Long-term functional capacity in patients with Extracardiac Fontan Circulation

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Objectives: thanks to advances in cardiac surgery life expectancy of patients with congenital heart disease has considerably grown. Aim of our study was to evaluate functional capacity in two groups of patients who underwent an extracardiac Fontan operation in the last 25 years (N=135). There are few available data on long-term results of these patients.

Methods: we have divided the sample into two subgroups on the basis of months of follow-up from Fontan procedure. All patients were tested with maximal cardiopulmonary exercise test (CPET) with maximal oxygen uptake (VO2 max). Data from time of effort (TE), maximal heart rate (HR), maximal blood pressure (BP) and VO2max were also calculated as a percentage of normal predicted values. Lung function test to measure forced vital capacity (FVC), forced expiratory volume in 1 sec (FEV1), and peak expiratory flow (PEF) was also performed.

Results: Group A (N= 77, male =48, female=29) months after surgery: 180-60. Group B (N =58, male 32, female 26) months after surgery: 180-300. No statistically significant differences were found in TE% (58% group A vs 56% group B). We found a tendency, although not statistically significant, in decreasing in VO2 max and in PEF between groups (group A: VO2 max 28.5 ml/kg/min + 5.6. PEF 84%. Group B VO2 max 25. ml/kg/min + 5.3. PEF 79%).

Conclusions: We can speculate that, although exercise tolerance in patients underwent extracardiac Fontan is greatly reduced compared with healthy controls, there is no evidence, however, of a significant deterioration of functional capacity over the years. It seems rather possible that the completion of this surgical procedure is leading to a gradual improvement of cardiorespiratory parameters of these patients. Further studies are needed to confirm our hypotheses.