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Anesthesia for noncardiac surgery in children with congenital heart disease

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A 4.5 year old, 12 kg female with pulmonary and tricuspid atresia (functionally a single ventricle), transposition of the great arteries, Down syndrome, and radiologic evidence of cervical spine hypermobility needs extensive dental restorative surgery under general anesthesia. She has a functioning right Blalock Taussig shunt, performed at five days of age. Her hematocrit is 55% and the child is visibly cyanotic. She has a history of congestive heart failure for which she takes digoxin.

QUESTIONS FOR CONSIDERATION

1. What are the key issues in this patient's perioperative management?
2. What are the important pathophysiologic effects of her congenital heart disease? How will they affect your anesthetic plan?
3. Should you do anything about the hematocrit of 55%? If so, what?
4. Draw a diagram of this child's heart. How will changes in heart rate, blood pressure, myocardial contractility, or systemic vascular resistance affect this child's hemodynamic status? Prepare a cardiac grid to demonstrate how to optimize this child's circulation during the perioperative period.
5. How will you anesthetize this child?

GENERAL DISCUSSION

The anesthesiologist providing care to a child with congenital heart disease (CHD) must be fully aware of the child's specific intracardiac and extracardiac defects, the effects of hemodynamic changes on those defects, and the cardiovascular effects of the anesthetic agents to be administered. The major pathophysiologic effects of CHD are: shunting, hypoxemia, heart failure, dysrhythmias, pulmonary hypertension, and outflow track obstruction.

Shunting: The excessive pulmonary blood flow associated with shunting may cause congestive heart failure or produce impaired pulmonary function, small airway obstruction, left mainstem bronchial obstruction, increased interstitial/alveolar lung water, and vascular obstructive disease. Shunt reversal or hypercyanotic episodes may occur in children with intracardiac shunts. Shunt reversal can occur with any communication between the atriae or ventricles while hypercyanotic episodes are most commonly associated with Tetralogy of Fallot. Shunt reversal occurs when systemic vascular resistance (SVR) decreases and pulmonary vascular resistance (PVR) increases due to anesthetic effects of increased airway pressures. Hypercyanotic episodes during anesthesia may result from surgical stimulation, dynamic right ventricular outflow track obstruction, decreased pulmonary blood flow associated with hypovolemia, increased airway pressures, or decreased SVR.i. Therapy includes increasing intravascular volume, oxygen, increasing SVR with alpha agonists such as phenylephrine, and decreasing outflow track obstruction with beta blockade (Table I).

Hypoxemia: In response to chronic hypoxemia, the body attempts to normalize oxygen transport by: 1) Polycythemia; 2) Increased blood volume; 3) Neovascularization; 4) Alveolar hyperventilation. The consequences of these adaptive mechanisms include decreased cardiopulmonary reserve, increased systemic vascular resistance from increased blood viscosity, cerebral or renal thrombosis especially with dehydration, and coagulopathy. Symptoms of hyperviscosity (headache, fatigue, paresthesias, dizziness, depressed mental state) are most common with hematocrits > 65%. However, isovolumic hemodilution should not be performed on the basis of the hematocrit, but rather to relieve symptoms. Plebotomy of iron-deficient patients with excessive hematocrits should be avoided. The major risks of polycythemia are pulmonary and cerebral thrombosis. Chronic hypoxemia impairs global ventricular function, particularly in the immature myocardium. In the hypoxic child, systemic blood

flow is redistributed to the brain and heart with decreased perfusions of splanchnic organs, skin, muscle, and bone.

Pulmonary Hypertension: Transient or permanent increases in pulmonary vascular resistance (PVR) complicate the perioperative care of many infants with CHD such as endocardial cushion defects, patent ductus arteriosus, ventricular septal defects, and aortic outflow anomalies, among others. An increase in endothelin (endothelial constricting factor) is present in children with pulmonary hypertension secondary to CHD. The increase in endothelin may result from hemodynamic shear stresses resulting from high pressure in the pulmonary artery. Increased beta receptor density in the lung is also present as is an increase in Factor VIII von Willebrand factor.

Table I. Factors affecting PVR

Increase PVR	Decrease PVR
Hypoxia	Oxygen
Hypercarbia	Hypocarbia
Acidosis	Alkalosis
Hyperinflation	Normal function residual capacity
↑ Hematocrit	↓ Hematocrit
Sympathetic stimulation	Sympathetic block

Hyperventilation is the most reliable way to decrease PVR but it is pH, not pCO_2 that controls pulmonary vasoconstriction. Prostacyclin and nitric oxide will also decrease PVR in children with CHD. Nitrous oxide does not increase PVR in most children with CHD; however, dramatic increases may occur in a few children.

Perioperative management: The approach to any congenital heart lesion involves knowledge of three aspects:

- A written description of the cardiac lesions
- Diagram of the cardiac lesions
- Hemodynamic grid of changes affecting cardiac performance

This information can easily be obtained from the echocardiogram or cardiac catheterization reports. With this information plus a knowledge of the cardiovascular effects of the proposed anesthetic drugs, a detailed perioperative plan can be made.

The medical history should include questions about cyanosis, fatigability, frequency of infections, particularly respiratory and whether the patient can keep up with his/her peers. The need for pharmacologic intervention to maintain cardiovascular performance should be noted. On physical examination, the stage of growth and development in relation to age should be noted. The absence of peripheral pulses may indicate prior performance of a subclavian flap angioplasty for coarctation, use of the subclavian artery for a Blalock Taussig shunt or prior cannulation with thorombosis. Symptoms of heart failure include wheezing, respiratory distress, hepatosplenomegaly, regarded growth and development, diaphoresis, cardiomegaly, and decreased perfusion. The child should also be carefully evaluated for other congenital anomalies such as Down syndrome, Treacher Collins, Pierre Robin, etc.

Morray and coworkers demonstrated increased anesthetic risk in children with Down syndrome undergoing cardiac surgery. Children with Down syndrome often have airway problems such as a small mandible, small larynx, large tongue, high arched palate, and may require a smaller tracheal tube owing to congenital subglottic stenosis. The need for careful positioning, handling, and stabilization of the neck is essential if spinal cord involvement is associated with the cervical spine hypermobility. Inquiries about gait changes, clumsiness, spasticity should be made and the patient evaluated for hyperreflexia and Babinskis that would suggest cord involvement. Other problems relevant to the anesthesiologist in patients with Down syndrome are retardation, increased gastroesophageal reflux and decreased central/peripheral nervous system activity.

Laboratory evaluations should include a chest X-ray, electrocardiogram (ECG) hematocrit, electrolytes, echocardiogram (if not performed recently), arterial blood gases (if not available from prior catheterization or other evaluation). The chest X-ray should be inspected for heart size, atelectasis, acute respiratory infection, and elevated hemidiaphragms. The ECG should be evaluated for rate, rhythm, ventricular strain (ST-T changes), and hypertrophy. Echocardiography is usually adequate for evaluation of most CHD.

Risk is often difficult to assess in these patients but probably depends upon the severity of the cardiac lesion, age, type of correction if any (palliative *vs* definitive), presence of other defects, and the status of the pulmonary vasculature. In 1990 Strafford and colleagues at the Boston Children's Hospital surveyed 110 patients with CHD (41 corrected, 50 uncorrected and 19 palliated) for adverse events during noncardiac surgery requiring 135 anesthetics. Several of the patients had more than one cardiac diagnosis and more than one adverse event during anesthesia. There were 71 adverse events in 52 patients (47% of patients had an adverse event). Adverse events included airway problems (22), bronchospasm (4), dysrhythmias (17), circulatory instability (16) with 9/16 patients needing inotropic support and other lesser problems. Factors associated with adverse events included uncompensated congestive heart failure, cyanosis with an $SpO_2 < 85\%$ and uncorrected CHD.

Preoperative preparation of the pediatric patient concludes with a concise, yet detailed discussion of the cardiac conditions and anticipated perioperative plan of action with the child and parents. Adequate perioperative hydration is essential in children with cyanotic CHD so preoperative fasting should be minimized (clear liquids up to two hours before surgery in infants 0-12 months; clear liquids up to four hours preop in children 12 months or older) or an intravenous infusion started. Prophylactic antibiotics should be administered according to the AHA guidelines. Premedication is usually desirable in children over six months of age with CHD since crying and generalized emotional upset worsen oxygenation and shunting in many lesions. Intramuscular premedications are rarely used and have been largely replaced by oral, nasal, or rectal routes of administration. The table II provides premedication options:

Table II. Premedications

Oral	Parenteral
Midazolam 0.5-0.75 mg/kg OR	Morphine 0.1-0.2 mg/kg plus
Diazepam 0.15 mg/kg plus	Pentobarbital 2 mg/kg plus
Meperidine 1.5 mg/kg	Scopolamine 0.01 mg/kg
Nasal	Rectal
Midazolam 0.2-0.3 mg/kg	Methohexital 25 mg/kg

The arrival of a quiet, sleeping child in the operating suite allows a choice of anesthetic induction techniques, ranging from inhalation of sevoflurane or halothane, through intramuscular ketamine, to placement of an intravenous catheter and use of narcotics, propofol, or barbiturates. Specific induction techniques depend upon the cardiac lesion and the child's general condition.

The speed of induction is influenced by the presence of an intracardiac or extracardiac shunt. Theoretically, one would expect a faster induction with a volatile agent in patients with a left-to-right shunt due to augmented pulmonary blood flow. With a left to right shunt, blood that has already picked up anesthetic recirculates through the lungs picking up even more anesthetic and causing a higher anesthetic concentration to the leave the heart. However, induction time is essentially unchanged. The distribution and onset of action of the intravenous agents is slower with a left to right shunt. With a right to left shunt, intravenous induction is rapid and can produce sudden, dramatic effects.

Inhalation induction is prolonged with a right to left shunt due to bypass of the lungs, although the prolongation is only moderate. Theoretically, blood that has left the lungs with a concentration of anesthetic has that concentration diluted by blood which has bypassed the lungs. Thus, the blood leaving the heart has a lower concentration of anesthetic and induction is slowed. Such effects are most marked for insoluble anesthetic gases such as nitrous oxide and less marked for more soluble gases. The temptation to increase the concentration too rapidly in children with right to left shunts must be carefully resisted. Another possibility is a change in shunting pattern during use of a volatile inhalation anesthetic. Finally, if a very large shunt is present, increasing the FiO_2 will not increase arterial pO_2.

Monitoring devices for noncardiac surgery in patients with CHD are chosen according to the requirements of the surgical procedure and the severity of the patient's disease. Usually only the standard American Society of Anesthesiologists' monitors such as the pulse oximeter, blood pressure cuff, and electrocardiogram are applied prior to anesthetic induction with more invasive monitors such as intra-arterial, central venous, or transesophageal echocardiographic (TEE) transducers applied following induction.

CASE SPECIFIC DISCUSSION

Important anesthetic and perioperative management principles in this child include: 1) Avoidance of air bubbles that might cross through her Blalock shunt into the systemic circulation; 2) Maintenance of systemic arterial pressure because

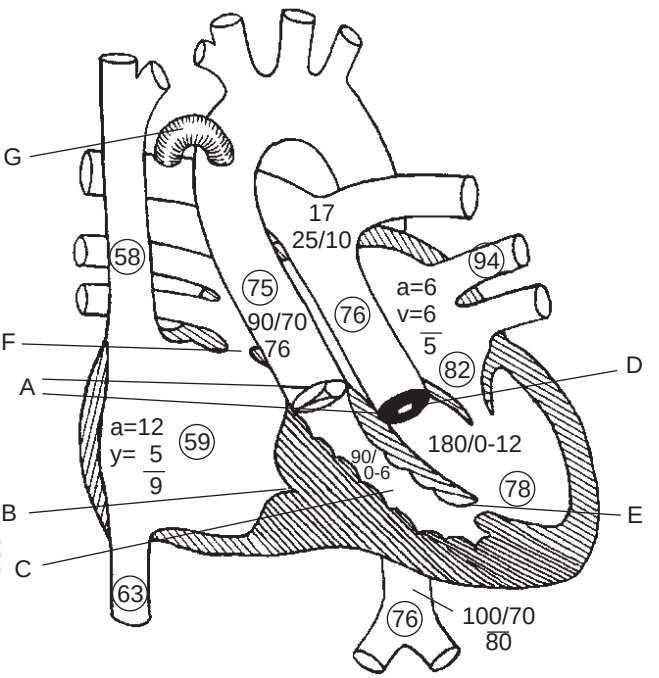

Figure 1. Cardiac diagram.

systemic perfusion will control pulmonary blood flow and oxygenation in this patient; 3) Avoidance of drugs or events that might increase PVR and worsen systemic oxygenation; 4) Avoidance of decreased myocardial contractility that might worsen systemic perfusion and heart failure. This child has undergone only a palliative procedure and continues to have both cyanosis and heart failure, major risk factors for adverse events during a noncardiac procedure. The compromise to the arterial circulation of the right arm should be recognized, necessitating measurement of blood pressure and establishment of intravenous access at alternative sites. The function of the patient's systemic to pulmonary shunts can be monitored with pulse oximetry. Capnography will detect serious alterations in pulmonary blood flow and cardiac output. However, the curvilinear negative correlation between the arterial-end tidal carbon dioxide gradient and arterials oxygen saturation in a child with CHD must be recognized. This gradient increases 2-3 mmHg for each 10% decrease in SpO_2. Although TEE would be useful in evaluating this child's intraoperative cardiac function, it is probably not feasible during a dental procedure. Nevertheless, a transthoracic echocadiographic evaluation could be performed if an adverse event occurs. While there are many satisfactory anesthetic approaches for this child, one possibility is oral midazolam premedication, inhalation induction with sevoflurane, careful nasotracheal intubation with in-line neck stabilization, anesthetic maintenance with low dose sevoflurane supplemented with narcotic and neuromuscular blocker.

Cardiac diagrams: Cardiologists often display the echocardiographic or cardiac catheterization findings using a diagram of the atria, ventricles, and great vessels that shows the septal defects, valvular stenoses, ventricular outflow obstruction, intracardiac/extracardiac shunts (Figure 1). An example of such a diagram for this patient is:

Cardiac grids: The physiologic needs of any particular patient with CHD can be analyzed using a cardiac grid, a table in which the hemodynamically desirable changes in heart rate, preload, cardiac output or myocardial contractility, and systemic and pulmonary vascular resistances are plotted against the anatomic components of any congenital lesion. Any example of a cardiac grid for a patient with a hypoplastic right heart, transposition of the great arteries, tricuspid and pulmonic atresia is shown below:

Anatomy	Preload	PVR	SVR	HR	Contractility
ASD/VSD	N	↓	↑	N	N
S→P Shunt	→	↓	↑	↑	↑
TGA	N	↓	N-↑	N	N

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