Exercise Physiology and Testing in Adult Patients with Congenital Heart Disease

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KEYWORDS

• Exercise physiology • Adults • Congenital heart diseases • Exercise testing • Exercise training

KEY POINTS

- Adult patients with congenital heart diseases comprise a heterogeneous patient population whose altered cardiovascular, pulmonary, and skeletal muscle physiology may contribute to their overall reduced exercise capacity.
- Formal exercise testing provides an important and excellent objective evaluation of patient physiology and exercise capacity.
- Specific congenital cardiac conditions and types of repair are associated with somewhat characteristic alterations of exercise physiology and the treating physician should be familiar with these distinctive patterns.
- In many conditions the degree of exercise intolerance can have significant prognostic implications.
- The role of exercise training to improve the exercise capacity and the overall survival of ACHD patients is an area of interest to many and may be part of the brighter future that awaits this patient population.

The outstanding improvement in the long-term survival of patients with congenital heart disease (CHD) is perhaps one of modern medicine's greatest triumphs. Many individuals with congenital cardiac conditions that were considered nontreatable merely 4 or 5 decades ago are now adults. As the number of individuals with CHD who survive into adulthood increases, the interest in their unique cardiovascular physiology and long-term outcomes grows and promotes vast research.

One of the early observations made by investigators of this field was that despite improvements in survival for adult patients with congenital heart disease (ACHD), these patients commonly had impaired exercise capacity. Many factors have been identified to explain this phenomenon, including residual cardiovascular defects, deconditioning, repeated surgical interventions, cardiac device implantations, multidrug therapy, and alterations in the anatomy and physiology of organ systems other than the cardiovascular system, such as the pulmonary and the musculoskeletal systems.

Regardless of the underlying mechanisms, exercise intolerance is perhaps the single most important factor responsible for the impairment in the quality of life of ACHD patients. Even simple lesions that are easily addressed at surgery or via transcatheter solutions may be associated with limited exercise capacity. Interestingly, it is common for ACHD patients' self-perception of their exercise intolerance to be unrealistically optimistic. This

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observation has been ascribed to the fact that most of them have impaired exercise tolerance that dates back to their early childhood, causing a distorted perception of what constitutes "normal" exercise capacity. Indeed, a recent study of asymptomatic young ACHD found that their exercise capacity, when assessed with formal cardiopulmonary exercise testing (CPET), was comparable to much older adults with equivalent functional class and congestive heart failure (CHF) secondary to acquired heart disease.[1] Formal exercise testing is therefore currently considered to be an important component of the routine evaluation of ACHD patients, because it can reliably and objectively assess exercise capacity and provide valuable risk stratification data.

IMPAIRED EXERCISE TOLERANCE IN ACHD: PREVALENCE AND SPECIAL CONSIDERATIONS

Effort intolerance is experienced by patients from across the CHD spectrum, ranging from simple lesions such as mild left ventricular outflow tract obstructive lesions to complex conditions such as single ventricle physiology (**Table 1**). When CPET was performed in a large cohort of ACHD patients with various conditions, on average, peak oxygen consumption (V_{O2}) was found to be reduced in all examinees regardless of the baseline disease as compared with healthy individuals of similar age.[1] As can be appreciated from **Table 1**, different degrees of exercise intolerance do exist within the CHD spectrum, and the general rule is that there is a good correlation between the severity of the underlying cardiac defect or the completeness of its repair and the degree of exercise intolerance.[1–4] ACHD patients with Eisenmenger physiology experience the most severe form of exercise intolerance, followed by patients with univentricular physiology and cyanosis.[4]

In normal adult subjects, exercise function tends to deteriorate with age; peak V_{O2} has been found to decline ~0.5% to 0.6% per year after age 21.[5–8] Studies in various ACHD populations have found that this decline tends to progress more rapidly, although observational data suggest that engaging in frequent physical activity may favorably alter this trend.[9]

This observation may be particularly relevant to patients with a systemic right ventricle, such as patients with L-loop transposition of the great arteries (L-loop TGA) or with D-loop TGA who underwent an atrial switch operation (either the Senning or the Mustard procedures). In these patients, severe exercise intolerance often manifests itself in the third or fourth decade of life.[10,11] In fact, almost two-thirds of patients with atrial repairs of D-loop TGA who have other associated defects or prior surgical intervention experience heart failure by the age of 45. The common explanation for this delayed presentation is failure of the systemic right ventricle and progressive insufficiency of the systemic atrioventricular valve, both of which tend to progress slowly over the years.[12] Chronotropic

Table 1
Cardiopulmonary test abnormalities encountered in patients with various congenital and acquired heart diseases

Defect	↓ Peak V_{O2}	↓ Peak HR	↓ O_2 Pulse	↑ V_E/V_{CO2}	↓ VT
Repaired TOF/Truncus	+++	++	+++	+++	++
Fontan	++++	+++	++++	++++	+++
PVOD	++++	+	++++	++++	++++
Ebstein anomaly	+++	++	+++	++	++
D-TGA S/P atrial switch	+++	++	+++	++	++
Aortic valve disease	++	+	++	+	++
Coarctation	++	+	++	+	+++
DCM	++++	+	++++	++	++++
HCM	++	+	++	+	++
Isolated PR	+	+	+	+	+

Note: This table assumes that the patient is not receiving beta-blocker or other anti-arrhythmic therapy that might impair the chronotropic response to exercise.

Abbreviations: +, implies rarely present; ++, sometimes present; +++, often present; ++++, usually present; DCM, dilated cardiomyopathy; HCM, hypertrophic cardiomyopathy; peak HR, heart rate at peak exercise; Peak V_{O2}, oxygen consumption at peak exercise; PR, pulmonary regurgitation, postvalvuloplasty; PVOD, pulmonary vascular obstructive disease; TOF, tetralogy of Fallot; Truncus, Truncus arteriosus; V_E/V_{CO2}, slope of the linear portion of minute ventilation vs carbon dioxide production curve.