Anesthesia for non-cardiac surgery in children with congenital heart disease

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INTRODUCTION

It has been estimated that 0.8% of children born in the United States will be diagnosed to have some form of congenital heart disease. (CHD) Many of these children will require surgery for noncardiac conditions that are commonly encountered during childhood (e.g. inguinal hernia, dental caries, or chronic tonsillitis). Moreover, many children with CHD have other associated noncardiac (e.g. orthopedic or genitourinary) congenital anomalies that require surgical repair. Many of these procedures are performed in general hospitals by noncardiac anesthesiologists. It is therefore essential that the anesthesiologist be knowledgeable, not only of the basics of pediatric anesthesia, but also of the pathophysiology of the cardiac lesions.

Knowledge of the physiological consequences of shunting or obstruction is essential to understanding the hemodynamic consequences of any congenital heart deformity. Application of these physiologic concepts permits the anesthesiologist to design a rational management plan for the child with CHD.

Congestive Heart Failure

Congestive heart failure is not a disease, but a symptom complex caused by an underlying cardiac condition. The clinical diagnosis of congestive heart failure rests upon the identification of its four cardinal signs: tachycardia, tachypnea, cardiomegaly and hepatomegaly. Upper respiratory infections are common in these children. The classic radiologic findings of cardiac failure are increased cardiac size and pulmonary congestion. Echocardiography is ideal as a non invasive tool to measure cardiac chamber sizes, ejection fraction and circumferential fiber shortening rate. Exercise tolerance (including feeding tolerance in infants) is the simplest and most informative method of clinically estimating cardiac reserve in patients with congenital heart disease. Heart failure must be controlled before any elective noncardiac surgery is scheduled. Control of congestive heart failure improves pulmonary function, and also reduces the possibility of perioperative hypoxemia and respiratory failure. Digoxin remains the therapy of choice for treatment of heart failure in infants and children. It is often used in combination with diuretics such as furosemide, spironolactone or chlorothiazide. Preoperative evaluation in these patients should include digoxin level measurements and serum electrolyte determinations.

In children whose congestive heart failure is caused by lesions that increase pulmonary blood flow (e.g., ventricular septal defect), the anesthesiologist must attempt to reduce the shunt and maximize systemic perfusion. This can be accomplished by manipulating the impedance to outflow of the respective ventricles. Increasing the ratio of pulmonary vascular resistance (PVR) to systemic vascular resistance (SVR) results in less blood being shunted into the lung. Worsening cardiac failure during anesthesia can be recognized by systemic hypotension, diminished heart sounds, decreased oxygen saturation, poor skin perfusion, decreased urine output, and the development of metabolic acidosis. Therapy should start with optimizing the determinant of myocardial performance (cardiac rhythm, preload, contractility, and afterload) that is the most impaired.

Cyanosis

Any child with a cardiac lesion that causes right-to-left shunting from obstructed pulmonary blood flow, venous admixture to the systemic circulation, or both, may ultimately develop cyanosis. Detection of cyanosis depends upon hemoglobin concentration, oxygen saturation, and cutaneous perfusion. Severe chronic hypoxemia results in polycythemia, with an increase in blood volume and viscosity, neovascularization, alveolar hyperventilation, and coagulopathy. Coagulation abnormalities include thrombocytopenia, platelet dysfunction, hypofibrinogenemia, and accelerated fibrinolysis with factor deficiency. Bleeding time may be prolonged. The increased sympathetic tone due to chronic hypoxemia leads to down-regulation of beta-receptors in the myocardium, in turn leading to global ventricular dysfunction. Increased blood viscosity increases the cardiac workload by elevating the SVR. If the hematocrit is high, ($\geq$ 60%) clotting of existing shunts (e.g., Blalock-Taussig) as well as cerebral and/or renal thrombosis may occur. In children with cyanotic heart disease, hyperviscosity is associated with thrombosis of intracranial veins and sinuses. This sometimes results in a stroke. Children under 5 years of age are at increased risk, especially during periods of dehydration and fever. Because of the risk of hemoconcentration, preoperative fasting (NPO) times should be held to a minimum in cyanotic children, or the patient should be hydrated intravenously. Scheduling these patients early in the morning will not only avoid prolonged fasting, but also provides a longer postoperative observation period during recovery. Aggressive use of antiemetics (e.g. ondansetron) will allow for early fluid intake following surgery.

Anesthetic manipulations in cyanotic children should be designed to avoid further increases in right-to-left shunting, and the consequent reductions in pulmonary blood flow. Right-to-left shunting may be decreased by reducing the ratio of PVR/SVR and by relieving the degree of right heart obstruction. Reductions in PVR are accomplished by hyperventilation with a high-inspired O_2 concentration, and avoidance of high mean airway pressure. Shunting of blood into the lungs can usually be improved by elevating the SVR. Maintaining an adequate depth of anesthesia and administering alpha-adrenergic agonists may be effective. For example, infusion of phenylephrine into children with tetralogy of Fallot will increase SVR, reduce intracardiac shunting, and results in decreased cyanosis. Decreasing SVR, by contrast,

promotes shunting of blood from the lungs and can create or intensify cyanosis. For that reason, anesthetic agents such as isoflurane and propofol, vasodilators, and deep levels of anesthesia should be avoided in cyanotic children. If a child with CHD that is obstructing pulmonary blood flow becomes hypovolemic, and systemic hypotension develops, both the right-to-left shunt and cyanosis are intensified. Coronary artery perfusion may also be compromised. If untreated, this physiologic sequence can lead to shock and cardiovascular collapse. Aggressive treatment with fluid resuscitation and vasopressors is essential.

Dysrhythmias
Dysrhythmias are the result of either altered cardiac impulse generation or conduction. In patients with congenital heart disease, disorders of the conduction system, both congenital and acquired, are more common than disorders of impulse generation. The etiologies for conduction system disturbance that produce dysrhythmias include one or more of the following:

1. Injury occurring at the time of surgery (S/P VSD, TGA, AV canal, TOF, Fontan repair)
2. Intrinsic anatomic or physiologic abnormalities.
3. Damage resulting from chronic hypoxia or hemodynamic stress.

Ventricular Outflow Obstruction
Pulmonic stenosis in children is usually the result of valvular obstruction. However, supravalvular, infundibular or peripheral arterial stenosis also occurs. Obstruction to right ventricular outflow leads to pressure overload of the right ventricle.

The most common variety of aortic stenosis in children is valvular. This results in left ventricular hypertrophy and increased diastolic volume. In severe cases, relative coronary insufficiency may be present. Significant symptoms of angina and/or syncope are seen in less than 10 percent of patients.

A major concern for the anesthesiologist is that right or left ventricular outflow obstruction can be present in children in the absence of significant symptoms. The first indication of any cardiac dysfunction may be a precipitous drop in arterial blood pressure, when potent inhalation anesthetics are used in an affected child. As an example, newborns undergoing general anesthesia for congential cataract surgery, may not have clinical evidence yet of significant ventricular outflow obstruction. Careful history and physical examination are essential to detect these lesions in their early asymptomatic stages.

Shunting
A shunt-producing defect is nonrestrictive if there is no pressure gradient across the defect, and restrictive if there is a significant resistance across it, thus producing a pressure gradient between the two chambers.

The obvious sequelae of intracardiac shunts include arterial desaturation (right-to-left), paradoxical embolization, pulmonary circulatory overload with associated vascular changes (left-to-right), volume overload of the right ventricle, and ventilatory changes.

Eisenmenger's Syndrome occurs in patients with increasing pulmonary hypertension, due to prolonged left-to-right shunting, resulting in shunt reversal, corpulmonale and worsening cyanosis. Excessive pulmonary blood flow and pressure contribute to the development of pulmonary vascular obstructive disease. Anesthetic management in these patients should include the avoidance of hypovolemia, and SVR decrease by anesthetic agents, or an increase in PVR due to cold, acidosis, hypercarbia, hypoxia, and catecholamines. Postoperative observation in a cardiorespiratory monitored bed, including continuous pulse oximetry, is indicated in these patients if they undergo general anesthesia.

Prior Surgical Intervention
A child's cardiac lesion may be virgin or in some stage of surgical repair (either palliated or corrected). By definition, palliative surgery neither creates a normal anatomic relationship between the cardiovascular structures, nor establishes normal blood flow pathway. It simply replaces a sequence of abnormal relationship with another more functional system. Corrective cardiac surgery, on the other hand, is designed to normalize the relationships between cardiovascular structures and create a normal route for blood flow. The true result of corrective surgery more often than not, is merely reparative; cardiovascular problems associated with the original anomaly quite often remain, and, ironically, new difficulties may develop. In one survey, only individuals with surgery for atrial septal defect (ASD), pulmonic stenosis, and patent ductus arteriosus (PDA) came close to being considered cured. Arrhythmias, congestive heart failure, and pulmonic hypertension often develop after surgery for tetralogy of Fallot, ventricular septal defect (VSD), coarctation of the aorta, aortic stenosis, and transposition of the great vessels. The cardiovascular physiology following correction of complex defects may pose a challenge for the anesthesiologist, as is evident in patients who undergo Fontan repair or cardiac transplantation.

CHOICE OF ANESTHETICS
The choice of anesthetic agents for the child with CHD depends on the type of operation to be performed, the anticipated duration of the surgery, the preference of the anesthesiologist, and the patient's cardiovascular status. The choice of anesthetic induction technique depends on the child's age, psychologic preparedness, cardiovascular status and whether an intravenous (IV) catheter is present, or can be easily placed. The recognized cardiovascular effects of the anesthetic agents must also be taken into consideration. Anesthetic induction may also be influenced by factors that alter the uptake and distribution of inhalational or IV agents.

The speed of induction of an inhalational anesthetic is determined by the rates of anesthetic inflow into the lungs, transfer of the agent from the lungs to the arterial blood, and anesthetic transfer from the arterial blood. Anesthetic equilibration between the alveoli and arterial blood and, in turn, the brain, is usually rapid. Under most circumstances, the speed of induction depends on those factors that determine the rate of rise of alveolar anesthetic level. In children with CHD with diminished pulmonary blood flow, the transfer of the agent from the lungs to the arterial blood is slowed. Inhalational induction in a patient with a right-to-left shunt can be prolonged, because the shunted blood decreases, or dilutes the partial pressure of the anesthetic in the blood that is reaching the brain, compared with the partial pressure in the blood leaving the lungs. A high inspired concentration of soluble anesthetic agents (e.g., halothane) combined with augmented ventilation, can be used to counteract this effect.

In patients with left-to-right shunting, the speed of inhalation induction is unchanged. Recirculation through the lungs of left-to-right shunt blood, which already carries a high concentration of anesthetic, reduces anesthetic uptake from the alveoli and promotes a more rapid rise in alveolar partial pressure. The augmented pulmonary blood flow, however, increases anesthetic uptake from the alveoli, thereby delaying the rise in alveolar anesthetic levels. So, while a left-to-right shunt increases

pulmonary blood flow, it does not substantially alter the rate of anesthetic induction.

There are two exceptions to this rule. Firstly, when systemic cardiac output (CO) is not maintained, the reduction in systemic venous return to the lungs results in a more rapid rise in alveolar anesthetic level. Secondly, in patients with a large right-to-left shunt, that by itself will slow the speed of induction, the coexistence of a left-to-right shunt will speed the rate of induction, thereby counteracting the initial delay. The effects of shunting on the speed of inhalation inductions are more pronounced for N_2O than for the more soluble volatile anesthetics.

The pharmacokinetics of IV agents may also be affected by shunting of blood, as can readily be seen by examining CO curves in children with CHD. In patients whose lesions are characterized by left-to-right shunting and increased pulmonary blood flow, an IV bolus of a given drug reaches the brain at the same time as it would if no shunt existed. Its initial peak concentration, however, is lower, and its effect is prolonged. In lesions with right-to-left shunting, where systemic venous blood bypasses the pulmonary circulation, the bolus reaches the brain sooner than predicted. Both anesthetic effects and cardiovascular depression may, therefore, appear more rapidly after IV administration of intravenous agents in these patients.

N_2O is often used as the carrier gas with other volatile anesthetics to facilitate inhalational induction in children. Of concern, however, is the potential hazard of air bubble enlargement in children with shunting lesions, and the fact that the use of N_2O may preclude the administration of high concentrations of inspired O_2. N_2O may also cause increases in PVR in non-cyanotic children.

Nevertheless, volatile anesthetic agents are widely used during cardiac and noncardiac surgery in children with CHD. Mask induction with sevoflurane (and to a lesser degree, halothane) is associated with minimal struggle, and is often selected when IV access is difficult. The use of volatile anesthetics permits the delivery of high oxygen concentration when appropriate. Although all of the volatile agents are myocardial depressants, the actual effect on cardiac output and arterial blood pressure is modified by other factors, such as the effect on SVR. Both halothane and isoflurane may cause a dose-dependent decrease in arterial blood pressure. The mechanism is different in each case. The decrease in blood pressure associated with the use of halothane is mainly due to a reduction in cardiac output. Systemic vascular resistance remains largely unchanged. The decrease in blood pressure with isoflurane, on the other hand, is due to a decrease in systemic vascular resistance, while the cardiac output remains normal. In children with balanced shunts, the difference in the effect of these two commonly used agents on systemic vascular resistance (particularly in those children in whom a drop in SVR occurs) can result in an increase in right-to-left shunting and hypoxia. Sevoflurane is associated with less bradycardia or dysrhythmias than halothane. Cardiovascular changes at equipotent concentrations of sevoflurane and halothane in healthy children have been measured by echocardiography. Sevoflurane resulted in a lesser decrease in cardiac output than halothane. Halothane caused a greater decrease in heart rate and cardiac index at all concentrations, than did sevoflurane.

It is important to recognize that the circulatory effects of the volatile agents are dose-related, titratable, and well tolerated by most children with CHD. Profound circulatory effects are generally related to relative anesthetic overdose. Myocardial depression resulting from an anesthetic overdose, will decrease CO and result in systemic hypotension in patients with severe aortic stenosis. In children with severe pulmonic stenosis, halothane may decrease pulmonary blood flow and compromise oxygenation. On the other hand, the hemodynamic effects of these drugs may be desirable in some children. For example, the negative chronotropic and inotropic actions of halothane may reduce the degree of outflow obstruction, and promote forward CO in patients with subvalvular aortic stenosis. It may also increase pulmonary blood flow in patients with pulmonic stenosis or tetralogy of Fallot.

The effect of volatile anesthetics on the heart rate is also dose dependent. Barash reported a dose-dependent depression of ventricular function in children during halothane anesthesia that produced a significant decrease in cardiac output, heart rate and blood pressure. The administration of atropine resulted in a rapid improvement in cardiac output and blood pressure.

The use of opioids in anesthesia for children with CHD is usually associated with excellent hemodynamic stability. High-dose narcotic techniques can be used in children undergoing major surgical procedures, where postoperative ventilatory support will be needed. Both fentanyl (25-75 mcg/kg) and sufentanil (5-20 mcg/kg) can be used in very sick infants and children with all forms of CHD. Both agents produce minimal pulmonary and systemic hemodynamic changes. Remifentanil can be used in children undergoing short surgical procedures, without prolongation of recovery. Low doses of narcotics may be used to supplement a reduced volatile anesthetic concentration in children undergoing less extensive procedures. They can also be combined with muscle relaxants and N_2O in a balanced IV technique. "Light" anesthetic techniques maintain sympathetic tone and maintain cardiac output and SVR, both of which may be desirable in children with severe valvular stenosis or ventricular failure. Most children with decreased pulmonary blood flow lesions, tolerate balanced anesthetic techniques, as long as excessive sympathetic tone does not cause PVR to rise.

Ketamine can be administered intramuscularly to induce anesthesia (4-8 mg/kg) or sedation (2-3 mg/kg) in an uncooperative child with CHD. The sympathomimetic effects of ketamine maintain contractility and SVR. In the absence of hypoventilation, a ketamine dose of 1-2 mg/kg IV does not increase PVR in children with CHD, including those with pulmonary vascular disease.

Thiopental (4-6 mg/kg IV) is well-tolerated in normovolemic children with compensated CHD. Reduced doses must be used in children whose circulatory function is compromised. The hemodynamic effects of propofol in children with CHD are similar to those of thiopental. Rectal methohexital may be used in children with CHD as long as the child is carefully monitored for possible hypoxia or hypoventilation during induction.
The choice of specific neuromuscular blocking agents in children with CHD is usually made on the basis of the drugs' predicted cardiovascular effects, as well as their duration of action. Pancuronium is a popular choice in major cases because its vagolytic effect supports HR and CO, especially in children who receive high-dose opioids. If tachycardia is not desirable, (or for shorter procedures), mivacurium, rapacuronium, atracurium, vecuronium, or rocuronium can be used, with minimal effects on hemodynamic functions. SAJAA

Suggested further reading

1. Hannallah RS, Verghese ST. The Pediatric Patient. In Estafanous, Reves, and Barash Eds. Cardiac Anesthesia 2/E Lippincott Williams & Wilkins, Philadelphia, 2001.