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## **Review**

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# Paediatrician's guide to post-operative care for biventricular CHD: a review

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#### **Abstract**

Importance: Paediatricians play an integral role in the lifelong care of children with CHD, many of whom will undergo cardiac surgery. There is a paucity of literature for the paediatrician regarding the post-operative care of such patients. Observations: The aim of this manuscript is to summarise essential principles and pertinent lesion-specific context for the care of patients who have undergone surgery or intervention resulting in a biventricular circulation. Conclusions and relevance: Familiarity with common issues following cardiac surgery or intervention, as well as key details regarding specific lesions and surgeries, will aid the paediatrician in providing optimal care for these patients.

Paediatricians care for patients with CHD, many of whom have undergone cardiac surgery. A wide range of issues may arise post-operatively, from wound care and sternal precautions to support for feeding and neurodevelopmental problems. While some of these are individually addressed in depth elsewhere, a concise and accessible summary for the paediatrician on common post-operative topics is underrepresented in the existing literature, particularly with regard for lesion-specific concerns and indications for surgery.<sup>1-3</sup>

The aim of this manuscript is to review essential principles for the optimal care of children who have undergone cardiac surgery or an interventional procedure. It builds on prior reports by providing additional pertinent lesion-specific context for patients who have undergone a biventricular operation. A separate manuscript in this edition of *Cardiology in the Young* similarly expands on the post-operative care of patients who have undergone a single ventricle palliation.<sup>4</sup>

# Universal issues following cardiac intervention or surgery

Paediatricians familiar with common issues among post-operative patients will be better positioned to help families navigate such medical, behavioural, and social concerns. The hospital discharge summary serves as a useful resource, especially for items such as dates of procedures, performed interventions, residual cardiac lesions, expected oxygen saturations, wound care, and return to activity. Though this is a valuable source for guidance, it may lack details on common topics elaborated upon below.

# Wound care and sternal healing

Appropriate management of the post-operative surgical or cardiac catheterisation site is essential to prevent infection. If signs of a surgical site infection arise – such as redness, warmth, swelling, unexplained fever, or purulent discharge – the cardiologist and/or cardiothoracic surgeon will want to be notified immediately. To prevent this, post-operative patients should avoid direct spraying of water onto the incision during showers and submersion or soaking of the incision in a tub bath until the entire eschar and any surgical glue have naturally fallen off, leaving nothing but scar underneath. Post-catheterization patients should be similarly cautious with surgical

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sites, even though there is no large incision/eschar. Most interventional cardiologists recommend avoiding submersion and baths for a few days after the procedure, though showers and short baths are generally considered acceptable. Incisions should be dried well after bathing. Lotion, ointments, or powder on the incisions should generally be avoided. Once healed, sunscreen while exposed outdoors during the first year following surgery may decrease the prominence of scarring. <sup>5,6</sup>

There are many types of sutures used to close post-operative incisions, and many or all may be removed prior to hospital discharge. A few extruding sutures may be left in place for certain patients and may need to be removed at a follow-up appointment. The cardiologist or surgical team often removes them, but in some cases, removal by the paediatrician may be necessary to save the family a long return trip for subspecialty services. Adolescent females should consider use of support bras to prevent tension on the incision. Occasionally, wound vacs may be used to minimise tension on the incision and promote healing; should wound vac concerns arise, they should be discussed with the cardiologist and/or cardiothoracic surgeon.

To prevent separation of the healing sternum, "sternal pre-cautions" are recommended for the first 6–8 weeks after sternotomy. The specific precautions can vary based on age and expected activities.<sup>7–9</sup> Often, they include activity limitations such as avoiding gym class, contact sports, weightlifting, and carrying heavy backpacks and bags. Infants should be scooped up in a cradle position rather than lifted from under the arms, as this places significant stress on the sternum. Infants may have tummy time while awake, if tolerated and not in apparent pain.

## Return to activity

Specific activity restrictions beyond those required for sternal precautions are determined on an individual basis and are typically outlined in the discharge summary or early post-operative cardiology follow-up note. Patients may feel tired for a couple of weeks after an operation and need to rest during the day. In general, patients can resume typical daily activities – such as walking, light chores – after hospital discharge. They can typically return to school within 1–3 weeks of discharge and should be able to return to their usual exercise activities within a few weeks. Adolescents and older children who change classrooms throughout the day may benefit from additional time allowance between classes for a few weeks.

Catheterisation patients should avoid activities that involve significant leg strain and motion, such as bicycle riding and weight-lifting, for at least 5 days after the procedure and potentially longer if large catheters were used.

# **Common medications**

Common medications prescribed in the post-operative period include diuretics such as furosemide, chlorothiazide, and spironolactone. These medications have the potential to cause electrolyte imbalances, necessitating caution if a gastrointestinal illness arises. Usually, these diuretics are weaned during the early post-operative period.

Patients who were exposed to prolonged sedative and analgesics (e.g., opioids and benzodiazepines) may require slow weans after hospital discharge. Often, a wean plan is formulated in collaboration with a hospital pharmacist. In some cases, a wean that is too aggressive may result in withdrawal symptoms such as irritability, yawning, sweating, gagging, and vomiting.

Some cardiac surgeries and interventions require anticoagulation to preserve the integrity of the repair and/or device. Commonly used medications include warfarin, enoxaparin, and aspirin, among others. The primary paediatric cardiologist will define the target therapeutic levels (e.g. international normalized ratio (INR), anti-Xa), in collaboration with the cardiac surgeon or interventional cardiologist. The cardiologist is often responsible for monitoring drug levels, though some centres utilise a haematology teambased monitoring program. Occasionally, the paediatrician may aid in drug monitoring as well.

# Neonatal feeding

Optimal growth is a crucial issue for babies after cardiac surgery, with many neonates experiencing feeding difficulties in the post-operative period. Caloric fortification of feeds may be required to achieve adequate weight gain. Occasionally, poor oral motor skills necessitate nasogastric feeds or placement of a gastrostomy tube. Ancillary services such as speech therapy and occupational therapy may be utilised to minimise the risk of oral aversion and maintain oral motor feeding skills. The paediatrician is an important partner for the paediatric cardiologist in optimising and monitoring weight gain in the post-operative period.

#### **Immunisations**

Immunisations have historically been deferred around the time cardiac surgery. One recent study demonstrated that cardiopulmonary bypass does not significantly lower existing titres of vaccine-related antibodies, and thus there may be no need to defer immunizations prior to surgery. However, fevers shortly before surgery or in the early post-operative period may cloud management, and as such, some degree of deferment may be appropriate. The primary paediatric cardiologist should specify if one or more vaccines need to be deferred from the routine immunisation schedule or if the patient qualifies for additional immunizations. For example, some post-operative and post-catheterization patients may still be deemed to have "hemodynamically significant congenital heart disease," so palivizumab (Synagis) may continue to be indicated. 13

# **Neurodevelopmental outcomes**

Infants and children who have undergone cardiac surgery are at risk for developmental delay and specific learning disabilities later in childhood. 14 The causes are multi-factorial including abnormal foetal circulation with potentially altered neurological development in utero, undergoing procedures that require the use of inhaled anaesthetics and cardiopulmonary bypass, periods of desaturation (either acute or chronic), and underlying genetic syndromes. Common developmental issues include language delay, learning difficulties, and attentional challenges. Adherence to the developmental screenings recommended by Bright Futures guidelines is essential to identify and treat such delays early in childhood.<sup>15</sup> Early intervention support should be initiated when indicated. Likewise, schools are well positioned to identify and provide support services for learning disabilities. The Cardiac Neurodevelopmental Outcome Collaborative has published recommendations for the care of these patients.<sup>16</sup>

# Dental care and SBE prophylaxis

Infective endocarditis can be a life-threatening complication, especially after a cardiac intervention. As such, routine dental hygiene

is essential to preventing caries and dental abscesses that can lead to bacteraemia and endocarditis. The paediatrician can help reiterate the importance of regular dental evaluations for patients with CHD. In addition, antibiotic prophylaxis to prevent spontaneous bacterial endocarditis may be indicated according to current guidelines. <sup>17,18</sup> Given rapidly evolving therapies, an individual patient's characteristics may not fall within guideline descriptors, so the paediatric cardiologist may specify if prophylaxis is indicated.

## **Lesion specific considerations**

Paediatricians familiar with common congenital heart lesions will be better equipped to care for their patients in the months following intervention and surgery.

# Atrial septal defects

Atrial septal defects that persist past toddlerhood are typically closed between three and five years of age to prevent damage to the pulmonary vascular bed and allow the right heart to remodel to normal size. Options for repair include device closure in the cardiac catheterisation laboratory and surgical closure with a patch or directly with sutures in the operating room. The decision for which approach is preferred depends on the size and location of the atrial septal defect.

Because of its minimally invasive nature and short recovery, transcatheter atrial septal defect device closure is typically preferred if there are adequate rims around the defect to keep the device in place. Potential complications following device closure include device migration in the short term and device erosion into the atrial wall or aorta in the longer term. Fortunately, these complications are quite rare, with the former occurring in roughly 0.55% and the latter in roughly 0.1% of cases. <sup>19</sup>

Surgical repair of atrial septal defects remains very safe with a mortality rate of approximately 0.1%. However, up to 40% of patients may develop post-pericardiotomy syndrome, which is characterised by pericardial inflammation and effusion in the first few weeks after surgery. Symptoms and signs of concern include fever, chest pain, abdominal pain, emesis, fatigue, and pericardial friction rub on exam. Evaluation typically includes an echocardiogram but may also include an electrocardiogram and chest X-ray. To prevent post-pericardiotomy syndrome, some centres place patients on non-steroidal anti-inflammatory therapies for 6–8 weeks following repair, though some data have challenged the efficacy of this approach. Supplementation of the supplementa

# Patent ductus arteriosus

When the patent ductus arteriosus does not close spontaneously, there are specific circumstances in which closure is indicated. The first is in the pre-mature neonate whose left to right shunt is haemodynamically significant. In these cases, the first-line treatment is often medical management with either non-steroidal anti-inflammatory drugs or acetaminophen. If medical therapy fails, then an intervention is often indicated, either surgical ligation or transcatheter closure. The second scenario is in the infant/toddler who does well clinically, but whose patent ductus arteriosus persists with resultant left-heart dilation from the volume load. In these cases, closure is warranted since the patent ductus arteriosus is haemodynamically significant. The third scenario is toddlers, children, and adults in whom the patent ductus arteriosus is small but remains audible on exam. Though based largely on expert opinion,

closure is typically recommended because the lifetime risk of endocarditis is thought to be greater than the one-time risk of closure.

Options for patent ductus arteriosus occlusion include transcatheter patent ductus arteriosus device placement and surgical ligation via thoracotomy or video-assisted thoracoscopic surgery. Because of its minimally invasive nature, the former is typically preferred, though both approaches carry a low procedural risk. Given the recent advent of devices for pre-mature infants, the indications and approaches are evolving and trending toward transcatheter closure. Potential complications following transcatheter patent ductus arteriosus device closure include device migration, with or without encroachment into the aorta and/or pulmonary artery, which rarely requires re-intervention and occasionally needs to be followed over time by a paediatric cardiologist.

# Ventricular septal defects

There are generally two instances where surgical closure of a ventricular septal defect is warranted. The first is in patients whose defects are large, unlikely to close spontaneously, and result in symptoms and/or chamber dilatation. In these cases, the congestive heart failure symptoms are typically managed medically until surgical closure between three and six months of age. The second common indication for ventricular septal defect closure is for defects of any size adjacent to the aortic valve, resulting in valvular distortion, prolapse, and/or aortic insufficiency.

Ventricular septal defect surgery typically involves sewing a patch of fabric or pericardium over the defect, though occasionally the ventricular septal defect can be closed directly with sutures. The operation carries a low mortality rate of approximately 0.2%. Up to 8% of patients may develop complete heart block during the operation, though most resolve spontaneously within a few days. A right bundle branch block is commonly seen on electrocardiogram after ventricular septal defect closure; this is typically of little haemodynamic significance but may result in a widely split S2 on examination. Post-operative follow-up includes monitoring for any residual defects and/or aortic insufficiency.

# Atrioventricular septal (canal) defects

An atrioventricular septal defect is an endocardial cushion defect in the crux (centre) of the heart, resulting in an abnormal atrioventricular valve and septal defects at the atrial and/or ventricular levels. Endocardial cushion defects are commonly associated with trisomy 21. A "complete" atrioventricular septal defect includes both an atrial septal defect and a ventricular septal defect. Cases in which the ventricular septal defect is partially closed by membranous tissue may be referred to as a "transitional" atrioventricular septal defect occurs when the ventricular septal defect is completely closed by fibrous tissue, though the left atrioventricular valve remains abnormal.

Surgical repair of complete atrioventricular septal defects is typically undertaken around three to six months of age, while transitional and partial atrioventricular septal defects can often be repaired at older ages, typically around three to five years of age. The repair involves closing the atrial and ventricular septal defects and dividing the common atrioventricular valve into functional left and right atrioventricular valves. Like isolated ventricular septal defect closures, there is a risk of acquired heart block with operation. The most common post-operative complications include left and right atrioventricular valve stenosis and/or regurgitation, development of left ventricular outflow tract obstruction, and

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residual atrial septal defects and ventricular septal defects, necessitating reintervention in up to 20% of patients.<sup>27,28</sup>

# Tetralogy of Fallot

Neonates with tetralogy of Fallot and severe cyanosis may require surgical or catheter-based intervention prior to hospital discharge to ensure adequate pulmonary blood flow. However, the "complete repair" of tetralogy of Fallot is generally undertaken around three to six months of age and consists of ventricular septal defect closure and relief of any obstruction of flow from the right ventricle to the distal pulmonary arteries. A pulmonary valve-sparing repair is preferred, but in cases where the pulmonary valve is too hypoplastic, a transannular patch may be required, sacrificing the pulmonary valve's competency. While pulmonary insufficiency following transannular patch repair is universal, it can also develop in patients who have undergone valve-sparing repair.

Over time, this insufficiency leads to progressive right ventricular dilatation, eventually necessitating pulmonary valve replacement. The timing of pulmonary valve replacement varies considerably based on numerous factors, including size and function of the right ventricle, presence of symptoms, and arrhythmias. Some children may also develop recurrent pulmonary artery stenoses necessitating intervention, including dilation and/or stent placement via cardiac catheterisation or arterioplasty at the time of a subsequent operation. Cardiac MRI serves as an important complementary diagnostic modality to echocardiography and catheterization in patients with history of tetralogy of Fallot repair. In the cardiac dilatation and the catheterization in patients with history of tetralogy of Fallot repair.

#### D-looped transposition of the great arteries

Neonates born with transposition of the great arteries require surgical repair within the first few weeks of life to survive. Typically, this is accomplished with an arterial switch operation, in which the great arteries are reversed to re-establish normal arterial connections to the ventricles. This involves transferring the coronary arteries as well. In addition, a LeCompte manoeuvre is often required, draping the branch pulmonary arteries over the front of the ascending aorta, rather than having the right pulmonary artery pass under the aortic arch.

Common residual lesions include aortic or pulmonary stenoses and/or insufficiency, as well as neo-aortic root dilatation. <sup>32,33</sup> Recurrent branch pulmonary artery stenoses, more likely when a LeCompte procedure is performed, may require one or more subsequent percutaneous interventions. <sup>34</sup> Coronary insufficiency, though rare, may develop over time and warrant follow-up by the primary paediatric cardiologist. <sup>35,36</sup>

# Total anomalous pulmonary venous connection

Most new-borns with total anomalous pulmonary venous connection undergo surgical intervention prior to hospital discharge, though exceptions exist. Important post-operative complications include pulmonary vein stenoses, which can present as tachypnoea or worsening shortness of breath and warrant urgent evaluation by the cardiologist.<sup>37</sup>

# Truncus arteriosus

Surgical repair of truncus arteriosus is generally performed early in infancy due to high output heart failure from torrential pulmonary blood flow and to minimise resultant damage to the pulmonary vascular bed. The Rastelli repair consists of closing the ventricular

septal defect, removing the pulmonary arteries from the aorta, and placing a valved conduit from the right ventricle to the pulmonary arteries. Truncal valve stenosis and insufficiency may complicate the repair and prognosis. Furthermore, about one-third of babies born with truncus arteriosus have 22q11.2 deletion syndrome (DiGeorge Syndrome), which can result in important hypocalcaemia and immunodeficiency. Pulmonary conduit stenosis and/or insufficiency ultimately necessitates pulmonary valve replacement. Additional complications include branch pulmonary artery stenoses and truncal root dilatation.<sup>38</sup>

#### Coarctation of the aorta and interrupted aortic arch

Aortic arch obstruction is typically identified and surgically repaired in the neonatal period, though diagnoses in later child-hood do still occur. Recurrent laryngeal nerve injury after manipulation of the aortic arch can result in vocal cord paralysis, which may complicate post-operative feeding. In the months to years following repair, recurrent coarctation may develop and necessitate balloon angioplasty and/or stent placement. Some repair techniques utilise the subclavian artery, so cognizance of subclavian artery origins and patency guides interpretation of non-invasive upper and lower extremity blood pressures and pulses. A gradient of greater than 20 mmHg (upper extremity higher than lower extremity) is considered haemodynamically significant and may warrant intervention.

Systemic hypertension is common in patients following aortic arch repairs and should be treated if detected.<sup>39,40</sup> Hypertension that is resistant to multiple medications may be due to recurrent coarctation as well.<sup>41</sup> Additional important complications include aortic and cerebral aneurysms. Because significant morbidity and mortality arise from cardiovascular causes in patients who have undergone coarctation repair, it is imperative that comorbid hypertension is adequately treated.<sup>42</sup>

# Aortic stenosis

Abnormalities of aortic valve development (e.g., bicuspid aortic valve) can result in aortic stenosis. When severe and not appropriate for balloon valvuloplasty, one of several surgical options is chosen based on the patient's size, potential for growth, and tolerance for chronic anticoagulation. The Ross operation utilises the patient's native pulmonary valve to replace the dysfunctional aortic valve, and a pulmonary homograft or valved conduit is placed in the pulmonary position. This has the advantage of a neo-aortic valve that grows with the patient but typically necessitates one or more future pulmonary valve replacements for stenosis and/ or insufficiency. Alternatively, the aortic valve can be replaced with either a bioprosthetic or mechanical valve. Bioprosthetic valves do not typically require chronic anticoagulation, though they have a limited lifespan. Mechanical valves have the potential to last a lifetime but require chronic anticoagulation, carrying roughly a 1–3% risk of thrombosis or bleeding per valve-year. 43,44 Neither of these valves have potential to grow with the child. Paediatricians can serve as important allies in encouraging adherence to anticoagulation, oral hygiene, and endocarditis prophylaxis recommendations, as non-adherence can rapidly lead to complications including aortic stenosis, aortic insufficiency, and endocarditis.

# **Pulmonary stenosis**

Pulmonary valve stenosis can be relieved by balloon angioplasty or surgical valvotomy. Potential complications include residual

pulmonary stenosis or insufficiency. In some cases, these can ultimately necessitate pulmonary valve replacement.

# Ebstein anomaly

In Ebstein anomaly, the tricuspid valve is displaced apically into the right ventricle and can have varying degrees of dysplasia and regurgitation. In milder cases requiring surgery, tricuspid valve repair or tricuspid valve replacement may be performed, resulting in a biventricular repair. However, in more severe cases, the functional right ventricle may be too small to handle all the systemic venous return, necessitating at least partial progression down the single ventricle pathway. Complications of biventricular repair include residual tricuspid stenosis and/or regurgitation, as well as supraventricular tachycardia.

# Vascular ring

Several embryologic substrates result in a vascular ring encircling the trachea and oesophagus. When this substrate results in respiratory insufficiency, frequent respiratory infections, or swallowing difficulties, vascular ring division is indicated. This is typically undertaken via lateral thoracotomy or video-assisted thoracoscopic surgery. Residual tracheal hypoplasia and/or malacia typically improves with patient growth after vascular ring division. Swallowing difficulties also typically resolve, though in some cases, the presence of a retroesophageal vascular structure continues to cause symptoms.

# Congenitally corrected transposition of the great arteries

Children with congenitally corrected transposition of the great arteries have both atrioventricular discordance and ventriculoarterial discordance. This results in deoxygenated blood appropriately being pumped to the pulmonary arteries and oxygenated blood appropriately being pumped to the aorta. Children who have associated lesions such as ventricular septal defects or pulmonary stenosis may require surgery early in life. Complete heart block commonly complicates post-operative recovery, in some cases necessitating placement of a pacemaker.

Children without associated lesions may not require surgery for many years. However, the morphologic right ventricle (typically the anatomically left-sided ventricle) is not designed to pump against systemic vascular resistance for a lifetime, ultimately leading to pre-mature heart failure in early to mid-adulthood if unaddressed. Some centres pre-empt this by performing a "double switch" operation, in which an atrial switch is performed simultaneously with an arterial switch. In addition to the above complications associated with arterial switch, the atrial switch introduces additional risks of atrial arrhythmias and atrial baffle stenoses and/or leaks. Baffle stenoses and leaks can often be addressed in the catheterisation laboratory.

# **Special Circumstances**

# Pulmonary valve replacement

As described above, many forms of repaired and unrepaired CHD require one or more pulmonary valve replacements as children grow. Surgical pulmonary valve replacement carries a low procedural risk. Hospital stay for surgical pulmonary valve replacement is typically around 2–5 days, with a post-operative mortality less than 1%. Most surgically placed pulmonary valves

last between 4 and 15 years before requiring replacement, either due to valve dysfunction or patient growth. 46

Transcatheter pulmonary valve replacement utilises a percutaneous approach and requires a shorter hospital stay, typically under 24 hours. Though data are relative sparse and still evolving, there has been question as to whether some transcatheter valves have a higher risk of bacterial endocarditis, approaching 2.5% per patient-year. As mentioned earlier, proper dental hygiene is critically important for these patients. Similarly, aspirin has been shown to decrease this risk, so adherence is of particular importance. The paediatrician should be aware that an unexplained fever lasting greater than 5 days should prompt consideration of obtaining a blood culture.

# Balloon and stent arterioplasty and venoplasty

Numerous congenital heart lesions, whether unrepaired or due to a complication of repair, benefit from percutaneous angioplasty and/ or stent placement. Reperfusion injury to the affected lung can occur following intervention for branch pulmonary artery stenoses; this typically occurs the night of the procedure and is treated with diuretics and oxygen. Re-stenosis is common following intervention for acquired pulmonary vein stenoses, often necessitating multiple repeat interventions. Balloon dilation of recurrent aortic coarctation carries a risk of aortic aneurysm, dissection, embolisation, and stroke. Percutaneous right ventricle to pulmonary artery conduit interventions may result in worsening pulmonary insufficiency. In general, stents require endocarditis prophylaxis for 6 months following placement, with aspirin therapy at the discretion of the cardiologist.

## Pacemaker and defibrillator placement

Patients with documented sinus node dysfunction, atrioventricular block, and life-threatening arrhythmias may have a pacemaker or implanted cardioverter-defibrillator placed. Such devices can be placed either via a transvenous or epicardial approach, depending on patient size and anatomy. Complications following placement include surgical site infection and endocarditis. Battery life in a pacemaker or defibrillator is limited and generally necessitates replacement of the device. The wires typically last much longer and only need to be replaced when no longer conducting effectively; these are routinely tested during device interrogation.

MRI in patients with implanted electronic devices was previously regarded as an absolute contraindication. Recent studies demonstrating safety have resulted in guideline revisions that allow some patients with devices to undergo MRI. <sup>49,50</sup> Such studies are typically performed under the guidance of a supervising radiologist or cardiologist in collaboration with the device manufacturer.

# Conclusion

Paediatricians are integral in the lifelong care of children with CHD. Familiarity with common issues following cardiac surgery or intervention, as well as key details regarding specific lesions and surgeries, will aid the paediatrician in caring for these patients well.

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