



November 7, 2016

## **ChemoCentryx Announces Presentations of Positive Results from Phase II ANCA-Associated Vasculitis Trials ('CLEAR' and 'CLASSIC') of Orally Administered Complement 5a Receptor Inhibitor CCX168 ('Avacopan')**

*-- Oral presentation at the American Society of Nephrology (ASN) Kidney Week 2016 to highlight Phase II AAV CLEAR trial results --*

*-- Oral presentation at the American College of Rheumatology (ACR) 2016 Annual Meeting to highlight Phase II AAV CLASSIC trial results --*

MOUNTAIN VIEW, Calif., Nov. 07, 2016 (GLOBE NEWSWIRE) -- ChemoCentryx, Inc., (Nasdaq:CCXI), today announced oral presentations at two upcoming medical meetings discussing the positive results obtained from the Phase II CLEAR and CLASSIC trials of CCX168 (newly designated 'avacopan') in patients with anti-neutrophil cytoplasmic auto-antibody (ANCA) - associated vasculitis (AAV). Avacopan is a potent orally-administered small molecule that is a selective inhibitor of the complement C5a receptor, or C5aR, and is the lead drug candidate in the Company's orphan and rare disease program.

The current standard of care in AAV uses high doses of chronic glucocorticoids (steroids such as prednisone or prednisolone), which can cause significant safety issues, including premature death. The Phase II CLEAR trial was designed to assess whether avacopan could provide highly effective control of AAV disease while also eliminating the need for steroids. Separately, the Phase II study known as CLASSIC was designed primarily as a safety study to inform eventual labeling requirements for avacopan. As previously reported, the CLEAR and CLASSIC trials both successfully met their objectives.

"The positive results from the CLEAR and CLASSIC trials mark the successful culmination of our Phase II development program with avacopan in AAV. We believe the data show strongly that avacopan, via a novel mechanism for the treatment of AAV, provides rapid and effective control of the disease. Importantly, the data also show that avacopan eliminates the need for chronic high doses of steroids currently used in the standard of care," said Thomas J. Schall, Ph.D., President and Chief Executive Officer of ChemoCentryx. "It follows that the robust datasets to be presented at these two upcoming medical meetings support our advancing avacopan into Phase III development, which we are now preparing to do."

Presentation information is as follows:

### **AAV Phase II CLEAR Oral Presentation:**

Conference: American Society of Nephrology (ASN) Kidney Week 2016  
Title: Rapid Onset of Action of Orally Administered C5aR Inhibitor CCX168 in Randomized Clinical Trial in ANCA-Associated Vasculitis (CLEAR)  
Presenter: Prof. Vladimír Tesař, MD, PhD, MBA, FASN, Department of Nephrology, Charles University and Principal Investigator of the CLEAR trial  
Session: CKD and AKI Clinical Trials  
Date & Time: Thursday, 11/17/2016, Presentation Start Time: 5:42 PM. CT  
Location: Session room S103, McCormick Place, Chicago, IL.

### **AAV Phase II CLASSIC Oral Presentation:**

Conference: American College of Rheumatology (ACR) 2016 Annual Meeting  
Title: A Randomized Clinical Trial of CCX168, an Orally Administered C5aR Inhibitor for Treatment of Patients with ANCA-Associated Vasculitis  
Presenter: Dr. Peter A. Merkel, Chief of Rheumatology and Professor of Medicine at the University of Pennsylvania  
Session: Vasculitis I: Novel Approaches to Therapy  
Date & Time: Sunday, November 13, 2016, 3:15 PM - 3:30 PM CT

Location: Walter E. Washington Convention Center, Washington, D.C.

## **About CCX168 (avacopan)**

CCX168 (avacopan) is an orally-administered small molecule that is a selective inhibitor of the complement C5a receptor, or C5aR, and is the lead drug candidate in the Company's orphan and rare disease program. The U.S. Food and Drug Administration granted orphan-drug designation for avacopan for the treatment of patients with AAV, (which includes Wegener's granulomatosis, microscopic polyangiitis, and Churg-Strauss syndrome) and also for the treatment of patients with atypical hemolytic uremic syndrome (aHUS). The European Commission has granted orphan medicinal product designation for avacopan for the treatment of microscopic polyangiitis and granulomatosis with polyangiitis (formerly known as Wegener's granulomatosis). Both conditions are forms of AAV. 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 ANCA-Associated Vasculitis**

Anti-neutrophil cytoplasmic auto-antibody (ANCA)-associated vasculitis, or AAV, is a type of rare autoimmune inflammation caused by auto-antibodies. AAV encompasses granulomatosis with polyangiitis (GPA, formerly known as Wegener's granulomatosis), microscopic polyangiitis (MPA), eosinophilic polyangiitis (formerly Churg-Strauss syndrome) and renal limited vasculitis.

AAV represents a severe and often fatal autoimmune disease that is characterized by inflammation that can destroy different organ systems. AAV is the lead indication in the Company's orphan and rare disease program which has the objective of eliminating chronic high dose steroids, which are associated with significant safety issues including death, from the standard of care (SOC) regimen in AAV and replace steroids with avacopan.

AAV affects approximately 40,000 people in the U.S. (with approximately 4,000 new cases each year) and greater than 75,000 people in Europe (with at least 7,500 new cases each year), and is currently treated with courses of immunosuppressants (cyclophosphamide or rituximab) combined with high dose steroid administration. Following initial treatment, up to 30 percent of patients relapse within six to 18 months, and approximately half of all patients will relapse within three to five years.

Current SOC for AAV is associated with significant safety issues. First year mortality is approximately 11 to 18 percent. The single major cause of premature mortality is not disease-related adverse events, but rather infection that is thought largely to be a consequence of steroid administration. Indeed, the multiple adverse effects of courses of steroid treatment (both initial courses and those that are repeated as a consequence of relapse) are major causes of both short-term and long-term disease and death. Such therapy related adverse events contribute significantly to patient care costs, as well as to the diminution of quality of life for patients.

By damaging the body's small blood vessels, AAV affects many organ systems, mostly the kidneys, eyes, lungs, sinuses and nerves. This damage is caused by the destructive activity of inflammatory leukocytes in the body, with neutrophils considered to be the terminal effector cell. In AAV, neutrophils are attracted to sites of vascular destruction as well as activated at those sites by the activity of the complement system product known as C5a and its receptor, C5aR, which is the target of avacopan. By blocking the C5aR, avacopan is thought to reduce vasculitis by reducing neutrophil activation, accumulation, and adhesion, as well as vascular permeability.

## **About ChemoCentryx**

ChemoCentryx, Inc. is a clinical-stage biopharmaceutical company focused on discovering, developing and commercializing orally-administered therapeutics that target the chemokine and chemoattractant systems in order to treat autoimmune diseases, inflammatory disorders and cancer. The chemokine system is a biological network that regulates inflammation via a collection of secreted chemokine molecules, or ligands, and their specific cell surface receptors. Based on its proprietary drug discovery and drug development platform, ChemoCentryx has generated multiple clinical and preclinical-stage programs, each targeting distinct chemokine and chemoattractant receptors with different small molecule compounds. Avacopan, a C5aR inhibitor, is in Phase II development for the treatment of anti-neutrophil cytoplasmic auto-antibody associated vasculitis (AAV). Avacopan appears to be safe, well tolerated and successful in allowing reduction and elimination of high-dose steroids, part of standard of care for AAV patients, while providing effective control of the disease in clinical studies to date. Avacopan is also in Phase II studies for the treatment of atypical hemolytic uremic syndrome (aHUS) and immunoglobulin A nephropathy, or IgA nephropathy (IgAN). ChemoCentryx has licensed exclusive rights to Vifor Pharma to commercialize Avacopan in Europe and certain other markets outside of the U.S. and most of Asia. CCX872, a CCR2 inhibitor, successfully completed Phase I development and is in development for the treatment of non-resectable pancreatic cancer. CCX140, a distinct CCR2 inhibitor, successfully completed a Phase II clinical trial where it was shown to be safe and well tolerated while demonstrating statistically significant improvement in albuminuria in patients with diabetic

nephropathy. Other clinical programs include CCX507, a next generation CCR9 inhibitor, which has successfully completed Phase I development, vercirnon (also known as Traficet-EN or CCX282) a specific CCR9 inhibitor for the treatment of inflammatory bowel disease, and CCX354, a CCR1 inhibitor which successfully completed a Phase II clinical trial for the treatment of rheumatoid arthritis. ChemoCentryx also has several programs in advanced preclinical development.

### **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 regarding timing of initiating Phase III development in AAV for avacopan and whether avacopan will be shown to be effective in Phase III clinical trials in the treatment of AAV and other orphan and rare diseases. 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, 2016 and its other reports which are available from the SEC's website ([www.sec.gov](http://www.sec.gov)) and on ChemoCentryx's website ([www.chemocentryx.com](http://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.

**Source:** ChemoCentryx (CCXI-G)

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