

Dr. Guerra, *et al* reply

To the Editor:

Zhong and colleagues offer an interesting view¹ on our paper on subclinical cardiac impairment in patients with idiopathic inflammatory myopathies (IIM)².

First, Zhong suggests that the reported difference in right ventricular global longitudinal strain (GLS) between patients with and without lung involvement (−17.1% vs −20.9%) could be due to the endothelial dysfunction and systemic inflammation related to interstitial lung disease (ILD) rather than to the primary heart involvement by IIM. Albeit the reported difference was not statistically significant ($p = 0.118$), subgroup analyses were greatly underpowered, so we can neither exclude nor confirm association and we recommend caution in extracting clinical messages. While the idea is surely compelling, unfortunately very little data support the hypothesis suggested by Zhong. First, indirect markers of pulmonary circulation involvement, such as estimated pulmonary arterial pressure (sPAP), and right atrial volume (iRAV), were not different between our patients with IIM and controls, and well within the cutoff range for normality (sPAP 23.6 mmHg vs 21.5 mmHg, $p = 0.385$; iRAV 14.3 ml/m² vs 14.8 ml/m², $p = 0.734$). Second, an increase in right atrial pressure usually precedes overt systolic impairment of the right ventricle, because they both decline in the last stages of decompensated right heart failure³. Third, an abnormal cardiac uptake during scintigraphy⁴ and late gadolinium enhancement⁵ has been reported for the right as well as the left ventricle in patients with IIM. Fourth, diffuse perivascular and interstitial mononuclear infiltrates, muscle fiber degeneration, and widespread fibrosis have been commonly reported in IIM autopsies⁶. As mentioned, however, available data are scarce and a synergy between direct heart involvement and endothelial function dysregulation cannot be excluded.

Regarding the association of IIM with the more traditional CV risk factors, we agree that systemic hypertension, diabetes mellitus, and dyslipidemia are strongly represented in patients with IIM. Therefore, even though our patients with IIM showed impaired GLS when compared with controls that were matched for age, sex, and CV risk profile, we agree that IIM could be worsened by these comorbidities. In fact, it has been estimated that one-fifth of all hospital admissions in patients with IIM are related to a CV issue, and that IIM patients with atherosclerotic disease are twice as likely to die during hospitalization⁷.

As Zhong and colleagues rightly suggest, well-executed, prospective, observational studies focused on heart involvement progression and clinical and echocardiographic response to treatment represent the next step in unraveling the deep connection between IIM and the heart.

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