Stockholm, Sweden **October 1, 2018**

Presentation at International IgA Nephropathy Network meeting highlighting supportive post-hoc analysis of the NEFIGAN study

Calliditas Therapeutics AB (publ) ("Calliditas") today announced supportive post-hoc results from its clinical Phase 2b study NEFIGAN of the company's lead candidate Nefecon, presented by Prof. Bengt Fellström at the International IgA Nephropathy Network meeting (IIgANN) in Buenos Aires, Argentina, on 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* – highlighted among other things, that Nefecon has a potential to become an effective IgAN specific treatment, targeting intestinal mucosal immunity, upstream of disease manifestation.

The new post-hoc results, presented by Prof. Bengt Fellström, MD, PhD, from the Department of Medical Sciences, Uppsala University, Uppsala, Sweden, 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 end-stage renal disease (ESRD). In 2017, the primary results from nine months' treatment with either Nefecon or placebo were published in The Lancet.

The new study results further support the effectiveness of Nefecon in reducing proteinuria and stabilizing the kidney function. The NEFIGAN study supports the prevailing understanding of the disease mechanism, by demonstrating that local treatment of the mucosal immune system in a specific part of the GI tract is effective in reducing renal leakage and preventing loss of kidney function.

Key results from the post-hoc study, as per the presentation by Prof. Bengt Fellström:

- Nefecon added to optimized RAS blockade led to a significantly and consistently reduced UPCR[1], 24 hr urinary protein, UACR[2] and 24 hr urinary albumin.
- UACR was reduced by 43% in the 16 mg treatment group at 12 months compared to baseline, with UPCR showing a reduction of 35%.
- The effects were independent of baseline UPCR, baseline eGFR and time from diagnosis to randomization.
- eGFR was stabilized among NEFECON treated patients but declined in placebo patients.

"These additional results strengthen previously released data, supporting that Nefecon by local action in the gut has the potential to reduce the risk of progression to ESRD", says Prof. Bengt Fellström.

Based on this successful Phase 2B study, Calliditas will initiate a clinical Phase 3 study in H2 2018. All abstracts and summaries of the sessions are 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 08:00 CET on October 1, 2018.

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1. Urinary protein creatinine ratio
2. Urinary albumin creatinine ratio

About Calliditas Therapeutics
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. Over 200 participants participated in this global meeting focused exclusively on IgAN.