## Comment on: Microscopic polyangiitis: clinical characteristics and long-term outcomes of 378 patients from the French Vasculitis Study Group Registry

by Nguyen *et al*.

Sirs,

We read the article by Nguyen *et al*. with great interest (1). In this study, they presented the French Vasculitis Study Group microscopic polyangiitis (MPA) experience including 378 patients between 1966 and 2017. They found older age, high creatinine levels, severe gastrointestinal involvement and presence of mononeuritis multiplex were found of predictors of mortality and also severe renal impairment was found as protector against relapse. However, there are raising concerns to be answered.

MPA-associated fibrosing interstitial lung disease (ILD) is an emerging area of AAV vasculitis. In German MPA cohort, a higher mortality [hazard ratio (HR) 4.04 (95% CI 1.21, 13.45), $p$=0.02] was reported (2). Even though fibrosing ILD is not an item in 2009 FFS, subgroup analysis of this larger cohort could give more information about the impact of ILD in MPA.

Ear, nose, and throat (ENT) symptoms were present in a minority of MPA patients (13.2%). ENT involvement is not considered as a significant marker of prognosis for MPA patients. In the methods section, the authors described 1996 FFS and the revised FFS 2009. Absence of ear, nose, and throat symptoms was also written in the text. This can lead to misunderstandings of the impact on ENT in MPA. The frequency of ENT absence can be calculated from Table I (86.8%). We believe ENT symptoms should be a part of the FFS used only for patients with granulomatous polyangiitis (GPA) or eosinophilic granulomatosis with angiitis (EGPA) as mentioned in 2011 paper (3). This concern should be clarified in the article.

Neurological involvement was reported in 191 (50.5%) patients reported; 119 patients had mononeuritis multiplex and 10 had CNS involvement. It would be better to know which involvements the other patients had.

As reported, 49 (13.9%) patients were ANCA negative. Although relapse and mortality were reported similar irrespective from ANCA status, it would be great to know if there were any differences between ANCA positive and negative groups regarding demographic data, disease presentation and treatment choices.

There were some missing and conflicting points about treatment. It was reported that 97.3% of patients received glucocorticoids and/or immunosuppressants as induction therapy. In other words, 2.7% of patients did not have any kind of immunosuppressive treatment. Management protocol of these patients would be great to know. Also, did any patient receive intravenous immunoglobulin as induction therapy? Of 46 patients who underwent plasmapheresis, indications were severe renal involvement in 28 patients, alveolar haemorrhage in 3 patients, both indications in 5 patients. It would be good to know the indications of plasmapheresis in 4 remaining patients. In addition, in page 3, it was reported that 278 patients had immunosuppressants, however, in Table I the number was 277. There is a contradiction here.

There are some concerns regarding the technical and statistical issues. In Table I, "Neurological involvement" should be written in line with other subheadings like "Gastrointestinal involvement" and "Eye involvement" etc. There is duplicate information regarding data presentation style in the footer note of Table II. In the univariate analysis of predictors of relapse, $p$ was 0.03 for immunosuppressant use in the induction regimen. It would be great to know why this variable was not included in final model.

Nevertheless, with longer duration of follow-up and large number of participants, this study is a pioneer study in the area of small-vessel vasculitidies.

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*Competing interests: none declared.*

*Clin Exp Rheumatol 2021; 39 (Suppl. 128): S8.*



### References

1. NGUYEN Y, PAGNOUX C, KARRAS A *et al*.: Microscopic polyangiitis: Clinical characteristics and long-term outcomes of 378 patients from the French Vasculitis Study Group Registry. *J Autoimmun* 2020: 102467.
2. SCHIRMER JH, WRIGHT MN, VONTHEIN R *et al*.: Clinical presentation and long-term outcome of 144 patients with microscopic polyangiitis in a monocentric German cohort. *Rheumatology* (Oxford) 2016; 55: 71-9.
3. GUILLEVIN L, PAGNOUX C, SEROR R *et al*.: The Five-Factor Score revisited: assessment of prognoses of systemic necrotizing vasculitides based on the French Vasculitis Study Group (FVSG) cohort. *Medicine* (Baltimore) 2011; 90: 19-27.