Clinical applications of exercise pathophysiology in patients with congenital heart disease

Abstract

The aim of this thesis is to evaluate the data from cardiopulmonary exercise testing and their interpretation in relation to novel predictors of morbidity and mortality in patients with complex congenital heart disease (CHD). Of the entire broad spectrum of CHD, patients with total cavopulmonary connection have the most altered exercise capacity, which results form a plethora of exercise pathophysiological components characteristic of this form of circulation. The theoretical part focuses on stress testing in patients with complex congenital heart disease and summarizes the main pathophysiological mechanisms affecting exercise tolerance in patients with TCPC. The practical part is devoted to the evaluation of the clinical profile of TCPC patients with excellent functional outcomes (Super-Fontan), the possibilities of retrospective evaluation of skeletal muscle mass from computed tomography and cardiac magnetic resonance imaging in these patients. It also discusses the relationship between oxygen delivery, systemic ventricular function measured by magnetic resonance imaging and peak oxygen utilization during exercise stress testing. In the cohort of patients with the Super-Fontan phenotype, we found a lower mortality risk, lower body mass index, larger preoperative pulmonary artery dimensions, a greater proportion of women and patients with tricuspid atresia. We demonstrated a higher mortality risk in TCPC patients with low skeletal muscle mass as measured by muscle cross-sectional area on computed tomography chest examination. There was positive correlation between muscle cross-sectional area on magnetic resonance imaging and exercise tolerance. Evaluation of muscle mass index should be transferred to routine clinical practice in the long-term follow-up of patients with congenital heart disease. New insights into the role of contractile function of the systemic ventricle may help in the differential diagnosis and management of TCPC patients with exercise intolerance.

Keywords: Congenital heart disease, exercise tolerance, magnetic resonance imaging, muscle mass, oxygen delivery, oxygen uptake, total cavopulmonary connection, cardiopulmonary exercise testing, Super-Fontan