The benefits and safety of pulmonary rehabilitation in patients with lymphangioleiomyomatosis (LAM)

Lymphangioleiomyomatosis (LAM) is currently considered a low-grade neoplasm that mainly affects young women, and is characterized by the presence of diffuse pulmonary cysts. Several studies have shown that patients with LAM have reduced exercise capacity secondary to multiple mechanisms, which contributes to decreased quality of life. In addition, there is a concern, even among some respiratory physicians, that these patients should not perform exercise due to a higher risk of developing complications, mainly pneumothorax.

Pulmonary rehabilitation (PR) is an intervention that involves exercise training, education, including information about physiopathology, treatment and self-management, and behaviour changes, which aims to improve the physical and psychological conditions of patients with different chronic respiratory diseases. However, the impact of PR has not been described in patients with LAM, and we hypothesized that such intervention would be beneficial and safe for these patients.

We performed a clinical trial in 40 patients with LAM (21 and 19 patients were assigned to PR and control groups, respectively). The PR program consisted of 24 sessions of 1 h duration (twice weekly) divided into 30 min of aerobic exercise on a treadmill and 30 min of muscle strength training. Patients also received education on different themes related to LAM, including physiopathology, risk of complications, treatment and self-management.

We evaluated several variables before and after PR, including exercise capacity, quality of life, daily physical activity (steps per day), pulmonary function tests, dyspnea intensity, anxiety and depression symptoms, and muscle strength. We also examined the safety of exercise in this population by the occurrence of adverse events, including pneumothorax.

Our study has shown that PR determined benefits in several outcomes in patients with LAM, including a significant improvement in exercise capacity and in daily physical activity (steps per day), reduction in breathlessness intensity and in depression symptoms, and also improvement in quality of life, and in muscle strength. However, there was no change in pulmonary function tests. Our findings suggested that the increase in exercise tolerance related to PR in LAM was due to an improvement in peripheral muscle function. In addition, our program had a high rate of adherence and PR was safe and no pneumothorax or other severe complication was found during the study.

Respiratory physicians should consider the enrollment of patients with LAM with reduced exercise capacity in a PR program or consider to stimulate exercise practice for those with less severe disease, if there is no contraindication. Besides the benefits in pulmonary function tests and exercise capacity, we reinforce the importance that interventions for patients with LAM also demonstrate impact on other relevant outcomes, such as quality of life, breathlessness intensity, and depression and anxiety symptoms.

Bruno Guedes Baldi, Carlos Roberto Ribeiro Carvalho
Pulmonary Division, Heart Institute (InCor), University of São Paulo Medical School, Sao Paulo, Brazil

Publication

Pulmonary rehabilitation in lymphangioleiomyomatosis: a controlled clinical trial.
Araujo MS, Baldi BG, Freitas CS, Albuquerque AL, Marques da Silva CC, Kairalla RA, Carvalho CR, Carvalho CR
Eur Respir J. 2016 May