

MP2-4

A restrictive ventilatory pattern is common in patients with univentricular heart after Fontan palliation and associated with a reduced exercise capacity and a reduced quality of life. A European multicenter retrospective cross-sectional study with 232 patients.

Callegari A. (1, 2), Neidenbach R. (2), Milanese O. (1), Castaldi B. (1), Ono M. (3), Mueller J. (2), Ewert P. (2), and Hager A. (2)

(1) Department of Women's and Children's Health, Pediatric Cardiology, University of Padova, Italy

(2) Department of Pediatric Cardiology and Congenital Heart Disease, Deutsches Herzzentrum München, Technische Universität München, Munich, Germany

(3) Department of Cardiovascular Surgery, Deutsches Herzzentrum München, Technische Universität München, Munich, Germany

Objectives: The Fontan circulation is highly dependent on ventilation, which improves pulmonary blood flow and cardiac output. A reduced ventilatory function was reported in these patients. This analysis investigates the extent of this impairment and its relation to exercise capacity and quality of life.

Methods: This multicenter retrospective cross-sectional study included 232 patients (140 females, 92 males, 25.6±10.8 years old) with univentricular heart after Fontan palliation (19.8% atrioventricular connection; 20.3% atriopulmonary connection; 59.9% total cavopulmonary connection). Resting spirometry, cardiopulmonary exercise tests, and health-related quality-of-life assessment (SF-36) in those 14 years or older were performed between November 2003 and May 2015.

Results: FEV1 was 74.7±17.8%pred. and FVC 71.1±16.9%pred. In those 58.3% of patients with a FEV1 <80%pred., all had a normal FEV1/FVC>70% suggestive of a restrictive ventilatory pattern. The reduced FEV1 was associated with a reduced peakVO₂ of 67.0±17.6%pred. (r=0.43, p<0.0001), even if analyzed together with possible confounding factors (sex, BMI, age, years after Fontan palliation, total number of interventions, presence of scoliosis, diaphragmatic paralysis, and PLE).

Synergistically to exercise capacity, FEV1 was weakly associated to quality of life in the aspects of physical functioning (r=0.25, p=0.008), bodily pain (r=0.22, p=0.02), and general health (r=0.16, p=0.024).

FEV1 itself was associated to diaphragmatic paralysis (p=0.001), scoliosis (p=0.001), high total number of interventions (p=0.002), and reduced BMI (p=0.01). No correlation was found with ventricular morphology, timing/type of surgeries, catheter and echo data, and others peri-operative or long-term complications.

Conclusions: The common restrictive ventilatory pattern in patients with a Fontan circulation has a substantial impact on exercise capacity and quality of life. Risk factors are diaphragmatic paralysis, scoliosis, a high total number of interventions, and cachexia.