



## EDITORIAL COMMENT

# The role of cardiopulmonary exercise testing in decision-making in adults with congenital heart disease



## O papel do teste de exercício cardiopulmonar na tomada de decisão em adultos com cardiopatia congénita

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Worldwide, there are now more adults affected with congenital heart disease (CHD) than children.<sup>1</sup> Surgical advances and improvements in diagnosis and medical care have reduced early mortality and most patients now reach adulthood.<sup>2</sup> The increasing longevity of this population has led to concerns about their well-being and quality of life.<sup>2</sup> Long-term complications are common, with heart failure becoming a significant cause of exercise intolerance, morbidity and mortality.

Assessment of CHD patients' functional state is important for prognostic reasons and, as expected, individuals with CHD have lower functional capacity than their peers, even including patients who have undergone corrective surgery. However, such assessments should be treated with caution, as they may underestimate the degree of limitation in adult patients with long-standing disease who have adapted their lifestyle to their ability.

Cardiopulmonary exercise testing (CPET) is an accurate tool for quantifying exercise capacity and can be used to assess abnormal cardiovascular response to increased

pressure or volume demands. However, the ideal tool for risk stratification would provide early signs of insidious subclinical heart failure, when the progressive disease is still reversible.<sup>3</sup>

In this issue of the *Journal*, Rosa et al.<sup>4</sup> review the role of CPET as a potential tool for risk stratification, aiming to identify adults with different types of CHD and impaired cardiovascular function. Patients with left and right pressure or volume overload were selected. The aim of the study was to assess and compare functional capacity in the different patient groups and to investigate a possible association between CPET parameters and outcome. There were significant differences in CPET parameters between the study groups. Cyanosis and pulmonary hypertension conferred poor exercise tolerance and an increased risk of hospitalization and mortality. The progressive failure of a systemic right ventricle has deleterious effects on short- and long-term outcomes for all patients. The right ventricle, with its complex geometry and varied adaptive mechanisms in CHD when in the systemic position, must by itself sustain the systemic load.<sup>5</sup> This is the physiology of transposition of the great arteries with previous atrial switch repair, mostly seen in older patients, or congenitally corrected transposition of the great arteries, operated or native, double-inlet right ventricle and hypoplastic left heart syndrome.

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The outcome of patients with CHD has improved in recent years due to advances in early diagnosis and treatment, particularly cardiac surgery and intensive care procedures. Earlier interventions may reverse ventricular remodeling and improve long-term benefits. CPET has diagnostic and prognostic value in CHD. However, before randomized trials can be designed, further research is necessary. More routine use will play an important role in improving risk stratification of adults with CHD.

### Conflicts of interest

The author has no conflicts of interest to declare.

### References

1. Marelli AJ, Ionescu-Ittu R, Mackie AS, et al. Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010. *Circulation*. 2014;130:749–56.
2. Coelho R, Teixeira F, Silva AM, et al. Psychosocial adjustment, psychiatric morbidity and quality of life in adolescents and young adults with congenital heart disease. *Rev Port Cardiol*. 2013;32:657–64.
3. Dallaire F, Wald R, Marelli A. The role of cardiopulmonary exercise testing for decision making in patients with repaired tetralogy of Fallot. *Pediatr Cardiol*. 2017;38:1097–105.
4. Rosa SA, Agapito A, Soares RM, et al. Congenital heart disease in adults: assessment of functional capacity using cardiopulmonary exercise testing. *Rev Port Cardiol*. 2018;37:399–405.
5. Brida M, Diller G, Gatzoulis M. Systemic right ventricle in adults with congenital heart disease: anatomic and phenotypic spectrum and current approach to management. *Circulation*. 2018;137:508–18.