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ChemoCentryx Granted EU Orphan Drug Designation for Avacopan in the Treatment of Debilitating Kidney Disease C3 Glomerulopathy (C3G)

MOUNTAIN VIEW, Calif., May 23, 2017 (GLOBE NEWSWIRE) -- ChemoCentryx, Inc., (Nasdaq:CCXI), a biopharmaceutical company developing new medications targeted at inflammatory and autoimmune diseases and cancer, announced today that the European Medicines Agency (EMA) has granted orphan medicinal product designation for avacopan (formerly CCX168) in the treatment of patients with C3 glomerulopathy (C3G).

In March, ChemoCentryx announced that the U.S. Food and Drug Administration (FDA) had granted orphan drug designation for avacopan in the treatment of C3G. Orphan drug designation can help expedite a drug candidate's path to market and provide certain exclusive marketing advantages after approval. ChemoCentryx plans to initiate a multi-center clinical endpoint study of avacopan for the treatment of C3G in mid-2017.

C3G is a severe kidney disease characterized by deposits of proteins from the body's complement system into and around the kidney's filtration units, the glomeruli. The deposition of complement protein disrupts kidney function and also triggers a profoundly destructive inflammation of the kidney. The disease process ultimately leads to renal failure, with the need for dialysis and kidney transplant.

There is currently no approved treatment for C3G, and relapse is common even after kidney transplant.

Avacopan targets the complement C5a receptor (C5aR) which drives the destructive inflammatory cells in C3G and other kidney diseases. In this way, avacopan is designed to arrest the ongoing process of kidney damage in C3G, allowing kidney function to stabilize.

"Securing both U.S. and EU orphan drug designations in quick succession for avacopan in the treatment of C3G provides further validation of the drug candidate's potential," said Thomas J. Schall, Ph.D., President and Chief Executive Officer of ChemoCentryx. "There are no approved treatments for C3G, a disorder caused by dysregulation of the complement system. We believe the ability to selectively target the C5a receptor with a patient-friendly small molecule such as avacopan could be a significant breakthrough in the treatment of C3G and other kidney disorders."

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 in addition, 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 FDA has granted avacopan orphan-drug designation for all three of these diseases: C3G, AAV, and aHUS. The EMA has now 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 EMA's PRIority MEdicines (PRIME) initiative**,** which supports accelerated assessment of investigational therapies addressing unmet medical need.

About Complement 3 Glomerulopathy (C3G)

C3 glomerulopathy is characterized by evidence of alternative complement activation based on C3 deposition in the glomeruli. This C3 deposition is a manifestation of inappropriate complement activation which also leads to complement 5a (C5a) production. Patients with C3 glomerulopathy often have high levels of protein in the urine (proteinuria) and experience progressive deterioration in renal function. Without treatment, C3G leads to kidney failure, with the need for dialysis and kidney transplant. Even after transplantation, the new kidney will frequently manifest the disease.

There is no approved effective standard therapy for C3G; non-specific immunosuppressants are frequently employed. The estimated prevalence of C3G is two-to-three per million people.

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 C3 glomerulopathy and other rare diseases, whether orphan drug designation will expedite a drug candidate's path to market and provide certain exclusive marketing advantages after approval and the Company's statement regarding the timing of initiating additional clinical trials to further investigate avacopan in the treatment of C3G. 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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