

Response to: 'Inflammation in SLE-PAH: good news or not?' by Junyan Qian *et al*

Thank you very much for your interest in our article 'Two distinct clinical phenotypes of pulmonary arterial hypertension secondary to systemic lupus erythematosus', and we are glad to answer the question 'Inflammation in SLE-PAH: good news or not?' as below.

First, referral bias may exist and contribute to the different outcome. As an example, the Peking Union Medical College Hospital (PUMCH) cohort has more patients with pulmonary arterial hypertension secondary to systemic lupus erythematosus (SLE-PAH) with vasculopathic subtype.¹ Among these patients, the 3-year survival is precisely in line with our two cohorts,^{1,2} suggesting those SLE with 'pure' PAH are more homogeneous. Further analysing the more heterogeneous vasculitic subtype, the baseline manifestations and SLE disease activity index (SLEDAI) of our data vary. We totally agree that such patients with more systemic inflammatory components would be a better responder to immunosuppressive therapies, and some patients may even experience a reversible PAH course.³⁻⁵ Nevertheless, when talking about all-cause mortality, it makes sense that a patient who had a pronounced lupus nephritis (LN) or neuropsychiatric systemic lupus nephritis (NPSLE) or haematological manifestations on top of PAH will have a higher probability of poorer outcome.⁶ Another scenario is that when such patients underwent aggressive immunosuppressive therapy and complicated with infection, they tend to be more vulnerable to haemodynamically unstable. In other words, for those more heterogeneous vasculitic SLE-PAH subtype, baseline parameters and other confounders such as comorbidities, complications and treatment protocols should be scrutinised before a definitive conclusion can be made. It seems indisputable that clinical trials should be carried out on such patients in order to justify the appropriate combination of immunosuppressive therapy and PAH-targeted treatment, and improve the overall outcome.

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