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Title: Exercise training in pulmonary arterial hypertension associated with connective tissue diseases

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Body: Background: The objective of this prospective study was to assess short-and long-term efficacy of exercise training(ET) as add-on to medical therapy in patients with connective tissue diseases-associated pulmonary arterial hypertension(CTD-APAH). Patients with invasively confirmed CTD-APAH received ET in-hospital for 3 weeks and continued at home for 15 weeks. Efficacy parameters have been evaluated at baseline and after 15 weeks by blinded-observers. Survival rate has been evaluated in a follow-up period of 2.9±1.9 years. Results: Twenty-one consecutive patients were included and assessed at baseline, and after 3 weeks, 12 after 15 weeks. Patients significantly improved the mean distance walked in 6 minutes compared to baseline by 67±52 meters after 3 weeks(p<0.001) and by 71±35 meters after 15 weeks(p=0.003), scores of quality of life(p<0.05), heart rate at rest and maximal workload. Systolic pulmonary artery pressure and diastolic systemic blood pressure improved significantly after 3 weeks of ET. The 1- and 2-year overall-survival rates were 100%, the 3-year survival 73%. In one patient lung transplantation was performed 6 months after ET. Conclusion: ET as add-on to medical therapy is effective in patients with CTD-APAH to improve work capacity, quality of life and prognostic parameters and improves the 1-, 2- and 3-year survival rate. Further randomized controlled studies are needed to confirm

these results.