

## Response to: 'Correspondence on: 'Dupilumab as a novel steroid-sparing treatment for IgG4-related disease' by Simpson *et al*' by Ebbo *et al*

We thank Ebbo *et al* for their interest in our case report<sup>1</sup> and for providing their case of a patient with IgG4-related disease (IgG4-RD) whom they have also treated with dupilumab.<sup>2</sup> Notably, Ebbo *et al* reported variability in the response to dupilumab compared with our patient. Our patient did have different clinical manifestations than that of Ebbo *et al*'s patient, as our patient had profound retroperitoneal and genitourinary fibrosis along with atopic dermatitis, asthma, allergic rhinoconjunctivitis, parotitis, and sinusitis while Ebbo *et al*'s patient presented with dacryoadenitis and sialadenitis in conjunction with allergic manifestations such as chronic rhinosinusitis with nasal polyps, asthma and urticaria. We are curious also to ask if the relationships between the variables presented in Figures C, D and E were statistically significant from their data.

In response to Ebbo *et al*'s belief that the response we have seen is confounded by initial use of prednisone, we would like to clarify that it has come to our attention post publication (29 December 2019), that the patient in our case did not take a single dose of prednisone due to undisclosed non-compliance, therefore making dupilumab the first-line treatment that was used. Due to the fact that the patient Ebbo *et al* described underwent rituximab therapy prior to dupilumab therapy, it is also possible that this added to the variability in the observed patient's response compared with ours. It should also be noted that because Ebbo *et al*'s patient was on glucocorticoids for 3 months and then approximately 1 month a year before dupilumab treatment, the same argument can be made that perhaps this confounds their results. However, due to the variability in both clinical manifestations and prior treatments that can be seen in the IgG4-RD patient presented in comparison to ours, it can be understood why perhaps Ebbo *et al*'s patient may have not shown such complete resolution to their disease after dupilumab treatment.

We do find it interesting that Ebbo *et al* have reported their case with respect to a suboptimal clinical and radiological response to dupilumab 8 months after treatment. We believe it would be useful to provide information with respect to the 2019 American College of Rheumatology/European League Against Rheumatism classification criteria for IgG4-related involvement post-dupilumab treatment to compare to the pre-treatment score. We do believe Ebbo *et al*'s report shows valuable data with regards to the variability in responses to dupilumab that may take place in IgG4-RD and we wonder if it would be valuable to study IgG4-RD response to dupilumab in two separate groups through a large randomised clinical trial: dupilumab as

a first-line treatment for IgG4-RD and dupilumab for patients that are refractory to conventional treatments such as glucocorticoids, immunosuppressants and/or rituximab for IgG4-RD. By studying both groups fully using well-detailed inclusion and exclusion criteria, we believe that the optimal place for dupilumab in the IgG4-RD treatment algorithm will be able to be identified, as well as dupilumab's safety and efficacy for this condition will be thoroughly investigated.

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### REFERENCES

- 1 Simpson RS, Lau SKC, Lee JK. Dupilumab as a novel steroid-sparing treatment for IgG4-related disease. *Ann Rheum Dis* 2020;**79**:549–50.
- 2 Ebbo M, De Sainte-Marie B, Muller R, *et al*. Comment on article: 'Dupilumab as a novel steroid-sparing treatment for IgG<sub>4</sub>-related disease' by Simpson *et al*. *Ann Rheum Dis* 2022;**81**:e26.