

**The Fontan circulation over time: attrition or natural decline of aging? A cross sectional study of functional status in Fontan patients**

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**Introduction:** The nature of the impaired functional status of Fontan patients is far from unraveled, but is believed to be related to impaired cardiac function due to abnormal ventricular loading conditions, and to restricted pulmonary function. Furthermore, most studies reported the absolute value of peak oxygen uptake (VO<sub>2</sub>index) during exercise, whereas the peak VO<sub>2</sub> as percentage of predicted could provide important information on how functional performance of the Fontan patients over time compares to healthy individuals. This study aims to investigate the functional status in a cohort of Fontan patients, and to identify its determinants, including cardiac characteristics, pulmonary characteristics and time since Fontan completion.

**Methods and results:** Eighty-five consecutive Fontan patients  $\geq 10$  years who performed adequate cardiopulmonary exercise testing (respiratory exchange ratio  $> 1.01$ ) in 2012 or 2013, were included. Mean time since Fontan completion was  $15 \pm 9$  years (range 2 - 37 years). New York Heart Association Functional Class (NYHA-FC) was I in 36 patients (42%), II in 41 patients (48%) and III in 8 patients (9%). Peak oxygen uptake during exercise (VO<sub>2</sub>index) was  $25.7 \pm 7.9$  ml/min/m<sup>2</sup> ( $58 \pm 14\%$  of predicted). NYHA-FC and peak VO<sub>2</sub>index both correlated with time since the Fontan operation, but peak VO<sub>2</sub> as percentage of predicted did not. In multivariate analyses, peak VO<sub>2</sub> as percentage of predicted was independently associated with maximum heart rate, oxygen pulse at peak exercise and forced expiratory volume in 1 second (Rsquare=0.579), but not with cardiac output in rest.

**Conclusions:** In Fontan patients, functional status is restricted compared to normal values, already early after completion of the Fontan circulation. The decrease in peak exercise capacity with longer time since the Fontan operation appears to be comparable to natural decline of aging. Additionally, functional status in Fontan patients appears to be related with pulmonary function and cardiac functional parameters during exercise, but not with conventional cardiac measurements at rest.