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Title: Is pulmonary rehabilitation (PR) an effective therapy in lymphangioleiomyomatosis (LAM)?

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Body: Background: LAM is an orphan lung disease affecting young women. There is an estimated prevalence of 200 patients in Germany. Beside supportive treatment of respiratory distress there are only few drug therapies (progesterone, sirolimus) of unknown effectiveness. In end-stage LAM lung transplantation (LTx) may be the only remaining therapeutic option. The role of PR is not yet defined. Methods: In a prospective open clinical trial data of 22 LAM patients prior to LTx (LAM-P) (45,9±9y., FEV1=31,4±17%pred, diffusion capacity=35,8±13%, paO_2=59,1±9 mmHg, LTOT n=19) and 24 LAM patients after LTx (LAM-Tx) (42,8±10y., FEV1=65,7±19%pred) were evaluated. All patients underwent a specialized multidisciplinary inpatient PR for 45±9,6 (LAM-P) and 35,4±18 (LAM-Tx) days. Results: We found significant and clinically relevant changes for both PR approaches. For the LAM-P group we saw a significant improvement in 6-min walking distance (6MWD) 59±50m ($p<0,001$). In LAM-Tx patients 6MWD increased significantly by 103±85m ($p<0,001$). Thus the benefit of post LTx-PR in LAM-patients is comparable to the results of lung transplant patients with other underlying disease. Health-related quality of life (HRQL) (SF36) improved significantly for the mental summary score in LAM-Tx. Lung function parameters did not change for LAM-P but FEV1 improved significantly for LAM-Tx patients. Conclusion: PR in LAM patients before and after LTx is a safe therapeutic approach that leads to significant increases of exercise capacity and tends to improve HRQL. In view of a progressive disease with only a few therapeutic options rehabilitation should be considered early in the treatment of LAM, especially when LTx is necessary.