Stockholm, Sweden **September 29, 2023**

Calliditas Presents Data from the NefIgArd Phase 3 trial at the 17th International Symposium on IgA Nephropathy (IIgANN) Tokyo 2023

Calliditas Therapeutics AB (Nasdaq: CALT, Nasdaq Stockholm: CALTX) ("Calliditas"), today announced the presentations of new biomarker and subgroup analyses from the Phase 3 NefIgArd study with Nefecon (TARPEYO® (budesonide) delayed release capsules/Kinpeygo®) in adults with Primary IgA nephropathy (IgAN). The data was presented in both posters and oral presentations at the 17th International Symposium on IgA Nephropathy (IIGaNN), held in Tokyo, Japan, on September 25-27, 2023.

"We are proud to have showcased new data at this year's International Symposium on IgAN in Tokyo," said Richard Phillipson, Chief Medical Officer at Calliditas. "Taken together, the positive findings from our biomarker and patient subgroup analyses from our Phase 3 NefIgArd study indicate that Nefecon treatment results in a coordinated immunological response with the potential to modulate the intestinal immune network responsible for IgA production. These additional findings further reinforce the potential of TARPEYO in enabling physicians to proactively manage and potentially mitigate the impact of the disease by targeting the source and slowing kidney function decline."

Biomarker Analyses

IgA-containing immune complexes (IgA-IC) have previously been shown to accumulate in renal tissue, where they trigger inflammation and scarring in the glomeruli. Investigation of the effect of Nefecon on circulating levels of IgA-IC in the Part A population of the NefIgArd clinical trial (n=160) revealed that patients treated with Nefecon 16mg/day exhibited a statistically significant decrease in IgA-IC levels compared to patients who received placebo when evaluated at the 3-, 6- and 9-months post randomization. Suppression by treatment with Nefecon of both IgA-IC formation and galactose-deficient IgA1 as previously reported offers an unprecedented opportunity to target the fundamental immune abnormalities that drive IgA deposition in the kidney and development of IgAN.

An additional analysis of the 160 patients enrolled in Part A of the Phase 3 NefIgArd clinical study was conducted to assess the levels of three soluble factors known for modulating B cell maturation in gut-associated lymphoid tissue (GALT) BAFF, APRIL and sBCMA. Levels of BAFF were decreased at the 3-, 6-, and 9-months post treatment mark compared to placebo. Levels of APRIL were significantly reduced at the 3- and 6-months timepoints and levels of sBCMA were decreased at the 6-months timepoint. These data further support the potential disease-modifying effect of Nefecon in IgAN and reinforce the central role of the gut–kidney axis in the pathogenesis of IgAN.

To further understand the impact of Nefecon on chemokine signaling in IgAN patients, the levels of three circulating chemokines known to be present in the intestinal tract at the site of IgA production, CXCL5, CCL13 and CCL11, were assessed in the Part A patient population portion of the NefIGArd trial (n=160). Dr. Barratt and colleagues reported that treatment with Nefecon 16 mg/day resulted in a statistically significant decrease in the levels of CXCL5 and CCL13 and an increase in levels of CCL11 at 3-, 6- and 9-months post-treatment when compared to placebo. These biomarker data provide additional evidence for a mucosal mechanism of action of Nefecon in IgAN and support previous observations that disordered lymphocyte trafficking and mucosal dysregulation likely contribute to the pathogenesis of IgAN.

Patient Subgroup Analyses

The full two-year results of the Phase 3 NefIgArd trial (n=364 patients) were further analyzed to assess potential differences in response to Nefecon treatment based on self-reported Asian (n=83) or White (n=275) ancestry in patients with IgAN. Treatment with Nefecon 16 mg/day over a 9-month period resulted in clinically meaningful preservation of kidney function in both subgroups, as evidenced by ameliorated eGFR and proteinuria in these two subgroups when compared to placebo subgroups.

NefIgArd Sub-Analyses

Full analysis of the Phase 3 NefIgArd study over the entire 2-year study period of the 364 patients (1:1 patient randomization to Nefecon 16 mg/day or placebo) enrolled in the study revealed clinically meaningful impact on three key markers of IgAN disease progression as evidenced by:

- A statistically significant benefit of Nefecon over placebo in estimated glomerular filtration rate (eGFR) over the two-year study period, which consisted of nine months of treatment with Nefecon or placebo, followed by a 15-month follow-up period off the study drug.
- A clinically significant reduction in proteinuria was found to be durable for the entire 15-month follow-up period off the study drug.
- Relevant decrease in microhematuria was also observed during Part B of the study, with fewer patients in the Nefecon treatment group exhibiting microhematuria at follow up visits compared to the placebo group.

Indication

TARPEYO® (budesonide) delayed release capsules is a corticosteroid indicated to reduce proteinuria in adults with primary immunoglobulin A nephropathy (IgAN) at risk of rapid disease progression, generally a urine protein-to-creatinine ratio (UPCR) ≥1.5 g/g.

This indication is approved under accelerated approval based on a reduction in proteinuria. It has not been established whether TARPEYO slows kidney function decline in patients with IgAN. Continued approval for this indication may be contingent upon verification and description of clinical benefits in a confirmatory clinical trial.

Important Safety Information

Contraindications: TARPEYO is contraindicated in patients with hypersensitivity to budesonide or any of the ingredients of TARPEYO. Serious hypersensitivity reactions, including anaphylaxis, have occurred with other budesonide formulations.

Warnings and Precautions

Hypercorticism and adrenal axis suppression: When corticosteroids are used chronically, systemic effects such as hypercorticism and adrenal suppression may occur. Corticosteroids can reduce the response of the hypothalamus-pituitary-adrenal (HPA) axis to stress. In situations where patients are subject to surgery or other stress situations, supplementation with a systemic corticosteroid is recommended. When discontinuing therapy *[see Dosing and Administration]* or switching between corticosteroids, monitor for signs of adrenal axis suppression.

Patients with moderate to severe hepatic impairment (Child-Pugh Class B and C, respectively) could be at an increased risk of hypercorticism and adrenal axis suppression due to an increased systemic exposure to oral budesonide. Avoid use in patients with severe hepatic impairment (Child-Pugh Class C). Monitor for increased signs and/or symptoms of hypercorticism in patients with moderate hepatic impairment (Child-Pugh Class B).

Risks of immunosuppression: Patients who are on drugs that suppress the immune system are more susceptible to infection than healthy individuals. Chicken pox and measles, for example, can have a more serious or even fatal course in susceptible patients or patients on immunosuppressive doses of corticosteroids.

Avoid corticosteroid therapy in patients with active or quiescent tuberculosis infection; untreated fungal, bacterial, systemic viral, or parasitic infections; or ocular herpes simplex. Avoid exposure to active, easily transmitted infections (eg., chicken pox, measles). Corticosteroid therapy may decrease the immune response to some vaccines.

Other corticosteroid effects: TARPEYO is a systemically available corticosteroid and is expected to cause related adverse reactions. Monitor patients with hypertension, prediabetes, diabetes mellitus, osteoporosis, peptic ulcer, glaucoma, cataracts, a family history of diabetes or glaucoma, or with any other condition in which corticosteroids may have unwanted effects.

Adverse reactions: In clinical studies, the most common adverse reactions with TARPEYO (occurring in ≥5% of TARPEYO patients and ≥2% higher than placebo) were hypertension (16%), peripheral edema (14%), muscle spasms (13%), acne (11%), dermatitis (7%), weight increase (7%), dyspnea (6%), face edema (6%), dyspepsia (5%), fatigue (5%), and hirsutism (5%).

Drug interactions: Budesonide is a substrate for CYP3A4. Avoid use with potent CYP3A4 inhibitors, such as ketoconazole, itraconazole, ritonavir, indinavir, saquinavir, erythromycin, and cyclosporine. Avoid ingestion of grapefruit juice with TARPEYO. Intake of grapefruit juice, which inhibits CYP3A4 activity, can increase the systemic exposure to budesonide.

Use in specific populations

Pregnancy: The available data from published case series, epidemiological studies, and reviews with oral budesonide use in pregnant women have not identified a drug-associated risk of major birth defects, miscarriage, or other adverse maternal or fetal outcomes. There are risks to the mother and fetus associated with IgAN. Infants exposed to in utero corticosteroids, including budesonide, are at risk for hypoadrenalism.

Please see **Full Prescribing Information**.

About TARPEYO

Calliditas has introduced TARPEYO, the first FDA-approved therapy for the treatment of the autoimmune renal disease primary IgA Nephropathy, or IgAN, to reduce proteinuria in adults with primary IgAN who are at risk of rapid disease progression, generally a UPCR≥1.5g/g. This indication is approved under accelerated approval based on a reduction in proteinuria. It has not been established whether TARPEYO slows kidney function decline in patients with IgAN. Continued approval for this indication may be contingent upon verification and description of clinical benefit in a confirmatory clinical trial.

TARPEYO is an oral, delayed release formulation of budesonide, a corticosteroid with potent glucocorticoid activity and weak mineralocorticoid activity that undergoes substantial first pass metabolism. TARPEYO is as a 4 mg delayed release capsule and is enteric coated and designed to remain intact until it reaches the ileum. Each capsule contains coated beads of budesonide that target mucosal B-cells present in the ileum, including the Peyer's patches, which are responsible for the production of galactose-deficient IgA1 antibodies (Gd-Ag1) causing IgA nephropathy. It is unclear to what extent TARPEYO's efficacy is mediated via local effects in the ileum vs systemic effects.

About the NefIgArd Study

The global clinical trial NefIgArd is a Phase 3, randomized, double-blind, placebo-controlled, multicenter study to evaluate the efficacy and safety of TARPEYO 16 mg once daily vs placebo in adult patients with primary IgAN (N=364), as an addition to optimized RAS inhibitor therapy. Part A of the study included a 9-month blinded treatment period and a 3-month follow-up period. The primary endpoint was UPCR, and eGFR was a secondary endpoint. Part B included a 12-month observational period off drug and assessed eGFR over the entire 2-year period for patients who were treated with the TARPEYO or placebo regimen in Part A. The full NefIgArd trial met its primary endpoint. Topline data from the full NefIgArd study were reported on March 12, 20

About Primary Immunoglobulin A Nephropathy

Primary immunoglobulin A nephropathy (IgA nephropathy or IgAN or Berger's Disease) is a rare, progressive, chronic autoimmune disease that attacks the kidneys and occurs when galactose-deficient IgA1 is recognized by autoantibodies, creating IgA1 immune complexes that become deposited in the glomerular mesangium of the kidney. This deposition in the kidney can lead to progressive kidney damage and potentially a clinical course resulting in end- stage renal disease. IgAN most often develops between late teens and late 30s.

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The information was sent for publication, through the agency of the contact person set out above, on September 29, 2023, at 14.00 p.m. CET.

About Calliditas

Calliditas Therapeutics is a commercial stage biopharma company based in Stockholm, Sweden focused on identifying, developing and commercializing novel treatments in orphan indications, with an initial focus on renal and hepatic diseases with significant unmet medical needs. Calliditas' lead product, developed under the name Nefecon, has been granted accelerated approval by the FDA under the trade name TARPEYO® and conditional marketing authorization by the European Commission under the trade name Kinpeygo®. Kinpeygo is being commercialized in the European Union Member States by Calliditas' partner, STADA Arzneimittel AG. Additionally, Calliditas is conducting a Phase 2b clinical trial in primary biliary cholangitis and a Phase 2 proof-of-concept trial in head and neck cancer with its NOX inhibitor product candidate, setanaxib. Calliditas' common shares are listed on Nasdaq Stockholm (ticker: CALTX) and its American Depositary Shares are listed on the Nasdaq Global Select Market (ticker: CALT).

Forward-Looking Statements

This press release contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995, as amended, including, without limitation, statements regarding Calliditas' strategy, commercialization efforts, business plans, regulatory submissions, clinical development plans, revenue and product sales projections or forecasts and focus. The words "may," "will," "could," "would," "should," "expect," "plan," "anticipate," "intend," "believe," "estimate," "predict," "project," "potential," "continue," "target," and similar expressions are intended to identify forward-looking statements, although not all forward-looking statements contain these identifying words. Any forward-looking statements in this press release are based on management's current expectations and beliefs and are subject to a number of risks, uncertainties, and important factors that may cause actual events or results to differ materially from those expressed or implied by any forward-looking statements contained in this press release, including, without limitation, any related to Calliditas' business, operations, continued and additional regulatory approvals for TARPEYO and Kinpeygo, market acceptance of TARPEYO and Kinpeygo, clinical trials, supply chain, strategy, goals and anticipated timelines, competition from other biopharmaceutical companies, revenue and product sales projections or forecasts and other risks identified in the section entitled "Risk Factors" in Calliditas' reports filed with the Securities and Exchange Commission. Calliditas cautions you not to place undue reliance on any forward-looking statements, which speak only as of the date they are made. Calliditas disclaims any obligation to publicly update or revise any such statements to reflect any change in expectations or in events, conditions, or circumstances on which any such statements may be based, or that may affect the likelihood that actual results will differ from those set forth in the forward-looking statements. Any forward-looking statements contained in this press release represent Calliditas' views only as of the date hereof and should not be relied upon as representing its views as of any subsequent date.