedical and local communities;
- Research and Development investment of US$463 million; with 35 clinical studies in operation across its pipeline. Achieved 14 product registrations or new indications for serious diseases in various markets;
- 237 Good Manufacturing Practice regulatory audits of CSL’s manufacturing facilities and plasma collection centres with no impact on the company’s product marketing licences. Across an extensive portfolio of products, CSL responded to three safety related product recalls in the reporting period;
- A global workforce of 14,874 employees as of June 30 2015, representing a 10% increase over the previous year – this increase follows two consecutive years of 7% increases per annum;
- Health and safety performance saw lost time due to injury (LTIFR), days lost (DLFR) and serious injury/illness (SIIFR) reduce by 47%, 37% and 54% respectively, while the medical treatment incident rate (MTIFR) increased by 8%;
- Across global operations, CSL maintained compliance with all environmental laws and regulations, and with an expanding facility footprint, CSL experienced increases in total numbers across environmental indicators.
- Successful acquisition of the Novartis influenza vaccine business to form, Seqirus – the world’s second largest influenza vaccine business.

In addition, this year CSL finalised an enterprise-wide climate change risk assessment concluding it is not exposed to climate change risks that have a potential to generate a substantive change in its business operations.


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operates in over 30 countries with more than 14,000 employees. Our unique combination of commercial strength, R&D focus and operational excellence enables us to identify, develop and deliver innovations so our patients can live life to the fullest.

###
CSL Behring Submits New Drug Application to Japan’s Pharmaceuticals and Medical Devices Agency for rIX-FP for Hemophilia B Patients

rIX-FP, CSL Behring’s long-acting recombinant albumin fusion protein, demonstrates the company’s promise to develop innovative specialty biotherapies that help people with serious medical conditions live full lives

TOKYO — 21 December 2015

Global biotherapeutics leader CSL Behring announced today that the company has submitted its new drug application to Japan’s Pharmaceuticals and Medical Devices Agency (PMDA) for its investigational fusion protein linking recombinant coagulation factor IX with recombinant albumin (rIX-FP). rIX-FP is a long-acting recombinant albumin fusion protein for people with hemophilia B, a congenital bleeding disorder characterized by deficient or defective factor IX. Based on the 2011 National Survey of Coagulation Disorder, hemophilia B occurs in approximately 1 to 2 of every 100,000 male births in Japan.

“CSL was formed nearly 100 years ago with a promise to develop and deliver innovative specialty biotherapies that help people with serious medical conditions live full lives,” said Dr. Andrew Cuthbertson, Chief Scientific Officer and R&D Director, CSL Limited. “rIX-FP demonstrates our longstanding commitment to innovation and patient care and we are pleased to submit our new drug application to PMDA. We look forward to, upon approval, bringing this innovative specialty biotherapy to patients with hemophilia B in Japan.”

About PROLONG 9-FP
The submission is based on the PROLONG-9FP clinical development program. PROLONG-9FP includes Phase I through Phase III open-label, multicenter studies evaluating the safety and efficacy of rIX-FP in adults and children (ages 1 to 61 years) with hemophilia B (FIX ≤ 2%) who were previously treated with other factor IX products.

The data from PROLONG-9FP showed median annualized spontaneous bleeding rates (AsBR) of 0.00 in patients using rIX-FP prophylactically. This result was achieved for both 14-day dosing and 7-day dosing. The data for on-demand therapy showed that 93.6 percent of bleeds were controlled with one infusion, while 98.6 percent were controlled with one or two infusions. Across the completed Phase III studies, none of the patients developed inhibitors to factor IX or antibodies to rIX-FP. The most common adverse reaction in clinical trials was headache.

About rIX-FP
CSL Behring engineered rIX-FP to extend the half-life of recombinant factor IX through genetic fusion with recombinant albumin. CSL Behring selected recombinant albumin as its recombinant genetic fusion partner for its coagulation factor proteins due to its long physiological half-life.

The U.S. Food and Drug Administration, European Medicines Agency and Swissmedic are currently reviewing CSL Behring’s license applications for rIX-FP. For more information about CSL Behring’s recombinant coagulation factor products in development to treat hemophilia, visit http://www.cslbehring.com/products/bleeding-disorders/novel-recombinant-hemophilia-treatments.

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For more information about CSL Behring visit www.CSLBehring.com or follow us at www.Twitter.com/CSLBehring.
CSL Behring Submits Marketing Authorization Application to the European Medicines Agency for rVIII-SingleChain for Patients with Hemophilia A

**rVIII-SingleChain demonstrates CSL Behring’s promise to develop and deliver innovative biotherapies that improve the well-being of patients with serious medical conditions**

MARBURG, Germany — 22 December 2015

Global biotherapeutics leader CSL Behring announced today that the company has submitted its Marketing Authorization Application (MAA) to the European Medicines Agency (EMA) for its novel investigational recombinant factor VIII single-chain (rVIII-SingleChain) for the treatment of hemophilia A. In the pivotal clinical trial, rVIII-SingleChain met all primary endpoints.

Hemophilia A is a congenital bleeding disorder characterized by deficient or defective factor VIII; nearly all affected patients are male. People with hemophilia A may experience prolonged or spontaneous bleeding, especially into the muscles, joints, or internal organs. According to the World Federation of Hemophilia, about 1 in 10,000 people are born with hemophilia, the majority of whom have hemophilia A.

“Our focused research and development and strong relationship with the bleeding disorders community led us to seek further advancements in the care and treatment of hemophilia,” said Dr. Andrew Cuthbertson, Chief Scientific Officer and Director of R&D, CSL Limited. “Today, we have the only recombinant single-chain factor VIII product in late-stage development for the management of hemophilia A. We look forward to, upon approval, providing this innovative specialty biotherapy to patients in the EU and European Economic Area.”

The MAA is based on the AFFINITY clinical development program, which includes a phase I/III open-label, multi-center trial examining safety and efficacy. The pharmacokinetics of rVIII-SingleChain compared with recombinant human antihemophilic factor VIII (octocog alfa) was also studied. Study design details for rVIII-SingleChain are available at clinicaltrials.gov (NCT01486927).

Results from the phase I/III study presented earlier this year at the International Society on Thrombosis and Haemostasis (ISTH) congress in Toronto showed that patients treating prophylactically had a median annualized bleeding rate (ABR) of 1.14 and a median annualized spontaneous bleeding rate (AsBR) of 0.00. The data also showed that, of 848 bleeds treated in the study, 94 percent were controlled with no more than two infusions of rVIII-SingleChain, with 81 percent controlled by one infusion. Moreover, hemostatic control of a bleeding event treated with rVIII-SingleChain was assessed by the investigator as excellent or good 94 percent of the time (835 assessed bleeding events).

The results presented at ISTH included data on more than 14,000 exposure days in 146 patients on prophylaxis and 27 patients treating on demand for a bleeding event. In total, 120 patients were treated for more than 50 days of exposure; 52 had more than 100 days of exposure. In the prophylaxis group, 32 percent of patients were dosed twice weekly and 54 percent received treatment three times per week; the regimen was determined by the investigator. The most common adverse events were naso-pharyngitis, arthralgia, and headache. No inhibitors were reported.

**About rVIII-SingleChain**

Specifically designed for greater molecular stability, rVIII-SingleChain is the first and only recombinant single-chain factor VIII (FVIII) product in late-stage development for the treatment of hemophilia A. rVIII-SingleChain has a strong affinity for von Willebrand factor, leading to greater stability and integrity of factor VIII in circulation. The stability of rVIII-SingleChain leads to a longer-lasting therapeutic effect with reduced dosing frequency. In July 2015, the U.S. Food and Drug Administration accepted for review CSL Behring’s Biologics License Application for rVIII-SingleChain.
For more information about CSL Behring’s recombinant products in development to treat hemophilia, visit http://www.cslbehring.com/products/bleeding-disorders/novel-recombinant-hemophilia-treatments.

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