

Post-hoc results from NEFIGAN study to be presented at the International IgA Nephropathy Network meeting (IIgANN)

Calliditas Therapeutics AB (publ) ("Calliditas") today announced that post-hoc results from its clinical Phase 2b study NEFIGAN will be presented in an oral session at the International IgA Nephropathy Network meeting (IIgANN) in Buenos Aires, Argentina, on Friday September 28, 2018.

The oral presentation of the abstract – with the title *Treatment of IgA nephropathy with Nefecon, a targeted-release formulation of budesonide – extended posthoc results from the Nefigan trial* – will be presented by Prof. Bengt Fellström, MD, PhD, from the Department of Medical Sciences, Uppsala University, Uppsala, Sweden.

The new results stem from extended post-hoc analyses of the randomized, double-blind controlled Phase 2b study NEFIGAN which Calliditas conducted in 149 IgA nephropathy (IgAN) patients at risk of developing ESRD. In 2017, the primary results from nine months' treatment with either Nefecon or placebo were published in *The Lancet*.

The study results demonstrated the effectiveness of Nefecon in reducing proteinuria and stabilizing the kidney function as well as confirming that the treatment was safe and tolerable for the patients. The NEFIGAN study strongly supports that local treatment of the mucosal immune system of the GI tract with budesonide is effective in reducing renal leakage and preventing loss of kidney function. Based on this successful Phase 2B study, Calliditas will initiate a clinical Phase 3 study in H2 2018.

In total, five abstracts based on the NEFIGAN study were accepted by the 15th International IgA Nephropathy Network meeting (IIgANN):

Title: *Extent of segmental glomerulosclerosis in IgA nephropathy is associated with the level of eGFR response to TRF-budesonide (Nefecon)*

Authors: Maria Soares, Andrew Stone, Jonathan Barratt and Ian Roberts

Title: *Treatment of IgA nephropathy with Nefecon, a targeted-release formulation of budesonide – extended posthoc results from the Nefigan trial*

Authors: Bengt Fellström, Jonathan Barratt, Jürgen Floege et al, on behalf of NEFIGAN investigators

Title: *Targeted Release-Budesonide modifies mucosal IgA responses and possibly gut permeability in IgA nephropathy*

Authors: Masahiro Muto, Jasraj Bhachu, Jeremy Brown, Karen Molyneux, Rosanna Coppo and Jonathan Barratt

Title: *Targeted Release-Budesonide modifies circulating IgA-IgG immune complex levels and levels of poorly O-galactosylated IgA in IgAN*

Authors: Jasraj Bhachu, Katrin Scionti, Masahiro Muto, Karen Molyneux and Jonathan Barratt

Title: *Nefecon, an oral disease modifying treatment for progressive IgA nephropathy. The strategy behind developing proteinuria as surrogate endpoint for accelerated approval*

Authors: Johan Häggblad, Ann-Kristin Myde and Jens Kristensen

All abstracts and summaries of the sessions will be published in a special number of the scientific journal *Kidney Diseases*.

The information was submitted for publication, through the agency of the contact person set out below, at 8:00 am CEST on September 25, 2018.

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About Calliditas

Calliditas Therapeutics is a specialty pharmaceutical company based in Stockholm, Sweden, focused on developing high quality pharmaceutical products for patients with a significant unmet medical need in niche indications, in which the Company can partially or completely participate in the commercialization efforts. The Company is focused on the development and commercialization of the product candidate Nefecon, a unique formulation optimized to combine a time lag effect with a concentrated release of the active substance budesonide, within a designated target area. This patented, locally acting formulation is intended for treatment of patients with the inflammatory renal disease IgA nephropathy. Calliditas Therapeutics aims to take Nefecon through a global Phase 3 study to commercialization. The company is listed on Nasdaq Stockholm (ticker: CALTX). Visit www.calliditas.com for further information.

About Nefecon

Nefecon is a potential treatment for patients with IgAN at risk of developing ESRD. It is a proprietary oral formulation of budesonide, designed to deliver budesonide to the ileum where the so-called Peyer's patches, which harbor the majority of B-cells producing IgA antibodies, are found. By delivering budesonide locally instead of systemically, Nefecon greatly reduces the side-effect burden observed with high dose steroid treatment while optimizing the effective dose level of the drug where it is required. Budesonide has been used to treat patients with asthma, inflammatory bowel disease and allergic rhinitis for over 35 years. It is rapidly degraded soon after entering the circulatory system, making it an ideal basis for drugs such as Nefecon because local delivery to disease tissue minimizes the systemic effects seen with other corticosteroids. Nefecon has been granted orphan drug designation for IgAN by the US Food and Drug Administration (FDA) and the European Medicines Agency (EMA).

About IgA Nephropathy (IgAN)

IgA nephropathy (IgAN) – also known as Berger's disease – is the most common form of glomerulonephritis, a chronic inflammatory condition of the kidney, in the Western world. IgAN is a serious autoimmune, progressive disease that leads to decreasing kidney function over the course of 10 to 20 years. Up to 50 percent of patients diagnosed with IgAN will progress to end-stage renal disease (ESRD), a disease state requiring dialysis or kidney transplant for survival due to insufficient kidney function within 20 years. IgAN is an orphan disease, designated as an orphan indication in both the US and Europe. IgAN affects approximately 130,000–150,000 people in the US and about 250,000 people in Europe. Today, there are no approved treatments for IgAN. Today's standard of care treatment regimens entails primarily established, generic drugs such as blood pressure lowering agents to alleviate symptoms, complemented by off-label use of systemic corticosteroids.

About IIgANN

The International IgA Nephropathy Network (IIgANN) was established in 2000 based on The International IgA Nephropathy Club that started in 1987. The purpose with IIgANN is to increase the awareness of the disease since it was felt that the clinical impact of IgAN was underappreciated by nephrologists and general physicians in many countries. This year's IIgANN meeting, which is held September 27-29, will also mark the 50th anniversary of the initial description of IgAN by Dr. J Berger and Dr. N. Hinglais in 1968. Approximately 175 participants are expected at this global meeting focused exclusively on IgAN.