Post-operative Care of Common Congenital Heart Defects

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Introduction
Children with acquired and congenital heart disease account for 30 – 40% of all admissions to Pediatric Intensive Care Units (PICU). Children with heart disease may require care in PICUs for many reasons including management of cardiac failure, management of rhythm issues, and recovery following surgery for congenital heart disease. Thus, knowledge of assessment and management of children with cardiac disease is essential to the practice of Pediatric Intensive Care Medicine. Congenital heart disease (CHD) occurs in 4/1000 live births and is the leading cause of birth defect associated mortality in children. One third have critical CHD and require intervention within the first year of life. Successful repair of CHD requires accurate anatomical diagnosis, skilled surgical and perfusion teams, excellent post-operative care, and long-term follow-up and care.

Post-operative care of children undergoing congenital surgery
Post-operative care of children undergoing surgery for CHD requires the coordinated effort of a team of caregivers including medical, surgical, nursing, allied health care professionals, and family members. The post-operative care requires knowledge of pre-operative condition, anatomy and physiology of the CHD. At the time of ICU admission a detailed sign-out of information from the operating room team to the ICU providers is of paramount importance. Intra-operative course and information from intra-operative imaging at the end of the operation should be obtained from the operating room team. Post-operative assessment should include physical examination, review of hemodynamic data, laboratory data, ECG, and CXR. Evaluation of the post-operative patient should be focused on diagnosing residual heart defects, assessing cardiac output (CO), surgical bleeding, and end-organ function. A plan for management of the post-operative patient, type of frequency of laboratory assessment, and expected time course to extubation from mechanical ventilation should be made with the time of admission to the PICU.

Post-operative management should aim to optimize CO and tissue oxygen delivery by ensuring adequate preload, myocardial contractility, and afterload, ensuring sinus rhythm, and reducing oxygen consumption (Table 1). Low Cardiac output syndrome (LCOS) resulting in a Cardiac Index (CI) of < 2L/min/m² occurs in about 25% of infants undergoing cardiac surgery. LCOS increases risk of mortality, morbidity, and resource utilization and should be promptly diagnosed and managed. Post-operative LCOS resolves over the first 24-48 hours in most cases. Hoffman et al showed that prophylactic use of milrinone infusion reduced the incidence of post-operative LCOS and improved clinical outcomes in children undergoing cardiac surgery. Persistent LCOS or a post-operative course deviating from the expected course for type and physiology of CHD should promptly trigger investigation for residual heart defects that may impair recovery. Other post-operative considerations include adequate pain control, nutrition, and care of the patient’s family. Weaning from mechanical ventilation and extubation should be considered once hemodynamic stability and good hemostasis has been achieved.

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Specific issues pertinent to the post-operative management of 3 common forms of CHD following surgical repair are discussed below.

**Ventricular Septal Defect (VSD)**

Defects in the inter-ventricular septum are the commonest form of congenital heart defects (20% of all defects) with an incidence of 0.38 - 2.3/1000 live births\(^3\). Ventricular Septal Defects (VSD) can be single or multiple and can occur in the outlet (conoventricular), membranous, inlet, or the muscular portions of the inter-ventricular septum\(^7\). VSD cause a left-to-right shunt at the ventricular level and symptoms depend on the size of the defect. Large defects (approximately the size of the aortic valve) cause increased pulmonary blood flow and equalize pressures in the left (LV) and right ventricles (RV). Small defects may be asymptomatic and present with murmurs, however large defects present with symptoms of congestive heart failure (CHF) due to excessive pulmonary blood flow. Diagnosis and location of VSDs can be confirmed using an echocardiography (ECHO). Cardiac catheterization can be used to quantitate the magnitude of left-to-right shunt.

Indications for surgical intervention in patients depend on the size of VSD, presence of CHF, and response to CHF therapies. Because some defects can close spontaneously, patients with small - moderate sized VSD responding to CHF therapy could be followed for spontaneous closure up to 5 years of age. Simple and single VSD can be successfully closed in the operating room. Post-operative recovery following simple VSD closure should be generally uneventful.

**Tetralogy of Fallot**

Tetralogy of Fallot is a common form of cyanotic congenital heart disease with an incidence of 0.21 – 0.37/1000 live births\(^3\). The characteristics features of TOF include 1. VSD (conoventricular type), 2. Right Ventricular Outflow Tract (RVOT) obstruction (consisting of a combination of obstruction at, below, or above the level of the pulmonary valve), 3. Dextroposed Aorta that overrides the ventricular septum, and 4. Right Ventricular Hypertrophy (RVH)\(^9\). Van Praagh et al have proposed that the fundamental problem underlying the development of TOF is hypoplasia of the sub-pulmonary infundibulum\(^10\). The physiology of TOF is determined the severity of obstruction of the RVOT\(^7\). When obstruction to the RVOT is minimal the predominant physiology is one

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**Table 1: Post-operative Management following Surgical Repair of Congenital Heart Defects**

<table>
<thead>
<tr>
<th>Step</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Receive Information from Cardiac Surgical Team: “Sign-out” and Assess patient&lt;br&gt; Review operative course&lt;br&gt; Review information post-operative hemodynamic assessment in OR</td>
</tr>
<tr>
<td>2.</td>
<td>Post-operative Assessment and Post-Operative Management Plan&lt;br&gt; Physical Exam and Review of Laboratory Data&lt;br&gt; Post-operative management plan</td>
</tr>
<tr>
<td>3.</td>
<td>Ensure Adequate Pre-Load&lt;br&gt; Use CVP or atrial filling pressure to titrate preload</td>
</tr>
<tr>
<td>4.</td>
<td>Support Myocardial Contractility&lt;br&gt; E.g. Dopamine or Epinephrine</td>
</tr>
<tr>
<td>5.</td>
<td>Reduce Afterload&lt;br&gt; Vasodilators or Inotodilators&lt;br&gt; Positive pressure ventilation</td>
</tr>
<tr>
<td>6.</td>
<td>Manage Rhythm&lt;br&gt; Treat Arrhythmia&lt;br&gt; Temporary Pacing</td>
</tr>
<tr>
<td>7.</td>
<td>Provide Mechanical ventilation&lt;br&gt; Maintain Functional Residual Capacity to lower PVR&lt;br&gt; Using minimal Mean Airway Pressure</td>
</tr>
<tr>
<td>8.</td>
<td>Reduce Oxygen Consumption&lt;br&gt; Mild Hypothermia&lt;br&gt; Mechanical ventilation&lt;br&gt; Pain management, sedation, and neuromuscular blockade</td>
</tr>
<tr>
<td>9.</td>
<td>Monitor for Recovery&lt;br&gt; Follow heart rate, urine output, mixed venous oxygen saturation, or serum lactate levels</td>
</tr>
<tr>
<td>10.</td>
<td>Rule out Residual Heart Defects&lt;br&gt; Physical exam, Echocardiography, or Cardiac Catheterization</td>
</tr>
<tr>
<td>11.</td>
<td>Mechanical Circulatory Support&lt;br&gt; Consider ECMO for failure of conventional medical therapies</td>
</tr>
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CVP: Central Venous Pressure; PVR: Pulmonary Vascular Resistance; ECMO: extracorporeal Membrane Oxygenation
of a left-to-right shunt across the VSD. When RVOT obstruction is severe, then RV pressure rises and a right-to-left shunt across the VSD ensues causing cyanosis and pulmonary blood flow is reduced. The obstruction to the RVOT may be “dynamic” or increase under circumstances such as crying or agitation resulting in marked worsening of cyanosis resulting in “hypercyanotic” or “Tet Spells”.

As previously stated the clinical manifestation of TOF vary based on the degree of RVOT. Cyanosis, RVOT ejection systolic murmur, RVH on Electrocardiogram (EKG), and a boot shaped heart indicating RV enlargement on CXR may be present and indicate a diagnosis TOF. ECHO can be used to demonstrate the anatomical details of VSD, degree and site of RVOT obstruction. Other anatomical details may be important for surgical planning include presence or absence of additional VSDs, size of the proximal and branch pulmonary arteries, presence or absence of Aorto-Pulmonary collateral vessels, coronary anatomy, and presence of atrial communication. TOF may be associated with other chromosomal (DiGeorge Syndrome) and non-chromosomal (VATER) syndromes. These issues should be evaluated as they can complicate the post-operative course and management.

“TET Spells” can be caused both by increased RVOT obstruction brought on by catecholamine induced spasm of the pulmonary infundibulum or as result of a decrease in systemic vascular resistance encouraging increased right-to-left shunting at the VSD. Management includes calming an agitated child, supplemental oxygen administration and placing the infant in knee chest position. If a spell cannot be terminated, then intravenous access should be obtained, narcotics should be administered, and preload to the RV should be increased by volume expansion should be used. Bolus doses of Phenylephrine to increase systemic vascular resistance and decrease right-to-left shunting may be needed. Some children may not respond to these measures and may require endotracheal intubation, mechanical circulatory support, or emergency surgical intervention.

All patients with TOF require surgical intervention to correct the underlying anomalies. Timing of intervention for patients with TOF is somewhat controversial with some groups pursing a full TOF repair by 3 -4 months of age and other delaying repair to later age (6 months – 1 year) using a Systemic to Pulmonary Artery (Blalock-Taussig Shunt) to provide pulmonary blood flow which awaiting full repair. Severe RVOT obstruction or “TET Spells” is an indication for early surgical intervention including in the neonatal period. Full repair of TOF includes relief of RVOT obstruction and VSD closure. Most children undergoing TOF repair make an uneventful post-operative recovery. Clinical assessment following admission to the ICU after TOF repair should focus on detection of residual anatomical abnormalities including the presence of a residual VSD or significant RVOT obstruction.

LCOS due to diastolic dysfunction of the right ventricle also called “restrictive RV physiology” is common in patients undergoing repair of TOF. Myocardial injury and edema resulting from incision of the RVOT, RV hypertrophy, and post-operative pulmonary regurgitation are thought to contribute to development of restrictive RV physiology. These patients have a non-compliant RV thus need increased venous pressure for RV filling in diastole. LCOS in these patients can arise from decreased RV output and adverse inter-ventricular interaction due to RV diastolic dysfunction both resulting in decreased LV output and CO. Patients with “restrictive RV physiology” present with signs of LCOS, poor perfusion, elevated central venous pressures, metabolic acidosis, oliguria, hepatomegaly, ascites, and pleural effusion. A decompressing atrial communication (e.g. Patent Foramen Ovale) can help maintain LV preload and CO by allowing a right to left atrial shunt, and offset the high RA pressure by allowing decompression into the LA via the communication. Although an atrial communication can make post-operative management of patients with RV physiology easier, it comes at the expense of lower oxygen saturation because of the right-to-left shunt. The right-to-left shunt will decrease and oxygen saturation will increase as RV compliance improves over time in the post-operative period.

The diagnostic features of restrictive RV physiology were described by Cullen et al using Doppler
Echocardiography in patients with Tetralogy of Fallot and includes demonstration of antegrade flow into the pulmonary artery in atrial systole. Antegrade flow into the pulmonary artery during atrial systole contributes to cardiac output and can be decreased by the use of positive pressure mechanical ventilation often used during recovery following cardiac surgery. Thus mechanical ventilation in patients recovering from TOF should be provided with the lowest mean airway pressure possible. Early extubation from mechanical ventilation in the postoperative may be beneficial in the patients.

A number of cardiac arrhythmias requiring intervention have been described following TOF repair. Junctional ectopic tachycardia (JET) is a common (4n–22%) rapid, automatic, and catecholamine sensitive tachycardia seen following TOF repair. Early detection and aggressive management of JET is essential because JET often results in loss of atrioventricular (a-v) synchrony and can decrease cardiac output. Management of JET includes, use of sedation and neuromuscular blockade, decreasing vasoactive support where possible, induced hypothermia (usually 34–35°C), use of antiarrhythmic agents (procainamide, amiodarone) and temporary atrial pacing to provide a-v synchrony. Recently administration of intravenous magnesium following weaning from cardiopulmonary Bypass has been shown to reduce the incidence of JET and use of Intravenous Dexmedetomidine have been shown to be effective in the management of JET.

**D-Transposition of Great Arteries** (also see p161)

The estimated incidence of D-Transposition of the Great Arteries (D-TGA) is 0.21 – 0.38/1000 live births and accounts for 7 – 8% of all congenital cardiac malformations. D-TGA is thought to arise from abnormal rotation and septation of the arterial trunci however the exact embryological origin of the defect has not been clearly defined. In D-TGA the aorta arises from the right ventricle and the pulmonary artery from the left ventricle. This results in a parallel systemic and venous circulation where desaturated venous blood returning to the heart is ejected to the systemic circulation resulting in profound cyanosis and saturated pulmonary venous return is ejected back into the pulmonary circulation. Survival and oxygenation depends on mixing of desaturated systemic venous and saturated pulmonary venous return via a Patent Ductus Arteriosus (PDA), Atrium (Patent Foramen Ovale or Atrial Septal Defect) or Ventricle (Ventricular Septal Defect).

The diagnosis of D-TGA in any cyanotic newborn is made using 2-dimensional echocardiography (ECHO) because physical exam, chest radiography, and electrocardiography may not be contributory to making the diagnosis. In addition to confirming the diagnosis of D-TGA, ECHO should survey the presence of other cardiac abnormalities as these issues may have implications surgical planning. These include VSD (present in 20% of cases), Aortic arch obstruction, coronary anatomy, and the presence left ventricular outflow tract obstruction.

Pre-operative management includes maintaining adequate oxygen saturation and cardiac output. Patient with profound cyanosis require resuscitation with use of prostaglandin E1 (PGE1) infusion, mechanical ventilation, and an emergent Balloon Atrial Septostomy (BAS) to ensure adequate mixing and oxygen saturation.

The Arterial Switch Operation (ASO) is the procedure of choice in patients with D-TGA. The procedure is performed via median sternotomy and cardiopulmonary bypass and includes transection of the great vessels and restoring their connection to the appropriate ventricle, translocation of coronary arteries to the neo-aorta and closure of atrial communications (and VSD when present). The optimal timing of ASO is though around 5 – 10 days of age, however a recent study showed increased morbidity and cost for when ASO is undertaken after 3 days of age. Because the myocardium of left ventricle in D-TGA pumping to a low resistance pulmonary circulation can involute over time, infants presenting late (> 8 weeks) with DTGA are at risk of LV failure after ASO and require special consideration with regards to LV training or post-operative support of the LV prior to ASO. Strategies for management of children presenting late with D-TGA and LV involution include a 2-staged procedure where a pulmonary artery band and Blalock-Taussig shunt are used to train the LV prior to ASO or recovery of
the LV function with ECMO support. All general principles of Intensive Care monitoring and management for children undergoing cardiac surgery apply to patients recovering following ASO. Special considerations include managing Post-Operative Low Cardiac Output Syndrome (LCOS) and recognition and management of coronary ischemia. Wernovsky et al in a cohort of 170 neonates (122 had cardiac index measurements) undergoing ASO demonstrated that a decrease in cardiac index (CI) occurred during the 9 to 12 post-operative hours following cardiac surgery and approximately 24% of patients in this had a CI < 2.0 L/min/m² during the first 24 post-operative hours. Thus LCOS is common following ASO and management strategies should include ensuring adequate pre-load, enhancing myocardial performance using inotropes and inodilators, afterload reduction, ensuring sinus rhythm, mechanical ventilation, and therapies aimed at reducing oxygen demand/consumption (hypothermia, sedation, and neuromuscular paralysis). Careful monitoring of heart rate, systolic blood pressure, peripheral perfusion, urine output and serum lactate can help assess cardiovascular function. Progressive deterioration should prompt further evaluation with ECHO and consideration of mechanical circulatory support. Myocardial ischemia may be suggested by LCOS, hypotension, increasing Left Atrial (LA) pressure, ST-T abnormalities on ECG, and regional LV wall motion abnormalities on ECHO should prompt investigation of coronary anastomosis and possible revision. Survival to hospital discharge and long-term outcomes appear to be good for neonates undergoing ASO.

Summary
Successful management of children undergoing surgery for CHD requires detailed understanding of anatomy and physiology of the CHD both in pre and post-operative periods, and coordination of a multidisciplinary team of experts and anticipation and management of complications that arise in the post-operative period.

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References