Adult Congenital Interventions in Heart Failure

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KEYWORDS

• Adult congenital heart disease • Heart failure • Congenital interventions

KEY POINTS

- The advances in medical and surgical therapies for children with congenital heart disease (CHD) have resulted in a growing population of patients reaching adulthood, with survival rates exceeding 85%.
- Many of these patients, especially if they are managed inappropriately, face the prospect of future complications, including heart failure and premature death.
- Transcatheter interventions have evolved over the past decades to become the primary treatment for many forms of CHD.
- In this article, we discuss the role of transcatheter interventions in the treatment of heart failure in adults with CHD.

INTRODUCTION

Congenital heart disease (CHD) is present in 0.8% to 1.0% of live births, and the vast majority of these patients are diagnosed and treated in infancy or childhood. The advances in medical and surgical therapies for children with CHD have resulted in a growing population of patients reaching adulthood, with survival rates exceeding 85%[1]. As a result, there are now an estimated 1.2 million patients included in this group in the United States.[2] This has created a well-established field on its own, the adult with CHD (ACHD), that serves as a fundamental component of any center providing care for these patients.[3] Further, board certification is now offered in this unique subspecialty.

ACHDs form 3 distinct groups: those with lesions that have not been previously diagnosed (new diagnoses); those who were treated appropriately (corrected), however, they require lifelong follow-up; and those who have had palliative procedures and also require lifelong follow-up. The first group is a rapidly diminishing group, as most forms of CHD are routinely diagnosed and treated in infancy or childhood. The most common diagnosis in this group is secundum atrial septal defect (ASD). The second group is rapidly expanding, as the children with corrected CHD reach adolescence and adulthood. Examples of patients in this group include patients following tetralogy of Fallot repair and arterial switch operation. The third group fortunately is also diminishing and

Disclosure Statement: H.S. Suradi: Nothing to disclose. Z.M. Hijazi: Consultant for Occlutech & Venus Medtech.
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Intervent Cardiol Clin 6 (2017) 427–443
http://dx.doi.org/10.1016/j.iccl.2017.03.011

includes patients who had palliative procedures for single ventricle and are not candidates for the Fontan pathway.

In ACHD, heart failure is the ultimate expression of the sequelae and complications that patients with ACHD often face even after successful repair of their primary defect. Exercise intolerance is the main feature of heart failure, affecting more than a third of patients in the Euro Heart Survey, a large registry of patients with ACHD across Europe.[4] Patients with cyanotic lesions tend to be with the highest prevalence of exercise intolerance. Within the cyanotic population, those with significant pulmonary arterial hypertension (Eisenmenger syndrome) tend to be most severely limited. Patients with the right ventricle in the systemic position, either as a result of congenitally corrected transposition of the great arteries or after atrial switch operation (Mustard or Senning procedure) for transposition of the great arteries also tend to become severely limited in their exercise capacity, especially after the third decade of life. Patients with univentricular circulation and a Fontan-type operation are also typically limited in their exercise capacity, especially in the presence of ventricular dysfunction, atrioventricular valve regurgitation, or a failing Fontan circulation. Even patients with simple lesions, such as ASD, often present with reduced exercise capacity, even though often at a later stage.

Myocardial dysfunction is common in ACHD and can be caused by multiple factors. Hemodynamic overload of 1 or both ventricles due to obstructive or regurgitant lesions, shunting, and pulmonary or systemic hypertension is common in ACHD. This long-standing overload can eventually lead to severe ventricular dysfunction. Right ventricular systolic dysfunction is common in patients with significant volume overload, such as those with large ASDs or patients with tetralogy of Fallot and severe pulmonary regurgitation. Ventricular dysfunction also can result from repeated cardiac surgeries, anomalous coronary circulation, and abnormal myocardial perfusion. Ventricular-ventricular interaction is not uncommon in ACHD, with right-sided lesions often affecting the left ventricle and vice versa, such as in Ebstein anomaly. Acquired coronary and noncoronary heart disease superimposed to the congenitally abnormal heart also may cause deterioration of myocardial dysfunction, such as coronary atherosclerosis, infective endocarditis, systemic hypertension, myocarditis, and substance abuse. Coronary artery disease always should be suspected in ACHD when ventricular dysfunction is encountered and should be managed accordingly. Medications, permanent pacing, and arrhythmias are other important causes of heart failure in this population. Therefore, identification of the mechanisms responsible for heart failure is essential in the management of patients with ACHD because these can become targets for therapies (Box 1).

Over the past decades, interventional pediatric and adult cardiologists have become increasingly experienced with transcatheter interventional therapy for CHD. The rapid development of successful transcatheter procedures has led to interventional procedures becoming the primary treatment for many forms of CHD. These same techniques are currently being applied to ACHDs with excellent results. In this article, we focus in a defect-specific approach on the role of catheter-based interventions in the treatment of ACHDs.

PREPROCEDURAL ASSESSMENT

All patients with CHD should undergo a comprehensive, multidisciplinary evaluation with close collaboration between the clinical cardiologist, interventionalist, and surgeon. Details of the procedure and the benefits and risks of any anticipated intervention should be discussed with the patient. Alternative options, including surgical treatment, should be discussed and a meeting with a surgeon should be offered if the patient desires. A thorough review of the patient's complete history and physical examination, including all previous pertinent noninvasive studies, cardiac

Box 1
Potential mechanisms of heart failure in adults with congenital heart disease

- Valve disease
- Outflow obstruction
- Shunting
- Volume/pressure overload
- Residual lesions
- Coronary anomalies
- Coronary artery disease
- Prior surgery
- Arrhythmias
- Pericardial disease
- Pacing
- Medications
- Chronotropic incompetence