Current Interventional and Surgical Management of Congenital Heart Disease

Specific Focus on Valvular Disease and Cardiac Arrhythmias

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Abstract: **Successful outcome in the care of patients with congenital heart disease depends on a comprehensive multidisciplinary team. Surgery is offered for almost every heart defect, despite complexity. Early mortality for cardiac surgery in the neonatal period is ≈10% and beyond infancy is <5%, with 90% to 95% of patients surviving with a good quality of life into the adult years. Advances in imaging have facilitated accurate diagnosis and planning of interventions and surgical procedures. Similarly, advances in the perioperative medical management of patients, particularly with intensive care, has also contributed to improving outcomes. Arrhythmias and heart failure are the most common late complications for the majority of defects, and reoperation for valvar problems is common. Lifelong surveillance for monitoring of recurrent or residual structural heart defects, as well as periodic assessment of cardiac function and arrhythmia monitoring, is essential for all patients. The field of congenital heart surgery is poised to incorporate new innovations such as bioengineered cells and scaffolds that will iteratively move toward bioengineered patches, conduits, valves, and even whole organs. (*Circ Res*. 2017;120:1027-1044. DOI: 10.1161/CIRCRESAHA.117.309186.)**

Key Words: arrhythmia ■ congenital heart disease ■ heart failure ■ intervention ■ surgery

Advances in Surgery for Congenital Heart Disease

The first 50 years of cardiovascular surgery were characterized by the development of new operations, with progressive improvement in outcomes. The early era of cardiovascular surgery (1950s–1970s) was marked by the application of cardiopulmonary bypass (CPB) for intracardiac repair of congenital heart defects (eg, tetralogy of Fallot). Imaging included cardiac catheterization and echocardiography. Anatomic detail was unsophisticated, and relatively few institutions performed cardiovascular surgery, and operative mortality was high (25%–50%). In the mid 1970s, outcomes improved with the development of cardioplegia for myocardial protection and improvements in 2-dimensional echocardiography-tenable accurate diagnosis. Also important was the emergence of Prostaglandin to maintain ductal patency. The 1980s

Original received February 14, 2017; revision received February 20, 2017; accepted February 20, 2017.

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Nonstandard Abbreviations and Acronyms

3D	3-dimensional
AF	atrial fibrillation
AV	aortic valve
CHD	congenital heart disease
CPB	cardiopulmonary bypass
HLHS	hypoplastic left heart syndrome
IART	intra-atrial reentry tachycardia
ICD	implantable cardioverter-defibrillator
LV	left ventricle
MR	mitral regurgitation
MRI	magnetic resonance imaging
MS	mitral stenosis
RV	right ventricle
STS-CHSD	Society of Thoracic Surgeons Congenital Heart Surgery Database

were notable for neonatal corrective procedures (eg, arterial switch). Imaging improved with the use of transesophageal echo, and operative mortality was <10% for most lesions. As the pace of innovation plateaued, emphasis shifted to improving early outcome. Procedures were available in most metropolitan areas as children's hospitals proliferated, and surgery was the norm for most patients with complex defects. Today, cardiovascular surgery uses sophisticated imaging, with virtual or printed models available on request for surgical planning and education. Surgery is almost always offered even for the most complex defects. There is now pressure for perfection, with results to further reduce operative mortality to <2%, and public reporting of outcomes are becoming universal. In addition, late survival into the adult years is expected for almost all congenital heart lesions, and the need for some type of reintervention or reoperation is inevitable for many. The goal of this review is to provide an appraisal of surgical problems that commonly occur in patients with congenital heart disease (CHD), with a focus on valvular heart disease and cardiac arrhythmias.

Diagnosis and Imaging

Fetal Diagnosis

Ongoing developments in prenatal imaging include advances in echocardiography that have improved rates of fetal diagnosis of CHD. The rate of fetal diagnosis has increased in neonates and infants (≤6 months) who require surgical intervention from ≈26% in 2006 to 42% in 2012.[1] Children who require operative intervention as neonates had increased rates of prenatal diagnosis compared with those who had operative interventions as infants, 43.2% versus 23.9%, respectively (P<0.0001), and rates of fetal detection varied significantly by geographic region.[1]

Published literature has not universally demonstrated improved outcomes after fetal diagnosis, although more and more investigations highlight improved coordination of cares, including planned deliveries, immediate postnatal cares, and perioperative and operative planning.[2] In addition, fetal detection was associated with decreased rates of emergent procedures and decreased incidence of perinatal adverse events.[3] The American Heart Association has developed guidelines outlining recommendations related to specialized delivery room care[4] because many congenital cardiac lesions can quickly result in neonate deterioration requiring multiple potential interventions (Table 1).[2]

Anatomic lesions that are readily detected on 4-chamber echocardiography views have better rates of fetal diagnosis than outflow tract abnormalities.[1,2] CHD lesions more readily detected on 4-chamber view include atrioventricular septal defects, large ventricular septal defects, hypoplastic left heart syndrome (HLHS), critical aortic stenosis, severe coarctation of the aorta, tricuspid atresia, pulmonary atresia with intact ventricular septum, Ebstein anomaly of the tricuspid valve (TV), and double-inlet left or right ventricle (RV).[2] Lesions that require extended echocardiography views of the outflow tracts are more difficult to diagnose prenatally; these include transposition of the great arteries, tetralogy of Fallot, common arterial trunk, some forms of double-outlet RV, and coarctation of the aorta.[2]

Fetal Interventions and Impact on Surgical Care

In addition to improved coordination of delivery and postnatal cares, fetal CHD diagnosis brings the possibility of fetal interventions, which may improve subsequent cardiac function. At present, lesions with the greatest potential benefit from fetal interventions are fetal arrhythmias, severe aortic or pulmonary stenosis, and HLHS. Although <5% of patients with a prenatal diagnosis are currently candidates for fetal intervention,[2] the prospect of altering fetal anatomy and physiology to improve subsequent outcomes is a new and exciting advancement in the care of patients with CHD.

The largest experience with fetal interventions is balloon dilation of the aortic valve (AV) in fetuses, with critical aortic stenosis and additional features of left heart hypoplasia.[5,6] The rationale for fetal intervention in HLHS is to promote development of the left heart.[5] The subset of patients potentially eligible for valvuloplasty is those with mitral and aortic stenosis with a relatively dilated left ventricle (LV). The initial attempt at fetal aortic valvuloplasty in 100 patients with critical aortic stenosis was successful in 77 patients at a median gestational age of 23.8 weeks.[6] Of the 77 successful aortic valvuloplasties, 45% were able to undergo a biventricular repair rather than single ventricular pathway.[6]

Image-Based Operative Planning

Improvements in cardiac imaging greatly contribute to optimal operative planning outside of the neonatal period, both related to surgical strategy and postoperative monitoring and assessments. Advancements in echocardiography, computed tomography, and magnetic resonance imaging (MRI) allow for detailed noninvasive cardiac assessment. Two-dimensional echocardiography remains the standard imaging tool used by surgeons to evaluate cardiac function and structural defects, such as presence of intracardiac shunts and valvar anatomy and pathology.

Advancements in 3-dimensional (3D) reconstructions have led to increasing utility of these models for operative planning, surgical training, simulation, and training of multidisciplinary teams. The impact of 3D reconstructions on operative outcomes is difficult to quantify, but its practical value for complex surgical planning is well appreciated by surgeons.

Table 1. Cardiac Lesions and Situations Requiring Immediate Perinatal Attention

Cardiac Lesion	Potential Complication	Potential Interventions
Simple transposition of the great arteries (associated with a restrictive atrial septum and arterial duct)	Metabolic acidosis	Urgent balloon atrial septostomy
	Profound hypoxemia	
Hypoplastic left heart syndrome or critical aortic stenosis (associated with an intact or restrictive atrial septum)	Profound hypoxemia	Urgent balloon atrial septostomy or surgical septectomy
	Metabolic acidosis	
Obstructed total anomalous pulmonary venous drainage	Profound hypoxemia	Early surgical intervention
Absent pulmonary valve syndrome	Airway compression	Mechanical ventilation (positive pressure) Plication of pulmonary arteries
	Respiratory compromise or failure	
	Air trapping	
Severe Ebstein anomaly of the tricuspid valve	Pulmonary hypoplasia	Oxygen therapy
	Respiratory failure	Mechanical ventilation (positive pressure)
	Severe hypoxemia	Inhaled nitric oxide
	Management of hydrops	Drainage of associated pleural effusions or ascites
Complete heart block with or without congenital heart disease	Cardiac failure	Medical therapy with chronotropic agents
		Temporary cardiac pacing
	Hydrops	Drainage of associated pleural effusions or ascites

Availability of perinatal intervention increases impact of fetal cardiac screening programs. Reproduced from Hunter and Simpson[2] with permission of the publisher. Copyright ©2014, Nature Publishing Group.

In addition, 3D-reconstructed models and images are valuable teaching tools.

Cardiac MRI is being increasingly used in CHD because of superiority in cross-sectional imaging without radiation exposure. MRI is particularly useful in quantification of right heart size and function and can be used to create 3D printed models for operative planning. MRI allows for high-resolution images with high spatial and temporal resolution to be obtained and can also be used to measure intracardiac flows.[7] The combination of anatomic and flow data provided from advanced imaging combined with advanced computer simulation allow for increased surgical simulation. Such simulations can facilitate optimal preoperative planning of surgical procedures, for example, optimal Fontan conduit geometry, to be determined virtually by simulating flow through multiple conduit geometries before actual surgery.[8] MRI does often require general anesthesia for younger patients, particularly those <7 to 8 years of age, and therefore, the risk of anesthesia must be weighed with the potential benefit as related to clinical and operative management.

Surgical Management of Patients With CHD

Surgical Planning for a Lifetime

Initial operative management for patients with some forms of CHD is palliative rather than reparative. For all such patients, and for many individuals who undergo repair of complex anomalies early in life, it is likely that they will subsequently require additional operations or percutaneous/hybrid interventions throughout their lifetimes, many of which are valve related (Table 2). Improvements in survival in CHD and survival of 80% to 95% of children born with a congenital cardiac lesion surviving to adulthood have resulted in an increase in the incidence of reoperation in CHD and operation on adults with CHD (Figure 1).[9] The need for multiple operations throughout an individual's lifetime and the additive risk of each surgical intervention must be considered with the initial operation and subsequent (re)operations. Operative mortality at time of reoperation is associated with increased number of previous operations in both national (Figure 2)[10] and institutional reports.[11]

The majority of reoperations are valvar, for both regurgitation and stenosis.[11,12] Operative planning that addresses the

Table 2. Congenital Heart Defects With Associated Valvar Abnormalities

Anomaly	Valve
Atrial septal defect	Tricuspid
Ventricular septal defect	Tricuspid, aortic
Atrioventricular septal defect	Mitral (LAVV), tricuspid (RAVV)
D-transposition of the great arteries	(Neo)aortic, pulmonary
L-transposition of the great arteries	Systemic atrioventricular (tricuspid), pulmonary
Truncus arteriosus	Truncal (aortic), pulmonary
Tetralogy of Fallot, pulmonary atresia	Tricuspid, pulmonary, aortic
Hypoplastic left heart syndrome	Tricuspid
Aortic coarctation	Aortic
Ebstein anomaly	Tricuspid
Anomalous pulmonary venous connection	Tricuspid

LAVV indicates left atrioventricular valve; and RAVV, right atrioventricular valve.

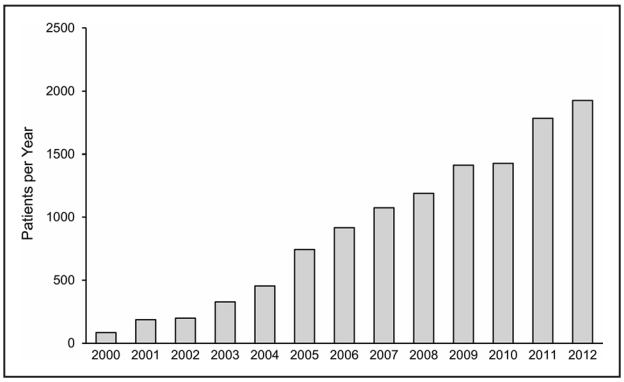

Figure 1. Adult congenital volume through time. Adult patients with congenital heart disease continue to increase through time. Adult patient volumes as reported to The Society of Thoracic Surgeons Congenital Heart Surgery Database from 2000 to 2012 from 116 congenital heart surgery centers. Reproduced from Fuller et al[9] with permission of the publisher. Copyright ©2015, The Society of Thoracic Surgeons.

goals of the current and the subsequent operations need to be considered prior to each operative intervention, with the ultimate goal of minimizing the cumulative number of operations over a lifetime by maximizing the longevity of each individual operation. The likelihood of long-term survival has been shown to be significantly decreased for patients requiring a fifth sternotomy (or more). Thus, it is essential that surgical planning and therapeutic strategies in general address a goal of achieving durable benefits, while minimizing the number of reoperations that are likely to be required.[11]

Indications for intervention for valve-related abnormalities are shown in Table 3.[13] In general, when symptoms are present in the setting of a hemodynamic abnormality secondary to a valve abnormality, intervention is advised. The challenge in patients with CHD (both children and young adults) is that symptoms are often absent, despite significant valvar stenosis or regurgitation in some circumstances. Table 3, which is based on valve abnormalities in acquired heart disease, serves as a guide as to when intervention should be considered because formal valve guidelines in the congenital population are lacking.

Current Operation

Outcomes of surgery for CHD, both primary and reoperative procedures, continue to improve, but are not without risk. Early mortality and morbidity are unique for each individual patient and lesion, and concerted efforts are being made to analyze operative risk at a national level, create standards for accepted risk, and identify patient- and institution-specific factors that increase or decrease observed operative mortality. National data related to the outcomes of surgery in patients with CHD are primarily analyzed through the STS-CHSD (Society of Thoracic Surgeons Congenital Heart Surgery Database). Overall, operative mortality for CHD was recently reported at 3.7% over 2010 to 2013. Mortality rates vary by patient age and were 10.1%, 3.0%, 0.9%, and 1.7% for neonates, infants, children, and adults, respectively.[14] The national database available through the STS-CHSD provides valuable insight for operative risk stratification based on preoperative and intraoperative factors and are largely interpreted through utilization of STS European Association for Cardiothoracic Surgery Congenital Heart Surgery Mortality Categories (The Society of Thoracic Surgeons Mortality Categories) that were developed in 2009.[15]

Outcomes and practices in CHD continue to be relatively heterogeneous; this is largely because of surgeon-specific practices, institutional preferences, and heterogeneity inherent to CHD. These practice differences, in combination with significant differences in case mix across centers, make it difficult to isolate the effect of individual institutional therapeutic policies or treatment decisions on outcomes. There are some well-established factors that reduce operative risk that include safe resternotomy, minimization of CPB and aortic cross clamp times, optimal myocardial protection, minimization of blood product transfusion, and maximizing the functional durability of a surgical intervention.

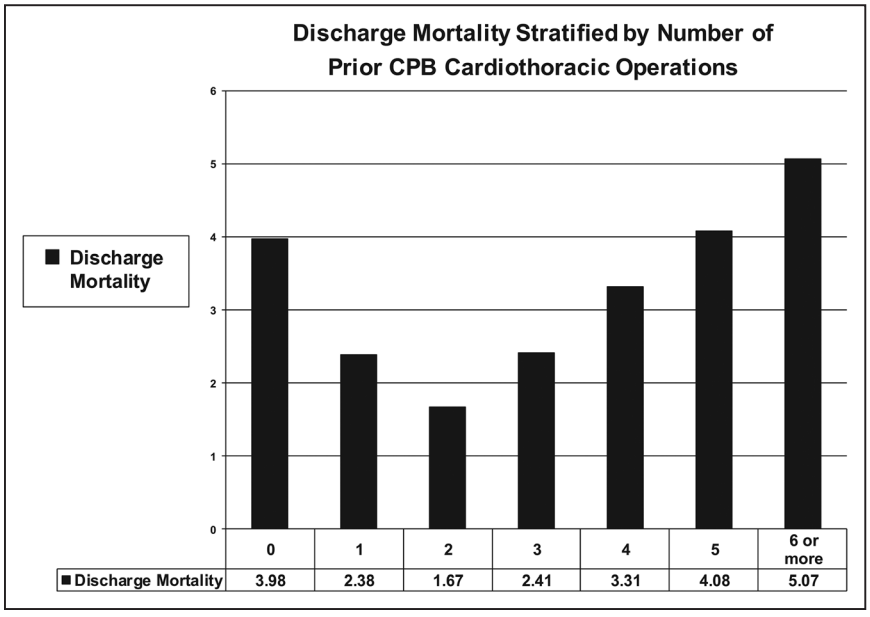

Figure 2. Discharge mortality stratified by number of previous cardiopulmonary bypass (CPB) operations. Discharge mortality of patients with congenital heart disease (CHD) stratified by number of previous cardiopulmonary operations from 2007 to 2011. Reproduced from Jacobs et al[10] with permission of the publisher. Copyright ©2014, Elsevier.

Table 3. Indications for Intervention for Valve-Related Abnormalities

Aortic stenosis	AV_{max} ≥4.0 m/s; mean gradient ≥40 mm Hg, AVAi ≤0.6 cm^2/m^2; SVi <35 mL/m^2
Aortic regurgitation	Doppler JW ≥65% of LVOT; VC >0.6 cm; Rvol ≥60 mL/beat; RF ≥50%; ERO ≥0.3 cm^2; LVESD >50 mm; holodiastolic aortic flow reversal
Mitral stenosis	MVA ≤1.5 cm^2, $T_{½}$ ≥150 ms
Mitral regurgitation	VC ≥0.7 cm; Rvol ≥60 mL; RF ≥50%; ERO ≥0.40 cm^2; LVESV ≥40 mm
Pulmonary stenosis	PV velocity >4 m/s, PIG >64 mm Hg
Pulmonary regurgitation	Color jet fills RVOT; RVEDVI >150 mm/m^2; holodiastolic pulmonary artery reversal
Tricuspid stenosis	$T_{½}$ >190 ms, tricuspid VA <1.0 cm^2
Tricuspid regurgitation	Central jet area >10 cm^2; VC >0.7 cm; hepatic vein flow reversal

Symptomatic patients may be advised intervention with lesser degrees of objective criteria above, when there is multivalvar involvement, depressed left ventricular systolic function, or when valve repair (as opposed to replacement) has high probability of success. These guidelines reflect indications for intervention for acquired heart disease and serve as a reference point for valve-related abnormalities in congenital heart disease. AV indicates aortic valve; AVAi, aortic valve area indexed; EF, ejection fraction; ERO, effective regurgitant orifice; LV, left ventricle; LVESV, left ventricular end-systolic volume; LVOT, left ventricular outflow tract; PV, pulmonary valve; PIG, peak instantaneous gradient; RV, right ventricle; RVEDVI, right ventricular end-diastolic volume indexed to body surface area; Rvol, regurgitant volume; SV, stroke volume; $T_{1/2}$, diastolic pressure half time; and VC, vena contracta.

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Reoperation

Computed tomography and MRI imaging are particularly valuable in preoperative evaluation of a patient with previous cardiac operation because of the superior visualization of anatomic structures in proximity to the sternum, which are at risk at time of resternotomy.[16]

Safe chest entry is of paramount importance in the setting of reoperation for CHD, and structures at risk may differ from those at risk during operations for acquired cardiac disease in which fundamental cardiac morphology is generally predictable.[11] In reoperations for CHD, the aorta and extracardiac conduits are at greatest risk of injury on re-entry, although there is not a clear association of structure at risk and underlying cardiac lesion.[11] Other structures at risk include the innominate vein, the right atrium, or RV depending on the primary diagnosis.

The amount of time since the most recent cardiac operation impacts safety of resternotomy.[11,16] Reoperation within the first 12 months from most recent operation is often more difficult secondary to edematous, vascular adhesions requiring a tedious dissection.[16] In contrast, increased time from the previous operation is associated with decreased risk of cardiac injury on sternal re-entry.[11]

In patients at high risk for cardiac injury at resternotomy, peripheral cannulation with institution of bypass prior to resternotomy can facilitate sternal re-entry by allowing decompression and dissection of structures at risk. In selected circumstances, CPB can be instituted for cardiac decompression and successful reentry, followed by separation from bypass so that a lengthy mediastinal dissection time can be performed off bypass, thus, minimizing total pump time, which has been shown to be a risk factor for early mortality.[11] After the lengthy mediastinal dissection has been completed, bypass is resumed for performance of the actual operation.

Importantly, previous invasive catheterization procedures or indwelling monitoring lines in both femoral arteries and veins may have resulted in occlusion/stenosis of these vessels. Assessment of peripheral vascular patency should be evaluated preoperatively if peripheral cannulation is being considered or required. Alternative peripheral cannulation sites include femoral arteries or veins, iliac arteries or veins, axillary arteries, innominate or carotid arteries, internal jugular vein (typically on the right), LV apex (via left thoracotomy), pulmonary artery (via left thoracotomy), and abdominal aorta.[16]

Intraoperative Techniques

Blood Conservation and Small Circuits

Blood transfusion in congenital heart surgery has been associated with increased morbidity and mortality. Blood product exposure has been associated with increased postoperative ventilation, longer intensive care unit stay, increased systemic inflammatory response, increased postoperative infections, and increased early and late mortality. In addition, there can be transfusion-related allosensitization[17] that can adversely affect future candidacy of transplantation in the event that it is needed. Many approaches to reducing blood product transfusion in cardiac surgery and congenital cardiac surgery specifically have been investigated, including minimizing blood dilution through use of small CPB circuits and protocol-driven transfusion practices based on threshold values of laboratory test values.[17,18]

CPB circuit miniaturization has been shown to be successful in reducing blood product transfusion through reduction of pump prime volume.[17] Strategies to reduce prime volume include minimizing the length of the cardiopulmonary tubing, using the smallest oxygenator possible, minimizing the size of the venous and arterial cannulae along with reducing the size of the tubing circuit, and smaller vacuum and venous reservoirs. Through utilization of these techniques, prime volumes can be reduced to 240 to 300 mL; however, the estimated blood volume of a 4-kg infant is 340 mL, and this still results in at least a 70% blood dilution.[17]

Use of ultrafiltration during the period of bypass can further hemoconcentrate by removing excess fluid. Ultrafiltration uses hydrostatic pressure to increase concentration of blood products through removal of excess fluid. Modified ultrafiltration provides another option for ultrafiltration after CPB has been discontinued. The extent of modified ultrafiltration strategies is institution dependent; generally, it is applied to children ≤20 kg, with a goal time interval (15–20 minutes), hematocrit (40%), or volume to be removed (750 mL/m^2) is established and achieved.[17]

In addition to reduction of prime volume and hemodilution of a patient's own blood, laboratory-based transfusion algorithms have proven effective in reducing the amount of transfusion compared with common/unstandardized transfusion

practices.[18] The following outcomes are from a prospective, randomized clinical trial, which compared transfusion practices based completely on laboratory testing compared with common practice of clinical testing with or without laboratory tests.[18] The outcomes of this study were substantial; the laboratory-based algorithm group had significantly less units of plasma (P=0.0002) and platelets (P=0.0001) transfused compared with the group under standard practice. The benefit of laboratory testing–based transfusion continued as patients transitioned to the intensive care unit, and multivariate analysis revealed that the transfusion algorithm decreased blood loss as measured by mediastinal chest tube output and surgical reoperation for bleeding while decreasing overall rates of transfusion.[18] Figure 3 outlines the transfusion algorithm used in this clinical trial, which is still being used by this group.[18]

Valve Surgery in CHD

Valvuloplasty Strategies

Valve repair is generally preferred to valve replacement in CHD; however, consideration must be given to the best approach to minimize the subsequent number of reoperations. This may lead a surgeon to consider offering valve repair(s) at a younger age before significant ventricular dysfunction has occurred to improve the result of the repair or alternatively may lead a surgeon to proceed with valve replacement when the repair was technically possible but suboptimal. In general, although these decisions are patient, surgeon, and valve specific, most studies have found improved late outcomes when valve repair is possible rather than valve replacement.

Tricuspid Valvuloplasty

TV disease in CHD is more common than historically appreciated and is present within a variety of congenital lesions. Ebstein anomaly and TV dysplasia are inherent lesions of the TV, yet other diagnoses associated with development of TV regurgitation are atrioventricular septal defects, pulmonary stenosis (or RV outflow tract obstruction), ventricular septal defect, or RV volume overload from other causes. Iatrogenic causes of TV disease include pacemaker lead–induced TV damage or TV injury during ventricular septal defect repair.[19]

Indications for operation because of TV dysfunction, most commonly regurgitation, in the CHD population are related to right-sided heart disease. Common symptoms include fatigue, decreased exercise tolerance, shortness of breath, edema, ascites, or failure to thrive. Importantly, symptoms are often absent. Other indications for intervention include the new onset of an atrial arrhythmia in the setting of right atrial enlargement or RV failure or progressive RV dilatation or deterioration in RV function on serial imaging studies. Surgery may be offered before the overt development of these symptoms if there are other cardiac lesions that need to be addressed, for example, pulmonary valve regurgitation causing RV enlargement and TV annular dilatation. Centers with high rates of successful TV repair may also consider offering surgery sooner rather than later if anatomy is favorable for repair.

Approach to valvuloplasty is dependent on the underlying pathogenesis of the TV dysfunction. When there is a structural problem beyond annular dilatation, a simple annuloplasty will not suffice, and the operative approach should include repair of the subvalvar apparatus or leaflet(s).[19] Surgical techniques to address structural abnormalities of the TV can be shared between Ebstein and non-Ebstein etiologies of TV disease. The cone repair and subsequent modifications have been widely adopted and have increased the spectrum of Ebstein anomaly that is amenable to repair without prosthetic replacement of the TV. The most readily applicable modifications to non-Ebstein TV disease include approximation of leaflet defect,

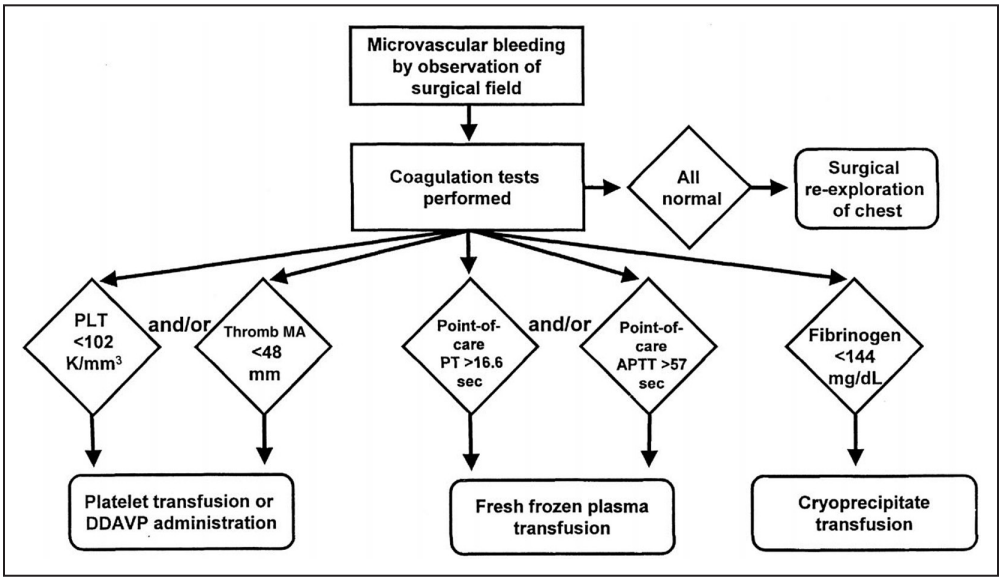

Figure 3. Intraoperative transfusion algorithm for nonerythrocyte component utilization after cardiopulmonary bypass. The intraoperative transfusion algorithm initiated after the clinical diagnosis of abnormal bleeding. After initial treatment, the applicable coagulation tests were repeated to evaluate the effectiveness of the therapy. APTT indicates activated partial thromboplastin time; DDAVP, desmopressin acetate; PLT, platelets concentrates; PT, prothrombin time; Thromb MA, thromboelastogram maximum amplitude. Reproduced from Nuttall et al[18] with permission of the publisher. Copyright ©2001, American Society of Anesthesiologists.

resection of excess leaflet tissue, creation of Gore-Tex artificial chordae, leaflet augmentation, and papillary muscle approximation.[20,21] Application of an annuloplasty band or ring may be required to stabilize the repair or further reduce the size of a dilated annulus.

Outcomes for TV repair versus replacement in the setting of CHD are not well established. The largest series to date[19] included over 500 patients undergoing TV surgery; repair was possible in 80%, while replacement was performed in the remaining 20%. Reoperation was not common; however, rates of successful TV repair were lower in the reoperative setting. Survival was significantly improved in those undergoing TV repair compared with those undergoing replacement. The need for reoperation for recurrent TV regurgitation was a significant risk factor for late mortality, which emphasizes the importance of trying to achieve good tricuspid competence at the initial tricuspid operation.

Iatrogenic causes of TV dysfunction secondary to pacemaker or defibrillator leads generally are related to leaflet perforation by the lead, entrapment of the lead in the TV apparatus, impingement of the leaflets by the lead, or adherence of the lead to the TV leaflet.[22] Repair techniques are dependent on the mechanism of injury but generally involve repositioning the lead, repair of the leaflet defect caused by the lead, and then moving the lead to either the inferoseptal or anteroinferior commissure so that leaflet excursion is not impeded. A banded annuloplasty is also generally performed to stabilize the repair and maintain a small annulus.[22] In adults, a 26 or 28 mm ring is used routinely. Rates of TV repair for lead-induced TV failure are lower than those for TV disease secondary to the congenital diagnosis. In a recent series of congenital patients with lead-induced TV regurgitation, the repair was possible in less than half of the patients.[22]

Mitral Valvuloplasty

Mitral disease in CHD can generally be divided into mitral stenosis (MS) and mitral regurgitation (MR). Both often present with other congenital diagnoses and age, and severity of presentation are dependent on the degree of stenosis or regurgitation at the mitral valve level and associated lesions. Pathological specimens of both MS and MR have led to detailed anatomic classifications; however, the more useful classification in the surgical setting is the functional classifications outlined by Carpentier et al.[23] MS is functionally subdivided into MS with normal papillary muscles, Type A, or MS with abnormal papillary muscles, Type B. Type A is then subdivided into stenosis at the supravalvar ring or because of leaflet fusion. Type B is broken down to parachute deformity or primary papillary muscle abnormality.

Classification of MR is slightly more complex; there are 3 initial groupings: normal leaflet motion (Type I), leaflet prolapse (Type II), and restricted leaflet motion (Type III). Type I, normal leaflet motion, is further subdivided into annular dilatation, cleft leaflet, and leaflet defect. Type II, leaflet prolapse, is subdivided into chordal elongation, papillary muscle elongation, or absence of chordae. Type III, restricted leaflet motion, is divided into restricted motion with normal papillary muscles, A, and subdivided into commissure papillary fusion or short chordae, or divided into restricted motion with abnormal papillary muscles, B, subdivided into parachute mitral valve, hammock mitral valve, or papillary muscle hypoplasia. The reported rates of associated congenital lesions is highly varied for both MS and MR, likely because of the inherently heterogeneous nature of this disease process; reported rates range from 25% to nearly 100%.[24–26]

Advancements in rates of repair compared with valve replacement, particularly in neonates and infants, is largely attributable to advancements in 2-dimensional and 3D echocardiography,[26] which have facilitated the understanding of these anatomically complex lesions and provide the platform for improved preoperative planning.

Techniques to approach MS includes initial examination at the supravalvar level, progressing to the valvar level, and subsequently transitioning to the subvalvar level, where most of the pathology is located.[26] Stenotic pathology at the supravalvar level is most commonly the supravalvar mitral ring, which can be resected with a combination of sharp and blunt dissection through creation of a plane between the abnormal connective tissue and the leaflet.[26] Valvar stenosis is generally related to decreased leaflet mobility, which may involve either or both the anterior and posterior leaflets; the abnormality generally started at the base of the leaflet and extends out toward the free edge of the leaflet. The thickened membrane covering the leaflet tissue is sharply resected without creating an injury to the leaflet tissue to remain in place. Despite complete resection of the fibrous membrane, the leaflet tissue may still be shortened and require patch augmentation for adequate leaflet coaptation. Autologous or processed bovine pericardium or other prosthetic patches can be used, but long-term outcomes comparing these are not well established. Subvalvar papillary muscles and chordal structures harbor the majority of pathology in MS. The structures are often fused with fibrous connective tissue and separation is difficult, although splitting and dividing these fused structures is often the goal. Posterior or anterior leaflet detachment may facilitate visualization for division of the numerous secondary chordal attachments. After connective tissue removal, leaflet coaptation should be assessed as augmentation may be required.[26]

Repair in the setting of MR is also dependent on the mechanism of regurgitation and most frequently involves the use of an annuloplasty ring, chordal or papillary muscle shortening, or leaflet augmentation.[24] The use of an annuloplasty ring in young patients has been controversial given the somatic growth potential of these children and future possibility for problems associated with an undersized mitral annuloplasty ring compared with the appreciation of benefits of annular remodeling.[24,25] The decision to use or not use an annuloplasty ring is particularly challenging in younger, smaller patients in comparison to older children. Partial (eccentric) bands are an option when growth is incomplete. Alternative solutions also include the use of a patch to increase the size of the anterior leaflet and allow for the use of an adult-sized ring; a smaller ring may be used and repercussions of the smaller ring addressed in the future, and most commonly in neonates and infants, annular reduction is performed without use of an annuloplasty ring.[25]

Aortic Valvuloplasty

AV pathology in CHD is divided into aortic stenosis and aortic regurgitation. Aortic stenosis is largely treated by percutaneous balloon dilation; however, some may require subsequent operation for residual stenosis or aortic regurgitation after balloon valvotomy. Aortic regurgitation in the setting of CHD is commonly attributable to bicuspid AV, previous intervention for aortic stenosis, or Marfan syndrome. Less common etiologies include rheumatic heart disease, Turner syndrome, aortic cusp prolapse secondary to ventricular septal defect, tetralogy of Fallot, or truncus arteriosus. Regardless of pathogenesis, aortic regurgitation often presents with limited symptomatology. Indications for intervention are usually determined by changes in LV size (dilation) from increased stroke volume or the development of progressive LV dysfunction. The decision to proceed to surgery is dependent on the pathogenesis of aortic regurgitation. If the patient has associated aortic dilatation from either bicuspid AV or Marfan's syndrome, the size or change in size of the aorta may likely dictate timing of the operation. Asymptomatic aortic aneurysms should be repaired when aortic size index (aortic diameter in centimeter/body surface area in meter squared) is ≥2.75 cm/m^2.[13,27,28] In a patient without aortic dilatation, development of symptoms, increasing ventricular size, or decreased ventricular function on imaging are indications for operative repair.

AV repair is generally possible in the setting of aortic regurgitation because of a bicuspid AV, including regurgitation after balloon valvotomy for aortic stenosis. Results of bicuspid AV repair are most thoroughly described.[29,30] Standard repair techniques include commissural sutures and plication of inadequately fused cusps. Patients with large roots require mobilization of the commissures and reattachment of the commissures distally in the tube graft being used to replace the ascending aorta.[29] Freedom from reoperation in a large series of AV repair for bicuspid AV disease was recently reported and was 95%, 87%, 78%, and 64% at 1, 5, 10, and 15 years, respectively. This averaged to a risk of reoperation of ≈2.6% per year. The most common pathogenesis of reregurgitation was cusp prolapse, and the majority of patients who required reoperation underwent AV replacement.

The role of valve sparing root replacement has also been reported in patients with CHD.[31] This includes patients who have previously undergone the arterial switch procedure, Ross procedure, and to a lesser extent, other conotruncal anomalies. This has the advantage of valve preservation while also addressing dilatation of the ascending aorta. In general, valve sparing root replacement is preferred when the AV is tricuspid and relatively competent (less than moderate regurgitation). There is also a role for valve sparing root replacement in the setting of bicuspid AV, but patient selection is essential in this group of patients, particularly, when aortic regurgitation is present and concomitant AV repair is needed at the time of valve sparing root replacement.[32]

Valve Replacement Strategies

All attempts to avoid valve replacement in neonates and infants should be pursued; however, valve replacement is becoming increasingly common in CHD secondary to increased survival of patients with CHD into adulthood and subsequent valve deterioration. Valve intervention is the most indication for reoperation in adults with CHD,[11] and pulmonary valve replacement is the most frequent operative procedure. In addition, ≈25% of adults with valvar problems secondary to CHD presenting for reoperation require intervention on multiple valves.[12]

The ideal prosthesis for a patient with CHD would be one that is available in multiple sizes, handles easily, has low risk of thrombosis or dysfunction, has good long-term function, and is of low cost.[33] Options include bioprosthetic (porcine or pericardial), autograft (Ross procedure), homograft, and mechanical. Selection is determined by multiple factors that include number of prior operations and the number of valves involved, patient age, sex and pregnancy considerations, patient preferences related to lifestyle, and issues related to warfarin anticoagulation and chronic international normalized ratio monitoring.

Bioprosthetic Valve Replacement

Valve selection is individualized for a patient at a particular point in time; however, generally bioprosthetic replacement is usually preferred in the right heart (tricuspid and pulmonary positions), given the relative good durability in these locations and ease of subsequent percutaneous valve-in-valve options in the future. Furthermore, while mechanical valves are an option in the pulmonary or tricuspid position, it does require higher international normalized ratio levels (3-4) because of higher thrombosis rates noted in right-sided locations of the heart.[34,35]

Pulmonary valve replacement may or may not be accompanied by RV outflow tract reconstruction. Potential nonmechanical substitutions for the pulmonary valve include aortic or pulmonary homografts, bovine jugular vein grafts, Dacron porcine-valved conduits, and percutaneous bioprosthesis.[33] Long-term results of the ideal bioprosthesis is not clear; some groups report superiority of the Dacron porcine-valved conduits compared with homografts, while others were not able to show a difference between these valve types.[33] Bovine vein conduits may be superior to homografts; however, late results are not completely clear. In general, stented bioprosthesis perform well in both the pulmonary and tricuspid positions. Often they are chosen in situations where subsequent valve-in-valve therapy by means of catheter intervention is anticipated because they provide a suitable landing zone for the future catheter-based approach.

Bioprostheses are often favored in the tricuspid position given, good valve durability, particularly in adult patients, and no need for chronic systemic anticoagulation[22] when rhythm is normal. This improved longevity may be related to decreased flow rates and trauma to the prosthetic valve on the right side. Pericardial bioprostheses, however, have been found to decrease durability in the tricuspid position that may be related to stiffer leaflets inherent to the pericardial structure.[22] Whether newer generation pericardial valves have improved durability remains to be demonstrated. Conduction tissue may be at risk at time of TV replacement and is dependent on the pathogenesis of tricuspid disease. When the TV is in the normal location, valve sutures are confined to native leaflet tissue,

and the risk of heart block is low. Conversely, in the setting of Ebstein anomaly, septal leaflet tissue is usually absent, and the conduction tissue is vulnerable to injury. In this setting, the prosthesis is usually positioned to the atrial side of the conduction tissue to avoid iatrogenic heart block. This may result in the need to leave the coronary sinus draining below the prosthesis into the RV if the coronary sinus is in close proximity to the conduction tissue that is generally marked by a small vein adjacent to the membranous septum.[20,21]

Postoperative management for bioprosthetic valve replacement (in any location) generally involves short-term warfarin anticoagulation for 3 to 6 months followed by aspirin therapy indefinitely. The use of novel oral anticoagulants has not been adequately investigated in the setting of prosthetic valves.

Mechanical Valve Replacement

Mechanical prostheses are generally preferred on the left side of the heart, in both the aortic and mitral position. Interestingly, the literature has demonstrated a survival benefit of mechanical valve replacement in both the aortic and mitral locations in adults <65 years of age.[36–41] This may be because of the constant hazard of the hemodynamic alterations related to a deteriorating bioprosthesis in addition to the operative risk related to reoperation. These issues must be considered when selecting a prosthesis in an individual patient, particularly in the current era of enthusiasm for future percutaneous valve-in-valve therapy.

Mechanical valves may also be considered in the right heart in selected individuals. Situations when mechanical valve replacement might be appropriate in either the tricuspid or the pulmonary positions would be in patients with multiple previous valve replacements, in those with documented short bioprosthetic valve longevity, or in patients requiring systemic warfarin anticoagulation for another reason.[33] Right ventricular function should be adequate to facilitate proper disc opening and closing.

Slight upsizing of the prosthesis should be considered in children to minimize or avoid patient–prosthesis mismatch. In the aortic position, the prosthesis may be upsized through utilization of posterior annular enlargement procedures, Manougian or Nicks, or anterior annular enlargement with a Konno–Rastan procedure. In the mitral position, positioning the prosthesis in the left atrium can also allow slight upsizing of the prosthesis, but caution is advised to avoid potential obstruction of the pulmonary veins (particularly in infants) and to insure satisfactory disc motion that can be compromised by contact with the left atrial wall or residual native leaflet tissue.

Mechanical valves are typically well tolerated in patients with CHD, and advancements in home anticoagulation monitoring have shown further improvements in reducing complications related to anticoagulation therapy.[42,43]

Ross Procedure

An alternative to bioprosthesis or mechanical AV replacement is the Ross procedure in which the AV is replaced with a pulmonic valve autograft, and the pulmonary root is reconstructed with a homograft (aortic or pulmonary). This technique is typically applied with either aortic stenosis or regurgitation. The Ross procedure is the procedure of choice in infants and young children when AV replacement is required, and the Ross–Konno procedure is the best option in infants and young children in the setting of complex LV outflow tract obstruction, including aortic annular hypoplasia.[44–46] However, the Ross procedure is more controversial in young and middle-aged adults because it places 2 valves at risk at time of surgery, the diseased AV and the normal functioning pulmonary valve, when results with both mechanical and bioprosthetic are satisfactory. In addition, both the neoaortic and pulmonary valves are at risk for needing reintervention, and the reoperation can be difficult depending on the total number of prior operations.[33,47] Benefits of the Ross procedure are clear in infants and children and include a superior hemodynamic profile of the pulmonary valve in the aortic position, avoidance of anticoagulation, and growth potential, and the role of the Ross procedure in the adult population should be individualized.

Valve-in-Valve Replacement

Percutaneous valve-in-valve therapy is being increasingly used in CHD to avoid or delay repeat operation. Percutaneous replacements have now been performed in all positions. In the younger population, most have been performed in the setting of prior bioprosthetic valve replacement secondary to prosthesis deterioration.[48–50] The advantage is a safe, solid landing zone with low probability of adjacent coronary compromise. The importance of slightly oversizing the stented bioprosthesis at the time of surgical implantation cannot be overemphasized to facilitate future valve-in-valve therapies, which can be more than once if there is enough space in the original surgically placed bioprosthesis. Percutaneous interventions are a powerful adjunct to operative management; this therapeutic option may alter the decision-making process for valve selection, avoiding anticoagulation with mechanical valves in favor of subsequent valve-in-valve replacement options.

Operative Arrhythmia Management

Arrhythmias constitute a major source of morbidity and mortality in short-term and long-term follow-up of patients with CHD. Arrhythmia management in patients with CHD is complicated by the heterogeneity of anatomic presentation, arrhythmia types, potential influence of myocardial scarring from previous surgical intervention, and potentially limited approach through percutaneous techniques. Etiologies of and therapies for arrhythmias in CHD are increasingly being elucidated through experimental models and clinical studies. Although detailed explanation of this is outside the scope of this review, the following is an overview of ablative techniques, pacing, and implantable cardioverter-defibrillator (ICD) utilization in CHD. It is extremely important to consider both existing and potential arrhythmias in deciding an operative plan for patients with CHD because many techniques to address arrhythmias need to be performed using an operative approach. Although atrial arrhythmias are most common, the potential for ventricular arrhythmias must also be considered.

Ablation

Intra-atrial reentry tachycardia (IART) is a form of macroreentrant atrial tachycardia related to atrial flutter and is one of the most common arrhythmias seen in long-term follow-up of

CHD. The circuit may revolve around any conduction barriers in the heart (atriotomy incisions, baffles, patches, etc) and frequently involves the typical atrial flutter isthmus of the right atrium or other areas of slow atrial conduction. The goal of ablative therapy is to eliminate areas with diseased conduction, thus, removing the slow conduction that promotes the reentrant arrhythmia.[51] The energy source of surgical ablations is generally radiofrequency or cryoablation, and both techniques have been shown to be relatively equivalent in retrospective studies. However, a transmural lesion causing a complete line of conduction block must be created in order for the lesions to be effective because any gap in the line may serve as a nidus for IART.[52] It is also important to minimize potential damage to adjacent structures when performing the ablation.

In regards to IART, the inferior vena cava, TV, coronary sinus, right atrial appendage, and fossa ovalis are the anatomic landmarks of interest (Figure 4).[53] Even in complex CHD, the IART circuit frequently involves the right atrial cavotriscupid isthmus, between the tricuspid annulus, coronary sinus, and inferior vena cava, and targeted ablation in this area can be highly effective. The goal for a surgical ablation is to create lines of block between these structures at time of surgical ablation (a right-sided maze procedure) because IART requires a critical mass of atria to maintain the reentrant circuit.[51] Anatomic variants present in CHD may complicate the ablative approach, for which the surgeon must create a personalized lesion set to accomplish a successful ablation incorporating scars, patches, and baffles. Either a traditional cut-and-sew approach, cryoablation energy, radiofrequency energy, or an alternative energy source (such as microwave energy) can be used to create the lesions, with the most important factor in success being creation of a transmural lesion connecting structures to create a line of complete conduction block.

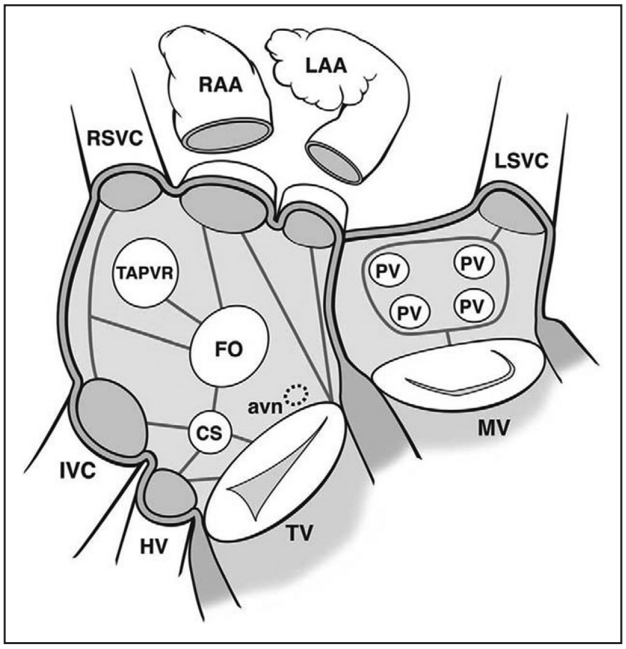

Figure 4. Schematic representation of the possible lines of ablation to treat macro reentrant atrial tachycardia in the presence of various atrial anomalies associated with complex congenital heart disease. avn indicates atrioventricular node; CS, coronary sinus; FO, foramen ovale; HV, hepatic vein; IVC, inferior vena cava; LAA, left atrial appendage; LSVC, left superior vena cava; MV, mitral valve; PV, pulmonary valve; RAA, right atrial appendage; RSVC, right superior vena cava; TAPVR, total anomalous pulmonary venous return; and TV, tricuspid valve. Reproduced from Mavroudis et al[53] with permission of the publisher. Copyright ©2008, The Society of Thoracic Surgeons.

Other types of tachycardia may play a role in the preoperative and postoperative management of patients with CHD. Focal or automatic atrial tachycardia (also known as ectopic atrial tachycardia) is seen. This type of tachycardia may resolve with resection and debulking of atrial tissue because atrial stretch and scarring may play a role in the pathogenesis of this type of tachycardia.[51] However, in some patients, medical therapy or catheter-based ablation therapy may be required for long-term control. Focal atrial tachycardias may be seen in the postoperative period after cardiac surgery, particularly in patients who have had extensive surgery on atrial tissue. These tachycardias may resolve spontaneously, especially, if they occur within 24 hours of the operation, but fast or symptomatic tachycardias occurring >3 to 4 days after surgery will likely be long-term conditions requiring intervention. Accessory pathways are an occasional cause of supraventricular tachycardia, particularly, in patients who have Ebstein's anomaly, with ≈15% to 30% of patients with Ebstein anomaly having an accessory pathway.[54] Although the initial therapy for tachycardias caused by an accessory pathway was surgical ablation, treatment approaches have largely transitioned to transcatheter radiofrequency ablation or cryoablation.[51] For this reason, it is important to consider ablation therapy prior to surgical intervention if the surgery will limit access to the heart (such as in a cavopulmonary anastomosis) or to the tricuspid or mitral valve annulus (such as in a cone repair or atrioventricular valve replacement). Some centers have recommended routine preoperative electrophysiology evaluation prior to surgery in patients at high risk for arrhythmias, such as patients with Ebstein anomaly.[55]

Atrial fibrillation (AF) is an arrhythmia seen in older patients who have had extensive atrial surgery or have poor ventricular function. When treating AF, the goal is to eliminate the potential substrates of AF, which frequently reside in the left atrium, maintain sinus node function, and sustain atrial function.[52] Surgical approaches traditionally include the Cox-Maze III, left atrial Maze, and biatrial Maze.[52] As sinus node dysfunction may play a role in the development of AF, pacemaker therapy is frequently used in conjunction with surgical ablation to decrease the incidence of AF. Transcatheter techniques are generally complex and require transseptal or transbaffle perforation, and surgical ablation techniques tend to have a higher success rate than catheter-based ablations in patients with complex CHD. Therefore, strong consideration should be given to placement of lesions for AF in patients who have clinically experienced the arrhythmia. As in IART ablations, multiple different techniques, including cut-and-sew techniques, as well as radiofrequency-, microwave-, or cryoablation-based approaches, are available, with each technique having advantages and disadvantages.[56] Left atrial Maze is focused on pulmonary vein isolation and division of reentry circuits from the left atrioventricular valve isthmus, coronary

sinus, and Bachmann's bundle.[52] Left atrial appendage ligation is commonly included in the left atrial Maze to remove it as a potential thromboembolic source.

Patients with known arrhythmias should have their arrhythmias addressed before or at the time of surgery. However, the appropriateness of prophylactic arrhythmia operations in CHD is dependent on the risk of an individual developing an arrhythmia, which is primarily based on the individual's underlying diagnosis. Importantly, considerations for prophylactic procedures must include the efficacy of the procedure; simplicity of the procedure to include in otherwise planned operative intervention; and ability to minimize and demonstration of low rates of complications or comorbidities. Although there are no uniform guidelines, some centers have recommended intraoperative prophylactic arrhythmia lesions be placed in adolescents and adults undergoing surgery for Ebstein anomaly or univentricular hearts (particularly in patients undergoing a Fontan revision) and in adults undergoing repair of an atrial septal defect or rerepair of tetralogy of Fallot.[52] Although these lesions may help in preventing arrhythmias, there is some concern about their potential proarrhythmic effect because ultimately electric connections are re-established across created lines of block, and there is also concern about disrupting atrial synchrony by creating lines of conduction block.

Pacemakers

Sinus node dysfunction is a common problem in patients with CHD either because of the intrinsic nature of the CHD or because of surgery that may disrupt the sinus node itself or the sinus node artery. With newer surgical techniques and a better understanding of cardiac anatomy, the incidence of surgically created atrioventricular (AV) block is low, but continues to be an issue after repair of CHD. In the modern era, the incidence of postoperative heart block requiring permanent pacemaker placement immediately after congenital heart surgery is low (1%), with the highest incidence being in patients undergoing a double switch operation or AV valve replacement (Table 4).[57] Postoperative AV block is a class I indication for pacemaker placement because of the high mortality rate (≤60%) in this population, regardless of the junctional escape rate or stability of the patient.[58] Pacemaker placement should be considered in patients with CHD and impaired hemodynamics because of sinus bradycardia or loss of AV synchrony and is routinely performed in patients at the time of Fontan conversion because of the high incidence of bradycardia and tachy-brady syndrome in these patients. A pacemaker can also be considered in adolescents with CHD, with resting heart rate <40 bpm or >3 second pauses.

Pacing leads may be placed transvenously through the venous system into the atrium or ventricle or placed epicardially on the outside of the heart. An epicardial approach is typically used in younger patients or in patients with limited venous access to the heart. After transvenous lead placement, 25% of all CHD patients have complete or partial obstruction 6.5 years after implant, with a risk of ≤40% in younger patients with transvenous leads, particularly those <6 years of age.[59] An epicardial approach preserves venous access to the heart when new leads need to be placed must be considered. Epicardial leads are also placed when there is need for a ventricular lead in the presence of a mechanical AV valve. As there is concern for long-term AV valve problems with a lead crossing the valve, epicardial lead placement may be considered in AV valve repairs or in patients with Ebstein anomaly. In addition, transvenous leads incur a >2-fold increased risk of systemic thromboemboli in patients with intracardiac shunts even if no documented right-to-left shunting occurs.[60] For this reason, the 2008 American College of Cardiology/American Heart Association guidelines[61] for the management of adults with CHD state that epicardial pacemaker and device lead placement should be performed in all cyanotic patients with intracardiac shunts who require devices (class I indication).

In young, active patients, pacemaker leads do not last as long as in adults because of the high level of activity in younger patients, as well as patient growth. Epicardial leads have a good longevity, although they tend to not be as durable as transvenous leads. In one study, epicardial atrial lead survival was 99% at 1 year and 72% at 10 years, while epicardial ventricular lead survival was 97% at 1 year and 60% at 10 years.[62] The number of prior surgeries and surgical approach (thoracotomy versus subxiphoid versus sternotomy) were not related to atrial or ventricular lead survival.

Table 4. Incidence of Surgical Complete Heart Block Requiring a Pacemaker by Operation

Operation	Complete Heart Block
Double switch operation	15.6%
Tricuspid valve replacement	7.8%
Mitral valve replacement	7.4%
Atrial switch with VSD	6.5%
Rastelli operation	4.8%
Ebstein repair (<30 d)	4.2%
Konno operation	4.1%
Arterial switch with VSD	3.4%
Aortic valve replacement	2.9%
Subaortic stenosis resection	2.8%
Ross operation	2.4%
Complete AV canal repair	2.2%
Mitral valvuloplasty	1.7%
Coarctation with VSD	1.7%

AV indicates aortic valve; and VSD, ventricular septal defect.

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Implantable Cardioverter-Defibrillators

Ventricular arrhythmias are a significant concern in patients with CHD. While atrial arrhythmias may cause symptoms, ventricular arrhythmias may potentially result in sudden cardiac death. Although antiarrhythmic therapies and ablation therapies may decrease the incidence of ventricular arrhythmias, by far, the most effective therapy for decreasing the incidence of sudden cardiac death is placement of an ICD. Although criteria for ICD placement in adults is well established from multiple large studies, pediatric data are limited, and indications

for ICD placement in patients with CHD are somewhat more controversial. Guidelines for ICD implantation in pediatric and CHD have been published, but as the recognition of patients at risk for sudden death increases, the indications for ICD placement will also likely expand. According to the 2008 American College of Cardiology/American Heart Association/Heart Rhythm Society Guidelines, an ICD is indicated for any survivor of a cardiac arrest because of a ventricular arrhythmia if there is no completely reversible cause.[58] An ICD is also indicated for patients with structural heart disease and spontaneous sustained ventricular tachycardia or for patients with syncope of undetermined origin with clinically significant sustained ventricular arrhythmias induced at electrophysiological study. An ICD should also be considered in patients with poor systemic ventricular ejection fraction because these patients are at risk for sudden death and in patients with syncope and advanced structural heart disease in whom thorough invasive and noninvasive investigations have failed to define a cause.

There are several different techniques available to place ICDs. A traditional transvenous route using the subclavian vein is the most common approach used to place the ICD lead into the ventricle. However, similar to pacemakers, the presence of intracardiac shunting or small patient size may necessitate placement of an epicardial system. It is important to consider the need for an ICD prior to a planned cardiac surgery because an epicardial approach may be the best device configuration in patients with complex CHD. Epicardial systems may be placed using a standard epicardial ventricular pace sense lead with a dedicated coil placed in the pericardial sac. The leads are then tunneled to a pocket in the abdomen where the ICD generator is placed. This technique is effective with low defibrillation thresholds and may even be achieved by a minimally invasive technique.[63]

Postoperative Care

Postoperative management begins in the operating room. During the separation from CPB, attention to both LV and RV performance is critical. Poor function of the RV is not uncommon for many congenital lesions, particular those lesions that require a right ventriculotomy or those lesions that result in pulmonary or tricuspid regurgitation. In these situations, it is critical that RV preload and RV afterload are optimized through various modalities. Modified ultrafiltration is routinely performed for 15 to 20 minutes in children ≤20 kg, and continuous ultrafiltration during the bypass run is used in older children and adults. Other strategies include optimal mechanical ventilation, appropriate inotropic support, careful fluid management, and avoidance of acidosis and arrhythmias.[64]

Selection of inotropic support at the time of discontinuation of CPB and in the early postoperative period is largely driven by institutional experience and practice algorithms. The combination of epinephrine and milrinone are commonly used as initial drug therapy after separation from bypass. The application of vasoconstrictors, such as vasopressin or norepinephrine, is reserved for those patients with systemic vasoplegia. Elevated serum lactate levels are common in the early postoperative period and related to longer duration of circulatory arrest (P=0.001), age at surgery <15 days (P=0.002), and preoperative intubation (P=0.002).[65]

The combination of epinephrine, milrinone, and selective use of nitric oxide optimizes oxygenation and minimizes RV afterload. It is not clear how inotropic-responsive a dilated and thinned RV can be, but there is evidence that milrinone and epinephrine can significantly improve cardiac index, SvO_2 (mixed venous oxygen saturation), CVP (central venous pressure), and increased RV contractility in animal models.[64] In the postoperative phase, once cardiopulmonary stability is established, it is important to wean off epinephrine and milrinone as timely as possible to avoid the potential side effects. For example, milrinone has been associated with tachyarrhythmias in the early postoperative phase after CHD surgery.[66] When RV failure is significant, especially if hypoxia is present during weaning from bypass, early initiation of nitric oxide may help with gas exchange; the duration of nitric oxide is short term and is typically weaned off within 24 to 48 hours with or without a bridge to another pulmonary artery vasodilator, for example, sildenafil. Furthermore, avoidance of respiratory and metabolic acidosis reduces pulmonary vascular resistance, which also aids in reducing RV afterload. To minimize RV dilatation when it is dilated and poorly functioning, increasing heart rates (100–120 bpm) with temporary atrial pacing may be needed.[20,21,67,68] This helps to minimize RV distention by reducing filling time and minimizes tension on a tricuspid repair, annular, and ventricular suture lines. Low-dose vasopressin can be helpful with right-sided heart failure because systemic vasoplegia is not uncommon and may be caused by the use of preoperative afterload reducing agents, liver congestion, or humoral factors from atrial natriuretic peptide secondary to atrial dilation. Optimal fluid management for these patients is challenging, and relative hypovolemia helps avoid RV distention. Fluid administration is slow and goal directed while avoiding transfusions. In general, a target right atrial pressure of <12 to 15 mm Hg is preferred. Importantly, this strategy of relative hypovolemia, relative tachycardia, and use of inotropes typically results in metabolic acidosis in the first 6 to 12 hours after surgery.

Mechanical ventilation strategies include minimizing end-expiratory pressure, decreasing mean airway pressure, decreasing inspiratory time, and choosing ventilation mode that promotes reduction of intrathoracic pressure because cardiac output is preload dependent. Early extubation is balanced with the need of mechanical ventilator support, particularly if there is significant pulmonary congestion compromising gas exchange. If hemodynamics can tolerate diuretics, early diuresis may improve pulmonary vascular congestion and pulmonary compliance and optimize gas exchange.

Atrial arrhythmias are common,[20,21,69] and the early and temporary use of amiodarone is usually effective in many patients with RV pathology, for example, Ebstein anomaly. The management of persistent arrhythmias can be challenging and requires advice and guidance from the electrophysiology team. It is debated whether early pre-emptive intervention and medical management with amiodarone versus other antiarrhythmics will require further research.

Medical therapy at hospital discharge for many patients undergoing valve surgery often includes β-blocker therapy.[67] In the setting of depressed systemic ventricular function, the addition of angiotensin-converting enzyme inhibitor is

common. Short-term sildenafil (6–8 weeks) can be considered in patients with poor RV function, although evidence for this strategy is lacking. The addition of amiodarone therapy for 2 to 3 months can also be considered when transient atrial or ventricular arrhythmias are present; input from the electrophysiology team is recommended in these extenuating circumstances to insure that the best medical regimen is selected. Because late arrhythmias, both atrial and, to a lesser extent, ventricular, are the most common late complication after surgery for almost every heart defect, proper lifelong arrhythmia surveillance (ECG, Holter, and selective electrophysiological studies) with appropriate exercise testing is essential.

Quality Metrics

There has been dramatic improvement in the perioperative care of children and adults with CHD in the recent era. Outcomes of surgery have improved, and patients' survival into adulthood is expected even with the most complex cardiac defects. The need to establish standard ways to report outcomes and measure quality with the overall goal of improving care is increasingly important.

There are several parameters that have to be followed to minimize interinstitutional variability and report meaningful outcomes[70]:

1. Use of a common language and standard nomenclatures
2. Collection of information using an established core data set within a database
3. Reliable system to evaluate the complexity of cases
4. Ensure the collected data are complete and accurate
5. Valid follow-up
6. Cooperation between the medical and surgical subspecialties
7. Systematic quality measurement and quality improvement

The STS-CHSD is currently being used for outcomes documentation[71,72] and quality measurement.[73] There are variations between center's quality and outcomes; however, the goal should be to consider high performance centers as a useful resource for the best practice and the low performing centers are where initiation of quality improvement should start.

The quality of care for patients with CHD can be evaluated with several metrics. These metrics emerged from the collaboration between the STS and the Congenital Heart Surgeons' Society.[74] There are several of these metrics (Table 5)[75] with the hope that these measures will serve to appropriately assess quality and eventually improve outcomes. These quality measures are also in harmony with the current nomenclature, standards, and rules currently used by the STS and the European Association for Cardiothoracic Surgery in their databases.[75,76]

These quality measures also need to be assessed in regards to reliability, validity, importance, scientific acceptability, usability, and feasibility. These measures also must span all the temporal, geographical, and subspecialty boundaries.[77]

Table 5. Quality Measures for Congenital and Pediatric Cardiac Surgery

1. Participation in a National Database for pediatric and congenital heart surgery
2. Multidisciplinary rounds involving multiple members of the healthcare team
3. Availability of institutional pediatric extracorporeal life support (ECLS) program
4. Surgical volume for pediatric and congenital heart surgery: total programmatic volume and programmatic volume stratified by the 5 STS-EACTS Mortality Categories
5. Surgical volume for 8 pediatric and congenital heart benchmark operations
6. Multidisciplinary preoperative planning conference to plan pediatric and congenital heart surgery operations
7. Regularly Scheduled Quality Assurance and Quality Improvement Cardiac Care Conference, to occur no less frequently than once every 2 months
8. Availability of intraoperative transesophageal echocardiography (TEE) and epicardial echocardiography
9. Timing of antibiotic administration for pediatric and congenital cardiac surgery patients
10. Selection of appropriate prophylactic antibiotics for pediatric and congenital cardiac surgery patients
11. Use of an expanded preprocedural and postprocedural time-out
12. Occurrence of new postoperative renal failure requiring dialysis
13. Occurrence of new postoperative neurological deficit persisting at discharge
14. Occurrence of arrhythmia necessitating permanent pacemaker insertion
15. Occurrence of paralyzed diaphragm (possible phrenic nerve injury)
16. Occurrence of need for postoperative mechanical circulatory support (IABP, VAD, ECMO, or CPS)
17. Occurrence of unplanned reoperation or unplanned interventional cardiovascular catheterization procedure
18. Operative mortality stratified by the 5 STS-EACTS Mortality Categories
19. Operative mortality for 8 benchmark operations
20. Index cardiac operations free of mortality and major complication
21. Operative survivors free of major complication

CPS indicates cardiopulmonary support system; EACTS, European Association for Cardio-Thoracic Surgery; ECMO, extracorporeal membrane oxygenation; IABP, intra-aortic balloon pump; STS, Society of Thoracic Surgeons; and VAD, ventricular assist device.

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Public Reporting

We live in a new era with increased transparency.[78] In early 2015, the STS began publicly reporting the outcomes of pediatric and congenital cardiac surgery. This was based on the 2014 STS-CHSD morality risk model (Table 6).[79] Centers need to be aware of the importance of complete and accurate data entry because this model is based on patient-level factors and procedural factors.[79]

For all participants, the STS reports the overall operative mortality rate over a 4-year period for all ages and for each of the 5 Society of Thoracic Surgeons mortality categories. This report also provides a summary in regards to number of observed patients included in the calculation of operative mortality and the observed and the expected rates as percentages.

The star rating of the centers is then based on the observed-to-expected (*O*/*E*) mortality ratio. There should be a concerted

Table 6. The STS Congenital Heart Surgery Database Mortality Risk Model: List of Included Variables for Which the Model Adjusts

Variable
Age group
Primary procedure*
Weight (neonates and infants)
Prior cardiothoracic operation
Any noncardiac congenital anatomic abnormality
Any chromosomal abnormality or syndrome
Prematurity (neonates and infants)
Preoperative factors
Preoperative/preprocedural mechanical circulatory support
IABP, VAD, ECMO, or CPS
Shock, persistent at time of surgery
Mechanical ventilation to treat cardiorespiratory failure
Renal failure requiring dialysis or renal dysfunction
Preoperative neurological deficit
Any other preoperative factor†

CHSD indicates congenital heart surgery database; CPS, mechanical cardiopulmonary support system; ECMO, extracorporeal membrane oxygenation; IABP, intra-aortic balloon pump; STAT, The Society of Thoracic Surgeons–European Association for Cardiothoracic Surgery Congenital Heart Surgery Mortality Categories; STS, Society of Thoracic Surgeons; and VAD, ventricular assist device.

*The model adjusts for each combination of primary procedure and age group. Coefficients obtained via shrinkage estimation with STAT mortality category as an auxiliary variable.

†Any other preoperative factor is defined as any of the other specified preoperative factors contained in the list of preoperative factors in the data collection form of the STS CHSD.

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effort to insure that what and how we report outcomes is relevant and to avoid confusion among physicians, administrators, and patients/families. Risk adjustment is critical to avoid confusion and ensure fair and proper assessment of programs and centers. There should be an avoidance of creating a system that will result in physicians and surgeons becoming risk averse, for example, avoiding the care of high-risk patients because it will adversely affect their mortality statistics and overall ranking.[80]

Cell-Based Therapies for CHD

There is over a decade of development experience with multiple types of cell-based products to mitigate the damaging effects of acquired heart diseases in adult populations. These efforts have largely focused on cell products derived from bone marrow–derived stem cells and have established confidence in the safety profile without evidence of increased arrhythmogenicity, cardiac toxicity, or tumor formation. These studies have been extensively reviewed in several published meta-analysis.[81–86] Although cell-based clinical trials have indicated promising results in younger adults, these studies have not established a definitive solution across a wide spectrum of CHD. Therefore, continued research and development are ongoing to customized solutions for specific patient populations depending on the pathogenesis of the underlying dysfunction whether the disease is primarily affecting the myocardium, valve, or vasculature and is either acquired or congenital.

The pediatric heart is more resistant to fibrosis in response to injury and is defined by increased density of progenitor cells or cardiomyocytes that retain proliferative potential. Notably, pressure overload in pediatric RV demonstrate a significant increase in cardiac stem cells.[87] The endogenous regenerative potential of the youthful heart may not be sufficient to overcome the challenges of severe CHD, such as HLHS, given the large number of individuals who develop progressive heart failure with time. Genetic, environmental, or metabolic challenges can accelerate progressive heart failure in severe forms of CHD within the first few years of life, despite the best supportive surgical and medical treatment, requiring a heart transplant eventually. To transform clinical outcomes, advanced regenerative technologies are required for CHD patients.[88]

Current experience with cell-based therapies in pediatric CHD clinical trials has focused on the single ventricle physiology. The largest study to date, PERSEUS (Cardiac Progenitor Cell Infusion to Treat Univentricular Heart Disease), is a randomized phase 2 study focusing on autologous cardiosphere-derived cells and intracoronary delivery of 3.0×10^5 cells/kg months after either stage 2 or stage 3 palliation.[89] The cardiosphere-derived cells require 3 to 4 weeks of expansion after right atrial samples are being processed from the prior surgery. The conclusions of this study with 41 patients in a 1:1 ratio demonstrates measurable improvements in cardiac function within 3 months after cell infusion without attributable adverse events other than transient ST-segment changes with cardiac enzyme leak and procedure-related hypotension. The statistical improvements are reported as ≈6% ejection fraction of the single ventricle by 3 independent measures, with nearly half of the cohort being HLHS. These results are consistent with the group's earlier study, TICAP (Transcoronary Infusion of Cardiac Progenitor Cells in Patients With Single Ventricle Physiology), that demonstrated persistent improvement after 36 months, with predominantly HLHS cohort of 14 patients randomly assigned to 2 groups.[90] Ongoing phase III study to include 40 patients in 3 centers in Japan is designed to provide further evidence for the scalability of this technology in single ventricle patients with ejection fraction <55% (NCT02781922).

Other cell types are also being examined in similar CHD patient cohorts using intracoronary delivery for Fontan patients with reduced cardiac function (RV ejection fraction <40%). Using autologous bone marrow–derived mononuclear cells, coronary infusions avoiding the use of heparin and avoiding stop-flow technique are being tested in patients with Fontan circulation from CHD (NCT02549625). Furthermore, intramyocardial delivery of autologous cells derived from either umbilical cord blood or bone marrow are being examined in 2 phase I studies during elective surgical procedures as adjunctive treatments. Patients with HLHS undergoing stage 2 surgical palliation and having autologous umbilical cord blood collected and processed may be eligible for direct injections into the myocardium during the surgical procedure (NCT01883076).

Additionally, autologous bone marrow–derived mononuclear cells are being delivered in a similar epicardial injection method during Ebstein anomaly repair (NCT02914171). The goal of these studies is to identify the most cost-effective cell-based product to augment cardiac function for heart failure–related CHD that may also require concomitant valvar repair to delay cardiac transplantation and prevent the development of latent heart failure because of persistent myopathic heart disease.

The field of CHD will also be poised to incorporate new innovations in 3D printing, bioengineered cells, and scaffolds that will iteratively move toward bioengineered patches, conduits, valves, and even whole organs. The surgical repair of CHD is continuously challenged by a growing heart with increased physiological demands. Therefore, customized regenerative products that can meet the emerging gaps of yesterday's treatments will afford new possibilities to preserve and extent the viability of each patient's suboptimal cardiac tissues. As these innovations improve and become incorporated into clinical care, outcomes will hopefully be realized by decreased demand for transplantation, decreased cost of palliative surgery, and increased wellness for individuals with CHD.

Summary

Successful outcome in the care of patients with CHD depends on a comprehensive multidisciplinary team. Early mortality for cardiac surgery in children is <5%, and the majority (>90%) of patients surviving surgery performed during infancy or childhood live well into the adult years with a good-to-excellent quality of life. When intervention is required after the initial repair, it is often for a valve-related problem. Advanced imaging facilitates accurate diagnosis and planning of interventional procedures. Advances in the perioperative medical management, particularly in intensive care, have also contributed to improved outcomes. Arrhythmias and heart failure are the most common late complication for most valvar lesions in CHD. Lifelong surveillance of recurrent or residual structural heart defects as well as arrhythmia monitoring is essential. Public reporting of institutional and surgeon-specific outcomes are becoming universal. Multi-institutional studies to further improve and standardize care pathways are currently underway. New innovations, such as bioengineered cells and scaffolds, will result in the successful development of bioengineered patches, conduits, valves, and even whole organs.

Acknowledgements

The authorship would like to acknowledge Rhonda Brincks for her significant contributions to manuscript preparation.

Disclosures

None.

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Circulation Research
JOURNAL OF THE AMERICAN HEART ASSOCIATION

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Circ Res. 2017;120:1027-1044
doi: 10.1161/CIRCRESAHA.117.309186
Circulation Research is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

Print ISSN: 0009-7330. Online ISSN: 1524-4571