June 6, 2017

# Oral Presentations at ERA-EDTA Congress Highlight Fourth Potential Indication for Avacopan and Potential for CCR2 Inhibition in Focal Segmental Glomerulosclerosis (FSGS)

*-- Clinical Study Shows Drop in Proteinuria in Patients Treated with Avacopan for IgA Nephropathy (IgAN) --*

*-- Potential of CCR2 Inhibitor as a Therapeutic Treatment Option for FSGS Validated --*

MOUNTAIN VIEW, Calif., June 06, 2017 (GLOBE NEWSWIRE) -- ChemoCentryx, Inc., (Nasdaq:CCXI), a biopharmaceutical company developing new medications targeted at inflammatory and autoimmune diseases and cancer, announced today the findings of two studies presented during the 54th European Renal Association - European Dialysis and Transplant Association (ERA-EDTA) Congress held June 3-6 in Madrid, Spain.

"The findings presented at ERA-EDTA illustrate the value of our chemoattractant inhibitor platform in the treatment of kidney disease," said Thomas J. Schall, Ph.D., President and Chief Executive Officer of ChemoCentryx. "The drop in proteinuria seen in the IgAN study presents a fourth potential application for our lead drug candidate avacopan and the animal model study supports the significant role of CCR2 inhibition in the treatment of FSGS."

Decrease in Proteinuria in Patients Treated with Avacopan for IgA Nephropathy (IgAN)

On June 6, 2017, researchers shared findings of a study investigating the tolerability and efficacy of avacopan in the treatment of IgA Nephropathy, also known as Berger's Disease, where immunoglobulin A (IgA) lodges in the kidney, causing proteinuria, or excess protein in the urine.  IgAN can lead to chronic kidney disease, kidney failure and End Stage Renal Disease (ESRD).

This open-label pilot Phase II trial in Sweden and the United States investigated the tolerability and efficacy of ChemoCentryx's complement 5a receptor inhibitor avacopan (formerly CCX168) in IgAN patients. After an 8-week run-in period on a maximum tolerated dose of a renin-angiotensin-aldosterone system (RAAS) inhibitor, patients started avacopan dosing, 30 mg twice daily for 12 weeks, with a 12-week follow-up period. The primary efficacy endpoint was change in slope of protein:creatinine ratio (UPCR) in the urine from the run-in period to the avacopan treatment period. Seven patients received avacopan treatment, and all 7 completed the study. At the end of 12 weeks, reduced levels of protein in the urine were seen in 6 of 7 patients, with 3 of the 7 patients showing significant improvement to UPCR < 1 g/g. At the end of the 12-week period following treatment, the protein ratio in 2 of these 3 patients returned to baseline levels, while the improvement was maintained in the third patient. One serious adverse event of unstable angina was observed, considered unrelated to avacopan treatment by the investigator. Longer avacopan treatment duration may be indicated for maximal benefit. Avacopan appeared to be safe and well tolerated.

"To see such a benefit on proteinuria, a key marker for IgA Nephropathy, on top of a maximum tolerated dose of a RAAS inhibitor, was quite encouraging," said Annette Bruchfeld, M.D., Ph.D., from the Karolinska Institute in Stockholm. "With no approved therapy for the disease available, avacopan presents a promising potential treatment option for patients."

Potential of CCR2 Inhibition as a Therapeutic Treatment Option for Focal Segmental Glomerulosclerosis (FSGS)

In an oral presentation on June 6, 2017 titled "CCR2 Antagonism Reduces Proteinuria and Glomerular Injury in Murine Models of Focal Segmental Glomerulosclerosis (FSGS)," researchers discussed the results of a study examining the efficacy of CCR2 inhibition in two in vivo models. FSGS is another disease that causes proteinuria and leads to ESRD. There is currently no approved treatment option for FSGS.

Researchers concluded that blocking CCR2 provides marked and rapid renal protection in two distinct models of FSGS, as measured by reduction in proteinuria and improvement in multiple histological parameters, and thus represents a novel and mechanistically distinct approach for the treatment of FSGS. The study support ChemoCentryx's plans to conduct a clinical

endpoint study with CCR2 inhibitor CCX140 for FSGS, which has already been shown to have good safety and tolerability in hundreds of patients in clinical trials completed to date.

Abstracts of both studies can be found in the May 2017 issue of Nephrology Dialysis Transplantation (NDT), the official journal of the ERA-EDTA: https://academic.oup.com/ndt/issue

**About Avacopan (CCX168)**

Avacopan (CCX168) is an orally-administered small molecule that is a selective inhibitor of the complement C5a receptor, or C5aR. Avacopan is in Phase III development for the treatment of anti-neutrophil cytoplasmic auto-antibody-associated vasculitis (AAV). In clinical studies to date, avacopan was shown to be safe, well tolerated and provided effective control of the disease while successfully allowing elimination of high-dose steroids, part of the standard of care for AAV patients. Avacopan is also being developed in patients with atypical hemolytic uremic syndrome (aHUS) and C3 glomerulopathy (C3G). In C3G, Avacopan targets the C5a receptor, blocking the effects of C5a which contributes to the inflammatory hypercellularity in the glomeruli, a main feature of C3G

The U.S. Food and Drug Administration has now granted avacopan orphan-drug designation for all three of these diseases: C3G, AAV, and aHUS. The European Commission has granted orphan medicinal product designation for avacopan for the treatment of two forms of AAV: microscopic polyangiitis and granulomatosis with polyangiitis (formerly known as Wegener's granulomatosis) and C3G.

Avacopan was also granted access to the European Medicines Agency's (EMA) PRIority MEdicines (PRIME) initiative**,** which supports accelerated assessment of investigational therapies addressing unmet medical need.

**About ChemoCentryx**

ChemoCentryx is a biopharmaceutical company developing new medications targeted at inflammatory and autoimmune diseases, and cancer. ChemoCentryx targets the chemokine and chemoattractant systems to discover, develop and commercialize orally-administered therapies. ChemoCentryx is currently focusing on its late stage drug candidates for patients with rare kidney diseases. Besides avacopan (described above), the Company's other late stage drug candidate is CCX140, an inhibitor of the chemokine receptor known as CCR2, which is currently being developed for patients with focal segmental glomerulosclerosis (FSGS), a debilitating kidney disease.

ChemoCentryx's Kidney Health Alliance with Vifor Pharma provides Vifor Pharma with exclusive rights to commercialize Avacopan and CCX140 in markets outside of the U.S. and China.

ChemoCentryx also has an immuno-oncology program, which includes a distinct CCR2 inhibitor, CCX872, currently in development for the treatment of advanced non-resectable pancreatic cancer.

**Forward-Looking Statements**
ChemoCentryx cautions that statements included in this press release that are not a description of historical facts are forward-looking statements. Words such as "may," "could," "will," "would," "should," "expect," "plan," "anticipate," "believe," "estimate," "intend," "predict," "seek," "contemplate," "potential," "continue" or "project" or the negative of these terms or other comparable terminology are intended to identify forward-looking statements. These statements include the Company's statements whether avacopan (CCX168) will be shown to be safe and effective in the treatment of IgA nephropathy and other rare diseases, whether CCX140 will be shown to be safe and effective in the treatment of focal segmental glomerulosclerosis (FSGS) and the Company's statement regarding the timing of initiating additional clinical trials to further investigate CCX140 in the treatment of FSGS. The inclusion of forward-looking statements should not be regarded as a representation by ChemoCentryx that any of its plans will be achieved. Actual results may differ from those set forth in this release due to the risks and uncertainties inherent in the ChemoCentryx business and other risks described in the Company's filings with the Securities and Exchange Commission ("SEC"). Investors are cautioned not to place undue reliance on these forward-looking statements, which speak only as of the date hereof, and ChemoCentryx undertakes no obligation to revise or update this news release to reflect events or circumstances after the date hereof. Further information regarding these and other risks is included under the heading "Risk Factors" in ChemoCentryx's periodic reports filed with the SEC, including ChemoCentryx's Annual Report on Form 10-K filed with the SEC March 14, 2017 and its other reports which are available from the SEC's website (www.sec.gov) and on ChemoCentryx's website (www.chemocentryx.com) under the heading "Investors." All forward-looking statements are qualified in their entirety by this cautionary statement. This caution is made under the safe harbor provisions of Section 21E of the Private Securities Litigation Reform Act of 1995.

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