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Review

Defining heart failure in adult congenital heart disease

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ABSTRACT

As the adult congenital heart disease (ACHD) population expands and ages, the incidence and prevalence of heart failure will rise. This poses several challenges, all complicated by our nascent understanding of heart failure epidemiology, pathophysiology and management in adults with congenital heart disease. Current definitions extrapolated from acquired heart failure often disregard the unique pathophysiology of heart failure in adults with congenital heart disease. Others have suggested that congenital heart disease is the 'original heart failure syndrome' implying that all ACHD patients are destined to manifest heart failure. Neither the adoption of acquired heart failure definitions nor the belief that heart failure is the common ultimate manifestation of ACHD has advanced the care of ACHD patients. The absence of a comprehensive definition that focuses on common themes while recognizing the unique manifestations of heart failure in ACHD stifles research progress and has translated to a paucity of ACHD specific recommendations in existing heart failure guidelines. Since many ACHD heart failure patients do not meet standard definitions of heart failure their access to potentially beneficial interventions such as cardiac rehabilitation is restricted by payers and regulators taking a narrow view of acquired heart failure guidelines. ACHD heart failure definitions that can be applied in the clinical and research setting are needed to guide treatment, facilitate communication between specialists, determine the prevalence and incidence of heart failure in AHCD, and improve ACHD patients' access to heart failure treatments. The purpose of this review is to understand how heart failure has been considered and defined in the existing ACHD literature and to highlight the need for a definition of heart failure applicable to ACHD.

1. Introduction

There are estimated to be more than 1 million adults in North America and 1.2 million in Europe with congenital heart disease [1–3]. In addition to an overall increase in ACHD hospitalizations over the last decade, there has been a substantial increase in the number of older (age >65 years) ACHD patients presenting with late complications such as heart failure [4]. Of ACHD hospitalizations, 20% will have a diagnosis of heart failure [5]. There are numerous causes for a high prevalence of heart failure in adults with congenital heart disease, including genetic predisposition, abnormal fetal and post-natal development, cyanosis, prior surgical incisions and scars, inadequate myocardial protection during cardiopulmonary bypass, pressure and volume overload, maladaptive hypertrophy, sub-endocardial ischemia, as well as the synergistic effects of these myocardial insults over time. The potential factors that predispose ACHD patients to heart failure have been expertly summarized elsewhere [6–9]. The purpose of this review is to understand how heart failure has been defined in the ACHD literature to date and to consider whether ACHD specific heart failure definitions are needed.

2. Defining heart failure in acquired heart disease

The American Heart Association/American College of Cardiology guidelines define heart failure as a clinical syndrome characterized by the symptoms of dyspnea and fatigue, as well as signs of congestion (e.g., peripheral edema, rales), resulting from any structural or functional cardiac disorder [10]. This definition encompasses a broad array of clinical manifestations of myocardial dysfunction, including systolic and diastolic left ventricular dysfunction, and significant valve disease. While the definition also seems to apply to other diagnoses such as pulmonary arterial hypertension and many forms of CHD, the recommendations which derive from this definition mostly apply to patients with left heart dysfunction. Younger ACHD patients may be found to have advanced ventricular systolic dysfunction without any 'classic' symptoms or signs of heart failure. Others may present with vastly different symptoms from their acquired heart failure counterparts. For instance, diarrhea or coughing of bronchial casts (plastic bronchitis) may be the first manifestation of a failing Fontan circulation whereas pre-

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syncope and worsening atrial arrhythmias may predominate in transposition patients seen after the Mustard/Senning operations.

The American College of Cardiology/American Heart Association classifies heart failure into Stages A to D (Fig. 1) [10]. Due to the universal presence of structural heart disease, ACHD patients are all classified as Stage B or higher, depending upon symptom severity and the need for hospitalization. According to current guidelines, designation of Stages B to D should encourage clinicians to initiate and up-titrate ACE inhibitors, beta-blockers and aldosterone antagonists in a step-wise manner leading to a consideration of advanced therapies (inotropes, ventricular assist devices, heart transplant) in patients with refractory heart failure. Yet there is little evidence to support this approach in ACHD patients and as such this classification schema is less helpful for directing treatment in this population. ACE inhibitors, beta-blockers and aldosterone antagonists have not been shown to improve survival in ACHD patients and questions remain surrounding outcomes of mechanical circulatory support and heart transplantation in high risk ACHD sub-groups.

Neurohormonal modulation constitutes the foundation of heart failure treatment in acquired heart failure and invasive hemodynamic assessment is usually reserved for patients with refractory heart failure. The reverse is true for most ACHD diagnoses. Indeed, invasive hemodynamic assessment is a common starting point in ACHD patients presenting with symptoms or signs suggestive of cardiac dysfunction and often leads to invasive structural (catheter or surgical) intervention early in the course of their condition (Fig. 2).

3. How is heart failure defined in the ACHD literature?

Most commonly, heart failure in ACHD is defined as a clinical endpoint marked by hospitalization for heart failure, initiation of heart failure medication, or death coded secondary to heart failure. Consequently we have few insights into how heart failure first presents in ACHD patients or current approaches to diagnosis and treatment. We also understand little about the trajectory of heart failure in ACHD or how ventricular function and other prognostic variables change over time.

In a large single center study of 2609 consecutive patients evaluated at the Toronto Congenital Cardiac Centre for Adults, Oechslin et al. [11] reported an overall mortality rate of 8% with a mean age of death at 37 years. Mortality varied by defect complexity, ranging from 0.6% for isolated pulmonic stenosis to greater than 20% mortality in adults with a single ventricle circulation and congenitally corrected transposition of the great arteries. Twenty-one percent of patients died secondary to heart failure, defined in this study as "progressive myocardial failure of either systemic or venous ventricle". Interestingly, 3 of 4 aortic coarctation patients who experienced sudden cardiac death had a LVEF < 40% and all 6 patients with congenitally corrected transposition and who died due to sudden death had a RVEF < 45%, depicting a clinical picture synonymous with heart failure. Along the same lines, in 42% (15/36) of perioperative deaths, cardiogenic shock was present. Combining deaths from heart failure as well as the sudden cardiac and perioperative deaths with significant ventricular systolic dysfunction, the proportion of heart failure deaths rises substantially from 21% to 35% (Fig. 3).

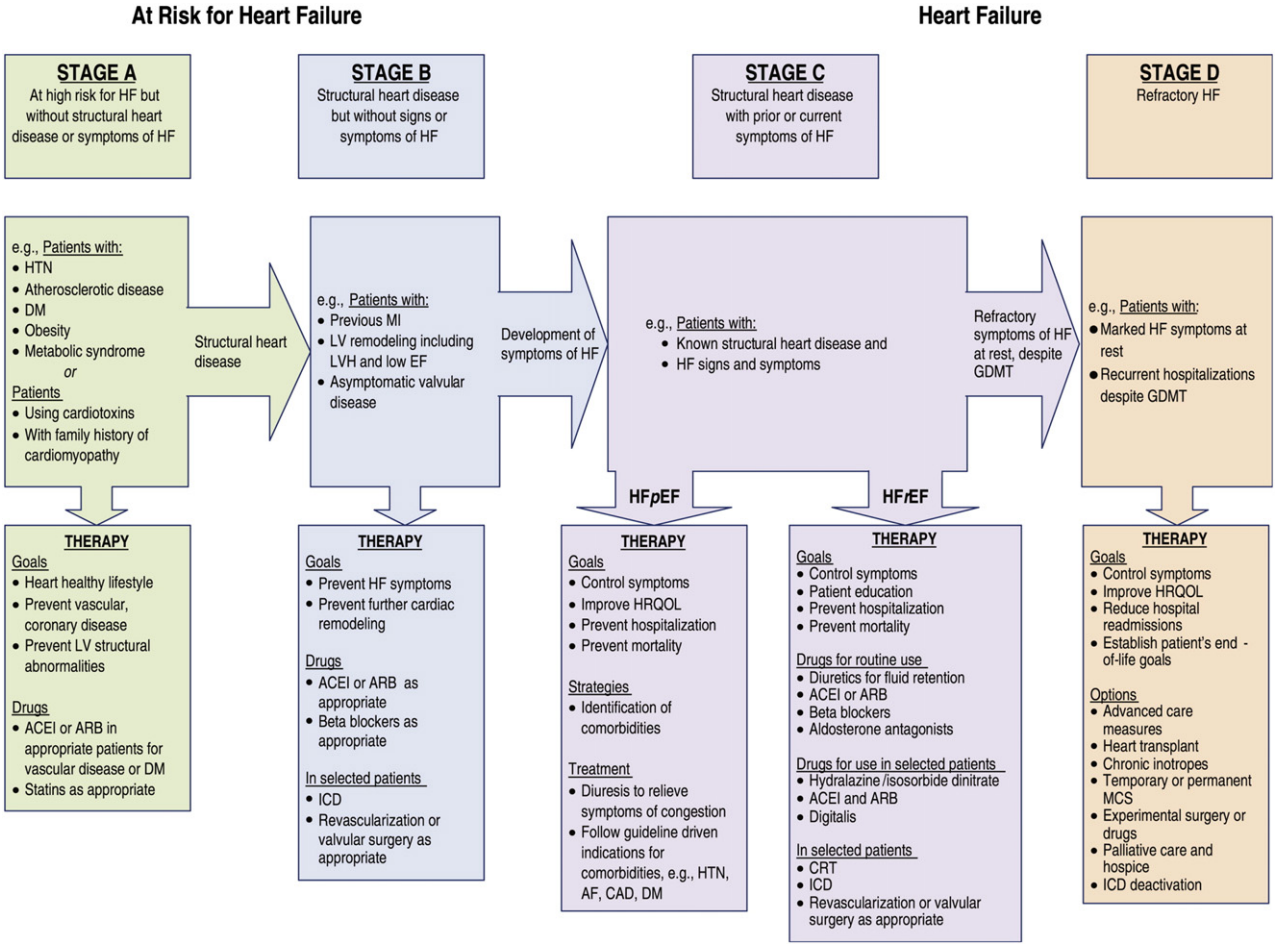

Fig. 1. American Heart Association classification of heart failure, stages A to D [32].