

Diastolic Heart Failure in ACHD

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With significant improvement in the long-term survival of congenital heart patients, the adult congenital heart population is increasing. Heart failure constitutes an important cause of morbidity and mortality and ascertaining the burden of systolic heart failure has been the focus in adult congenital heart disease (ACHD). Nonetheless, with recognition of ACHD patients having symptoms and signs of heart failure and yet with relatively preserved systemic ventricular ejection fraction, a better understanding of diastolic heart failure, the predominant underlying pathophysiologic process, in these patients is timely and of immense clinical relevance. In ACHD, the substrates that lead to impairment of myocardial relaxation, reduction of myocardial compliance, and poor filling of the hypoplastic ventricular chambers exist in various combinations to cause diastolic ventricular dysfunction. Diastolic ventricular dysfunction is recognized in functional single ventricles and the right and left ventricles in the setting of a biventricular circulation. The ACHD patients who are at risk of diastolic heart failure include those after Fontan-type procedure, tetralogy of Fallot repair, biventricular repair of pulmonary atresia with intact ventricular septum, and correction of coarctation of the aorta and those with hypoplastic right and left ventricles that restrict diastolic filling. Diagnostic evaluation should include assessment of conventional diastolic ventricular functional indices, ventricular myocardial deformation and stiffness, myocardial fibrosis, atrial function, and pulmonary hypertension. While evidence-based targeted treatment of diastolic heart failure in ACHD is lacking, results of recent trials on angiotensin-neprilysin inhibition and empagliflozin in non-congenital heart patients with heart failure with preserved ejection fraction have shed light on potential novel therapeutic strategies. Undoubtedly, challenges exist in the clinical recognition, diagnostic evaluation, and management of diastolic heart failure in ACHD and much more research works in this area are warranted.