

## President's Page

# Treating Heart Failure in Adults with Congenital Heart Disease: Lessons from the Left Ventricle

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**E**xercise intolerance and heart failure are frequent complications in adults with congenital heart disease (ACHD). Reduced exercise capacity, as assessed objectively, is prevalent even among those ACHD who consider themselves asymptomatic. When treating a patient with ACHD and symptoms of heart failure most cardiologists would apply their knowledge of acquired left ventricular heart failure. But is this strategy “evidence-based”?

Similarities in the pathophysiology of exercise intolerance in acquired heart failure and congenital heart disease suggest that established heart failure therapies might be beneficial to ACHD patients with exercise intolerance. For instance, renal dysfunction, anaemia and hyponatraemia are prevalent in ACHD, have a significant impact on clinical outcome and, therefore, should be taken into account when risk stratifying these patients.<sup>1-2</sup> Neurohormonal activation is also an important pathophysiological mechanism and levels of natriuretic peptides are a strong predictor of prognosis in ACHD.<sup>3</sup> When assessing symptomatic ACHD patients, special attention should be paid to any anatomical and haemodynamic corrections that may lead to improved haemodynamics and symptomatology. For example, severe pulmonary regurgitation causing right ventricular volume overload is an indication for pulmonary valve replacement in symptomatic patients with tetralogy of Fallot, while tricuspid valve replacement should not be delayed in congenitally corrected transposition of the great arteries with severe tricuspid valve regurgitation, before the systemic right ventricle starts to fail irreversibly.

Data regarding the pharmacological management of the dysfunction of a systemic right ventricle or a

single ventricle are limited, and current practice tends to incorporate traditional therapies that are known to improve systemic left ventricular dysfunction (angiotensin-converting enzyme inhibitors, beta-blockers, diuretics). Initial findings from clinical studies were discouraging; however, insights gained into potentially different pathophysiological mechanisms that underlie the failing systemic left ventricle versus right (or single) ventricle may ultimately lead to more appropriate tailored therapy. On the other hand, designing a prospective randomised clinical study to assess the impact of medical therapy on patients with complex congenital heart disease is greatly hampered by the limited number of such patients and the heterogeneity of their underlying congenital heart defects. In general, randomised trials in ACHD present difficulties at all stages, from design to recruitment to completion.<sup>4</sup>

Currently, in most expert centres the presence of systemic ventricular dysfunction with a prolonged QRS duration would suggest that multi-site pacing should be tried. While there is increasing evidence that ventricular dyssynchrony is present in ACHD, randomised trials of resynchronisation in this population are lacking. Finally, the scarcity of donors, the very slow clinical deterioration of ACHD and the often complex cardiovascular anatomy make the role of heart and/or lung transplantation relatively limited, resulting in the very small number of patients who finally receive a transplant.

ACHD represent a heterogeneous patient population, who have already outscored their paediatric counterparts. As these patients grow up, general cardiologists are likely to face with increasing frequency the complications of aging, such as cancers and coro-

nary artery disease.<sup>5</sup> Exercise intolerance and heart failure should be sought out and treated aggressively, and there is a great need for these patients to be enrolled in large multi-centre clinical trials.

## References

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