

## **Anesthesia for the Patient with Congenital Heart Disease Undergoing Non-cardiac Surgery**

### **Introduction**

The presence of congenital heart disease (CHD) increases the morbidity and mortality of children undergoing non-cardiac surgery. A review of 191,261 inpatient anesthetics administered to children revealed an increase in both short-term and 30-day mortality rates for patients with CHD; and in those patients with major cardiac anomalies, the observed 30-day mortality rate was nearly twice that of patients with minor cardiac anomalies. A 10-year study of patients with CHD undergoing inpatient and outpatient general surgery, showed mortality to be increased in patients with age less than 6 months, emergency status, complex cardiac lesions, and those undergoing major surgical procedures. Flick evaluated the incidence of perioperative cardiac arrest in over 92,000 pediatric patients between 1988 and 2005 and found 87.5% of patients with perioperative cardiac arrests had congenital heart disease. Although physiologically well-compensated patients may undergo non-cardiac surgery with minimal risk, certain patient groups have been identified as high risk: children less than 1 year of age, especially premature infants; patients with severe cyanosis, poorly compensated CHF or pulmonary hypertension; patients for emergency surgery and patients with multiple coexisting diseases.

### **Four Physiologic Categories, Two Major Surgical Categories**

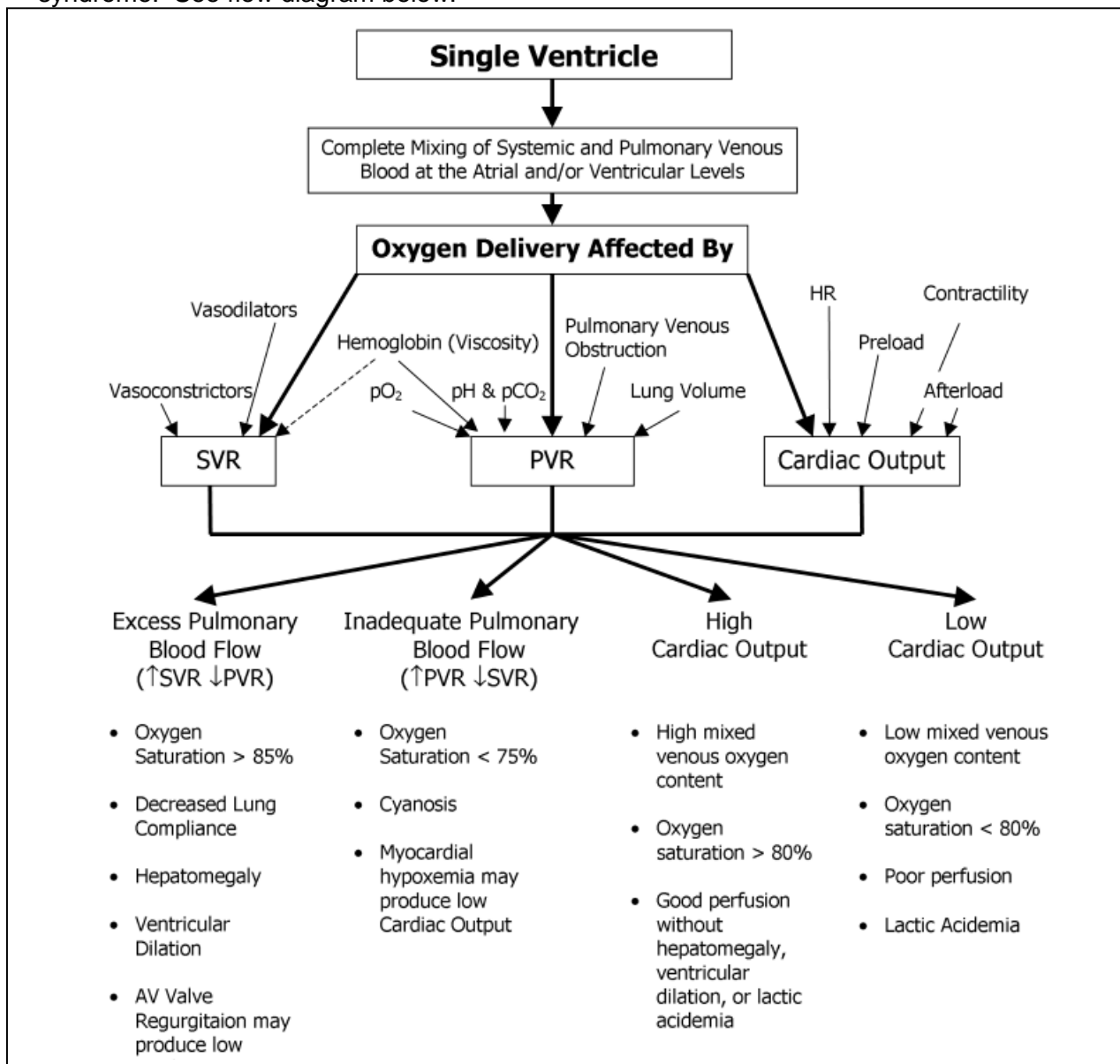
Shunts, Mixing Lesions, Single Ventricle Lesions, Obstructive Lesions, Regurgitant Lesions. Cardiac surgery has two pathways: biventricular repair and univentricular repair. Patients that undergo a biventricular repair include those with septal defects (ASD, VSD, and AV Canal), outflow tract obstruction (Pulm or aortic stenosis), and abnormalities of the great arteries (TGA, PDA, Truncus arteriosus, interrupted aortic arch and coarctation). Biventricular repair patients rarely undergo palliation, more commonly having a complete repair. In contrast, patients in the univentricular pathway typically undergo a three stage palliation (BT shunt or PA band in infancy, Glenn or Hemifontan in the first year of life, finally a Fontan procedure).

**L to R Shunts** are the most common lesions representing over 50% of children with congenital heart disease. Examples include ASD, VSD, PDA, and AV Canal. Children with L to R shunts present with signs and symptoms of congestive heart failure. For every aliquot of blood that is pumped into the systemic circulation, a portion of that blood is pumped back through the pulmonary circulation. Most children do not manifest heart failure unless their shunt produces a pulmonary to systemic fraction of three to one or greater,  $Q_p:Q_s$  of 3:1. In the situation of a child with a large VSD, for every four red cells in the left ventricle, three of the four will be shunted into the RV and one of the four will be ejected to the systemic circulation. Therefore, the child's heart must generate four times a normal cardiac output to maintain systemic blood flow. The physical signs and symptoms manifest by children are the same as those manifest by adults: tachypnea and dyspnea from left sided failure, and JVD and hepatomegaly from right sided failure; and poor exercise tolerance from decreased cardiac output. However, in order to evaluate exercise tolerance in an infant, one should enquire about feeding. Infants with significant CHF do not tolerate oral feeding, and also will manifest growth failure. Lastly, the same medications are used to treat CHF from infancy through adulthood: diuretics and ACE inhibitors.

**R to L Shunts** include TOF and Eisenmenger Complex. These patients will not manifest CHF; they are cyanotic. It is important to know baseline oxygen saturation, and also to know if the TOF patient has a history of hypercyanotic episodes "tet spell". Determine the

frequency and the severity of such episodes, and how they have been treated, so that you may anticipate and be prepared to treat a “tet spell” should it develop.

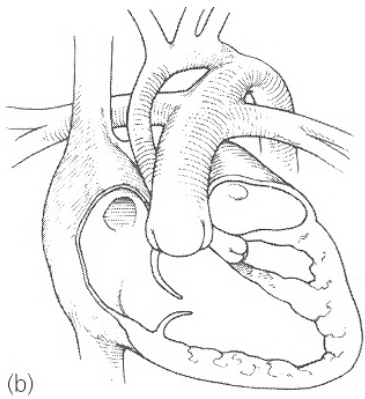
**Mixing Lesions** include patients with unrepaired TGA and truncus arteriosus and all patients in a single ventricle pathway such as tricuspid atresia, and HLHS. These lesions typically have more complex anatomy, rather than a simple hole between two chambers. Patients either have a single ventricle, or have 2 ventricles with near complete mixing of their systemic and pulmonary venous return. They typically will manifest cyanosis from mixing, but the degree of cyanosis is dependent on the pulmonary to systemic blood flow ratios. Most of these patients will be repaired or palliated before they undergo noncardiac surgery. In the palliated or unrepaired single ventricle patient, higher oxygen saturation may actually be associated with reduced oxygen delivery. Remember that oxygen is a pulmonary vasodilator and excessive pulmonary blood flow may actually “steal” flow from the systemic circulation; this situation is especially more likely in the patient with unrepaired hypoplastic left heart syndrome. See flow diagram below.



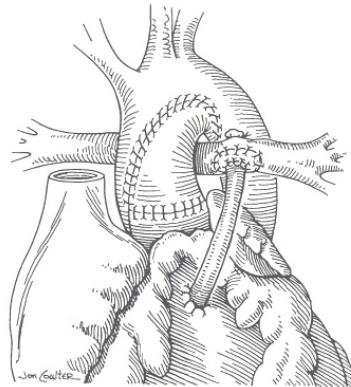
Almost all single ventricle patients undergo staged palliation, requiring a pulmonary artery band or Blalock-Taussig shunt as a newborn, followed by bi-directional cavopulmonary anastomosis (bidirectional Glenn or Hemi-Fontan), followed by the Fontan procedure. See diagram below. If these patients require elective surgery, such procedures are best performed during the second stage of palliation (bi-directional Glenn or Hemi-Fontan). After surgical palliation in the newborn period, these patients continue with a parallel pulmonary and systemic circulations producing a volume burden on their single ventricle. In the final stage (Fontan), all blood returning to the heart first passes through the pulmonary bed. Therefore, the conversion from spontaneous to positive pressure ventilation sometimes will produce significant reductions in cardiac output. Cardiologists do not always recognize this situation and may need the advice of the anesthesia team on appropriate referral of these patients for correction of other associated congenital abnormalities.

### HLHS

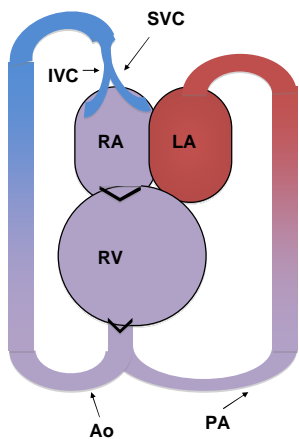
#### Unrepaired



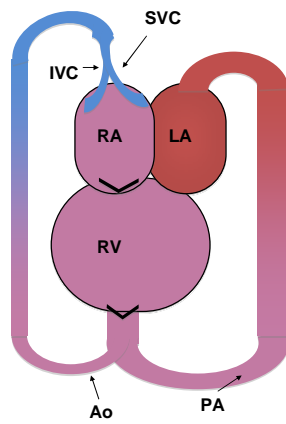
#### Norwood (Sano Mod)



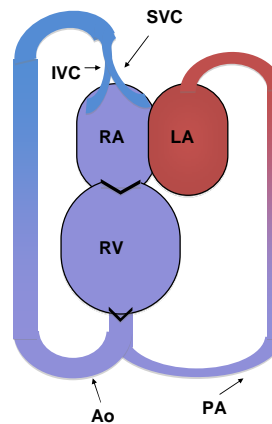
#### Balanced PBF



#### Excess PBF



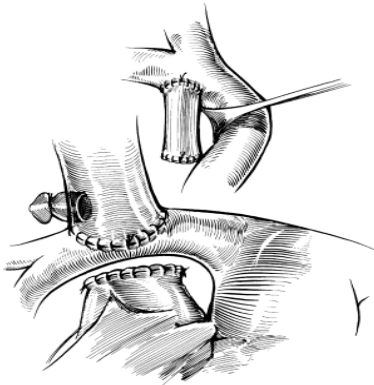
#### Inadequate PBF



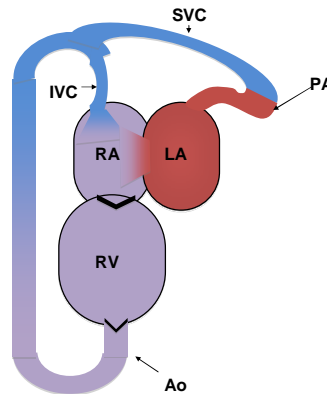
Patients with unrepaired HLHS or after the Norwood procedure have a similar physiology with one outflow from the right ventricle allowing blood to pass to the systemic or pulmonary circulations. Oxygenated blood returns to the left atrium, and mixes with deoxygenated blood in the right atrium. The classic Norwood and Sano modification limit pulmonary blood flow by providing fixed resistor for blood to enter the pulmonary circuit. The schematic shows the parallel circuit, and one can see that more blood entering the pulmonary circuit will increase the oxygen saturation, yet produce a greater volume burden on the single ventricle and may result in inadequate oxygen delivery. Inadequate PBF will produce worsening

cyanosis. The cavopulmonary shunt removes the volume burden by placing the pulmonary and systemic circulations into series circuit.

### Modified Glenn

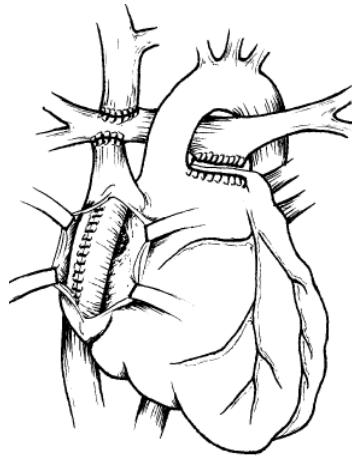


### Schematic

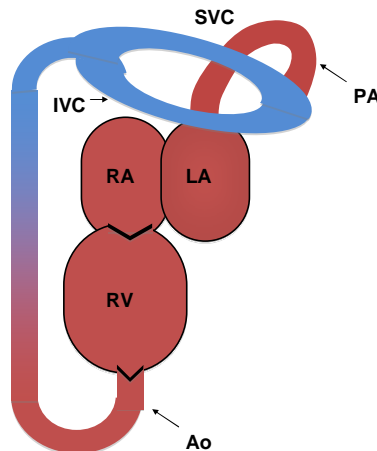


The Fontan procedures connects the IVC and SVC directly to the pulmonary arteries and the higher systemic venous pressures push blood through the pulmonary circuit. In this situation, spontaneous ventilation augments pulmonary blood flow.

### Lateral Tunnel Fontan



### Schematic



**Obstructive Lesions** include aortic stenosis, pulmonary stenosis, and coarctation of the aorta. The principles of optimal hemodynamic management are the same in children and in adults. There is limited flow across a stenotic outflow; thus increasing the preload, avoiding tachycardia, and maintaining contractility would optimize the cardiac output. A newborn may manifest critical aortic stenosis, pulmonary stenosis or coarctation with low cardiac output and requiring emergency intervention. Outside of the newborn period, the myocardium generally will compensate by developing hypertrophy without dilation when compromised by a pressure burden. Therefore, most patients are asymptomatic until the obstruction becomes very severe. However, decreased coronary blood flow from hypotension will produce endocardial ischemia in the hypertrophied ventricle and therefore it is critical to maintain arterial blood pressure in these patients.

**Regurgitant Lesions** are most commonly seen with a cleft mitral valve found in AV canal. Again, the same hemodynamic principles are used to manage both adults and children, avoid bradycardia that will increase the valvular regurgitation, reduce afterload to improve forward flow, and avoid excess myocardial depression. As opposed to patients with a pressure load on the ventricle, a regurgitant load will lead to hypertrophy and dilation that will produce symptoms of CHF as valvular regurgitation worsens.

## **Preoperative Assessment**

A complete history and physical examination, focusing on cardiac signs and symptoms, previous surgical and catheterization procedures is essential. Any and all cardiac imaging studies should be reviewed during the preoperative assessment. Echocardiography is non-invasive and the mainstay of diagnostic testing, and for many patients, it is the only cardiac imaging study performed. Today, cardiac catheterization is most often performed for interventional procedures, but valuable diagnostic data is also available from this modality. Cardiac MRI has assumed an increasingly important role to assess anatomy, function, and progression of pathophysiology. Abnormalities of cardiac rhythm are more common among older patients with residual defects and single ventricle patients. If there is any history of cardiac rhythm abnormality a recent ECG or Holter examination should be reviewed.

In general, all cardiac medications should be continued through the perioperative period. ACE inhibitors are commonly administered to single ventricle patients, or those with significant congestive heart failure (CHF) or mitral regurgitation; beta blockers for Tetralogy of Fallot patients or those with atrial arrhythmias; amiodarone for patients with significant atrial or ventricular arrhythmias; and diuretics for patients with CHF. Digoxin is rarely used in the modern era. Endothelin antagonists, phosphodiesterase-5 inhibitors, or prostaglandin analogs are used to treat pulmonary hypertension. Many patients with CHD will receive aspirin or other antiplatelet therapies to decrease risk of thrombus formation in shunts or conduits. Low dose ASA is not a contraindication for most simple, superficial surgeries, but if the surgery is major, a discussion with the patient's cardiologist and surgeon should take place and ASA is typically discontinued 7-10 days before surgery. Finally, some patients, like those with mechanical cardiac valves, will be on Coumadin therapy. These patients require careful planning and are frequently admitted before surgery while Coumadin is discontinued and heparin therapy initiated.

Standard NPO timing is used with the CHD population. It is critically important that patients with cyanotic lesions, shunt-dependent patients, and those with outflow tract obstruction do not remain NPO for long periods of time. Hypovolemia can be a critical problem for these patients, especially with induction of anesthesia and institution of positive pressure ventilation. These patients should be scheduled early in the day, and if there are delays should be fed clear liquids until 2 hours before induction.

A frequent question is whether the patient needs to be evaluated by their cardiologist prior to the procedure. In general, if the patient has a simple or moderately complex lesion that has been completely corrected and is well compensated, a standard preanesthetic visit without a cardiology consultation is appropriate. A patient with a moderately complex lesion who is not well compensated, any cyanotic or single ventricle patient or patient with a complex lesion needs a recent cardiology evaluation that usually includes an echocardiogram within 12 months of surgery. If the patient's condition has changed significantly since the last evaluation, the cardiologist should reevaluate them. Because cardiologists have limited knowledge of anesthetic effects and general surgical procedures, they should NOT be asked to clear the patient for surgery; but rather to determine if the patient's cardiac status is optimized. After consultation with the cardiologist, the anesthesiologist will clear the patient for surgery.

Many patients, particularly adults with CHD, will have implanted pacemakers and/or automated defibrillators. It is critical to understand the patient's underlying cardiac rhythm, the reason for placement of the device, and the current modes and settings of the device.

Recently published guidelines from the ACC/AHA advocate preoperative and postoperative interrogation of permanent pacemakers whenever possible. Previous recommendations to use a magnet to convert a pacemaker to asynchronous mode during surgery are no longer universally valid as most modern pacemakers are programmable and may be unpredictably affected by the placement of a magnet over the pacemaker. Unipolar devices should be programmed to an asynchronous mode, and special algorithms such as rate-adaptive functions should be suspended prior to surgery. Implanted cardioverter-defibrillators should have their anti-tachycardia and arrhythmia therapies disabled preoperatively and external methods of cardioversion or defibrillation should be available. Any necessary pacing function provided by the ICD should be re-programmed for the duration of the procedure. After surgery, pacemakers and implantable cardioverter-defibrillators should be reinterrogated and re-enabled.

### **Evaluation and care of the patient with a pacemaker or implantable cardioverter-defibrillator**

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- History
  - Indication for placement of device
  - Type of device
  - Pacing mode
  - Date device placed
  - Date of last evaluation
  
- Physical Examination/Laboratory
  - Evaluate underlying rate, rhythm and hemodynamic stability: Is patient device-dependent?
  - Review recent ECG
  - Determine anatomic position of generator
  - Examination of leads on CXR
  
- Interrogation of device by a trained individual
  - Evaluate lead integrity
  - Obtain current programming information
  - Determine frequency of initiated therapies with ICDs
  - Consult pacemaker representative if questions arise
  - Reprogram device if necessary prior to surgery
  - Disable antitachycardia therapies on ICDs preoperatively
  - Disable rate-responsive modes preoperatively
  
- Intraoperative management
  - Ensure electrical activity is converted to mechanical systole
  - Have temporary pacing support available: transvenous vs transthoracic
  - Ensure availability of trained personnel
  - Electrocautery to be delivered in short bursts
    - Consider use of bipolar electrocautery or ultrasonic scalpel
  - Assure defibrillator capability
  - Evaluate effects of anesthetic techniques on device function
  
- Postoperative interrogation and reprogramming of device

### **Stages of Repair**

Patients may present for noncardiac surgery before they have had cardiac surgical repair of their congenital heart defect, after they have had a palliative procedure, or after complete repair. There are two types of palliation - procedures used to augment or decrease pulmonary blood flow. Patients with left to right shunts and excess pulmonary blood flow, may have a pulmonary artery band placed to limit blood flow to the lungs, thus decreasing the left to right shunt. Patients with insufficient pulmonary blood flow will have a systemic to pulmonary shunt placed to augment their pulmonary blood flow, like a Blalock-Taussig shunt. Remember that a child who has a "repaired" heart may not have normal cardiac function. They may have residual shunts, poor myocardial function, and may be more susceptible to arrhythmias.

### **Conduct of Anesthetic**

A well equipped and staffed community hospital can readily care for the well compensated patient with a simple or moderately complex lesion. However, poorly compensated patients with complex or single ventricle physiology, should have their surgery in centers with expertise in CHD, where backup support is available should complications arise. In general, more complex patients can undergo outpatient surgery, but admission to the hospital must be readily available. For complex patients having major surgery, postoperative care in an ICU is essential.

Standard ASA monitors including are essential for all procedures, including diagnostic imaging procedures, where end-tidal CO<sub>2</sub> can be monitored via a nasal cannula for sedated patients. The decision to institute more invasive monitoring depends on the assessment for potential hemodynamic and respiratory instability due to the patient's pathophysiology and the effects of the planned surgery. Patients with significant baseline myocardial dysfunction, pulmonary hypertension, or cyanosis often benefit from preoperative intravenous access if possible.

With careful attention to pathophysiology and the desired hemodynamic goals, any anesthetic and sedation technique may be used. Premedication is well tolerated. Inhalation induction with sevoflurane is appropriate and well tolerated for most CHD patients. Propofol may be used for induction and maintenance, with careful attention to venous and arterial vasodilation produced by this agent. Ketamine, either IM or IV, is a very useful agent, preserving myocardial function and providing sedation and analgesia. Etomidate has little effect on myocardial contractility and hemodynamics, and is an excellent agent for the patient with impaired myocardial function. Dexmedetomidine is increasingly used for sedation in patients with CHD, and is usually well tolerated, however, bradycardia, and hypertension are commonly observed. Any opioid can be used and single shot caudal and nerve block techniques are also useful, even for patients receiving low dose aspirin. Major neuraxial techniques, i.e. lumbar/thoracic epidural and spinal are best avoided with patients receiving aspirin.

Airway management may range from sedation with spontaneous respiration, to mask or LMA general anesthesia, to endotracheal anesthesia, again being mindful of the effects of hyper- or hypocarbia, and positive pressure ventilation for the individual patient. With endotracheal anesthesia, the decision to extubate at the end of the procedure of course must take into account the severity of the underlying pathophysiology, and the magnitude of the surgical procedure. In shunted, single ventricle infants undergoing major abdominal procedures, for example, it is very often prudent to ventilate the patient in the early postoperative period.

### **Infective Endocarditis Prophylaxis**

The American Heart Association made significant changes to the guidelines for prevention of infective endocarditis (IE) in 2007, resulting in a narrowing of the indications for administering IE prophylaxis. This has resulted in confusion for patients, parents, surgeons, and even cardiologists. It is important to understand the new indications, which were based on an extensive review of the data and discussions among a large panel of experts. Patient must have BOTH a cardiac indication, and a surgical/procedural indication. The major cardiac indications are: 1. prosthetic cardiac valve or material; 2. previous IE; 3. congenital heart disease, but ONLY a) unrepaired or palliated cyanotic CHD; b) completely repaired CHD with prosthetic material or device during the first 6 months after the procedure; c) repaired CHD with residual defects at or near the site of a prosthetic patch or device; 4. cardiac transplant recipients who develop valvulopathy. The surgical/procedural indications are 1. all dental procedures that involve manipulation of the gingival tissue or perforation of the oral mucosa; 2. respiratory tract procedures or procedures on infected skin, or musculoskeletal tissue. IE prophylaxis is not recommended for simple gastrointestinal or genitourinary procedures where the mucosa is not incised, i.e. simple endoscopy and cystoureteroscopy, but is recommended for surgery where mucosa is incised. For dental prophylaxis, a single dose of ampicillin 30-60 minutes before the procedure, or as soon as IV access is obtained, is the recommended regimen, with clindamycin, cefozolin, or ceftriaxone acceptable for penicillin allergic patients.

### **Kids Who Scare Me**

Finally, I would like to review a few congenital heart lesions that are particularly high risk.

**Shunt dependent single ventricle patients** have a volume burden on their single ventricle from the parallel pulmonary and systemic circulations, they have reduced oxygen carrying capacity because normal oxygen saturation is 75 – 85%, and they have reduced diastolic pressure from the systemic to pulmonary shunt. This physiologic combination leaves them particularly vulnerable to circulatory insults. In fact, about 10% of patients with hypoplastic left heart syndrome who survive the Norwood procedure will die prior to their second stage of palliation. These patients commonly have feeding difficulties and a number of them will undergo gastrostomy tube placement, with or without fundoplication. A report from Stanford described good outcomes among 12 patients with HLHS (9 after stage 1 reconstruction, 3 after BDG undergoing laparoscopic surgery. At our institution these patients are admitted to the intensive care unit for postoperative care.

**Single ventricle patients with AV valve regurgitation.** Trivial to mild AV valve regurgitation is observed in essentially all single ventricle patients. However, patients with moderate to severe AV valve regurgitation commonly have a dilated valve annulus and reduced cardiac output. Even though these patients are most stable after their second stage of palliation, those with moderate or greater AV valve regurgitation have very limited cardiac reserve.

**Pulmonary hypertension with systemic or supra-systemic PA pressures** is a known risk factor for morbidity and mortality. Recent publications from the Children's Hospital in Denver report a significantly higher incidence of intraop cardiac arrest and death among patients with supra-systemic PA pressures. Pulmonary hypertensive crisis is defined by a sudden increase in PVR that results in pulmonary pressure exceeding mean arterial pressure. This will compromise cardiac output from RV failure and lower arterial pressure, producing arterial and/or venous oxygen de-saturation. Immediate treatment is indicated - increase FIO<sub>2</sub>, reduce CO<sub>2</sub>, increase pH and then institute inhaled nitric oxide.



**Severe aortic stenosis (gradient greater than 60 mmHg).** These children will be totally asymptomatic since a pressure burden on the ventricle will cause the ventricle to hypertrophy, but not dilate. However the hypertrophied ventricle is at risk for developing ischemia if the patient develops hypotension.

Up to two-thirds of patients with **Williams syndrome** develop supraaortic stenosis. The outflow tract obstruction frequently arises as a concentric narrowing of the ascending aorta at the superior margin of the sinuses of Valsalva, creating a typical hourglass deformity of the aorta. These patients may also have diffuse narrowing along the entire length of the aorta and their arteriopathy may also include the coronary arteries. Obstruction of the left coronary artery, reduction in the size of the ostia and diffuse coronary artery narrowing have all been described.

The annual mortality among patients with **hypertrophic cardiomyopathy** is 3-4% overall and up to 6% in children. Like aortic stenosis, these children often have few symptoms, but are also at risk for worsening LV obstruction with hypovolemia, tachycardia and increased contractility. Some patients, like those with Noonan's syndrome may manifest biventricular obstruction.

In about 50% of patients with **pulmonary atresia and intact ventricular septum** there are endothelial-lined blind channels within the RV myocardium known as sinusoids. These sinusoids are in direct communication with the RV cavity and can form coronary artery to RV fistulae. These fistulae send desaturated RV blood to the myocardium of the RV and in 20% of patients the myocardium is chronically ischemic having no antegrade flow into the right coronary.

The incidence of **coronary artery vasculopathy** is about 17% at 10 years **after heart transplantation** and because the heart is denervated, it is not associated with symptoms.

**Eisenmenger's syndrome** is fortunately rarely seen in this era.

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