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Mechanisms underlying abnormal cardiorespiratory response in Fontan patients

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BACKGROUND : Low peak oxygen consumption (peak VO₂) is a strong predictor of mortality in adults with congenital heart disease. In univentricular hearts after Fontan procedure, peak VO₂ is low at 59% of predicted value for the 50th percentile. But the mechanisms underlying this abnormal cardiorespiratory response to exercise are not well established.

OBJECTIVES : We aimed to determine which parameters can predict low peak VO₂ (<59% of predicted value) in Fontan patients.

METHODS : Twenty one patients (mean age 23 ± 8 years) including 13 univentricular hearts, 5 hypoplastic left heart syndromes, 2 tricuspid atresia and 1 congenitally corrected transposition of the great arteries prospectively underwent cardiopulmonary exercise testing (CPX) with arterial blood gas measurements.

RESULTS : Peak VO₂ was low as expected (24.4 ml/kg/min). All patients despite normal oxygen saturation, had shunt with PaO₂+PaCO₂ <120 mmHg (mean 96 ± 8 mmHg) at rest. Twelve patients had peak VO₂ <59% of predicted value, which is considered <50th percentile in Fontan population. In this group, resting oxygen saturation was lower (94.3 ± 1.8% vs 96.7 ± 1.4%, p=0.02), alveolar-arterial gradient was higher at rest (52 ± 9 mmHg vs 44 ± 5 mmHg, p=0.03) and at exercise (58 ± 10 mmHg vs 50 ± 6 mmHg, p=0.04). Secondly, these patients had lower exercise heart rate (122 ± 38 bpm vs 169 ± 25 bpm, p=0.002), lower chronotropic reserve (49 ± 31 bpm vs 79 ± 17 bpm, p=0.009) and had more often pacemakers (3 patients versus 0, p=0.05). Finally, in this group with altered peak VO₂, we found more Fontan procedures (or modified Fontan) than extracardiac total cavopulmonary connections (5 patients or 42% vs 1 or 11%, p=0.03), patients were older (27.6 ± 7 years vs 16.8 ± 6.4 years, p=0.0009), and converted to extracardiac cavopulmonary connection later (at 21.1 ± 13.9 years old vs 3.8 ± 2.4 years old, p=0.002).

CONCLUSION : Severe alteration of aerobic capacity evaluated by peak VO₂ measurement in Fontan patients is correlated to the severity of cyanosis, the incapacity to rise heart rate at exercise and the type of surgery. Assessment of aerobic capacity with arterial blood gas measurements seems worthwhile to understand abnormal cardiorespiratory response in this population.