Supplementary methods

A total of 11 patients received avacopan during the study period (Toulouse n=8, Bordeaux n=3) owing to an early-access program developed by Vifor Pharma and authorised by the French Authority for Health. The BVAS score was calculated as previously reported to allow comparison with the ADVOCATE study¹⁶. The Vasculitis Damage Index was calculated as recommended¹⁷. Remission was defined by a BVAS of 0. The estimated glomerular filtration rate (eGFR) was calculated using the CKD-EPI formula. Urinary levels of sCD163 were assessed using ELISA (R&D Systems, DuoSet, DY1607) after urine centrifugation and 1:4 dilution. Urinary creatinine measurement was performed on a ABX Pentra 400 analyser. usCD163 was given as a ratio to urine creatinine.

Ethical considerations: All patients gave informed consent to inclusion in the Nephrogene cohort, which was approved by the French national ethical review board (agreement number DC-2011-1388). The study was conducted according to the Declaration of Helsinki, as revised in 2004.

Acknowledgments: We acknowledge Mr Montbrun (INSERM UMR 1297 - Biochemical Platform) who performed the urinary creatinine dosage. This work was presented at the 6th congress of the French Society of Nephrology, Dialysis and Transplantation (Toulouse, October 2021).

Supplementary references

- S1. Geetha D, Jefferson JA. ANCA-Associated Vasculitis: Core Curriculum 2020. *Am J Kidney Dis*. 2020;75(1):124-137. doi:10.1053/J.AJKD.2019.04.031
- S2. Mukhtyar C, Flossmann O, Hellmich B, et al. Outcomes from studies of antineutrophil cytoplasm antibody associated vasculitis: a systematic review by the European League Against Rheumatism systemic vasculitis task force. *Ann Rheum Dis.* 2008;67(7):1004-1010. doi:10.1136/ARD.2007.071936
- S3. Ann Tan J, Dehghan N, Chen W, Xie H, Esdaile JM, Antonio Avina-Zubieta J. ExtEndEd rEport Mortality in ANCA-associated vasculitis: a meta-analysis of observational studies. *Ann Rheum Dis.* 2017;76:1566-1574. doi:10.1136/annrheumdis-2016-210942
- S4. Xiao H, Dairaghi DJ, Powers JP, et al. C5a receptor (CD88) blockade protects against MPO-ANCA GN. *J Am Soc Nephrol*. 2014;25(2):225-231. doi:10.1681/ASN.2013020143
- S5. Brilland B, Garnier AS, Chevailler A, Jeannin P, Subra JF, Augusto JF. Complement alternative pathway in ANCA-associated vasculitis: Two decades from bench to bedside. *Autoimmun Rev.* 2020;19(1). doi:10.1016/J.AUTREV.2019.102424
- S6. Kettritz R. Vasculitis: A CLEAR argument for targeting complement in ANCA vasculitis. *Nat Publ Gr.* 2017;13. doi:10.1681/ASN.2016111179
- S7. Merkel PA, Niles J, Jimenez R, et al. Adjunctive Treatment With Avacopan, an Oral C5a Receptor Inhibitor, in Patients With Antineutrophil Cytoplasmic Antibody-Associated Vasculitis. *ACR open Rheumatol*. 2020;2(11):662-671. doi:10.1002/ACR2.11185

Supplementary figure 1. Kinetics of urinary soluble CD163 (usCD163) concentration in 6 patients with ANCA-associated vasculitis who received avacopan.

sCD163 concentration in urine, normalized to urinary creatinine.

