

Factors associated with exercise capacity in patients with a systemic right ventricle

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Background: Systemic right ventricle (SRV) is a rare and complex congenital heart disease (CHD). Patients with SVR present a significant decrease of their exercise capacity but the prediction of this decline in clinical practice is still challenging.

Aims: We aimed at identifying clinical and paraclinical factors associated with maximum oxygen uptake (VO₂max) in adults with SRV.

Methods: We performed a multicentre cross-sectional study from January to December 2017 in three French tertiary CHD centres. All adults with D-transposition of the great arteries (d-TGA) and congenitally corrected TGA (cc-TGA) were included. Demographic, clinical, laboratory and imaging data were collected. Univariate and multivariate analyses were performed to identify predictors of impaired VO₂max, as measured by cardiopulmonary exercise test (CPET).

Results: A total of 111 patients were included in the study (85% d-TGA, median age 37.2 ± 8.2 years). Nearly 2/3 (n=70) of the patients presented with various conduction disorders and 1/3 (n=34) with cardiac rhythm disorders. Median B-type natriuretic peptide (BNP) was 58.5 [10-995] pg/ml. All echocardiographic parameters showed at least a mild SRV dysfunction and 17% of the patients presented with severe tricuspid valve regurgitation. VO₂max was impaired in all patients (mean 23.3 ± 6.9 ml/kg/min, representing 42.0 ± 7.4% of predicted values). In univariate analysis, NYHA functional class, BNP levels, right ventricular dysfunction, severity of tricuspid valve regurgitation, and the presence of a pacemaker and/or an implantable defibrillator, were correlated with VO₂max. In multivariate analysis, only the patient's self-assessment of functional status, as measured by NYHA functional class was correlated with VO₂max (P=0.005).

Conclusion: NYHA functional class is the strongest predictor of impaired exercise capacity in adult patients with SRV.

Keywords: systemic right ventricle, cardiopulmonary exercise test, NYHA functional class, congenital heart disease.