

Largest Ever CIDP Clinical Study Completed

PATH study evaluated subcutaneous immunoglobulin efficacy and safety for treating Chronic Inflammatory Demyelinating Polyneuropathy

KING OF PRUSSIA, Pa. – 01 March 2017 – Global biotherapeutics leader [CSL Behring](#) announced today that it has completed the largest ever Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) trial, known as PATH (Polyneuropathy And Treatment with Hizentra®). The Phase III clinical trial was designed to demonstrate the efficacy, safety and tolerability of two different doses of CSL Behring’s subcutaneous immunoglobulin (SCIG), Hizentra® (Immune Globulin Subcutaneous [Human]), compared with placebo, in the maintenance treatment of CIDP patients previously treated with intravenous immunoglobulin (IVIG).

Hizentra, the no. 1 prescribed immunoglobulin therapy in treating primary immunodeficiencies (PI), the most prescribed SCIG worldwide, and the only 20% SCIG designed with the natural stabilizer L-proline, was self-administered by patients. Throughout the study, subjects were allowed to use dose volumes up to 50 mL/site and infusion rates of up to 35 mL/hour, to provide them with greater flexibility and autonomy to infuse when and where they choose. A long-term open label extension study is ongoing and is expected to be completed in 2017.

“Patients living with CIDP experience different symptoms with varying severities. We value organizations like CSL Behring whose ongoing neurology research continues to look at bringing new options for patients, caregivers and physicians to choose the individualized treatment that’s right for each patient,” said Lisa Butler, Executive Director, GBS/CIDP Foundation International.

“The PATH trial, our second major neurology study, drives us towards unlocking the promise of immunoglobulins for treatment of neurological conditions and demonstrates our commitment in this important medical specialty,” says Andrew Cuthbertson, Chief

Scientific Officer and R&D Director. “We look forward to sharing the results of the PATH trial with colleagues at key neurology congresses this year.”

Investigators and patients in over 10 countries participated during the last five years to bring this groundbreaking prospective, multicenter, randomized, double-blind, placebo-controlled clinical trial to completion. A trial of this scale will contribute greatly to our understanding of these patients.

CIDP is a rare disorder of the peripheral nerves (those outside the brain and spinal cord). The condition is immune-mediated and the effects can worsen over time. The protective covering of the nerves is damaged, which may cause numbness or tingling, muscle weakness, fatigue and other symptoms. CIDP can occur at any age and is more common in men than in women. If left untreated, approximately 30 percent of CIDP patients will progress to wheelchair dependence.

For more information about the PATH study, visit <http://www.clinicaltrials.gov> and search with identifier: NCT01545076.

About Hizentra®

Hizentra (Immune Globulin Subcutaneous [Human]), the first 20 percent SCIG developed for subcutaneous use, is registered in over 46 countries and approved in North America, Europe and Japan. **Hizentra**, the world’s most prescribed SCIG, has a proven track record of safety, efficacy and tolerability and has over 3.6 million exposures worldwide since 2010. In the United States, **Hizentra** is indicated for the treatment of patients with primary immunodeficiency and contraindicated in individuals with a history of anaphylactic or severe systemic response to immune globulin preparations or components of **Hizentra**, as well as in persons with selective immunoglobulin A deficiency who have known antibody against IgA and a history of hypersensitivity. In all 29 European/European Economic Area member states and Japan, **Hizentra** is authorized for treating patients diagnosed with PI as well as secondary immunodeficiencies.

For country specific indication information, visit:

- Switzerland: <https://compendium.ch/mpro/mnr/22484/html/de> (German) or <https://compendium.ch/mpro/mnr/22484/html/fr> (French)
- Europe: http://www.ema.europa.eu/ema/index.jsp?curl=pages/medicines/human/medicines/002127/human_med_001440.jsp&mid=WC0b01ac058001d124
- United States: <http://www.hizentra.com/Professional/Prescribing-Information.aspx>
- Canada: <http://labeling.cslbehiring.ca/PM/CA/Hizentra/EN/Hizentra-Product-Monograph.pdf>
- Australia: <http://www.csl.com.au/productfinder/hizentraau>

Important Safety Information

Immune Globulin Subcutaneous (Human), Hizentra®, is indicated as replacement therapy for patients with primary humoral immunodeficiency (PI), age 2 and older. This includes but is not limited to the humoral immune defect in congenital agammaglobulinemia, common variable immunodeficiency, X-linked agammaglobulinemia, Wiskott-Aldrich syndrome, and severe combined immunodeficiencies.

WARNING: THROMBOSIS - Thrombosis may occur with immune globulin products, including Hizentra. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling vascular catheters, hyperviscosity, and cardiovascular risk factors.

For patients at risk of thrombosis, administer Hizentra at the minimum dose and infusion rate practicable. Ensure adequate hydration in patients before administration. Monitor for signs and symptoms of thrombosis and assess blood viscosity in patients at risk for hyperviscosity. See full prescribing information for complete boxed warning.

Hizentra is contraindicated in patients with a history of anaphylactic or severe systemic reaction to human immune globulin preparations or components of Hizentra, such as polysorbate 80. Because it contains the stabilizer L-proline, Hizentra is contraindicated in patients with hyperprolinemia. Hizentra is also contraindicated in patients with immunoglobulin A deficiency who have antibodies against IgA and a history of hypersensitivity.

Hizentra should be administered subcutaneously *only*. Do not administer intravenously.

IgA-deficient patients with anti-IgA antibodies may be at greater risk of developing potentially severe hypersensitivity and anaphylactic reactions with administration of Hizentra. If hypersensitivity occurs or anaphylactic reactions are suspected, discontinue administration immediately and treat as medically appropriate.

Monitor patients for aseptic meningitis syndrome (AMS), which has been reported with SCIg. In patients at risk of acute renal failure, monitor renal function, including blood urea nitrogen, serum creatinine and urine output. Also monitor patients for clinical signs of hemolysis or transfusion-related acute lung injury (TRALI).

Hizentra 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 most common adverse reactions (observed in 5% or more of study subjects receiving Hizentra) were local reactions (ie, swelling, redness, heat, pain, and itching at the injection site), headache, diarrhea, fatigue, back pain, nausea, extremity pain, cough, rash, pruritus, vomiting, upper abdominal pain, migraine and pain.

Ig administration can transiently impair the efficacy of live attenuated virus vaccines, such as measles, mumps and rubella. It can also lead to misinterpretation of serologic testing.

Please see full [prescribing information](#) for Hizentra including boxed warning.

About CSL Behring

CSL Behring is a global biotherapeutics leader which is driven by its promise to save lives.

Focused on serving patients' needs by using the latest technologies, we develop and deliver innovative therapies that are used to treat coagulation disorders, primary immune deficiencies, hereditary angioedema, inherited respiratory disease, and neurological disorders. 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. The parent company, [CSL Limited](#) (ASX:CSL), headquartered in Melbourne, Australia, employs more than 17,000 people, delivering its life-saving, life-changing therapies to people in more

than 60 countries. For more information visit www.cslbehring.com and follow us on www.Twitter.com/CSLBehring.

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