Management of Heart Failure in Adult Congenital Heart Disease

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KEYWORDS

• Congenital heart disease • Heart failure • Systemic ventricle dysfunction
• Single ventricle dysfunction • Fontan

KEY POINTS

- Children with congenital heart disease (CHD) are outnumbered by adults with CHD (ACHD).
- CHD–heart failure (HF) presentation differs based on anatomy and prior surgical repair.
- HF medical therapy is less well studied in CHD and needs to be considered in the context of CHD-related anatomy or physiology.
- The first step in evaluation of the adult CHD patient with HF is to examine the underlying anatomy for lesions with the possibility for intervention.
- Mechanical circulatory support and heart transplant in CHD is more complex secondary to anatomic limitations. These patients are at a disadvantage in the current allocation system.

INTRODUCTION

In the United States, children living with congenital heart disease (CHD) are outnumbered by adults with CHD (ACHD). Due to surgical and medical advances, it is now estimated that there are more than 1 million adults with CHD in the United States.[1,2] Advances in surgical technique, transcatheter intervention, imaging modalities, and focus on high-quality multidisciplinary care teams has contributed to improved CHD survival. Recent studies have shown that the median age of patients with severe CHF has increased from 11 years in 1985 to 17 years in 2000, and the overall age at death increased from 37 years in 2002 to 57 years in 2007.[3] Improved survival to adult age and late adulthood translates to a population with both cardiac and extracardiac disease, and specialized care needs. In particular, heart failure (HF) is common in the adult patient with CHD. Adults with CHD experience more hospitalizations, episodes of decompensation, and ultimately have higher mortality than non-CHD cohorts.[4–6] Therefore, it is critical to understand the heterogenetic nature of HF in ACHD population, assessment and management across the spectrum of CHD, and treatment options, as well as current gaps in treatments available to this unique group.

THE DISTINCT NATURE OF HEART FAILURE IN ADULT CONGENITAL HEART DISEASE

As CHD patients live to older age, the population becomes even more diverse because it includes

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patients who survived several percutaneous and surgical procedures in childhood, and adults who presented and were diagnosed later in life. Residual anatomic and hemodynamic lesions accompanied by acquired heart disease lead to an increasingly complex group of patients with varied presentation. HF, along with arrhythmia, sudden death, and late vascular complications are the most common late cardiac presentations in adults.[7]

Bolger and colleagues[8] described CHD as the original HF syndrome as "…characterized by a triad comprising cardiac abnormality, exercise limitation, and neurohormonal activation." The Heart Failure Society of America guidelines' define HF as "…a syndrome characterized by either or both pulmonary and systemic venous congestion or inadequate peripheral oxygen delivery, at rest of during stress caused by cardiac dysfunction."[9,10] In the CHD population, it is often difficult to stratify patients into common categories such as left-sided failure or right-sided failure. Standard functional class categorization is also difficult because it based on the premise that patients do not have structural abnormalities at baseline. For instance, the 2005 American College of Cardiology and American Heart Association guidelines recommend HF be divided into 4 subtypes: A, at risk for HF; B, structural heart disease without signs or symptoms; C, structural heart disease with previous or current symptoms; and D, refractory heart disease requiring advanced therapies.[11] One may wonder how CHD patients fit into this classification, which, despite updating in 2013, still did not account for the CHD patient with an underlying congenital cardiac defect. Guideline-level documents such as this are often not particularly helpful in the management of CHD patients because they amass data in the acquired HF population, which is often significantly different if not less well-studied than CHD patients.[9,12,13]

From an epidemiologic perspective, CHD patients do not fare as well as patients with HF from acquired forms of heart disease. Hospitalization rates in CHD patients with HF are higher (214 admissions/1000 adults) and the mean length of stay is longer (11.5 days in complex CHD vs 8 days in the acquired HF cohort).[14] The underlying anatomic defect and prior surgical interventions have been identified as independent risk factors for HF admission in CHD. When admitted for HF, CHD patients have a 5-fold higher risk of in-hospital mortality; death at 1 and 3 years post-HF admission was exceptionally high at 24% and 35%, respectively.[15]

Predictors of death due to HF include endocarditis, supraventricular tachycardia, ventricular tachyarrhythmia, conduction disturbances, pulmonary arterial hypertension, and myocardial infarction (hazard ratio 2–5; $P<.05$).[7] In 2 different European cohorts, HF has been shown to be the most common cause of mortality with the average age of death reported between 47 to 50 years of age.[7,16] As the CHD population continues to age, both outpatient and inpatient care for HF will continue to become among the most important aspects of managing these patients.[17–19]

ANATOMY DICTATES HEART FAILURE PHENOTYPE

HF in CHD is a broad topic that is often difficult to understand. It is easy to see why this may be the case, given the broad spectrum of CHD. Clinically, CHD is subdivided into categories based on the complexity of the structural lesions. Defects are classified as simple CHD, moderately complex CHD, or severely complex CHD. Published guidelines in the treatment of ACHD have indicated follow-up intervals for continued care based on the severity of underlying CHD[20] (**Table 1**). Prior interventions, including cardiac surgery, also play a role in the development of HF and late CVD risk; therefore, they are crucial to consider in caring for the ACHD patient. Given the heterogenous nature of CHD and palliative or surgical repair, the cause and presentation of HF in CHD is diverse. Some common themes in describing HF in this population include the side of the HF (subpulmonic ventricular vs subsystemic ventricular dysfunction),[21] cyanotic versus acyanotic HF, single ventricular failure, and pressure versus volume-mediated HF, among others.

ETIOLOGIC FACTORS OF CONGENITAL HEART DISEASE–HEART FAILURE

The mechanisms leading to HF in CHD are numerous and variable. Some potential causes include abnormal pressure or volume-loading of either the morphologic right ventricle (RV) or left ventricle, myocardial ischemia from either a supply demand mismatch or coronaries anomalies, ventricular hypertrophy, and constriction from prior sternotomy. Myocardial architecture must be considered in patients with CHD. Data suggest embryologic development of the right ventricular myocardium may be different from the left ventricle and more susceptible to dysfunction in lesions where the RV is the systemic ventricle.[9,22] Perfusion also seems to also be important as evidenced by the high prevalence of RV systolic function following