Heart Failure Caused by Congenital Left-Sided Lesions

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KEYWORDS

• Coarctation • Shone syndrome • Bicuspid aortic valve • Heart failure
• Left ventricular noncompaction • Cor triatriatum

KEY POINTS

- Depending on the location, left-sided lesions can obstruct left ventricular inflow, obstruct left ventricular outflow, cause left-sided valvular regurgitation, or affect ventricular myocardial performance directly.
- Left-sided lesions are often found in combination, whereas bicuspid aortic valve and coarctation often coexist and can be associated with more complex multilevel obstruction, which can markedly increase left ventricular workload.
- Left ventricular noncompaction is a difficult diagnosis because of marked phenotypic variability in left ventricular trabeculation. The prognosis of isolated left ventricular noncompaction is not well understood but seems to be poor in patients with systolic dysfunction and good in those with preserved ejection fraction.

INTRODUCTION

There are diverse mechanisms by which congenital left-sided cardiac lesions can precipitate heart failure. Left heart outflow obstruction can occur at, below, or above the aortic valve, imposing abnormal pressure load on the left ventricle, inducing adverse remodeling, hypertrophy, and diastolic and systolic dysfunction. Abnormalities in left ventricular inflow can occur in the pulmonary veins and within the left atrium, as well as in the region of the mitral valve, increasing pulmonary venous pressure and predisposing to pulmonary edema. In addition, inborn abnormalities in left ventricular myocardial structure and function can impair both systolic and diastolic function and manifest as heart failure later in life. In this article, the different mechanisms, outcomes, and treatments of heart failure in patients with congenital left-sided lesions are discussed.

Even with remarkable advancements in treatment of pediatric heart disease in the last 50 years, patients with repaired congenital heart disease (CHD) have reduced survival.[1,2] Most patients who die early with CHD die from cardiovascular causes. Heart failure continues to be the dominant mode of death in patients with repaired CHD, accounting for more than 40% of CHD-related death. Of the congenital left-sided lesions, coarctation of the aorta (CoA), and congenital aortic stenosis account for the highest proportion of premature mortality.[3,4]

AORTIC VALVE DYSFUNCTION

Bicuspid Aortic Valve

Bicuspid aortic valve (BAV) is present in 0.5% to 2% of the general population and represents the

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most common congenital heart lesion.[5–7] It is more common in males than females (3:1) and can be found in isolation or in association with a wide range of other congenital heart lesions.[8] BAV can be found in association with almost all types of CHD and is common in patients with other left heart obstructive lesions, with more than 50% of patients with aortic coarctation having a BAV.[9]

Most BAVs have 3 leaflets but only 2 well-developed commissures, resulting in 2 functional leaflets.[10–12] Fusion of the right coronary and left coronary leaflets (absence of the intercoronary commissure) is the most common morphology (70%) followed by fusion of the right coronary and noncoronary leaflets. Fusion of the left coronary and noncoronary leaflets is rare (1.4%).[9] A recent study by Fernández and colleagues[13] suggested that right coronary and left coronary leaflet fusion may have different developmental underpinnings from BAVs with fusion of the right coronary and noncoronary leaflets. This suggestion is supported by clinical findings that suggest that children with fusion of the right coronary and noncoronary leaflets are significantly more likely to have progression of aortic stenosis and regurgitation compelling intervention than children with fusion of the right coronary and left coronary leaflets.[14] However, studies in adults with BAV suggest that BAV morphology does not seem to be a strong predictor of valve dysfunction.[15] This distinction is likely related to the impact of calcific changes noted in adulthood, which are more likely to drive valve dysfunction, and the fact that right coronary and noncoronary leaflet fusion are less likely than right coronary and left coronary leaflet fusion to be without significant valve dysfunction until adulthood.[14]

Although patients with BAV can present in infancy with severe aortic stenosis, in most cases, the BAV functions well until later in life, when calcific changes cause progression of stenosis or regurgitation.[15] There is a well-documented association between BAV and progressive dilation of the ascending aorta, the cause of which is not completely understood and seems to be independent of the degree of aortic stenosis.[16,17] Several studies have suggested that not only do patients with BAV without significant valvular dysfunction have associated aortic dilation, but these patients also have abnormal left ventricular mechanics and performance.[18,19] This finding suggests that BAV is unlikely to represent isolated valvular disease, which may have significant implications on long-term ventricular function even in the absence of significant aortic stenosis or regurgitation.

Other Causes of Congenital Aortic Stenosis

BAV is the most common cause of aortic stenosis, but the valve morphology in rare cases can be quadricuspid (~0.005% of the general population)[20] or unicuspid (~0.02%) with only 1 functioning leaflet.[21] The latter often presents with severe obstruction in infancy.[22] In addition to obstruction at the level of the aortic valve, there can be obstruction below the aortic valve because of a discrete membrane or fibromuscular tunnel or ridge or there can be significant narrowing above the valve (supravalvar). Supravalvar aortic stenosis is commonly associated with William syndrome, and the level of the obstruction is typically above the aortic root.[23]

Regardless of the level of obstruction, compensatory left ventricular hypertrophy is required to meet the physiologic demands of pressure afterload caused by aortic stenosis. The increase in left ventricular mass can be inadequate for the degree of wall stress, resulting in left ventricular systolic dysfunction and decreased cardiac output.[24] Even when left ventricular hypertrophy seems adequate, chronically increased pressure load can lead to myocardial damage, negatively affecting both left ventricular systolic and diastolic function. Such deleterious effects often result in symptoms of angina, exercise intolerance, near syncope or syncope, and heart failure, which have been associated with significant morbidity and mortality in patients with aortic stenosis.[25,26] Once symptoms are present, the mean survival is only 2 to 3 years.[27] Continuous assessment of cardiac symptoms, aortic stenosis severity, and left ventricular performance as well as ensuring appropriate timing of intervention seems important to ensure long-term well-being.

Indications for intervention in symptomatic patients with severe valvar aortic stenosis and for asymptomatic patients with left ventricular dysfunction or exercise-induced symptoms or hypotension are well defined.[27–29] The timing of intervention in the remainder of asymptomatic patients with significant valvar aortic stenosis can be more difficult.[30] Although its role is not fully defined, stress echocardiography may be helpful in determining timing of intervention in this subset of patients. Studies suggest that a significant increase in the aortic valve gradient, the presence of exercise-induced pulmonary hypertension, and changes in ventricular function (increased left ventricular volume and stiffness) can identify those patients at increased risk of cardiac events.[29,31,32]

Aortic Regurgitation

Isolated congenital aortic regurgitation is rare. Typically, it is associated with abnormal morphology of