



PRESS RELEASE

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AVACOPAN CONDITIONAL MARKETING AUTHORIZATION APPLICATION ACCEPTED FOR REGULATORY REVIEW BY EUROPEAN MEDICINES AGENCY

- **Conditional marketing authorization application for treatment of patients with ANCA-associated vasculitis validated for start of procedure by EMA**
- **Significant milestone further strengthens Kidney Health Alliance**
- **ChemoCentryx to receive regulatory milestone payment from VFMCPR**

VIFOR FRESENIUS MEDICAL CARE RENAL PHARMA AND CHEMOCENTRYX ANNOUNCE THAT THE EUROPEAN MEDICINES AGENCY HAS ACCEPTED FOR REVIEW THE REGISTRATION DOSSIER IN SUPPORT OF A CONDITIONAL MARKETING AUTHORIZATION FOR AVACOPAN IN THE TREATMENT OF PATIENTS WITH ANCA-ASSOCIATED VASCULITIS.

ZURICH and MOUNTAIN VIEW, Calif., 4 January 2018 – Vifor Fresenius Medical Care Renal Pharma (VFMCPR) and ChemoCentryx, Inc., (Nasdaq: CCXI), announced today a significant milestone in their Kidney Health Alliance: ChemoCentryx's application for avacopan in the treatment of patients with anti-neutrophil cytoplasmic antibody associated vasculitis (ANCA-associated vasculitis or ANCA vasculitis) for regulatory review of its Conditional Marketing Authorization (CMA) application was accepted by the European Medicines Agency (EMA). The EMA's Committee for Medicinal Products for Human Use (CHMP) will now start to assess the CMA application. Under the terms of its kidney health alliance with VFMCPR, ChemoCentryx will receive a milestone payment triggered by this validation of the avacopan CMA application by the EMA.

Avacopan is an orally-administered small molecule that is a highly selective inhibitor of the terminal effector complement C5a receptor (C5aR). Avacopan is currently in late-stage clinical development for the treatment of orphan and rare renal diseases, including ANCA vasculitis. In a randomized, double-blind, placebo-controlled Phase II study in ANCA vasculitis patients, known as the CLEAR trial, avacopan demonstrated that blocking C5aR at the terminal effector pathway of the complement cascade provides therapeutic efficacy and a favourable risk/benefit profile with a rapid onset of action. Avacopan is currently being studied in the pivotal Phase III ADVOCATE trial for the treatment of ANCA vasculitis and is on track to complete enrolment by mid-2018. ChemoCentryx, which is responsible for the discovery and development of avacopan, owns and retains the commercial rights to the drug in the United States and China, and VFMCPR has licensed the rights to commercialize the drug in all other countries.

"The EMA's validation of the Conditional Marketing Authorization application for avacopan represents another critical step in Europe toward realizing our vision of becoming global leader in nephrology therapies. Avacopan is a much more selective, targeted means of treating ANCA than the current standard of care," said Stefan Schulze, Vifor Pharma President of the Executive Committee and COO. "We believe this treatment has real potential for transforming the way that ANCA vasculitis is treated and that it will be of tremendous value to patients who have to live with this rare renal inflammatory disease."

ANCA vasculitis is a systemic disease in which over-activation of the complement pathway further activates neutrophils to lead to inflammation and destruction of small blood vessels. This results in organ damage and failure, with the kidney as the major target, and is fatal if not treated. Currently treatment for ANCA vasculitis consists of courses of non-specific immuno-suppressants (cyclophosphamide or rituximab), combined with high-dose corticosteroid administration for prolonged periods of time, which can be associated with significant clinical risk including death from infection. The CLEAR data show that avacopan may markedly reduce the reliance on glucocorticoids in the treatment of ANCA vasculitis. Furthermore, avacopan by targeting the underlying inflammatory disease process while permitting otherwise normal functioning of other components of the immune system, may provide a basis for a totally new way of improving ANCA vasculitis outcomes.

“A new treatment paradigm for ANCA vasculitis has been a goal for clinicians and patients for decades,” said Professor David Jayne, Vasculitis Director, Addenbrooke’s Hospital, University of Cambridge, UK. “For half a century, we have used high-dose glucocorticoids as part of the treatment of ANCA vasculitis, despite their well-documented toxicities that contribute to the long term morbidity and incapacity associated with vasculitis. In the Phase II CLEAR study, it was demonstrated that inhibition of C5aR may make the chronic use of these toxic steroids obsolete, because the target of avacopan, the C5aR, is an important and specific driver of the destructive inflammation ANCA vasculitis. The current data are very promising and we look forward to the results from the ADVOCATE Phase III trial.”

“The value and promise of avacopan for patients with ANCA vasculitis has today been reinforced by the EMA’s validation of the avacopan CMA application,” said Thomas J. Schall, Ph.D., President and Chief Executive Officer of ChemoCentryx. “We at ChemoCentryx, together with our outstanding partners at Vifor Pharma, are unswerving in our devotion to creating new medicines for patients suffering from devastating kidney diseases. To ANCA patients, we say this: your cause is our cause; we are determined to succeed. Achieving this pivotal regulatory milestone is a big step toward our ultimate success.”

In June 2016, avacopan was granted access to the EMA’s Priority Medicines (PRIME) regulatory initiative for the treatment of patients with ANCA vasculitis. Access to the PRIME initiative is granted by the EMA to support the development and accelerate the review of new therapies to treat patients with unmet medical need.

FURTHER INFORMATION

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Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis, or ANCA vasculitis, is a group of highly inflammatory autoimmune and progressive rare diseases caused by the over-activation of the complement system, which activates neutrophils to destroy blood vessels through inflammation. Patients with granulomatosis with polyangiitis (GPA; formerly Wegeners) or microscopic polyangiitis (MPA), two forms of ANCA vasculitis have various presentations from asymptomatic abnormal lab results to relapsing-remitting states characterized with recurring flares, accruing into irreversible organ system damage, failure and death.

ANCA vasculitis is the lead indication in the ChemoCentryx's orphan and rare disease program which has the objective of making chronic high dose glucocorticoids irrelevant. In addition to being at major risk of severe necrotizing inflammation of vessel walls leading to multiple organ system failure, in particular end stage renal insufficiency as well as mortality, newly diagnosed patients as well as those in remission suffer from impaired quality of life, which amongst several general health-reported parameters, fatigue is major determinant. The disease affects people of working age and significantly impacts multiple aspects of their physical function, emotional well-being, and overall productivity.

ANCA vasculitis affects approximately 40,000 people in the U.S. (with approximately 4,000 new cases each year) and less than 50,000 people in Europe (with an estimated 5,000 new cases each year), and is currently treated with courses of immunosuppressants (cyclophosphamide or rituximab) combined with high dose steroid administration. These induction protocols achieve only partial sustained remission rates of approximately 70%, with up to 30 percent of patients relapsing within six to 18 months, and approximately half of all patients relapsing within three to five years. Patients with renal involvement have a worse prognosis than patients without renal involvement, and 23% of ANCA vasculitis patients who require dialysis or transplant at time of diagnosis die within six months. As early as six months after diagnosis, 8% of patients develop end-stage renal disease and a total of 42% of patients with renal involvement die or develop end-stage renal disease at two years.

The current standard of care for ANCA vasculitis is associated with significant safety issues, underscoring the need for new therapies that specifically target disease mechanisms more selectively. First year mortality is approximately 11% to 18%, with most of the deaths occurring within the first three months, during the time when glucocorticoids are used at

high doses. The single major cause of premature mortality is not disease-related, 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 morbidity and mortality. 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, ANCA vasculitis 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 ANCA vasculitis, 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.

Avacopan is an orally-administered small molecule that is a selective inhibitor of the terminal effector and neutrophil chemoattractant complement C5a receptor, or C5aR. It allows tempering of ANCA responses and thereby to prevent both complement C5a activation while leaving other host defense mechanisms (such as the membrane attack complex, being distinct from C5b) of the immune system unaffected. Avacopan is in phase III development for the treatment of anti-neutrophil cytoplasmic auto-antibody-associated vasculitis (ANCA vasculitis). 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 patients with ANCA vasculitis. 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 granted avacopan orphan-drug designation for all three of these diseases: C3G, ANCA vasculitis, and aHUS. The European Commission has granted orphan medicinal product designation for avacopan for the treatment of two forms of ANCA vasculitis: 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.

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 early stage drug candidates that target chemoattractant receptors in other Inflammatory and autoimmune diseases and in cancer.

Vifor Pharma Group, formerly Galenica Group, is a global specialty pharmaceuticals company. It aims to become the global leader in iron deficiency, nephrology and cardio-renal therapies. The company is the partner of choice for specialty pharmaceuticals and innovative patient-focused solutions. Vifor Pharma Group strives to help patients around the world with severe and chronic diseases lead better, healthier lives. The company develops, manufactures and markets pharmaceutical products for precision patient care. Vifor Pharma Group holds a leading position in all its core business activities and consists of the following companies: Vifor Pharma; Vifor Fresenius Medical Care Renal Pharma, a joint company with Fresenius Medical Care; Relypsa; and OM Pharma. Vifor Pharma Group is listed on the Swiss Stock Exchange (SIX Swiss Exchange, VIFN, ISIN: CH0364749348). For more information, visit www.viforpharma.com.

Vifor Fresenius Medical Care Renal Pharma Ltd., a common company of Vifor Pharma Group and Fresenius Medical Care, develops and commercialises innovative and high quality therapies to improve the life of patients suffering from chronic kidney disease (CKD) worldwide. The company was founded at the end of 2010 and is owned 55% by Vifor Pharma Group and 45% by Fresenius Medical Care. For more information about Vifor Fresenius Medical Care Renal

Pharma and its parent companies, please visit www.vfmcpr.com, www.viforpharma.com and www.freseniusmedicalcare.com.

ChemoCentryx 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" or "continue" or the negative of these terms or other comparable terminology are intended to identify forward-looking statements. These statements include statements regarding whether avacopan will be shown to be effective in the pivotal Phase III ADVOCATE trial in the treatment of ANCA vasculitis and other rare renal diseases, whether avacopan will receive conditional marketing approval by the EMA for the treatment of ANCA Vasculitis and whether patient enrollment of the ADVOCATE Phase III trial will be completed by mid-2018. 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 ChemoCentryx'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 14 March 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.