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**Home Based Long Term Cardiopulmonary and Inspiratory Muscle Training for Children and Adults with Fontan Circulation**

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**Objectives:** Cardiopulmonary capacity in patients with Fontan circulation is known to be reduced to around 60 % of that of healthy individuals (Kempny 2011). Short-term home-based physical training programs have been shown to improve physical function as well as quality of life in these patients (Jacobsen 2016). Inspiratory muscle training has also been shown to improve cardiac output (Laohachai 2017). Aim of this study is to combine these training modalities and show their long-term impact on cardiopulmonary capacity and quality of life.

**Methods:** Patient above 8 years of age with Fontan circulation were consecutively included in a 24-months training program. They were assessed by reviewing their medical history, routine cardiac examination, cardiopulmonary exercise testing, bodyplethysmography with measurement of inspiratory muscle strength and quality of life questionnaires. The training consisted of home-based bicycle-ergometer training 90 min per week in 3 to 6 training sessions; workload was individually set to 55% of the pre-training maximum values. The second training modality is daily inspiratory muscle training (using a PowerBreathe medic® device). Compliance, heart rate and activity are measured by Fitbit® wrist activity trackers and training journals. Follow-up visits are planned at 4 monthly intervals.

**Results:** 20 patients were included. After 4 months of training the follow-up data was analyzed: age 10- 43 [mean 20.8] years, 62.5 % male, 50 % adults. No complications occurred. There were no dropouts. Mean peakVO<sub>2</sub> improved from 24.8 to 27.6 ml/min/kg (+2.8ml/min/kg, +14.5 %), p=0.029. Mean maximum workload improved from 99.1 to 119.6 Watts (+20.5 Watts, +24.0%), p=0.001. Mean maximum inspiratory pressure (MIP) improved from 6.27 kPa to 8.53 kPa (+2.26 kPa, + 40.9 %) p=0.03.

**Conclusion:** After 4 months of training and having enrolled 20 patients, we can state that the interest in an individual home-based physical training program is high and that home-based training in Fontan patients is safe and shows good compliance. The data shows excellent effects on cardiopulmonary capacity and inspiratory strength.