# Heart Failure in Adult Congenital Heart Disease
## Nonpharmacologic Treatment Strategies

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**KEYWORDS**

• Adult congenital heart disease • Heart failure • Biomarkers • Arrhythmia • Cardiac surgery • Structural intervention

**KEY POINTS**

- Heart failure (HF) accounts for one-quarter of deaths in the adult congenital heart disease (ACHD) population; there has been an 80% increase in ACHD admissions involving HF.
- ACHD patients have lifelong adaptations to physiologic derangements related to their disease. Typical features of HF may not be present in this population.
- Whereas a diagnosis of acquired HF leads to standardized medical therapy, treating HF in ACHD relies on nonpharmacologic strategies, namely, transcatheter interventions and arrhythmias along with cardiac surgery.
- In addition to invasive treatment strategies, adjuvant palliative measures are important for reducing suffering associated with the condition.

### UNDERSTANDING HEART FAILURE IN ADULT CONGENITAL HEART DISEASE

Heart failure (HF) accounts for one-quarter of deaths in the adult congenital heart disease (ACHD) population.[1] In parallel with a dramatic increase in the number of hospitalizations for ACHD, there has been an 80% increase in the number of ACHD admissions involving HF during the last decade.[2] HF in ACHD patients is both multifactorial and lesion specific.[3] Those at greatest risk for developing HF are patients with a systemic right ventricle (RV; d-transposition of the great arteries postatrial switch and congenitally corrected transposition of the great arteries), residual valvular dysfunction (ie, pulmonary regurgitation after tetralogy of Fallot repair, congenital aortic stenosis), patients with a single ventricle circulation with or without Fontan palliation and patients with atrial or ventricular level shunt complicated by pulmonary hypertension. The observation that "repaired" ACHD patients also present with late-onset HF is a reminder that intrinsic myocardial structural and functional abnormalities predispose to HF even after the removal of the original anatomic or hemodynamic substrate.[4]

ACHD patients have had lifelong adaptations to physiologic derangements related to their underlying cardiac pathology. Thus, typical features of HF may not be present in this population. Chronic exercise intolerance is well-recognized, but is

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frequently underestimated. Cardiopulmonary exercise testing confirms universal reductions in peak oxygen uptake and circulatory dysfunction across the spectrum of congenital heart disease complexity.[5]

Whereas a diagnosis of acquired HF leads to standardized medical therapy for improvement of symptoms and prognosis, HF management in ACHD frequently rests on nonpharmacologic strategies. Clinicians caring for ACHD patients consider a wide array of factors that conspire to cause HF in this context. This article reviews emerging perspectives on nonpharmacologic treatment strategies for ACHD-related HF, including transcatheter interventions for structural heart disease, invasive electrophysiology strategies, and cardiac surgery. Whereas heart transplantation is beyond the scope of this review, the importance of palliative care in patients with end-stage disease is discussed.

## INVASIVE HEMODYNAMIC EVALUATION AND TRANSCATHETER INTERVENTIONS IN ADULT CONGENITAL HEART DISEASE–RELATED HEART FAILURE

Invasive hemodynamic evaluation plays a crucial role in the evaluation and treatment of patients with ACHD. Accurate assessment of HF patients' hemodynamic state by physical examination often underestimates severity,[6] particularly in younger ACHD patients who may not demonstrate classic physical findings of increased ventricular filling pressures, such as peripheral and pulmonary edema, until late in the course of their HF trajectory. The initial assessment of ACHD patients presenting with newly diagnosed HF begins with noninvasive investigations including electrocardiographs, laboratory tests, echocardiography, and imaging (chest x-ray, MRI, CT) directed by the history and physical examination. Depending on the results of these initial investigations, invasive hemodynamic assessment may be considered (**Fig. 1**).

In addition to facilitating tailored (diuretic and other) therapy, cardiac catheterization identifies residual lesions that may be contributing to HF pathophysiology. Residual cardiac lesions suitable for catheter-based interventions include[1] pressure and/or volume overload lesions,[2] intracardiac and extracardiac shunting and,[3] lesion-specific interventions for unique ACHD subgroups, namely d-transposition of the great arteries after atrial switch (Senning or Mustard) and those with failing Fontan physiology.

Mustard/Senning patients are at increased risk of HF owing to systemic ventricular dysfunction, systemic atrioventricular (AV) valve regurgitation, atrial arrhythmias, and pulmonary hypertension. Baffle complications are common with leaks reported in up to 25%[7] and varying degrees of baffle stenosis (superior vena cava > inferior vena cava) in 5% to 15% of patients.[8,9] Baffle leaks and stenoses often coexist. Depending on shunt volume and direction, baffle leaks may lead to ventricular volume overload (left-to-right shunt), systemic arterial desaturation, and systemic embolization (right-to-left shunt). When significant, systemic venous baffle stenosis can increase systemic venous pressure and impede venous return leading to reduced cardiac output. However, superior vena cava baffle stenosis is often asymptomatic owing to venous collaterals and azygous flow reversal developing as an alternative route for venous return to the heart. Inferior vena cava baffle stenosis or obstruction is less well-tolerated and may lead to hepatic venous congestion, hepatomegaly, and ascites. Pulmonary venous baffle obstruction leading to increased pulmonary venous pressure is a potentially reversible cause of pulmonary edema and hypertension that should be considered in Mustard/Senning patients presenting with these conditions.

Transthoracic echocardiography with saline contrast injection is helpful for confirming the presence of a baffle leak, although additional imaging with MRI and/or transesophageal echocardiography is usually needed to localize the leak. Cardiac catheterization remains the gold standard for evaluating hemodynamic status and should be undertaken in Mustard/Senning patients presenting with HF. Owing to high perioperative mortality for surgical baffle reintervention,[10] transcatheter procedures are now widely preferred as a lower risk alternative. For some patients, relief of baffle leaks and/or stenosis can translate to significant improvements in functional class and hemodynamics.[11] Percutaneous interventions include balloon angioplasty for baffle stenosis with or without stent placement. Baffle leaks can also be treated with deployment of a covered stent.

Adults with failing Fontan physiology typically demonstrate evidence of chronic systemic venous hypertension, low cardiac output, and ventricular diastolic dysfunction.[12] These changes progress over time and contribute to additional manifestations of HF, including atrial arrhythmia, hepatic venous and splanchnic congestion, ascites, protein-losing enteropathy (PLE), and plastic bronchitis. Other contributors that should be sought in patients presenting with failing Fontan physiology include pulmonary venous obstruction secondary to right atrial dilation, ventricular outflow tract obstruction, AV valve regurgitation, and residual