Predictors of morbidity and mortality in contemporary Fontan patients: results from a multicenter study including cardiopulmonary exercise testing in 321 patients.

Abstract:
Previous studies have established an association between exercise intolerance and increased morbidity and mortality in congenital heart disease patients. We aimed to clarify if exercise intolerance is associated with poor outcome in Fontan patients and to identify risk factors for mortality, transplantation, and cardiac-related hospitalization. A total of 321 Fontan patients (57% male, mean age 20.9 ± 8.6 years) who underwent cardiopulmonary exercise testing (CPET) at four major European centres between 1997 and 2008 were included. During a median follow-up of 21 months, 22 patients died and 6 patients underwent cardiac transplantation (8.7%), resulting in an estimated 5-year transplant-free survival of 86%. Parameters of CPET were strongly related to increased risk of hospitalization, but-with the exception of heart rate reserve-unrelated to risk of death or transplantation. In contrast, patients with clinically relevant arrhythmia had a 6.0-fold increased risk of death or transplantation (P < 0.001). Furthermore, patients with atrioventricular Fontan had a 3.7-fold increased risk of death or transplantation compared with total cavopulmonary connection patients (P = 0.009). The combination of clinically relevant arrhythmia,
atriopulmonary/-ventricular Fontan, and signs of symptomatic or decompensated heart failure was associated with a particularly poor outcome (3-year mortality 25%). On short-term follow-up, most parameters of CPET are associated with increased risk of hospitalization but not death or transplantation in contemporary Fontan patients. Only decreased heart rate reserve and a history of clinically relevant arrhythmia, atrio pulmonary/-ventricular Fontan, and/or heart failure requiring diuretic therapy are associated with poor prognosis, potentially identifying patients requiring medical and/or surgical attention.

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