Surgery for Adult Congenital Heart Disease

Tracy Geoffrion, MD, MPH[a,*], Stephanie Fuller, MD, MS[a,b]

KEYWORDS

• Surgery • ACHD • Fontan • Transplant • Congenital

KEY POINTS

- Describe the primary lesions found in adults with congenital heart disease requiring surgical intervention in adulthood.
- Review adult congenital heart disease diagnosis and treatment relating to surgery.
- Summarize surgical therapies used for adults with congenital cardiac disease.

INTRODUCTION

Technical and medical improvements for congenital cardiac disease in children have resulted in an increasing population of patients who survive into adulthood. Many of these patients are subject to progression of their native palliated disease or suffer from sequelae of their childhood repair and require repeat surgical intervention. In 2018, the American College of Cardiology/American Heart Association updated their published guidelines regarding the surgical management of adult congenital heart disease (ACHD).[1] An overview of surgical techniques, surgical decision-making strategies, and controversies in care when operating on the patient with ACHD is detailed here.

BASICS OF ADULT CONGENITAL CARDIAC SURGERY

There is no consensus as to the best location in which adult congenital cardiac operations should be performed. However, there are many who promote using adult hospital resources for these patients given their comorbid conditions.[2] Each facility (adult vs pediatric) has unique benefits for this patient population, including the availability of adult specialists, adult-sized equipment, psychosocial support, and cardiologists and surgeons well-versed in congenital heart disease lesions.[2] The Adult Congenital Heart Association has delineated comprehensive program features and services that constitute excellence in care of this unique population.[3]

Regardless of location, appropriate personnel are heavily emphasized. There are data supporting improved surgical outcomes when procedures are performed by surgeons specifically trained in congenital cardiac surgery.[4]

Preoperative Evaluation

Preoperatively, patients should receive a thorough evaluation by a multidisciplinary team that specializes in ACHD. In addition, adult specialists should be consulted as needed to address acquired health issues as a part of preoperative clearance, such as renal insufficiency, hepatopathy, and restrictive lung disease. Once a surgical plan is developed, medical optimization should be performed before undertaking surgical intervention. In some cases, this goal requires inpatient admission for medical management and a comprehensive dental evaluation. Significant consideration should be given to those patients with known or suspected commonly associated genetic syndromes such as trisomy 21 and DiGeorge syndrome, because they may pose unique issues for

[a] The Division of Cardiothoracic Surgery, Children's Hospital of Philadelphia, 3401 Civic Center Boulevard, Philadelphia, PA 19104, USA; [b] Division of Cardiothoracic Surgery, Children's Hospital of Philadelphia, The Perelman School of Medicine, University of Pennsylvania, 3401 Civic Center Boulevard, Philadelphia, PA 19104, USA
* Corresponding author.
E-mail address: Geoffriont@email.chop.edu

Cardiol Clin 38 (2020) 435–443
https://doi.org/10.1016/j.ccl.2020.04.013

perioperative care. Preoperative assessment by anesthesiologists should include a detailed history of prior anesthetic exposures, airway concern, and cervical spine issues when immobilized and positioned for surgery.

Multimodal imaging should be performed to clearly evaluate the current anatomy. This imaging includes chest radiography, transthoracic echocardiography, and cardiac catheterization with evaluation of left and right heart pressures and coronary anatomy. Additionally, cardiac MRI can be very useful in evaluating patients for complex repairs. Emerging modalities such as 3-dimensional echocardiography of computed tomography scans are advocated as well. Most surgeons request computed tomography with scans or without angiography and/or vascular ultrasound examinations of the neck and groin to determine the potential for injury on reoperative sternotomy and evaluate prospective sites of vascular access. In the operating room, all patients should undergo a preoperative transesophageal echocardiogram, ideally performed by a cardiologist who specializes in ACHD.

Reoperative surgery

Although there are many technical considerations in the conduct of ACHD operations, for most of these patients this operation is not their first. The reoperative sternotomy carries an increased risk at baseline secondary to the formation of scar tissue in the mediastinum. Many remotely placed patches and baffles become calcified, creating a hostile operative environment for safe mediastinal reentry, dissection, and cannulation to cardiopulmonary bypass. Additionally, reoperative patients have atypical anatomy that may be unfamiliar to the operating surgeon. Frequently, it is challenging to identify and access the necessary structures, further increasing the risk of injury to the heart, great vessels, and lungs during redo sternotomy, chest opening, and dissection.

Cannulation for cardiopulmonary bypass in the patient with ACHD varies by operative procedure with the intent of providing optimal exposure and safety.[5] The most commonly used strategies are ascending aorta and right atrial, ascending aorta and bicaval, femoral artery and femoral vein, right axillary artery, and femoral vein or right atria. In addition, partial bypass or left sided bypass may be used in unique instances. Aortoatrial cannulation is typically used for closed cardiac and great vessel operations. Aortobicaval cannulation is the most commonly used procedure for congenital cardiac surgery because it provides exposure of the intracardiac structures and is particularly protective when atrial or ventricular shunting lesions exist. In situations where the process of entering the chest is likely to be difficult or risky, femoral bypass can be initiated before sternotomy to support the patient should a major structure be entered. When the ascending aorta is unable to be cannulated for anatomic or technical reasons, the axillary artery may be used for antegrade arterial bypass inflow. The femoral vein, right atrium, or vena cavae can then be used for venous cannulation. Alternative arterial access via the innominate or carotid arteries can also be used if needed.[6] Hence, it is always necessary to document the patency of peripheral vessels given that these patients often have multiple surgeries, catheterizations, and central access, all of which may lead to occlusion of these vessels. It is always necessary to proceed safely and ensure the availability of appropriately typed and crossed blood products. Peripheral cannulae must always be readily available on in reoperative sternotomy in case of mediastinal injury. In the cases of catastrophic injury upon sternal reentry, surgeons should be prepared to initiate deep hypothermic circulatory arrest for cerebral protection during low flow states to repair the most serious of injuries, such as those to the ascending aorta, aortic arch, and innominate vein.

Multiple strategies are used for myocardial protection during the operative procedure. Primarily, the heart should be nondistended on cardiopulmonary bypass. Many procedures that are exclusively right sided (ie, pulmonary valve replacement) may be performed with the support of cardiopulmonary bypass without cardiac arrest, given confirmation that there are no intracardiac communications at the atrial or ventricular levels. For those cases requiring cardiac standstill, the goal is to achieve both electrical and mechanical arrest of the myocardium. Multiple commercially available cardioplegia solutions are available and used. They are all isotonic and hyperkalemic to induce a polarized cardiac arrest. Again, it is important to note that some procedures require the use of deep hypothermic circulatory arrest. Thus, effective and safe cooling and rewarming strategies must be used together with a cardiopulmonary bypass plan that allows for increased duration of bypass to allow for temperature regulation.

The decision to proceed with surgery is not always straightforward. Certain anatomic or physiologic findings have proven to be relatively prohibitive. These include anatomic risks of reentry into the chest cavity, both venovenous and arterial collateral burden in the mediastinum and pleural cavities, the risk of bleeding during or

after surgery, and the severity of ventricular dysfunction such that there is concern about the ability to wean from cardiopulmonary bypass. Recent guidelines have established that, for shunt lesions, in patients with severe pulmonary artery hypertension (greater than 2/3 systemic) or a net right-to-left shunt, surgery is contraindicated.[1] Additionally, severe comorbid conditions or profound frailty can make recovery from any operation challenging. These considerations illustrate the benefit of a multidisciplinary approach to the treatment of these complex patients. Although a number of previous sternotomies is often cited as a concern for sternal reentry, there is no prescribed number of previous sternotomies at which sternal reentry alone becomes prohibitive. These factors should be carefully considered in deciding whether an operation will improve the quality or length of a patient's life.

SHUNTING LESIONS

Atrial Septal Defect and Partial Anomalous Pulmonary Venous Return

Although commonly performed as de novo procedures in adults, these may also be reoperative owing to residual shunts. The repair of an atrial septal defect (ASD) and partial anomalous pulmonary venous return is most commonly performed through a median sternotomy, although alternative access sites include a right minithoracotomy or robotic-assisted surgery through the right chest. Small defects can be closed primarily, but the larger defects typically require patch closure using either autologous pericardium, bovine pericardium, or homograft. The repair of partial anomalous pulmonary venous return with sinus venosus ASD requires using the patch to direct anomalous flow into the left atrium. If there is no associated ASD, one must be created such that the veins can be tunneled to the left atrium without obstruction. For cases in which the right pulmonary veins are high on the superior vena cava and a baffle cannot be created to redirect flow, the Warden procedure is used. This procedure involves the division of the superior vena cava just above the highest anomalous pulmonary vein and anastomosis to the right atrial appendage. The orifice of the superior vena cava is then baffled to the right atrial appendage, which has been cleared of any trabeculations. Anomalous left-sided veins can be rerouted to the left atrium by connecting the draining vertical vein to the left atrial appendage either directly or with the use of a polytetrafluoroethylene graft. The risks of closure of an ASD or partial anomalous pulmonary venous return include residual ASD, pleural effusions, heart block, air embolism, atrial dysrhythmias, pulmonary venous obstruction, and superior vena cava obstruction.[7]

Ventricular Septal Defect

Isolated ventricular septal defects (VSDs) are rare in adults, but may present as a small restrictive VSD detected in a patient who has been lost to follow-up, develops infective endocarditis, or meets surgical indications as an adult. The surgical approach to the lesion is based on the specific type of defect. The transatrial approach uses a right atriotomy to access conoventricular, canal-type, and some muscular defects through the tricuspid valve. Other methods include access through a right ventriculotomy or through the pulmonary valve for conoseptal hypoplasia defects. Small VSDs can be closed primarily with pledgetted sutures, but larger defects should be closed with a patch of Dacron, Gore-Tex, or glutaraldehyde treated pericardium. The specific risks associated relate to the surrounding structures including the conduction system, aortic valve, tricuspid valve, or pulmonary valve.

Atrioventricular Septal Defect

An atrioventricular septal defect may present across the spectrum from incomplete atrioventricular canals to complete. Although it is unlikely that a patient with a complete canal will undergo their first surgical intervention as an adult, the less severe forms of disease are often seen de novo in adulthood. More commonly, previously repaired atrioventricular septal defect lesions can develop complications or disease progression in adulthood. Most commonly, this involves residual intracardiac atrial or ventricular shunting lesions, atrioventricular valve regurgitation often owing to a residual cleft, atrioventricular valve stenosis, or the development of left ventricular outflow tract obstruction. Surgical techniques are tailored to the specific lesions such as closure of residual ASD or VSD lesions as described elsewhere in this article, valve repair or potentially replacement, and resection of subaortic obstruction. The risks of these operations can include any of those mentioned previously for ASD or VSD. If valve repair or replacement are performed, these patients can also be at risk for valve stenosis, prosthetic valve infection or thrombosis, patient–prosthesis mismatch, and complications associated with systemic anticoagulation for mechanical valves.

Patent Ductus Arteriosus

Closure of a patent ductus arteriosus during adulthood can be complicated. Historically, these patients were managed surgically. The duct was ligated and potentially divided or patch closed through a pulmonary arteriotomy via sternotomy or thoracotomy, with or without the use of cardiopulmonary bypass.[8] However, minimally invasive and catheter-based techniques are now often being used.[9] Contraindications to these techniques include wide and short ducts, or chests that are otherwise inaccessible via video-assisted thoracoscopic surgery owing to scarring or pleural disease. Additionally, patent ductus arteriosus associated with aneurysms may not be closeable with these methods.[10] Last, those with extensive calcification also pose a high risk for surgical ligation.

LEFT-SIDED OBSTRUCTIVE LESIONS

Congenital Mitral Stenosis

Current American Heart Association guidelines for isolated mitral stenosis support percutaneous balloon valvotomy as first-line procedural therapy for mitral stenosis with appropriate valve morphology as assessed by the Wilkins score. For those who are not candidates for balloon valvuloplasty, mitral valve surgery with repair or replacement is indicated.[10] The mitral valve can be accessed directly into the left atrium via the interatrial groove or through the atrial septum. Minimally invasive techniques such as right minithoracotomy port access and robotic mitral surgery are increasingly popular and have been shown to have equivalent outcomes.[11] Repair techniques for mitral stenosis vary based on the etiology and are limited. These include commissurotomy, chordal splitting, leaflet augmentation with patch material, and papillary muscle division. When valve replacement is required, a mechanical, or bioprosthetic prosthesis can be used. The primary risks associated with surgery include heart block, circumflex coronary injury, coronary sinus injury, and perivalvular leak or dehiscence.

Subaortic Stenosis

Surgery for subaortic stenosis is typically performed with aortic and right atrial cannulation and cardioplegic arrest. The lesions creating the stenosis are a fibrous membrane below the valve and/or septal hypertrophy. The subaortic membrane is approached through a transverse incision in the ascending aorta, just above the valve. The fibromuscular ring is visualized through the aortic valve and excised in its entirety. The risks include damage to aortic valve leaflets, resulting in valve insufficiency, injury to conduction tissue causing heart block, and injury to the anterior leaflet of the mitral valve.[7] A myectomy is performed for hypertrophy of the ventricular septum. Transaortic exposure is typically used and the hypertrophied septum is sharply excised from just below the valve toward the apex. In addition to these risks, there is the additional risk of iatrogenic VSD from excess septal muscle excision. Diffuse tunnel-like subvalvar obstruction of the left ventricular outflow tract may require augmentation with more complex aorticoventriculoplasty operations potentially in combination with aortic valve replacement.[12] For example, the modified Konno procedure involves complete resection of the subaortic ventricular septum through an incision in the right ventricle. The defect or newly created VSD is then closed with a patch.[5] Often, the right ventricle requires a patch enlargement at the incision site as well. This technique has additional risk of damage to the pulmonary and tricuspid valves during the enlargement of the ventricular septum.

Aortic Stenosis

Adult congenital aortic stenosis can be related to calcific or degenerative disease of otherwise normal valves or be secondary to a bicuspid valve. Surgery for aortic stenosis is often direct aortic valve replacement with bioprosthetic or mechanical valve. Some centers employ minimally invasive access through a right minithoracotomy or hemisternotomy for valve replacement. Risks specific to surgical valve replacement include heart block, damage to or occlusion of the coronary arteries, injury to the anterior leaflet of the mitral valve, paravalvular leak, and patient–prosthesis mismatch. In cases where the aortic annulus is too small to fit an adequately sized prosthesis, the aortic root must also be enlarged. There are several aortic annular enlargement techniques used. In addition, assuming there is a normal pulmonic valve, the Ross procedure can be used in adult patients requiring a valve replacement. The Ross procedure involves an autologous root replacement from the pulmonary outflow and requires the use of a homograft to replace the right ventricular outflow tract (RVOT), thus creating both neoaortic and neopulmonic replacements. Patients often require revision of the homograft. The autograft in the aortic position may be prone to dilation or neoaortic valve regurgitation. There are increasing data to support the use of transcatheter aortic valve replacement for bicuspid aortic

valve stenosis.[12–14] However, caution should be used in young patients because there are no long-term data regarding durability and complications.

Supravalvular Aortic Stenosis

Repair of supravalvular aortic stenosis involves patch enlargement of aorta at the level of the sinotubular junction.[15] Additional options in the adult population include aortic root and/or ascending aortic replacement. The integrity of the aortic valve determines whether a concomitant valve replacement is indicated. If the coronary ostia are involved in the stenosis, the patient may require ostioplasty or ostial reimplantation. If this is not feasible and the patient has concern for ischemic events, coronary bypass may be necessary. During patch aortoplasty of the aortic root, there is an inherent risk to the aortic valve leaflets or otherwise normal coronary ostia.

Coarctation of the Aorta

Transcatheter balloon dilation and stenting is commonly used to treat coarctation in the adult.[16] If unsuccessful, surgical repair of coarctation is similar other open surgical intervention on the descending thoracic aorta. This intervention is typically performed through a left posterolateral thoracotomy. An end-to-end anastomosis is used if technically feasible; otherwise, a Dacron interposition graft is placed.[17] Often left heart bypass can be used to support the distal circulation while the aorta is clamped. The primary risks are neurologic, the most severe being spinal ischemia and ensuing paralysis.

Cor Triatriatum Sinister

Surgical repair for cor triatriatum involves excision of the membrane that divides the left atrium. This procedure is performed on bypass with bicaval cannulation. The membrane is accessed through the atrium or the interatrial septum[7] and the specific technical risk is damage to the mitral valve, resulting in mitral regurgitation. Confirmation that all 4 pulmonary veins return normally to the left atrium is necessary.

RIGHT-SIDED LESIONS

Pulmonary Valve Disease

In adults with moderate or severe pulmonary stenosis, balloon valvuloplasty has shown to be an efficacious first-line therapy with good short- and long-term results.[18] For those who fail percutaneous intervention or are not candidates for catheter-based interventions, surgical pulmonary valve repair or replacement is indicated. Pulmonary valve replacement in adults is most often a reoperation after childhood RVOT reconstruction. Bioprosthetic valves are commonly used with an extension of pericardium or graft material onto the proximal portion of the RVOT to prevent mechanical distortion.[5] Alternatively, for patients with a severely dilated RVOT, plication of the RVOT is advocated. If there are no residual intracardiac defects, the operation can be performed safely on cardiopulmonary bypass with the heart warm and beating.

Branch Pulmonary Artery Stenosis

Branch pulmonary artery stenosis frequently accompanies valve disease and can sometimes be alleviated with percutaneous balloon dilation and ensuing stent placement. If that is unsuccessful or if the patient is undergoing another procedure, they can be augmented surgically with a patch. These patches are typically bovine pericardium, autologous pericardium, synthetic pericardium, or homograft.

Tetralogy of Fallot

Patients with repaired tetralogy of Fallot often require reintervention on the RVOT in adulthood. Pulmonary regurgitation, stenosis, or a combination of both are the most commonly seen morphologies and the type of adult intervention required depends on the type of repair that was performed previously.[19] Isolated pulmonary valve disease can be repaired as described elsewhere in this article. RVOT reconstruction with a right ventricle-to-pulmonary artery conduit in childhood carries the risk of developing valve incompetence or conduit obstruction. Catheter-based intervention of conduits with stenting or placement of a transcatheter valve inside previously placed conduits is used,[20,21] but surgical replacement of the pathway is often warranted. This procedure is performed with cardiopulmonary bypass and the previously implanted conduit is removed from the pulmonary artery and right ventricular surface. Patch augmentation or repair of the pulmonary artery is often needed to reconstruct an appropriately sized confluent pulmonary artery bifurcation on which to attach the distal end of the new conduit. Removal of the old conduit from the ventricle is performed with focus on damaging as little native right ventricular myocardium as possible. Options for conduit include homograft and the Hancock Dacron conduit with a porcine bioprosthetic valve.[22] The proximal anastomosis is often augmented with a patch of homograft or Gore-Tex to avoid

distortion. Additionally, annular dilation often causes some degree of tricuspid regurgitation in adults with repaired tetralogy of Fallot. This finding is often addressed at the time of conduit exchange or valve replacement and the technical details are discussed in the section on Ebstein's Anomaly and Tricuspid Valve Regurgitation. The addition of arrhythmia surgery can also be used for patients with atrial arrhythmias. Other lesions seen in adults with repaired tetralogy of Fallot that could require operations include branch pulmonary artery stenosis, residual VSD, and RVOT aneurysm.

Ebstein's Anomaly and Tricuspid Valve Regurgitation

Regurgitation of a dysplastic and regurgitant tricuspid valve can be surgically treated with valve repair (annular reduction, leaflet resection, or repair in the cases of perforation) or replacement. For Ebstein's anomaly, there are multiple repair techniques. These included the Hardy, Danielson, Carpentier, and Cone techniques, which involve plication of the atrialized right ventricle, reconstruction of the valve to address the downward displacement of the septal leaflets with potential rotation of the anterior and posterior leaflets, reduction of the right atrium, and closure (total or subtotal) of any atrial communications.[23] Additionally, replacement can be considered if there is not a viable reconstructive option. Although mechanical valves provide superior durability, they are not used frequently in the tricuspid position owing to thrombosis. Porcine bioprosthetic valves have demonstrated a survival advantage in some series.[24]

There is a risk of right heart failure after tricuspid repair after these operations and some surgeons support leaving or creating a small atrial-level communication to unload the right ventricle. If right heart failure is profound and refractory, a bidirectional cavopulmonary anastomosis can be added, creating a functional 1.5 ventricle repair. Hence, all these patients should undergo thorough cardiac catheterization with calculation of pulmonary vascular resistance before surgery. This process can provide better tolerance of residual tricuspid dysfunction, right ventricular dysfunction, and decreased operative mortality.[25] Additional risks of tricuspid valve surgery include damage to the conduction system or coronary arteries and thromboembolism. Atrial tachyarrhythmias are also commonly found in this population either before or after surgery and some investigators advocate a full electrophysiologic assessment in the preoperative period.[25]

OTHER LESIONS

Repaired Transposition of the Great Arteries

Patients with ventricular arterial discordance who underwent repair of transposition in infancy with either atrial or arterial switch operations can often develop sequelae in adulthood and require surveillance. For those patients who underwent atrial switch, they carry a significant risk of systemic atrioventricular (tricuspid) valve regurgitation, right ventricular systolic failure, baffle obstruction or baffle leak, and atrial arrhythmias. Many of these patients require pacemakers, which lead to baffle obstruction.[26–28] Patients with history of arterial switch operations are at risk of coronary artery stenosis, neoaortic root dilation with valve regurgitation, pulmonary artery stenosis, and RVOT obstruction.[29] These complications can often be managed percutaneously in some cases, but may require tricuspid valve repair or replacement, coronary artery bypass, or aortic valve or root replacement.[30] For those patients with atrial switch who develop systemic ventricular failure, orthotopic heart transplantation is recommended.

Coronary Anomalies

The most common coronary anomalies discovered in adulthood are anomalous aortic origin of the coronary artery. Owing to the abnormal take off of these coronaries, patients may present with coronary ischemia. Anomalous aortic origin of the left coronary should be repaired, whereas the right-sided coronary lesions should be repaired when evidence of ischemia exists. The coronary unroofing procedure maintains the same location of the coronary, but allows for relief to the intimal obstruction of the coronary along the intramural path along the aorta. In adults, ischemia from other congenital coronary pathologies such as anomalous coronary artery from the pulmonary artery and ostial stenosis are rare. These pathologies can be addressed with coronary reimplantation, ostial patch, coronary unroofing, or coronary artery bypass grafting depending on the anatomy of the lesions.

Fontan Physiology

Fontan physiology predisposes adult patients to both cardiac and noncardiac complications. They are at risk for developing protein losing enteropathy, plastic bronchitis, Fontan-associated liver disease and various forms of cardiac failure. Surgical strategies that have been used to improve symptoms of these conditions include Fontan conversion to extracardiac Fontan, cardiac pacing, Fontan fenestration, thoracic duct ligation,

surgical rerouting of the innominate vein to the left atrium, and heart or combined heart–liver transplantation. The use of mechanical circulatory support in this population is controversial and there is not currently a device approved specifically for Fontan circulation.[31,32] However, promising case reports and single center data supports the use of ventricular assist devices for isolated ventricular failure and total artificial heart devices for Fontan failure.[33]

Acquired Disease in Congenital Patients

As patients with congenital cardiac disease become adults, they are susceptible to the same acquired diseases as those in the general population. Specific acquired pathologies in the ACHD population include degenerative valve disease, arrhythmias, great vessel and aortic root aneurysms, coronary atherosclerosis, and endocarditis. For some of these conditions, indications for surgery are not always clear and extrapolated from the general population. Thus, the traditionally accepted guidelines for patients without ACHD should be used as a guide.

Ascending Aorta and Aortic Root Pathology

The American Heart Association guidelines for the diagnosis, surveillance, and management of aortic root and ascending aorta pathology focus on the size of ascending or arch pathology, the presence of symptoms, and aortic valve competence.[34] These guidelines should be used to aid in decision making on the timing of surgical intervention for patients with ACHD with similar pathology. Exceptions include for those patients with known genetic syndromes and connective tissue disorders such as Turners disease or Marfan syndrome, patients with conotruncal defects who have a predisposition to aortopathy such as tetralogy of Fallot and truncus arteriosus, and those with a reconstructed neoaorta such as those patients with a Norwood or who have undergone the arterial switch and have a neoaortic root. The surgical management of these disease processes varies widely by centers, but includes replacement of the aortic valve and replacement of the aortic root with or without the aortic valve, ascending aorta, and/or aortic arch. When possible, valve-sparing procedures are preferred. These operations can be performed with a variety of different cannulation and myocardial protection strategies based on the location of the disease and the operative procedure planned.

Coronary Artery Atherosclerosis

Coronary artery bypass can be used in patients with ACHD who have reversible ischemia related to coronary artery atherosclerosis. Accepted indications for bypass for patients without ACHD should be followed. These cases can be challenging owing to variations in coronary anatomy and prior operative intervention.

MECHANICAL SUPPORT AND TRANSPLANTATION

Mechanical circulatory support is becoming more commonly used in adult patients with congenital heart disease to support the left ventricle, right ventricle, or both. Implantable devices can be used in patients with low output heart failure as bridge to recovery, destination therapy, or bridge to transplantation.[35] All devices require anticoagulation. Technical considerations in patients with ACHD include the size of the ventricular cavity, access to the necessary structures for cannula implantation in the reoperative setting, associated comorbid conditions (renal or hepatic failure, elevated pulmonary venous pressures), the presence of semilunar valve regurgitation, and the presence of intracardiac shunts. In the Fontan population, atrial device positioning with excision of the atrioventricular valve has been reported with success.[36]

Extracorporeal membrane oxygenation is widely used in the ACHD population. Venovenous or venoarterial circuits can be used depending on the underlying pathology.[37] Often, the cannulation strategies must be adjusted given anomalies of systemic venous drainage. Some options include central cannulation of the aorta and atrium, femoral vein and femoral artery, axillary artery and femoral vein, or femoral artery and internal jugular artery. Although the carotid artery is frequently used in pediatric cannulation for extracorporeal membrane oxygenation, it is not typically used for adults. For those patients requiring long term support, cannulation style should support ambulation and recovery.

For patients with ACHD with severe refractory heart failure and no other surgical options, orthotopic heart transplantation may be indicated.[38] Transplantation in the ACHD population is often higher risk than patients without ACHD because they have undergone multiple prior operations and could have developed aortopulmonary and venovenous collaterals that increase the risk of massive bleeding during repeat operations. Additionally, these patients often require more thoughtful approaches to the operation owing to anatomic variations, venous drainage, and great vessel anatomy.[39,40] In patients who have been on a single ventricle pathway, the elevated venous pressures lead to liver dysfunction or failure that also

affects coagulation. In some instances, combined heart and liver transplantation is being used.[31] The problem of increased pulmonary vascular resistance from longstanding heart failure also poses additional questions regarding the decision to transplant. In fact, combined heart and lung transplantation may be feasible. Overall, the outcomes for thoracic transplantation in patients with ACHD have been favorable with a median survival of 15 years for patients with ACHD.[41] Given the high risk, it is important to select carefully those patients with ACHD who will benefit from transplantation.

SUMMARY

There are an increasing number of patients with congenital cardiac disease surviving into adulthood. The complex anatomy and physiology pose unique challenges to surgeons from both a technical and perioperative management standpoint. The management of these patients should be performed by a specialized multidisciplinary team in an experienced center.

DISCLOSURE

The authors have nothing to disclose.

REFERENCES

1. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: executive summary: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. Circulation 2019;139(14):e637–97.
2. Kogon BE, Plattner C, Leong T, et al. Adult congenital heart surgery: adult or pediatric facility? Adult or pediatric surgeon? Ann Thorac Surg 2009;87:833–40.
3. ACHA ACHD PROGRAM CRITERIA. Adult congenital heart association. Available at: https://www.achaheart.org/provider-support/accreditation-program/accreditation-program-documents/. Accessed February 28, 2020.
4. Karamlou T, Diggs BS, Person T, et al. National practice patterns for management of adult congenital heart disease. operation by pediatric heart surgeons decreases in-hospital death. Circulation 2008;118:2345–52.
5. Kaiser L, Kron IL, Spray TL, editors. Mastery of cardiothoracic surgery. Philadelphia: Lippincott Williams & Wilkins; 2013.
6. Brown ML, Dearani JA, Burkhart HM. The adult with congenital heart disease: medical and surgical considerations for management. Curr Opin Pediatr 2009;21(5):561–4.
7. Khonsari S, Sintek C. Cardiac surgery: safeguards and pitfalls in operative technique. Phildelphia: Lippincott Williams & Wilkins; 2008.
8. Djukanovic BP, Micovic S, Stojanovic I, et al. The current role of surgery in treating adult patients with patent ductus arteriosus. Congenit Heart Dis 2014;9(5):433–7.
9. Jacobs JP, Giroud JM, Quintessenza JA, et al. The modern approach to patent ductus arteriosus treatment: complementary roles of video-assisted thoracoscopic surgery and interventional cardiology coil occlusion. Ann Thorac Surg 2003;76:1421–8.
10. Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of cardiology/American Heart Association Task Force on practice guidelines. J Am Coll Cardiol 2014;63(22):e57–185.
11. Dogan S, Aybek T, Risteski PS, et al. Minimally invasive port access versus conventional mitral valve surgery: prospective randomized study. Ann Thorac Surg 2005;79(2):492–8.
12. Silversides CK, Kiess M, Beauchesne L, et al. Canadian Cardiovascular Society 2009 Consensus Conference on the management of adults with congenital heart disease: outflow tract obstruction, coarctation of the aorta, tetralogy of Fallot, Ebstein anomaly and Marfan's syndrome. Can J Cardiol 2010;26(3):e80–97.
13. Sridhara S, Gavhane PU, Pandya B, et al. Meta-analysis comparing safety and outcomes after transcatheter aortic valve replacement in bicuspid aortic stenosis. Circulation 2019;140(Suppl_1):A13208.
14. Makkar RR. Outcomes of transcatheter aortic valve replacement with balloon expandable SAPIEN 3 valve in bicuspid aortic stenosis: an analysis of the STS/ACC TVT registry. Oral Presentation at the American College of Cardiology Annual Meeting 2019. New Orleans, Louisiana. Available at: http://www.clinicaltrialresults.org–www.clinicaltrialresults.org/Slides/ACC2019/Makkar_STSTVT.pdf.
15. Coskun TS, Coskun OK, El Arousy M, et al. Surgical repair of congenital supravalvular aortic stenosis in adult. ASAIO J 2007;53(6):e5–6.
16. Carr JA. The results of catheter-based therapy compared with surgical repair of adult aortic coarctation. J Am Coll Cardiol 2006;47(6):1101–7.
17. Bouchart F, Dubar A, Tabley A, et al. Coarctation of the aorta in adults: surgical results and long-term follow-up. Ann Thorac Surg 2000;70(5):1483–8.
18. Stanger P, Cassidy SC, Girod DA, et al. Balloon pulmonary valvuloplasty: results of the valvuloplasty and angioplasty of congenital anomalies registry. Am J Cardiol 1990;65(11):775–83.

19. Tretter JT, Morello M, Chaudhry A, et al. The management of tetralogy of Fallot after corrective surgery. Cardiovasc J S Afr 2018;15(1):6–15.
20. Peng LF, McElhinney DB, Nugent A, et al. Endovascular stenting of obstructed right ventricle-to-pulmonary artery conduits. Circulation 2006;113:2598–605.
21. Ong K, Boone R, Gao M, et al. Right ventricle to pulmonary artery conduit reoperations in patients with tetralogy of Fallot or pulmonary atresia associated with ventricular septal defect. Am J Cardiol 2013;111(11):1638–43.
22. Burri M, Lange R. Surgical treatment of Ebstein's anomaly. Thorac Cardiovasc Surg 2017;65(8):639–48.
23. Brown ML, Dearani JA, Danielson GK, et al. Comparison of the outcome of porcine bioprosthetic versus mechanical prosthetic replacement of the tricuspid valve in the Ebstein anomaly. Am J Cardiol 2009;103(4):555–61.
24. Chauvaud S, Fuzellier JF, Berrebi A, et al. Bi-directional cavopulmonary shunt associated with ventriculo and valvuloplasty in Ebstein's anomaly: benefits in high risk patients. Eur J Cardiothorac Surg 1998;13(05):514–9.
25. Dearani JA, Mora BN, Nelson TJ, et al. Ebstein anomaly review: what's now, what's next? Expert Rev Cardiovasc Ther 2015;13(10):1101–9.
26. Tobler D, Williams WG, Jegatheeswaran A, et al. Cardiac outcomes in young adult survivors of the arterial switch operation for transposition of the great arteries. J Am Coll Cardiol 2010;56(1):58–64.
27. Love BA, Mehta D, Fuster VF. Evaluation and management of the adult patient with transposition of the great arteries following atrial-level (Senning or Mustard) repair. Nat Clin Pract Cardiovasc Med 2008;5(8):454–67.
28. Dobson R, Danton M, Nicola W, et al. The natural and unnatural history of the systemic right ventricle in adult survivors. J Thorac Cardiovasc Surg 2013;145(6):1493–503.
29. Kempny A, Wustmann K, Borgia F, et al. Outcome in adult patients after arterial switch operation for transposition of the great arteries. Int J Cardiol 2013;167(6):2588–93.
30. Haeffele C, Lui GK. Dextro-transposition of the great arteries: long-term sequelae of atrial and arterial switch. Cardiol Clin 2015;33(4):543–58.
31. Reardon LC, DePasquale EC, Tarabay J, et al. Heart and heart–liver transplantation in adults with failing Fontan physiology. Clin Transplant 2018;32(8):e13329.
32. Rychik J, Atz AM, Celermajer DS, et al. Evaluation and management of the child and adult with Fontan circulation: a scientific statement from the American heart association. Circulation 2019;140(6):e234–84.
33. Woods RK, Ghanayem NS, Mitchell ME, et al. Mechanical circulatory support of the Fontan patient. In Seminars in Thoracic and Cardiovascular Surgery: Pediatric Cardiac Surgery Annual 2017 Jan 1 (Vol. 20, pp. 20-27). WB Saunders.
34. Hiratzka LF, Bakris GL, Beckman JA, et al. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM guidelines for the diagnosis and management of patients with thoracic aortic disease. J Am Coll Cardiol 2010;55(14):e27–129.
35. Steiner JM, Krieger EV, Stout KK, et al. Durable mechanical circulatory support in teenagers and adults with congenital heart disease: a systematic review. Int J Cardiol 2017;245:135–40.
36. Mascio CE. Unique cannulation technique and atrioventricular valve excision for HeartWare HVAD in the small Fontan patient. Oper Tech Thorac Cardiovasc Surg 2016;21(4):322–9.
37. Derk G, Laks H, Biniwale R, et al. Novel techniques of mechanical circulatory support for the right heart and Fontan circulation. Int J Cardiol 2014;176(3):828–32.
38. Ross HJ, Law Y, Book WM, et al. Transplantation and mechanical circulatory support in congenital heart disease: a scientific statement from the American Heart Association. Circulation 2016;133(8):802–20.
39. Gupta D, Reid J, Moguillansky D, et al. Heart transplantation for adult congenital heart disease: overview and special considerations. Cardiovasc Innov Appl 2018;3(1):73–84.
40. Fynn-Thompson F. Heart transplantation in adults with congenital heart disease. Methodist DeBakey Cardiovasc J 2019;15(2):145.
41. Khush KK, Cherikh WS, Chambers DC, et al. The International thoracic Organ transplant registry of the International Society for heart and lung transplantation: thirty-fifth adult heart transplantation report—2018; focus theme: multiorgan transplantation. J Heart Lung Transplant 2018;37(10):1155–68.