Author’s response to reviews

Title: Characteristics of exercise capacity in female systemic lupus erythematosus associated pulmonary arterial hypertension patients

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Reviewer 1 (Maria Grau): We have already addressed the questions of reviewer 1 during the earlier revision process. We thank you for your valuable inputs towards improving this manuscript. We have rephrased the objective and improved the abbreviations as per your suggestions.

Reviewer 2 (Athanassios Manginas): Thank you for your earlier suggestions. We improved the manuscript as per your suggestions. We have removed two references and shortened the discussion as you had asked us to shorten the discussion by at least 30%.
Reviewer 3 (Yoshihiro Fukumoto)

1. What was the clinical question in this study? From this study, what clinical benefits are there in the clinical situation? 2. There were some differences in cardiac and pulmonary function between SLE PAH and IPAH. Then, there were some difference in CPET data. Then, what would the authors like to say? Also, they should check frailty. 3. The authors should examine the time course of CPET in the aspects of each therapy. For example, they should examine if CPET can predict the efficacy of therapies, or prognosis. In the current paper, there is no clinical impact. 4. Next, the authors should also examine the time course of pulmonary function. 5. How about the evaluation of cardiac function on MRI?

CPET has been used in other patient cohorts and we wanted to use CPET along with right heart catheterization data to better understand the gas exchange efficiency and contrast that with hemodynamics in two patient groups. Since the RHC is invasive and not feasible to be performed regularly we wanted to understand if certain CPET parameters would adequately reflect disease state/progression. We agree that we do not have repeat results or follow up data, which are significant limitations of the present study.

We fully agree with your idea to examine the time course of CPET based on individual patient’s treatments. We initially tried to do that but since SLE PAH patients are so few, to appropriately stratify them for statistical analysis proved an ultimate challenge. But when we have more such patients in our hospital registry we will do that. In fact preliminary work for this has already been started. We hope to be able to have sufficient data soon so that we can write a follow up article.

In all patients and control subjects the pulmonary function test was performed prior to performing the CPET. Patients did not have Cardiac MRI and is not routinely done in our center, plus some patients have been lost to follow up. We appreciate your invaluable suggestion and we will improve hence, but to have a decent sample of SLE PAH patients is proving very challenging in a single tertiary level pulmonary hospital such as ours.