Surgical Device Therapy for Heart Failure in the Adult with Congenital Heart Disease

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KEYWORDS

• Adult congenital heart disease • Ventricular assist device • Heart failure • Systemic right ventricle

KEY POINTS

- Individuals with congenital heart disease (CHD) are at a great risk for heart failure, and the underlying anatomic features are important predictors of heart failure.
- As the adult with CHD (ACHD) population grows older, multiple events, including years of an altered physiology, the neurohormonal cascade, and many still unknown, culminate in ventricular failure.
- As the ACHD population continues to grow in number and complexity, those with systemic right ventricle or single ventricle are at an increased risk of ventricular failure following surgical palliation.
- Ventricular assist devices have been used with success in bridging ACHD patients to heart transplantation or destination therapy.
- As the ACHD population continues to increase and technological advancement continues, surgical devices will play a significant role in the future.

Congenital heart disease (CHD) has an incidence of approximately 8 per 1000 live births. Fifty years ago only 25% of infants with complex CHD survived beyond their first year of life, but today more than 95% will survive to adulthood.[1] Approximately 15% of children born with CHD have potentially life-threatening defects, and many have complex lesions.[2] With the advent of neonatal repair for complex lesions, modern surgical mortality rates are less than 5%.[3] Today there are more than 1 million adults with CHD, outnumbering pediatric patients with CHD.[4] Individuals with adult congenital heart disease (ACHD) are at a great risk for heart failure, and the underlying anatomic features are important predictors of heart failure. As the ACHD population grows older, multiple events, including years of an altered physiology, the neurohormonal cascade, and many still unknown, culminate in ventricular failure. Surgical device therapy is an effective method in supporting patients with heart failure.

INCIDENCE OF HEART FAILURE IN ADULTS WITH CONGENITAL HEART DISEASE

Adults with CHD have a myriad of primary underlying conditions: tetralogy of Fallot, Ebstein anomaly, single right or left ventricles palliated with a Fontan procedure, or systemic right ventricle anatomy resulting from congenitally corrected transposition of the great arteries (CC-TGA) or D-transposition of the great arteries (D-TGA) after atrial switch

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palliation. In addition, each surgical technique may have varied clinical outcomes depending on the era. The probability of heart failure in ACHD by congenital diagnosis is illustrated in Fig. 1.

Patients with D-TGA born before the mid-1990s are likely to have undergone an atrial switch operation (described by Senning in 1959 and Mustard in 1964), leading to a systemic right ventricle. However, in the last 2 decades the arterial switch (first described by Jatene in 1983) has become the preferred surgical operation because it results in a systemic left ventricle. After follow-up of 15 to 18 years, there has been a documented decrease in the systemic right ventricular (RV) function in 32% to 48% of Mustard and Senning patients.[5] Today, most patients with D-TGA undergo the arterial switch, and many are just now entering their second decade of life.

In CC-TGA the right ventricle is the systemic ventricle, and many patients first present with clinical heart failure in adulthood. The anatomic variations are varied and can include pulmonic stenosis, ventricular septal defect, or tricuspid valve abnormalities. Systemic atrioventricular valve regurgitation may be a harbinger of heart failure, owing to worsening ventricular failure and RV volume overload. Two percent of patients per year can develop complete heart block. Presbitero and colleagues[6] reported that 24% of the patients in the fifth decade of life had heart failure, increasing to 77% by the sixth decade of life. In study by Piran and colleagues,[7] incidence of heart failure in patients with D-TGA subsequent to Mustard procedure was 22%, and 32% in patients with CC-TGA.

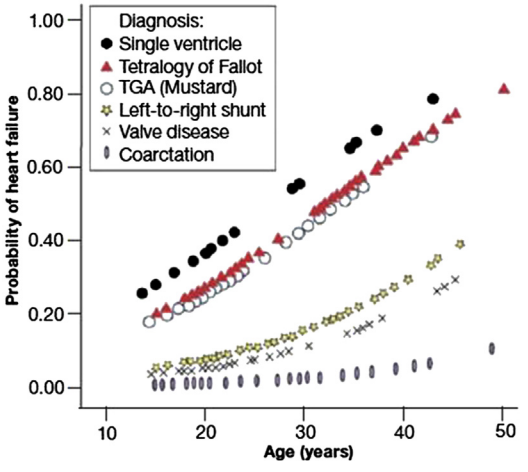

Fig. 1. Incidence of heart failure in adult congenital heart disease. TGA, transposition of the great arteries. (*From* Norozi K, Wessel A, Alpers V, et al. Incidence and risk distribution of heart failure in adolescents and adults with congenital heart disease after cardiac surgery. Am J Cardiol 2006;97(8):1238–43; with permission.)

In patients born with a single ventricle, many historically underwent the classic atriopulmonary connection procedure, a modification of the approach described in 1971 for tricuspid atresia by Fontan and Baudet.[8] The Fontan procedure, or total cavopulmonary connection, leaves the single-ventricle patient with abnormal venous circulation whereby the venal caval blood returns to the lung passively, with a subpulmonary ventricle. Today the Fontan operation has evolved to being a lateral tunnel through the right atrium, or an extracardiac conduit that connects directly to the branch pulmonary arteries. It is often the last surgical palliation in children born with a functional single ventricle. These patients can do well and survive into adulthood, but long-term follow up past the fifth or sixth decade is not known. In one study, 40% of patients who were palliated via a Fontan procedure developed systolic heart failure.[7]

Tetralogy of Fallot (TOF) is the most common cyanotic heart disease, for which Lillehei performed the first intracardiac repair in 1954. This procedure consisted of repair of a ventricular septal defect and resection of the infundibular region of the RV outflow tract.[9] Today the repair minimizes ventricular incision and infundibular resection, and incorporates a transannular patch. The surgical intervention ultimately leads to distortion of pulmonary valve apparatus and pulmonary regurgitation, which is well tolerated for many years but has an effect on the RV size and function.[3] Norozi and colleagues[10] reported on 94 patients with TOF, of whom 44 had heart failure defined by a brain natriuretic peptide (BNP) level of greater than 100 pg/mL and a maximal oxygen uptake (V_{O_2}max) of less than 25 m/kg/min, although most were asymptomatic or minimally symptomatic, of New York Heart Association (NYHA) functional class I or II. Complications of progressive RV dilation include right heart failure with decreased exercise tolerance, atrial and ventricular arrhythmias, and sudden cardiac death. The lifetime incidence of sudden cardiac death is 8.8% in postoperative TOF patients in adulthood.[1] Pulmonary valve replacement is considered the treatment of choice, and timing is still debated, with mitigating factors including exercise intolerance, ventricular arrhythmias, and/or RV dysfunction or dilation (RV end-diastolic volume >150 mL/m^2).

NEUROHORMONAL ACTIVATION

Neurohormonal activation is an important factor in adults with heart failure. Data have shown that the degree of neurohormonal activation in adults with heart failure is correlated with functional capacity, left ventricular (LV) dysfunction, and mortality.