



## Media Information

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Subject Vifor Pharma expands license agreement for avacopan to include additional territories

### **Vifor Pharma and ChemoCentryx Announce Expansion of Avacopan Agreement for Rare Renal Diseases**

*-- ChemoCentryx retains all rights in the United States and China; Vifor Pharma gains additional marketing rights in Asia, including Japan and Middle Eastern markets --*

*-- ChemoCentryx to receive additional USD 20 million upfront cash commitment plus royalties on potential net sales --*

*-- ChemoCentryx remains responsible for worldwide development of avacopan --*

Vifor Pharma, a company of the Galenica Group, and ChemoCentryx, Inc., (Nasdaq: CCXI), announced today that Vifor Pharma has gained rights to commercialize avacopan in Asia, including Japan and the Middle East. ChemoCentryx retains all rights in the United States and China. Avacopan (CCX168) is an orally-administered inhibitor of the complement 5a receptor (C5aR) and is currently in Phase III development for rare renal diseases.

The expanded agreement effectively gives Vifor Pharma rights to commercialize avacopan for orphan and rare renal diseases in all markets outside the United States and China, building on the original licensing agreement signed in May 2016.

ChemoCentryx will receive an upfront cash commitment of USD 20 million in return for the new rights, plus tiered double-digit royalties on potential net sales. This is in addition to the USD 85 million upfront paid under the original May 2016 licensing agreement.

Avacopan is being developed by ChemoCentryx for the treatment of orphan and rare renal conditions including but not limited to anti-neutrophil cytoplasmic auto-antibody (ANCA)-associated vasculitis (AAV). AAV is a systemic disease which most commonly presents as kidney dysfunction. AAV is currently treated with courses of non-specific immuno-suppressants (cyclophosphamide or rituximab), combined with high-dose corticosteroid administration, which is associated with significant safety issues, including premature death. Avacopan is in Phase III development (the ADVOCATE trial) for the treatment of AAV and is also in development for other orphan and rare renal diseases, including C3 glomerulopathy (C3G) and atypical hemolytic uremic syndrome (aHUS).

“The expansion of avacopan territory rights with renal care leader Vifor Pharma, and with ChemoCentryx retaining all US and China rights, expertly positions our rare renal disease portfolio for global commercialization,” said Thomas J. Schall, Ph.D., President and CEO of ChemoCentryx. “We have now harmonized both the avacopan and the subsequent agreement which we signed in late December for another rare renal asset, CCX140, in our highly productive Kidney Health Alliance with Vifor Pharma. ChemoCentryx also has greater than USD 200 million in proforma reserves, based on our last filings, providing for an ambitious development plan for avacopan and CCX140 in

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orphan and rare renal diseases. We and Vifor Pharma believe these two unique clinical assets have the potential to transform lives of those with devastating rare renal diseases, and to build extraordinary value for both of our enterprises.”

“The acquisition of global commercialization rights outside the US and China demonstrates our strong belief in the potential of avacopan in a wide range of rare and orphan renal diseases,” said Gianni Zampieri, CEO of Vifor Pharma. “This agreement further strengthens our growing partnership with ChemoCentryx, and underlines our commitment to bring highly innovative therapies to patients with serious renal conditions around the world.”

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**Avacopan** is an orally-administered complement inhibitor which specifically targets the receptor for the complement C5a receptor (C5aR). This receptor is known to activate destructive cells in certain autoimmune diseases including AAV. Avacopan is the lead drug candidate in ChemoCentryx'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. Avacopan successfully completed Phase II development where it was shown to be safe and effective in eliminating chronic high dose steroids, which are associated with significant safety issues including death, from the standard of care (SOC) regimen in AAV and is now in Phase III development. Avacopan is also being developed for other autoimmune disorders including C3 Glomerulopathy (C3G) and atypical hemolytic uremic syndrome (aHUS).

**ChemoCentryx** is a clinical-stage biopharmaceutical company primarily focused on developing new medicines for patients with rare renal diseases. ChemoCentryx targets the chemokine and chemoattractant systems to discover, develop and commercialize orally-administered therapies to treat orphan and rare diseases. Avacopan (CCX168), an inhibitor of the complement 5a receptor (C5aR), is in Phase III development for the treatment of anti-neutrophil cytoplasmic auto-antibody-associated vasculitis (AAV). Avacopan was 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 being developed in patients with atypical hemolytic uremic syndrome (aHUS) and C3 glomerulopathy (C3G). CCX140, an inhibitor of the chemokine receptor known as CCR2, successfully completed a Phase II clinical trial where it was shown to be safe and well tolerated while demonstrating statistically significant improvement in proteinuria in patients with diabetic nephropathy and is currently being developed in a rare kidney disease known as focal segmental glomerulosclerosis (FSGS). Both avacopan and CCX140 are part of a Vifor Pharma-ChemoCentryx Kidney Health Alliance which provides Vifor Pharma with exclusive rights to commercialize avacopan and CCX140 in certain markets outside of the U.S. and China. ChemoCentryx has an immuno-oncology program, which includes a distinct CCR2 inhibitor, CCX872, currently in development for the treatment of advanced non-resectable pancreatic cancer.

**Vifor Pharma**, a company of the Galenica Group, is a world leader in the discovery, development, manufacturing and marketing of pharmaceutical products for the treatment of iron deficiency. The company also offers a diversified portfolio of prescription medicines as well as over-the-counter (OTC) products. Vifor Pharma, headquartered in Zurich, Switzerland, has an increasingly global presence and a broad network of affiliates and partners around the world. For more information about Vifor Pharma and its parent company Galenica, please visit [www.viforpharma.com](http://www.viforpharma.com) and [www.galenica.com](http://www.galenica.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,"

Date 14 February 2017

Page 4/4

Subject Vifor Pharma expands license agreement for avacopan to include additional territories

*"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 CCX168 will be shown to be effective in Phase III clinical trials in the treatment of AAV and other rare renal diseases and whether eligible milestone payments or royalties on net sales of CCX168 will be attained and the potential of avacopan and CCX140 to build value for ChemoCentryx. 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.*