Impact of Pulsatile Pulmonary Blood Flow on Cardiopulmonary Exercise Performance long after the Fontan procedure

Objective: To evaluate the results of exercise capacity test in patients following atriopulmonary connections (Fontan–Kreutzer), atrioventricular connections (Fontan–Björk) and total cavopulmonary connection (TCPC).

Methods: A total of 229 patients who performed exercise capacity test at least one time after the Fontan procedure between 1979 and 2007 were included. Patients after Fontan-Björk procedure were divided into two groups according to the pulmonary blood flow (PBF) pattern: patients with pulsatile PBF and those without. Peak oxygen uptake (VO2) was measured and age- and sex-related reference values (percent-predicted peak VO2) were calculated.

Results: The types of the Fontan procedure included Fontan-Kreutzer in 50 patients, Fontan-Björk in 38 patients (11 with pulsatile PBF and 27 without pulsatile PBF), and TCPC in 141 patients. Median age at the Fontan procedure was 4.5 (IQR: 2.1-8.2) years (Fontan-Kreutzer: 7.8 (5.1-13.1), Fontan-Björk: 5.9 (2.2-7.9), and TCPC: 2.9 (1.9-6.5) years). Median follow-up was 20.0 (16.0-24.4) years. A total of 978 cardiopulmonary exercise tests were performed at median follow up of 17.7 (11.3-23.4) years postoperatively. Yearly distributions in percent peak VO2 in individual patients are shown in Figure. Percent-predicted peak VO2 was higher in patients with pulsatile PBF after Fontan-Björk (76.1 (64.0-83.9) %) compared to patients without pulsatile PBF after Fontan-Björk (59.0 (46.1-69.5) %, p<0.001), to patients after Fontan-Kreutzer (62.2 (51.5-71.3) %, p<0.001) and to patients after TCPC (66.0 (56.0-79.3) %, p=0.026). Percent-predicted peak VO2 was negatively correlated with the follow-up period in patients with pulsatile PBF after Fontan-Björk procedure, (R=0.410, p<0.01), and in patients after TCPC (R=0.228, p<0.01), but was not correlated with the follow-up period in patients without pulsatile PBF after Fontan-Björk (R=0.046, p=0.569) and in patients after Fontan-Kreutzer (R=0.115, p=0.084).

Conclusions: In long-term survivors after various types of Fontan procedure, patients with pulsatile pulmonary blood flow after the Fontan-Björk procedure demonstrated a better exercise performance, compared to those after TCPC, those after Fontan-Kreutzer procedure, and those after the Fontan-Björk procedure with non-pulsatile pulmonary blood flow. The results implicate the importance of pulsatile pulmonary flow to maintain the Fontan circulation.

Lukas Klemm (1), Paul Philipp Heinisch (1), Thibault Schaeffer (1), Helena Staehler (1), Alfred Hager (2), Peter Ewert (2), Jürgen Hörer (1), Masamichi Ono (1), (1) Department of Congenital and Pediatric Heart Surgery, German Heart Center Munich, Munich, Germany, (2) Department of Pediatric Cardiology and Congenital Heart Disease, German Heart Center Munich, Munich, Germany