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Title: Exercise in patients with lymphangioleiomyomatosis: Performance and the prevalence of dynamic hyperinflation

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Body: Introduction: Previous studies suggested multiple factors for exercise cessation in LAM. Although airflow limitation is frequent in LAM, the dynamic hyperinflation (DH) and its repercussions during exercise have not been evaluated yet. Objectives: To evaluate the prevalence and repercussions of DH in LAM and also the dynamic responses during maximal exercise, in comparison to controls. Methods: 42 patients performed pulmonary function tests (PFT) and symptom-limited incremental cycle exercise, in comparison to ten age-matched healthy women. Dyspnea intensity, inspiratory capacity, oxygen saturation (SpO2), cardiac, metabolic and respiratory variables were obtained during exercise. 6MWT was performed by LAM patients indeed. Results: Compared to controls, LAM had higher baseline dyspnea, obstructive pattern, air trapping and lower DLCO in PFT. In LAM, maximal exercise performance was reduced, associated with ventilatory limitation, greater dessaturation and dyspnea. DH occurrence was higher in LAM than controls (55 vs. 0%), with a significant correlation to airflow obstruction, air trapping and DLCO. Patients who developed DH had not only a ventilatory contribution to exercise cessation on cycling, but also greater dessaturation and dyspnea intensity during 6MWT, compared with non-DH subgroup. Conclusions: Ventilatory limitation is an important reason for exercise cessation in LAM, compared with controls. DH was prevalent and had association with severity of disease, higher dyspnea and lower SpO2. On 6MWT, dessaturation and increased dyspnea were greater in DH subgroup. Future interventions to reduce DH might contribute to improve dyspnea and exercise tolerance in LAM.