Congenital Cardiac Defects That Are Borderline Candidates for Biventricular Repair

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The goal of most surgical treatments for congenital heart disease is to produce septated circulation, in which pulmonary and systemic blood flow are divided. In borderline cases, the decision to attempt biventricular repair is made after comprehensive, multidisciplinary discussions among congenital heart disease surgeons, cardiologists, and the patient’s family members. These discussions often involve challenging decisions that require balancing short- and intermediate-term risks and may ultimately rely on subtle measurements and group consensus regarding the anatomic and physiologic feasibility of biventricular circulation. In this article I discuss congenital heart defects that are often marginal candidates for biventricular repair and highlight the anatomic considerations for these surgical decisions. I also review immediate postoperative patient treatment and criteria for considering biventricular repair of a marginal lesion to be a failure.
Diagnosis of a Marginal Ventricle

Over the past several decades, our understanding of the treatment of congenital heart disease has made considerable advances. Innovations in medical technology have provided a multidimensional perspective on cardiac anatomy and function, helping to determine the adequacy and candidacy for biventricular repair. Progressive developments in fetal echocardiography have clarified developmental influences on the maturation of the heart and its individual components. Detection of congenital heart disease in the fetus facilitates early discussions and strategic planning regarding the best surgical options.

The adequacy of a ventricle is determined by the atrioventricular valve (AVV) and the ventricle’s capacitance and compliance. Capacitance is a function of the effective orifice area of the AVV and the size of the ventricular chamber. The compliance of the ventricle refers to the ability of the ventricle to relax or the diastolic function of the ventricle. The AVV often determines the developmental morphology and adequacy of its associated ventricle. Compromised flow through an AVV in gestation can inhibit growth, limiting the size and altering the morphology of that valve.

Concomitantly, the development and maturation of the associated ventricle can also be affected. Simply stated, if there is no flow, there is no growth. The diastolic function and overall size of a ventricle are crucial in determining whether a ventricle is able to manage a full stroke volume. Echocardiography and cardiac catheterization yield essential, complementary information regarding the morphology of the AVV and the capacitance and compliance of its associated ventricle. Echocardiographic measurements of volume and length are used to assess ventricular size, and measurements of fractional shortening and ejection fraction are used to assess left ventricular function. Assessing the adequacy and function of the right ventricle by echocardiography is more challenging. Magnetic resonance imaging (MRI) and cardiac catheterization data are often more helpful in assessing the right ventricle. Cardiac MRI or computed tomography provides further quantitation of ventricular volumes and AVV function and yields a 3-dimensional perspective on ventricular morphology and capacitance.

Echocardiography provides measurements of AVV size, which are crucial in the decision-making process. These measurements are obtained in both the anterior-posterior and lateral dimensions and are compared with age-specific norms. Z scores are calculated to decide whether an AVV and ventricle are adequate. The z score is the number of standard deviations of a measurement above or below an age-specific mean. Z scores greater than -2 are minimally acceptable for consideration of a biventricular repair. Smaller AVVs are typically not amenable to surgical or transcatheter manipulation or augmentation and should give cause for reconsideration, given the risk for valvular stenosis and atrial hypertension.

Assessing the adequacy of ventricular size is more complicated. Measurements of volume and length, along with associated z scores, are used to assess ventricular size. Sufficient information can often be obtained from echocardiography. However, MRI provides additional quantitative 3-dimensional measurements that can be useful when considering septation of a marginal ventricle. Both echocardiography and MRI are useful in identifying endocardial fibroelastosis. The presence of endocardial fibroelastosis affects a ventricle’s diastolic function and ability to grow. This information is important during surgical decision-making because endocardial fibroelastosis may need to be addressed surgically. All of these complementary data influence the decision to perform 2-ventricle or single-ventricle palliation.

Candidate Lesions for Biventricular Repair

In this article, cardiac lesions to consider for biventricular repair are categorized as right- and left-sided lesions. The discussion includes a brief description of...
the defects and the important measurements to be considered when making the surgical decision.

**Right-Sided Lesions**

**Pulmonary Atresia With Intact Ventricular Septum.**

Pulmonary atresia with intact ventricular septum (PA/IVS) is a rare, cyanotic congenital heart anomaly characterized by an atrial septal defect, hypoplastic tricuspid valve, hypoplastic/hypertrophic right ventricle, and imperforate pulmonary valve (Figure 1). Because of the lack of antegrade flow in the right ventricle, the fetal right ventricle is often hypertensive in utero. The development of fistulous connections with the coronary circulation can heighten the complexity and severity of the anatomy, especially when the coronary circulation depends on those connections for antegrade flow (ie, right ventricle–dependent coronary circulation). Each individual component of this congenital lesion can present with varying severity in both function and structure.

Prognosis and treatment depend on the degree of tricuspid valve and right ventricle hypoplasia and the presence of right ventricle–dependent coronary circulation. Substantial research has investigated factors that affect the feasibility of biventricular repair in patients with PA/IVS. Ashburn et al reported that 85% of neonates born with PA/IVS are likely to reach a definitive surgical end point, with 50% receiving biventricular repair. This report and others have shown that tricuspid valve size ($z$ score) is a primary determinant of this type of repair. Management-decision protocols commonly use a tricuspid valve $z$ score of greater than -2 as a minimum requirement for 2-ventricle repair. Subsequent reports also show that the degree of tricuspid valve dysplasia (independent of size), right ventricular hypoplasia, and the presence of right ventricle–dependent coronary circulation are also important considerations. Another surgical option for patients with PA/IVS with a ventricle size marginal for biventricular repair is a 1 1/2-ventricle repair. This repair involves adding a Glenn procedure to offload the right ventricle. Further discussion of the decision-making process for this surgical procedure is beyond the scope of this article.

**Ebstein Anomaly.**

Ebstein anomaly consists of an atrial septal defect and tricuspid valve displacement into the right ventricle causing atrialization of the ventricle (Figure 2). The condition results in varying degrees of right ventricular hypoplasia. The important measurements for determining whether biventricular repair is feasible are the tricuspid valve annulus, degree of atrialization of the ventricle, and true right ventricle size. An alternative to biventricular repair is a 1 1/2-ventricle repair consisting of a tricuspid valve resuspension, atrial septal defect closure, and bidirectional Glenn procedure. This surgical procedure provides the right ventricle with roughly two-thirds of the anticipated cardiac output. The biventricular surgical repair for Ebstein anomaly consists of a tricuspid valve repair usually involving annuloplasty and
resuspension of the anterior leaflet to create an adequate orifice of the tricuspid valve.11

Double-Outlet Right Ventricle. Double-outlet right ventricle is a conotruncal defect involving abnormal conal tissue development, resulting in varying degrees of malposition of the great arteries, which leads to both great arteries arising from the right ventricle. Conotruncal ventricular septal defect (VSD) also exists in most cases. Double-outlet right ventricle is one of the most complicated defects, and the decision regarding the timing and appropriateness of repair is complex.12 However, many of the same principles of capacitance and compliance apply. In patients with this defect, the sizes of both ventricles and AVVs affect the surgical choice (Figure 3). Understanding the relationship of the great vessels to the VSD is also important for baffling the VSD if a biventricular repair is attempted. Other factors affecting the surgical decision are any variations in the anatomy of the tricuspid valve itself, the amount of tricuspid valve tissue within the VSD, the size of the baffle needed to make an adequate left ventricular outflow tract, and how much that baffle impinges on the size of the right ventricle cavity.

Unbalanced Atrioventricular Canal Defect. A complete atrioventricular canal defect can be categorized as either a right- or left-sided lesion. An atrioventricular canal defect consists of a primum atrial septal defect, a common AVV, and an inlet VSD with extension into the membranous septum. The balance of the atrioventricular canal is determined by the distribution of the common valve tissue over each ventricle. The choice of surgical correction for this defect depends on whether the AVV itself can be septated given the attachments of the AVV and the resulting size of each ventricle.13 As illustrated by the adage “no flow, no grow,” an imbalance in AVV tissue (more tissue committed to one ventricle than to the other) results in hypoplasia of the ventricle with less inflow. The measurements needed to decide whether an unbalanced atrioventricular canal can be septated are not clearly defined. The size of the ventricle, the size of the estimated orifice of the septated AVV, and color Doppler measurement of left-sided AVV inflow all contribute to the decision to perform biventricular repair.

Left-Sided Lesions
Shone Complex and Critical Aortic Stenosis. Shone complex comprises a supravalvar mitral ring, mitral stenosis, aortic stenosis, coarctation of the aorta, and borderline left ventricle size (Figure 4). The degrees of these lesions determine whether a biventricular repair is possible.14 The important factors are the size and morphology of the mitral valve and the sizes of the subaortic area, aortic valve, and left ventricle.

Critical aortic stenosis consists of obstruction at the valve and frequently at the subvalvar level. Important surgical considerations for this defect are the morphology of the aortic valve and the degree of obstruction. In utero critical aortic stenosis can lead to increased afterload on the ventricle, resulting in varying degrees of ventricular hypertrophy and myocardial dysfunction.
Stenosis at the valve level leads to decreased forward flow across the valve. This decreased forward flow causes an increase in end-diastolic filling pressure, affecting the flow across the mitral valve and thereby influencing the morphology of the mitral valve and left ventricle.

Multiple studies and reviews have examined values predictive of aortic valve adequacy and left ventricular size to guide the choice between 2-ventricle and single-ventricle repair. Hammon et al\textsuperscript{15} proposed that the threshold for successful biventricular repair was a left ventricular end-diastolic volume of 20 mL/m\textsuperscript{2} (determined by catheterization). Others have identified clinical risk factors for a successful biventricular repair, including mitral valve area, long-axis dimension of the left ventricle relative to the long-axis dimension of the heart, aortic root size, and left ventricle mass. Hemodynamic data obtained in a catheterization laboratory with the patient in different physiologic states often provide important information regarding the ventricle’s ability to handle a full cardiac output. Testing occlusion of a patent foramen ovale in the catheterization laboratory allows measurement of left atrial pressure and cardiac output across the left ventricle, which indicates whether the left ventricle is adequate. The decision-making process for this defect is complex, and the surgical decision must be made within the context of each patient’s unique presentation.\textsuperscript{16-18}

Interrupted Aortic Arch With VSD. Interrupted aortic arch with VSD is usually associated with a type B interruption of the aorta (interruption between the left subclavian and left carotid arteries) with an associated conotruncal abnormality. This condition results in normally related great arteries, a malaligned VSD, and a posteriorly deviated infundibulum leading to subaortic stenosis (Figure 5). The surgical decision for this defect is based on its 2 levels of shunting: the VSD and a patent ductus arteriosus supplying the left subclavian artery and descending aorta. Because the aortic valve never receives the full cardiac output, assessing whether the aortic valve can manage the full cardiac output is impossible. The surgical decision is based entirely on the sizes of the mitral valve, left ventricle, and aortic valve and the degree of left ventricular outflow tract obstruction.\textsuperscript{19,20}

Postoperative Management

Patients with borderline anatomy who have undergone biventricular repair present the cardiac intensive care unit with some of its most critical and challenging problems. Postoperative issues after biventricular repair of marginal lesions are related to ventricular compliance and capacitance. Some dominant postoperative problems for patients with left-sided disease are left atrial hypertension, residual mitral valve stenosis and/or regurgitation, ventricular diastolic dysfunction, and secondary pulmonary hypertension resulting in right ventricular dysfunction or failure. Left atrial hypertension is a challenging hemodynamic burden directly related to residual mitral valve lesions and left ventricular diastolic dysfunction resulting from an abnormal left ventricle size or intrinsic abnormalities in myocardial compliance. Secondary pulmonary hypertension and right ventricular failure are typically intermediate- and long-term postoperative issues caused by persistent pulmonary venous hypertension. These residual left-sided lesions can present significant hemodynamic burdens for the patient, often requiring aggressive diuretic therapy, prolonged positive-pressure ventilation, and ongoing intensive care hospitalization.

Postoperative issues related to compliance and capacitance of right-sided lesions are elevated right atrial pressure secondary to the size and function of the tricuspid
Patients with borderline anatomy who have undergone biventricular repair present to the cardiac ICU with challenging and critical problems.

valve and the diastolic properties of the right ventricle. Tricuspid valve morphology and residual stenosis or regurgitation play important roles in the degree of right atrial hypertension. Patients with right atrial hypertension may develop pleural effusion, hepatic dysfunction, cardiorenal syndrome due to systemic venous hypertension, and cerebral dysautoregulation. Right atrial hypertension that is significant in the postoperative period can be managed by creating an atrial septal defect and a right-to-left shunt across the atrial septum. This causes cyanosis in the patient but may alleviate some symptoms of right atrial hypertension. The degree of right-sided AVV disease and right ventricular diastolic dysfunction can also lead to significant atrial tachyarrhythmias.

Immediate postoperative treatment strategies focus on managing residual lesions. Management of fluid overload, ventricular dysfunction, residual valvar disease, and tachyarrhythmias are some of the important considerations following biventricular repair of a marginal lesion. Reducing afterload on a borderline left ventricle with agents such as milrinone or sodium nitroprusside optimizes ventricular filling and antegrade perfusion. Milrinone is a phosphodiesterase 3 inhibitor that has the theoretical advantage of optimizing ventricular relaxation, although its primary function is to increase myocardial contractility and peripheral vasodilation. For right-sided lesions, pulmonary vascular resistance (PVR) should be optimized by aggressive treatment. Ideal ventilation and oxygenation are crucial for avoiding respiratory acidosis that can provoke pulmonary hypertension. The goal of postoperative ventilation strategies is to ventilate patients at their ideal functional residual capacity, which helps manage PVR and decreases right ventricular afterload. Pulmonary vascular resistance increases when lung volume decreases, causing alveoli to collapse; PVR also increases with large lung volumes causing compression of vessels around the alveoli. Increased PVR leads to increased right ventricular afterload. The use of inhaled nitric oxide to decrease PVR can be a helpful adjunctive therapy to improve PVR in the immediate postoperative period if the patient shows evidence of right ventricular diastolic failure. Regulating intravenous fluid administration supports the diastolic function of the marginal ventricle. Fluid overload can exacerbate atrial hypertension in a volume-overloaded ventricle with diastolic dysfunction. Excessive diuresis can be deleterious for cardiac output given a ventricle’s preload sensitivity. Close attention to central venous pressure and its relationship to heart rate can be essential for optimizing fluid management. Aggressively managing tachyarrhythmias and preserving atrioventricular synchrony are important for maximizing cardiac performance.

In addition to monitoring the effects of residual postoperative lesions, bedside nurses also track a multitude of related issues in these critically ill children. Prolonged intubation secondary to cardiopulmonary compromise results in the need for extended exposure to narcotics and benzodiazepines, which in turn necessitates close attention to issues of tolerance and withdrawal. Feeding intolerance commonly occurs in these patients because of poor cardiac output, fluid overload, and narcotic or benzodiazepine exposure. Developing a comprehensive feeding plan and monitoring fluids and electrolyte levels are critical nursing interventions. Long-term failure to thrive and developmental delay require an interdisciplinary team including physical therapists, occupational and speech therapists, and others. The stress these patients and families experience is profound. Ensuring that they obtain support and follow-up is critical to their overall care.

Conclusions

The long-term outcomes of biventricular repairs of marginal lesions in complex congenital heart diseases vary and reflect current challenges in the accurate prediction of successful candidacy. It is not uncommon for a patient who has undergone biventricular repair of a borderline defect to face multiple surgical procedures to address residual anatomic lesions and/or valve replacement(s). Long-standing ventricular diastolic dysfunction can result in important sequelae. Patients with a marginal left ventricle with diastolic dysfunction may develop left atrial hypertension leading to secondary pulmonary hypertension. The development of pulmonary hypertension can affect a patient’s candidacy for heart transplant and may necessitate evaluation for heart and lung transplant. Right ventricular diastolic dysfunction can lead to hepatic congestion resulting in liver failure. Both right and left ventricular dysfunction may also lead to arrhythmias.
Over the last decade, the outcomes of single-ventricle palliation have improved significantly, providing an increasingly feasible alternative for treating complex congenital lesions. As discussed in this article, the decision to pursue biventricular repair in a patient with marginal anatomy must take into consideration at minimum the morphology and size of the AVV and the diastolic function of the borderline ventricle. Reversing a marginal 2-ventricle repair and converting to single-ventricle palliation should be an option when a marginal ventricle continues to fail. The exact timing of this decision must take into account secondary problems related to long-standing diastolic dysfunction and pulmonary vascular changes. Leaving a failing ventricle untreated for too long may leave heart or heart and lung transplant as the only viable option. Some congenital heart surgery programs are now considering conversion of single-ventricle repairs to biventricular repairs in patients who have demonstrated ventricular growth and improvement in AVV function. This procedure is high risk and can result in significant morbidity and mortality. A thorough investigation of each patient’s anatomy and physiology is key to making the right decisions for this complex patient population.

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None reported.

See also


References