Common Congenital Heart Defects and Perioperative Issues in Management

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Introduction

Incidence of congenital heart disease (CHD) is approximately one in every 1,000 live births however variation in incidence from 1-4 in every 1000 live births exists in reported literature1,2. Acyanotic congenital heart defects account for 70% of all congenital heart disease3. One third have critical CHD and require intervention within the first year of life4. Ventricular septal defect (VSD), Atrial septal defect (ASD) and Patent ductus arteriosus (PDA) account for a majority percentage of all congenital heart defects5. In these defects there is an abnormal communication between the high-pressure left side of the heart and the low-pressure right side of the heart allowing blood to shunt from left to right (Table 1).

Table 1 Acyanotic congenital heart defects:

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Blood flow through the shunt depends on the size of the shunt and the relative resistances on either side of the shunt. If the defect is large enough not to impede blood flow (i.e. non-restrictive defect), then the main determinant of the degree of shunting across this defect depends on the resistance on both sides of the shunt. If the defect opening is small (i.e. restrictive defect), then the degree of shunting will depend more on the size of the defect and less on the resistance across the defect. The drop in pulmonary vascular resistance (PVR) occurring shortly after birth creates the driving force for the left-to-right shunting of blood across the defect. The pressure difference resulting in left-to-right shunting of blood through the defect consequently leads to turbulence of abnormal blood flow producing a heart murmur in systole and sometimes in diastole, excessive blood flow into the lungs causing pulmonary vascular congestion leading to shortness of breath, increased volume overload of the myocardium resulting in hypertrophy and chamber dilation, and eventual congestive heart failure (CHF).

Development of CHF is the main concern in acyanotic shunt lesions, as opposed to cyanotic CHD in which hypoxia is the main concern. CHF presents as sweating, feeding difficulties, poor growth or failure to thrive (FTT), S3 gallop, and if left-sided, also tachypnea, substernal retractions, and/or pulmonary rales. Chest radiographs in CHF may demonstrate cardiomegaly and increased pulmonary vascular congestion and edema. Echocardiograms are the primary diagnostic modality, but magnetic resonance imaging provides excellent anatomic evaluation and often yields even more information than angiography.

Untreated defects with large shunts will eventually result in injury to the pulmonary arterioles, vascular obstruction, and pulmonary hypertension. The development of permanent injury to the pulmonary vessels is a function of the duration of exposure to this excessive blood flow, and occurs more rapidly in VSD and PDA than in ASD. If this process is not reversed, eventually the Eisenmenger’s complex of right to left shunting may occur as the elevated right-sided pressures (pulmonary hypertension) exceed left-sided pressures. Following is a critical review and discussion of specific lesion characteristics and perioperative issues6,9. A detailed lesion specific postoperative management is described in various subsequent sections of part 2 of Pediatric cardiac intensive care symposium in this issue.
Ventricular Septal Defect

VSD is the most common form of CHD at 15-20% of all cases of isolated CHD. Small defects account for about 85% of all VSDs. They are typed as per location in septum:
1. Perimembranous
2. Infundibular
3. Muscular
4. Inlet

Symptoms depend on the size of the defect. Symptoms can be related to mild to severe congestive heart failure.

Smaller defects (2-4 mm) are usually asymptomatic and more likely to spontaneously close (50% by 2 years). About 30-40% of small perimembranous and muscular defects spontaneously close within the first six months of life. Small defects usually do not require surgery or activity restrictions and patients are monitored with periodic follow-up.

Larger non restrictive VSDs are more likely to be symptomatic and less likely to spontaneously close. CHF can manifest around six to eight weeks of age. Early surgical repair may be indicated.

The physiology of the shunting results in volume overload of the right ventricle (RV), right atrium (RA) and the pulmonary circulation that is transmitted to the left side of the heart as well.

Initially, the increased blood volume returning to the left ventricle (LV) increases the stroke volume (Frank-Starling mechanism), however with time the volume overload can lead to LV dilatation, systolic dysfunction and symptoms of left-sided heart failure as early as 2-3 months of age (failure to thrive, tachypnea, lower respiratory tract infections). The volume overload of the pulmonary vasculature leads to increased pulmonary artery pressure (PAP) and pulmonary arteriolar hypertrophy with time with pulmonary hypertension setting in as early as the age of 2-3 months.

If Qp: Qs (pulmonary to systemic blood flow) ratio is greater than 2, accompanied by symptoms of right ventricular volume overload, tachypnea, poor feeding, failure to thrive, then surgery for VSD closure is indicated.

On cardiac auscultation one may hear grade II-VI harsh holosystolic murmur along the lower left sternal border and a prominent P2 as the pulmonic valve closes later than the aortic valve. A VSD murmur is not heard at birth till pulmonary vascular resistance (PVR) and pulmonary pressures are high. Once PVR falls, the left-to-right shunt increases resulting in a new emergence of increased severity of murmur on day 2 or day 3 of life.

Subacute bacterial endocarditis prophylaxis are recommended to children with VSD. In large defects with near systemic pressures in the right ventricle and pulmonary artery, surgical closure should be performed prior to 18 to 24 months of age even if heart failure control and adequate weight gain are present. Total surgical correction is currently recommended. If the congestive heart failure is difficult to control with the usual anticongestive measures or if failure to thrive is present, surgical closure should be undertaken earlier. At the present, transcatheter VSD occlusion is considered experimental.

Typically VSD closure involves a patch closure and common postoperative issues are related to post cardiopulmonary bypass status, low cardiac output syndrome, residual significant muscular VSD, rarely ventricular outflow tract obstruction caused by patch and aortic insufficiency if VSD is Subaortic. Usually post operative vascular lines include central and arterial line. Some units use left atrial lines. Pacing wires are kept for temporary pacing for conduction defects, such as AV conduction abnormalities and transient right bundle branch block. Conduction issues usually resolve in 2-3 days. In the event of an AV block persisting for longer than 7-10 days, a permanent pacemaker may be required. Pulmonary hypertension needs special attention as right ventriculat failure and tricuspid regurgitation may need to be monitored closely. Chest and mediastinal drain output is monitored closely. Most patients can be successfully extubated in 48 hours after surgery. Close monitoring of fluid therapy, inotropy as required directed to prevent pulmonary hypertension and optimal RV volume are the key issues in post operative period. Detailed lesion specific management issues are addressed in another review in this issue (page 127).

Atrial Septal Defect

ASD accounts for 10% of all CHD. It is more common in females.

Types based on different locations are:
1. Sinus venosus: Sinus venosus defects are located near the opening of the SVC.
2. Secundum: Most common is secundum defects which occur through the fossa ovalis.
3. Primum: Primum defects are a form of endocardial cushion defects involving the lowest part of the atrial septum between the atrioventricular valves. These are almost always associated with abnormal development of atrioventricular valves, most commonly a cleft mitral valve. Primum defects can be associated with Downs syndrome, Ellis-Van Creveld syndrome, and Holt-Oram syndrome.

Endocardial cushion separates the atrioventricular valves and forms the lower portion of the atrial septum and the upper portion of the interventricular septum. If ASD presents in infancy, then increased left atrial pressure, mitral valve stenosis, left ventricular dysfunction or left ventricular outflow obstruction should be suspected and investigated.

Most patients remain asymptomatic, but some children can develop right atrial and ventricular dilation, leading to atrial arrhythmias and ventricular dysfunction.

Patients may have a hyperactive precordium, right ventricular heave, fixed wide split S2, systolic ejection murmur at the second left intercostal space (increased flow across the pulmonic valve), or a mid-diastolic murmur at the lower right sternal border (increased flow across the tricuspid valve). Flow across the ASD is low velocity and not turbulent, so there is no audible murmur from the ASD itself.

Similar to VSDs, smaller defects are expected to spontaneously close while larger ones usually require surgical intervention with patch closure. Despite lack of symptoms at presentation, elective closure of the ASD is recommended around 4 to 5 years of age and is recommended so as to (i) prevent development of pulmonary vascular obstructive disease later in life, (ii) reduce probability for development of supraventricular arrhythmias and (iii) prevent symptoms during adolescence and adulthood.

Closure during infancy is not undertaken unless the infant is symptomatic. Right ventricular volume overload by echocardiogram and a Qp:Qs >1.5 (if the child had cardiac catheterization) are indications for early closure.

Device closure of ASD is discussed elsewhere in this issue (refer to page 143)

The Amplatzer Septal Occluder is rapidly becoming the device of choice by percutaneous right heart catheterization. Both immediate and mid-term follow-up results of Amplatzer Septal Occluder appear excellent with immediate complete closure rates varying from 62% to 96% which improved to 83% to 99% at six to 12 month follow-up.

Problems in the immediate postoperative period include:
   a. Low cardiac output
   b. Atrioventricular valve regurgitation [rapid fluid boluses may lead to annular dilatation and aggravate regurgitation, so rapid boluses should be avoided]
   c. Pulmonary hypertension
   d. AV block or supraventricular arrhythmias need to be monitored.
   e. Cardiac dysrhythmias and mitral valve prolapse may be late sequelae of a treated or untreated ASD. Atrial flutter or fibrillation may also occur in adults with a history of atrial septal defect, regardless of the treatment.

Patent Ductus Arteriosus

PDA results from retention of the fetal ductus arteriosus, which normally closes at about one to two weeks of age. This defect accounts for 5-10% of CHD and is more common in females. PDA can coexist with prematurity, VSD, coarctation of the aorta and rubella exposure during the first trimester of pregnancy.

As pulmonary vascular resistance decreases after birth, blood shunts from the aorta into the pulmonary artery, resulting in increased pulmonary artery blood flow and left atrial and ventricular overload. A large PDA results in large left to right shunt leading to pulmonary over circulation and low aortic diastolic pressure, leading to extensive aortic runoff and systemic end-organ hypoperfusion. Pulmonary vascular obstructive disease may occur as early as one year of age.

On clinical exam one finds bounding arterial pulses, a widened pulse pressure, an enlarged heart, a prominent apical impulse, a classic continuous machine-like murmur at the base and a mid-diastolic murmur at the apex.

Small defects are usually asymptomatic, while large
PDAs present with recurrent pulmonary infections, CHF, and failure to thrive. Closure may occur spontaneously. It is generally believed that the presence of an isolated ductus is an indication for closure, mainly to prevent bacterial endocarditis. Waiting until 6 to 12 months of age is generally recommended unless child is symptomatic with heart failure or pulmonary compromise. Indomethacin can be used for medical closure. Some require surgical closure with placement of an embolic device, such as an intravascular coil or PDA occluder, ligation, or division. PDA is the only CHD that is considered surgically “cured” without long-term sequelae. Post operative complications are rare but well known including opening of sutures (If no division performed after ligation), recurrent laryngeal nerve palsy (hoarseness and stridor), phrenic nerve injury(elevation of dome of diaphragm) and very rarely ligation of descending aorta (lower extremity pulses feeble or none along with upper extremity hypertension), left pulmonary arterial ligation(left lung gets oligemic)

Subacute bacterial endocarditis prophylaxis is recommended for all PDAs. There may not be any need for this prophylaxis three months following surgical or transcatheter closure, provided there is no residual shunt.

Aortic Stenosis (AS) AS is the obstruction of the left ventricle outflow tract. AS accounts for 7% of CHD and is described by location:
1) valvular (most common);
2) subvalvular or subaortic; or
3) supravalvular (least common, associated with Williams Syndrome).

As a word of caution critical aortic stenosis may present in newborn period as the ductus closes , in a similar way as the entire spectrum of left sided obstructive lesions such as aortic arch abnormalities, aortic atresia, hypoplastic left heart syndrome (Acidotic, Low perfusion, new born in shock at day5-7). Prostaglandin E1 therapy may be life saving temporizing measure to keep the ductus arteriosus patent to achieve systemic perfusion. When the child grows, the cardiac output increases resulting in an increased pressure gradient across the stenosis. Obstructed flow from the left ventricle results in increased pressure and hypertrophy. Mild AS is usually asymptomatic with some exercise intolerance and easy fatigability. Moderate AS may present with chest pain, dyspnea on exertion, dizziness, and syncope. Severe AS presents with weak pulses, left-sided heart failure, and chest pain and could lead to sudden death.

Clinical exam may reveal a left ventricular thrill at the apex, systolic thrill at the right base or suprasternal notch, ejection click, or III-IV/VVI systolic murmur at sternal border with radiation to carotids. AS can be treated by percutaneous balloon valvuloplasty. Surgical intervention with valvulotomy or valve replacement is indicated for symptomatic patients with high pressure gradients across the narrowed valve. There is no activity restriction in mild AS, but no competitive sports are allowed for moderate to severe AS. Lifelong anticoagulation therapy is required if a prosthetic valve replacement is performed.

Postoperative complications may include management of hypertrophic non compliant LV, significant residual stenosis and or regurgitation, coronary ischemia.

Coarctation of aorta
Coarctation of the aorta accounts for 6-8% of CHD. It is a narrowing of the aorta that may occur anywhere along its length, but 98% of cases occur distal to the left subclavian artery, where the PDA inserts into the descending aorta. Varying degrees of aortic arch hypoplasia may coexist with coarctaion of aorta, males are more commonly affected than females. Children with Turner syndrome are at increased risk compared to the general population. Other associated anomalies include a bicuspid aortic valve (85%) or an aberrant origin of the right subclavian artery (1%).

Clinical presentation depends on the severity of constriction and associated cardiac lesions. The classic clinical sign is a higher blood pressure and bounding pulses in the upper extremity, especially the right as most defects are distal to the right subclavian artery, compared to decreased blood pressure and diminished pulses in the legs. At birth, during the first new born detailed examination all upper and lower extremity pulses must be examined carefully. In the few defects occurring proximal to the right subclavian artery, the BP and pulses of the right arm
may be equal to the legs. It is useful to measure blood pressure in both arms and at least one leg to detect blood pressure differential. Older children may develop shortness of breath with exertion, leg pain with exercise, and chest pain with exercise. Obstruction of outflow from the left ventricle leads to left ventricular hypertrophy. In newborns, the ductus arteriosus usually allows for adequate lower body perfusion until it closes at its normal time. Severe obstruction leads to hypoperfusion, acidosis, heart failure, and shock. In severe cases, a ductus arteriosus patency should be maintained with a prostaglandin E1 infusion. A severe coarctation in association with a VSD causes increased left-to-right shunting across the VSD, leading to CHF within the first few months of life. In that case coarctation repair is recommended first followed by VSD repair. On examination, cardiac auscultation reveals a systolic murmur at the left sternal border, and especially on the back between the scapulae. Chest radiography may demonstrate cardiomegaly due to left ventricular hypertrophy, inferior rib notching due to erosion by collateral arterial circulation to bypass the obstruction, and a “reverse 3 sign” indicating the indentation of the aorta. The echocardiogram demonstrates narrowing of the distal aortic arch. The MRI produces a clearer picture than the echocardiogram of the anatomy of the coarctation. An angiogram is sometimes necessary to clarify the presence of associated cardiac lesions. Urgent surgical repair is performed in infants with circulatory shock, cardiomegaly, blood pressure extremes or severe CHF. Otherwise surgical repair is usually performed between the ages of one to two years. Post-operative complications may include, rebound systemic hypertension, due to catecholamine release and renin or residual coarctation due to narrow transverse arch. Rebound systemic hypertension usually resolves in first 24 hours. Persistent CHF may be seen if there is associated AS, mitral valve or left ventricular disease. Syndrome of mesenteric arteritis can occur. This is usually caused by reflex spasm of mesenteric arteries that are suddenly exposed to higher pressures after the coarctation is removed. The spasm can be severe enough to result in bowel ischemia. These patients are at risk for necrotizing enterocolitis. Typically a fever, leukocytosis, abdominal distension, vomiting or blood in stool 4-8 days post coarctation repair should make one suspect gut ischemia. Spinal cord ischemia due to anterior spinal artery syndrome (related to poor perfusion during clamping of aorta) is another known complication. Other complications related to thoracotomy and surgical field include injury to recurrent laryngeal nerve and phrenic nerve. Some common issues in preoperative care: Special health maintenance is indicated in patients with acyanotic CHD. Growth impairment is directly proportional to the severity of hemodynamic disturbance. Patient with acyanotic CHD tend to have more weight than height growth delay (versus both weight and height delay in cyanotic CHD). Contributing factors are caloric deprivation and reduced adipose stores, lower birth weight, increased caloric requirements, coexisting musculoskeletal, neurologic, renal, or gastrointestinal malformations, mild steatorrhea, and excess protein loss. Up 10% of children with CHD may have genetic syndromes. Poor nutrition results from anorexia, fatigability, vomiting, fluid restriction, and frequent respiratory infections. Cardiac drugs (i.e., diuretics) may exacerbate anorexia and cause early satiety. Systemic and respiratory illnesses can increase the body temperature and raise the metabolic rate by up to 13% for each degree centigrade above normal. Hypertrophic cardiac muscle can account for up to 30% of total oxygen consumption compared to the usual 10%. The caloric intake for catch-up growth is estimated at 140 to 200 calories per kg per day. In infants unable to gain sufficient weight with breastfeeding, supplementation can be achieved with a higher caloric density formula or tube feedings. In most patients, catch-up growth is largely complete within six to 12 months of surgery. As per American Heart Association (AHA) [2007], guideline on infective endocarditis prophylaxis includes: 1) prosthetic cardiac valve or prosthetic material used for valve repair, 2) previous infective endocarditis, 3) unrepaired cyanotic CHD (including palliative shunts and conduits),
4) completely repaired CHD with prosthetic material or device, whether placed by surgery or by catheter intervention, during the first six months after the procedure as endothelialization of prosthetic material usually occurs during that time period,
5) repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibits endothelialization) and
6) cardiac transplant recipients who develop cardiac valvulopathy.

Antibiotic prophylaxis is no longer recommended for any other form of CHD other than those listed above (11-12).

The CDC (Centers for Disease Control) recommends that the routine immunization schedule should be followed with some exceptions: 1) Varicella and MMR (measles, mumps, and rubella) vaccines are indicated at 12 months of age rather than at 15 months; 2) Polyvalent pneumococcal vaccine (the pneumococcal vaccine usually used in adults) is recommended at two years of age (this is in addition to pneumococcal conjugate vaccine given at 2, 4, 6, and 12 to 15 months); and 3) Influenza vaccine should be given yearly beginning at age six months in this higher-risk population (13).

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References