



ORIGINAL ARTICLE

Congenital heart disease in adults: Assessment of functional capacity using cardiopulmonary exercise testing



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KEYWORDS

Adult congenital heart disease;
Functional capacity;
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Abstract

Aim: The aim of the study was to compare functional capacity in different types of congenital heart disease (CHD), as assessed by cardiopulmonary exercise testing (CPET).

Methods: A retrospective analysis was performed of adult patients with CHD who had undergone CPET in a single tertiary center. Diagnoses were divided into repaired tetralogy of Fallot, transposition of the great arteries (TGA) after Senning or Mustard procedures or congenitally corrected TGA, complex defects, shunts, left heart valve disease and right ventricular outflow tract obstruction.

Results: We analyzed 154 CPET cases. There were significant differences between groups, with the lowest peak oxygen consumption (VO_2) values seen in patients with cardiac shunts (39% with Eisenmenger physiology) ($17.2 \pm 7.1 \text{ ml/kg/min}$, compared to $26.2 \pm 7.0 \text{ ml/kg/min}$ in tetralogy of Fallot patients; $p < 0.001$), the lowest percentage of predicted peak VO_2 in complex heart defects ($50.1 \pm 13.0\%$) and the highest minute ventilation/carbon dioxide production slope in cardiac shunts (38.4 ± 13.4). Chronotropic impairment was impaired in patients with complex defects. Eisenmenger syndrome ($n=17$) was associated with the lowest peak VO_2 (16.9 ± 4.8 vs. $23.6 \pm 7.8 \text{ ml/kg/min}$; $p=0.001$) and the highest minute ventilation/carbon dioxide production slope (44.8 ± 14.7 vs. 31.0 ± 8.5 ; $p=0.002$). Age, cyanosis, CPET duration, peak systolic blood pressure, time to anaerobic threshold and heart rate at anaerobic threshold were predictors of the combined outcome of all-cause mortality and hospitalization for cardiac cause.

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Conclusion: Across the spectrum of CHD, cardiac shunts (particularly in those with Eisenmenger syndrome) and complex defects were associated with lower functional capacity and attenuated chronotropic response to exercise.

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PALAVRAS-CHAVE

Cardiopatias congénitas do adulto; Capacidade funcional; Prova de esforço cardiorrespiratória

Cardiopatia congénita em adultos: avaliação da capacidade funcional por prova de esforço cardiorrespiratória

Resumo

Objetivo: Comparar a capacidade funcional nas cardiopatias congénitas, avaliada por prova de esforço cardiorrespiratória.

Métodos: Análise retrospectiva dos doentes adultos com cardiopatia congénita, submetidos a prova de esforço cardiorrespiratória. Os doentes foram divididos em tetralogia de Fallot operada, transposição de grandes artérias após cirurgia de Senning/Mustard, transposição de grandes artérias congenitamente corrigida, defeitos complexos, shunts, doença valvular esquerda e obstrução do trato de saída do ventrículo direito.

Resultados: Foram avaliadas 154 provas cardiorrespiratórias. Os valores mais baixos de consumo de oxigénio no pico foram observados nos doentes com shunt cardíaco (39% apresentavam síndrome de Eisenmenger) ($17,2 \pm 7,1 \text{ ml/kg/min}$, em comparação com $26,2 \pm 7,0 \text{ ml/kg/min}$ na tetralogia de Fallot; $p < 0,001$); o valor mais baixo da percentagem de consumo de oxigénio no pico relativamente ao previsto foi observado nos defeitos complexos ($50,1 \pm 13,0\%$) e o maior valor de rampa ventilação minuto/produção de dióxido de carbono nos shunts cardíacos ($38,4 \pm 13,4$). O cronotropismo foi menos eficaz nos doentes com defeitos complexos. A síndrome de Eisenmenger ($n = 17$) associou-se ao valor mais baixo de consumo de oxigénio no pico ($16,9 \pm 4,8$ versus $23,6 \pm 7,8 \text{ ml/kg/min}$; $p = 0,001$) e ao maior valor de rampa ventilação minuto/produção de dióxido de carbono ($44,8 \pm 14,7$ versus $31,0 \pm 8,5$; $p = 0,002$). Idade, cianose, duração da prova, pressão arterial sistólica no pico, tempo para o limiar anaeróbico e frequência cardíaca no limiar anaeróbico foram preditores do outcome combinado com mortalidade de todas as causas e hospitalização de causa cardíaca.

Conclusão: Os shunts cardíacos (particularmente com síndrome de Eisenmenger) e os defeitos complexos associaram-se a menor capacidade funcional e resposta cronotrópica atenuada ao exercício.

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Introduction

Nowadays most patients with congenital heart disease (CHD) are expected to reach adulthood. Because exercise intolerance has been documented at all ages of CHD, these patients need close follow-up and an objective assessment of functional capacity.^{1,2}

Due to long-term adaption, the majority of adult patients with CHD self-report their exercise capacity status as satisfactory, even in the presence of significantly depressed functional status. Cardiopulmonary exercise testing (CPET) is an accurate method for quantitative assessment of exercise capacity, including assessment of aerobic capacity, chronotropic response and arrhythmias.^{1,3–6} Quantifying exercise capacity by measuring parameters such as peak oxygen consumption (VO_2) is an established technique in the management of patients with chronic heart failure. However, in adult CHD patients its role has been much less studied, and interpretation of test results remains a

challenge. Previous studies have demonstrated that CPET data have an important influence on the treatment approach in CHD, including indication for cardiac transplantation, and on prognosis.^{2,7,8}

The aim of the present study was twofold: to assess and compare functional capacity in different CHD groups, measured objectively by CPET, and to investigate a possible association between CPET parameters and outcome.

Methods

Study design

A retrospective analysis was performed of consecutive adult patients with CHD who underwent CPET for assessment of functional capacity. The data were collected in a single tertiary center between March 2009 and June 2015.

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