Pediatric Cardiac Anesthesia and Surgery: The Heart of the Matter

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Pediatric cardiac anesthesia continues to evolve as an exciting and technically demanding subspecialty in which anesthetic management is based on sound physiologic principles. Congenital cardiovascular surgery and anesthesia are often performed under unusual physiologic conditions. Rarely in clinical medicine are patients exposed to such biologic extremes as during congenital heart surgery. Frequently, patients are cooled to 15–18°C, acutely hemodiluted by more than 50% of their extracellular fluid volume, and undergo periods of total circulatory arrest up to 1 h. The ability to manage patients under these physiologic extremes is a vital function of the pediatric cardiovascular anesthesiologist.

Clearly, the perioperative management of these complex patients requires a group of physicians (surgeon, anesthesiologist, cardiologist, and critical care specialist) and nurses working as a team. This team orientation is essential to achieve an optimal outcome. Although the quality of the surgical repair, the effects of cardiopulmonary bypass, and postoperative care are the major determinants of outcome, meticulous anesthetic management is imperative. Ideally, despite the complexity of the patients and the marked physiologic changes attributed to cardiopulmonary bypass (CPB) and the surgical procedures, anesthetic care should never contribute substantially to morbidity or mortality. The challenge is to understand the principles underlying the management of patients with congenital heart disease and apply them to clinical anesthesia.

General Anesthetic Considerations

Preoperative Management

Preoperative Evaluation. Caring for children with congenital heart disease presents the anesthesiologist with a wide spectrum of anatomic and physiologic abnormalities. Patients range from young, healthy, asymptomatic children having a small atrial septal defect (ASD) closed to the newborn infant with hypoplastic left heart syndrome requiring aggressive perioperative hemodynamic and ventilatory support. Intertwined with the medical diversity of these patients are the psychological factors affecting both the patient and the parents. Preparation of the patient and the family is time consuming, but omitting or compromising this aspect of patient care is a major deterrent to a successful outcome and patient/parental satisfaction. This approach mandates that cardiac surgeons, cardiologists, anesthesiologists, intensivists, and nurses work as a team in preparing the patient and the family for surgery and postoperative recovery. This team-oriented approach not only prepares the patient and family, but also serves as a safeguard to prevent errors and omissions in the exacting perioperative care necessitated by the complexity of cardiac surgery for congenital heart disease. The preoperative visit offers the family the opportunity to meet the surgeon and anesthesiologist and to begin preparing the patient and family for surgery. It is equally important to ascertain current medications, previous anesthetic problems, or family history of anesthetic difficulties. In the modern era of echocardiography and cardiac catheterization, physical examination rarely contributes additional anatomic information about the underlying cardiac lesion.

Premedication. For infants between 6 to 9 mo, pentobarbital (2–4 mg/kg) may be administered orally. A calm, cooperative, sedated child is the usual result. Children older than 9 mo of age receive a benzodiazepine or a barbiturate as an oral premedication. Our current preference is midazolam 0.3–0.7 mg/kg PO 10–20 min before induction. The higher dose is used in the smallest patients. Alternatively, pentobarbital 4 mg/kg is administered 1 h before induction. For infants <6 mo of age, no premedication is used.

Intraoperative Management

Physiologic Monitoring. The monitoring used for any specific patient should depend on the child's condition and the magnitude of the planned surgical procedure. Noninvasive monitoring is placed before induction of anesthesia. In the crying pediatric patient, the anesthesiologist may elect to defer application of monitoring devices until immediately after the induction of anesthesia. Standard monitoring includes

an electrocardiographic (ECG) system, pulse oximetry, capnography, precordial stethoscope, and an appropriate-sized blood pressure cuff (either oscillometric or Doppler). Additional monitoring includes an indwelling arterial catheter, temperature probes, and an esophageal stethoscope. Foley catheters are generally used when surgical intervention entails cardiopulmonary bypass, might produce renal ischemia, or when the anesthetic management includes a regional technique associated with urinary retention. Some centers routinely use central venous pressure monitoring for major cardiovascular surgery. Alternatively, we typically use directly placed transthoracic atrial lines to obtain that information for separation from cardiopulmonary bypass and the postoperative period. In that setting, the benefits of the information or access provided by percutaneous central venous pressure catheters in the pre-bypass period must be weighed against the risks they pose.

Induction and Maintenance of Anesthesia. The principles of intraoperative management of cardiothoracic surgical procedures are based on an understanding of the pathophysiology of each disease process and a working knowledge of the effects of the various anesthetic and other pharmacologic interventions on a particular patient's condition. Selecting an induction technique is dependent on the degree of cardiac dysfunction, the cardiac defect, and the degree of sedation provided by the premedication. In children with good cardiac reserve, induction techniques can be quite varied as long as induction is careful and well monitored. The titration of induction agents is more important than the specific anesthetic technique in patients with reasonable cardiac reserve. A wide spectrum of anesthetic induction techniques with a variety of agents has been used safely and successfully, e.g., sevoflurane, halothane, halothane and nitrous oxide, IV or IM ketamine, or IV propofol, fentanyl, midazolam, or thiopental. For neonates undergoing open-heart surgery, opioid-relaxant inductions are most prevalent whereas older children with sufficient cardiac reserve typically receive inhalational induction with either halothane or sevoflurane.

After anesthetic induction, IV access is established or augmented as appropriate. A nondepolarizing muscle relaxant is usually administered and an IV opioid and/or inhalation agent chosen for maintenance anesthesia. The child is preoxygenated with 100% oxygen and a lubricated nasal endotracheal tube is carefully positioned. A nasal tube is usually selected because most patients require a period of postoperative mechanical ventilation and the nasal route provides greater stability and patient comfort than the oral route.

Because of the diverse array of congenital heart defects and surgical procedures, an individualized anesthetic management plan is essential. The maintenance of anesthesia in these patients depends on the age and condition of the patient, the nature of the surgical procedure, the duration of cardiopulmonary bypass, and the need for postoperative ventilation. An assessment of the hemodynamic objectives designed to lessen the pathophysiologic loading conditions should be developed for each patient, taking advantage of the known qualitative effects of specific anesthetic agents and ventilatory strategies. These individualized plans must also integrate with the overall perioperative goals to configure the optimal anesthetic. In patients with complex defects requiring preoperative inotropic and mechanical ventilatory support, a carefully controlled hemodynamic induction and maintenance anesthetic with a potent opioid is generally chosen. In patients with a simple ASD or ventricular septal defect (VSD), a potent inhalation agent is preferred as the principal anesthetic agent. This allows for early postoperative extubation and a less prolonged period of intensive care monitoring. More important than the specific anesthetic techniques and drugs is the skilled execution of the anesthetic plan, taking into account patient response to drugs, the changes associated with surgical manipulation, and early recognition of intraoperative complications.

The use of potent inhalation agents as primary anesthetics should be reserved for the child with adequate cardiovascular reserve who is a candidate for early postoperative extubation. In these patients, the myocardial depression and hypotension associated with the use of inhalation agents is well tolerated. Examples include closure of an ASD or VSD, excision of a discrete subaortic membrane, pulmonic or aortic stenosis, ligation of a patent ductus arteriosus (PDA), and repair of coarctation of the aorta.

Cardiopulmonary Bypass

Differences Between Adult and Pediatric CPB. The physiologic effects of cardiopulmonary bypass CPB on neonates, infants, and children are significantly different from those in adults. During CPB, pediatric patients are exposed to different biologic extremes not seen in adults, including deep hypothermia (15–20°C), hemodilution (3- to 15-fold greater dilution of circulating blood volume), low perfusion pressures (20 to 30 mm Hg), wide variation in pump flow rates (ranging from highs of 200 mL $\cdot$ kg^{-1} $\cdot$ min^{-1} to total circulatory arrest), and differing blood pH management techniques (α-stat or pH stat, or both sequentially). These parameters differ significantly from normal physiology and affect preservation of normal organ function during and after CPB. In addition to

these prominent changes, patient age and subtle variations in glucose supplementation, cannula placement, and presence of aortopulmonary collaterals may also be important factors affecting organ function during CPB. Adult patients are infrequently exposed to these biologic extremes. In adult cardiac patients, temperature is rarely lowered below 25°C, hemodilution is more moderate, perfusion pressure is generally maintained at 50–80 mm Hg, flow rates are maintained at 50–65 mL · kg^{-1} · min^{-1} and pH management strategy is less influential because of moderate hypothermic temperatures and rare use of circulatory arrest. Variables such as glucose supplementation rarely pose a problem in adult patients as a result of large hepatic glycogen stores. Venous and arterial cannulae are larger and less deforming of the atria and aorta, and their placement more predictable. Although superficially similar, the conduct of CPB in children is considerably different from that in adults. One would therefore expect marked physiologic differences and sequelae in the response to CPB in the child.

Deep Hypothermic Circulatory Arrest. Neonates and small infants weighing <8–10 kg who require extensive repair of complex congenital heart defects may have their repair using DHCA. This technique facilitates precise surgical repair under optimal conditions, free of blood or cannulas in the operative field, providing maximal organ protection, and often resulting in shortened total CPB time. The scientific rationale for the use of deep hypothermic temperatures rests primarily on a temperature-mediated reduction of metabolism. Whole body and cerebral oxygen consumption during induced hypothermia decreases the metabolic rate for oxygen by a factor of 2 to 2.5 for every 10°C reduction in temperature. These results are consistent with in vitro models, which relate temperature reduction to a decrease in the rate constant of chemical.

Discontinuation of CPB. When weaning from CPB, blood volume is assessed by direct visualization of the heart and monitoring right atrial or left atrial filling pressures. When filling pressures are adequate, the patient fully warmed, acid/base status normalized, heart rate adequate, and sinus rhythm achieved, the venous drainage is stopped and the patient can be weaned from bypass. The arterial cannula is left in place so that a slow infusion of residual pump blood can be used to optimize filling pressures. Myocardial function is assessed by direct cardiac visualization and a transthoracic left or right atrial catheter, a percutaneous internal jugular catheter, or by the use of intraoperative echocardiography. Pulse oximetry can also be used to assess the adequacy of cardiac output. Low systemic arterial saturation or the inability of the oximeter probe to register a pulse may be a sign of very low output and high systemic resistance.

After the repair of complex congenital heart defects, the anesthesiologist and surgeon may have difficulty separating patients from cardiopulmonary bypass. Under these circumstances, a diagnosis must be made and includes 1) an inadequate surgical result with a residual defect requiring repair, 2) pulmonary artery hypertension, and 3) right or left ventricular dysfunction. Two general approaches are customarily used, either independently or in conjunction with one another. An intraoperative "cardiac catheterization" can be performed to assess isolated pressure measurements from the various great vessels and chambers of the heart (i.e., catheter pullback measurements or direct needle puncture to evaluate residual pressure gradients across repaired valves, sites of stenosis and conduits, and oxygen saturation data to examine for residual shunts). Alternatively, echo-Doppler may be used to provide an intraoperative image of structural or functional abnormalities to assist in the evaluation of the postoperative cardiac repair. If structural abnormalities are found, the patient can be placed back on CPB and residual defects can be repaired before leaving the operating room. Leaving the operating room with a significant residual structural defect adversely affects survival and increases patient morbidity. For the anesthesiologist, echo-Doppler can rapidly identify right and left ventricular dysfunction and suggest the presence of pulmonary artery hypertension. In addition, echo-Doppler can identify regional wall motion abnormalities resulting from ischemia or intramyocardial air that will direct specific pharmacologic therapy and provide a means of assessing the results of these interventions.

Specific Anesthetic Considerations

Anesthesia for Simple Open Heart Procedures

Simple open-heart procedures include those defects that involve relatively straightforward surgical repair, uncomplicated hemodynamics, minimal to moderate postoperative intensive care and mechanical ventilation. Examples include repairs of ASD, VSD, and some cases of tetralogy of Fallot in otherwise healthy children with good cardiopulmonary reserve. Patients with congestive heart failure and minimal reserve will require a different level of care despite the relatively "simple" nature of the surgical procedure.

Specific anesthetic considerations for these patients, in addition to the general guidelines previously discussed, are predicated on the use of CPB, occasional use of deep hypothermic circulatory arrest (DHCA), and the specific perioperative issues associated with these defects. The anesthetic management can be divided into three phases: precorrection, immediate postcorrection, and postoperative. During the precorrection phase, the plans for premedication, induction

and maintenance of anesthesia, and selection of monitors are based on the patient's physiology and plans for postoperative care. Premedication without opioid is selected for patients who will be extubated at the end of the procedure; opioid is added for those who will remain on mechanical ventilation. Inhalation induction is typically tolerated well, but special considerations should be made for patients with right-to-left shunting. The rise in arterial anesthetic concentration is slowed in patients with right-to-left shunting because arterial (anesthetic-saturated) blood is diluted with blood that has not passed through the lungs; this effect is more pronounced with insoluble agents. Patients with particularly high RVOT obstruction benefit from particular vigilance to adequate airway and ventilation, to prevent pulmonary hypertension, impaired pulmonary blood flow, and diminished left-side filling. Aside from necessary preparation for airway management, agents to maintain SVR (phenylephrine) and reduce PVR (oxygen, halothane, opioid) should be readily available throughout the precorrection period, particularly during induction.

The level of monitoring also reflects the nature of the procedure: adequate IV access for fluid or blood transfusion anticipates the potential for bleeding; invasive pressure monitoring is established in preparation for CPB. Transthoracic atrial lines are placed before discontinuing CPB if the need for central pressure monitoring, or for delivery of inotropic/hemodynamic infusions is anticipated. Simple PFO/secundum ASD closures do not typically require central catheters. Anesthetic maintenance comprises opioid, 10–50 μg/kg fentanyl, supplemented with inhalation anesthetic, depending on the anticipated time of extubation.

ASDs and VSDs require the use of CPB for access to the surgical site. Right atriotomy allows visualization of most ASDs and, with a trans-tricuspid approach, some VSDs (membranous). Depending on the size of the defect, suture (for PFO), pericardial patch, or prosthetic material may be used for closure. This technique may be associated with atrial arrhythmias and conduction delay after repair. Some VSDs (such as malalignment, supracristal, and muscular) might not be visible through the tricuspid valve; right or left ventriculotomy may be required for their closure. Consequences of ventricular incision include atrioventricular node dysfunction with various degrees of heart block, and ventricular myocardial dysfunction.

The numerous varieties of right ventricular outflow obstruction in tetralogy of Fallot necessitate an individualized approach to each patient. Generally, right ventriculotomy enables repair of the VSD and excision of excess muscle proximal to the pulmonic valve; extension of the incision permits the placement of a patch across the annulus of the pulmonary valve to enlarge the RVOT. Insufficiency or regurgitation of the pulmonic valve may result. More complex cases of tetralogy of Fallot may require more extensive surgical intervention, and are considered separately in the following section.

With these relatively straightforward repairs, separation from CPB is achieved with minimal or no inotropic support. The potential for conduction disturbances, particularly after ventriculotomy, supports the placement of temporary epicardial pacing wires. Other immediate postcorrection issues include residual shunt and bleeding. Significant residual shunts, as assessed by intraoperative transesophageal echocardiography, potentially necessitate return to CPB for reevaluation or re-repair. Bleeding can be monitored after sternal approximation with the use of a mediastinal drain placed to wall suction.

Factors to be evaluated for determining the level of postoperative care required include the need for mechanical ventilation, the need for inotropic support, and the presence of other complications. After simple ASD repair, in many patients the trachea can be extubated immediately. Patients who have undergone VSD repair and simple tetralogy of Fallot repair typically remain on mechanical ventilation for 2–12 h, depending on the type of repair. Dopamine at 3 $\mu g \cdot kg^{-1} \cdot min^{-1}$ or less is used to assist recovery from mild ventricular dysfunction.

Anesthesia for Complex Open-Heart Procedures

Except for the correction of left-to-right shunts and uncomplicated tetralogy of Fallot repair, most open-heart procedures for congenital heart defects are considered complex for a number of reasons. The defects themselves involve more than one cardiac structure; the hemodynamics that ensue necessitate more complex medical and surgical approaches defects. Despite the development of newer diagnostic techniques that allow for early detection of congenital heart disease, many patients present only when significant cardiovascular compromise has occurred. Although the framework of considerations used for the simple procedures can be applied here, patients undergoing complex repairs tend to require higher levels of perioperative monitoring and hemodynamic and ventilatory support.

Despite these complexities, children who have only moderate symptoms undergo premedication and inhalation induction as for simple procedures, with IV placement after induction. Patients with cardiovascular compromise undergo IV induction without premedication to allow for greater hemodynamic control. Nasotracheal intubation is selected over the oral route to provide more postoperative comfort for the patient and to minimize the likelihood of dislodgement. Invasive arterial monitoring is necessary for continuous blood pressure and frequent monitoring of arterial blood gases, hematocrit, and electrolytes, both in the

operating room and in the intensive care unit. Although the procedures share many of the same potential postoperative complications, such as bleeding, ventricular dysfunction, and arrhythmias, each type of case raises its own set of anesthetic issues. The encyclopedic list of complex defects precludes individual discussion; however, the unique physiologic and surgical features of the following lesions present considerations that can be extrapolated for similar defects. These unique defects are complex tetralogy of Fallot with abnormal pulmonary vasculature, univentricular heart, truncus arteriosus, and transposition of the great arteries.

Of the diagnostic criteria for tetralogy of Fallot (malalignment VSD, overriding aorta, right ventricular (RV) hypertrophy, and RV outflow obstruction), the RV outflow obstruction is the most variable in both degree and location. The spectrum ranges from dynamic subpulmonic obstruction with normal anatomy distally to small pulmonary valve to absent pulmonary valve and arteries. In the latter cases, with diminutive or absent pulmonary arteries, pulmonary blood flow occurs either through a patent ductus arteriosus, or by aortopulmonary collateral circulation. These phenomena can be demonstrated by echocardiography, but are frequently evaluated by angiography before surgery. In addition, the 5–6% incidence of abnormal coronary anatomy, with the anterior descending artery originating from the right coronary artery and traversing the RV outflow tract, necessitates careful preoperative evaluation before RV outflow tract incision is planned.

This physiology often results in "protection" of the pulmonary vasculature from overcirculation, and the subsequent development of pulmonary vascular occlusive disease. Cyanosis and compensatory polycythemia are frequent findings in these patients. Depending on the size and inflow source of the pulmonary circulation, surgical options range from augmentation of small pulmonary arteries to reimplantation of aortopulmonary collaterals onto an RV-pulmonary artery conduit. Although these plans theoretically provide pulmonary blood flow, the small caliber of the vessels can result in high pulmonary vascular resistance. The decision to close the VSD may also impact postcorrection myocardial function. Right ventricular failure may result from elimination of systolic "popoff," if VSD is closed, inducing a pressure load on the RV, which must now pump against potentially high pulmonary vascular resistance (PVR). Mechanisms to minimize PVR through a reconstructed pulmonary circulation include mild hypocapnia, high inspired oxygen concentration, adequate anesthesia/analgesia, and pharmacologic therapy. Agents useful for this purpose include inhaled nitric oxide, opioids, milrinone, dobutamine, and isoproterenol.

Mixing lesions present a distinct set of challenges to the anesthesiologist. Precorrection management is similar for these two defects, the differences in surgical goals for each is reflected in the postcorrection care. The parallel, rather than series, nature of the systemic and pulmonary circulations necessitates medical therapy to balance flow through each. Examples include univentricular heart and truncus arteriosus. Surgery for univentricular heart is typically staged over time, to allow development of the pulmonary vasculature and maintain ventricular function. After stage I palliation, the parallel arrangement of systemic and pulmonary circulations is maintained. Repair of truncus arteriosus, by contrast, achieves repair in one operative encounter, and results in separation of the circulations into the normal serial arrangement. Particularly in the neonatal period, when a majority of these patients present, the reactivity of the pulmonary vasculature provides a useful way, in the precorrection period, to achieve balanced circulation (i.e., Qp/Qs of 1). Because of this arrangement, pulmonary overcirculation implies systemic hypoperfusion; this may eventually result in diminished perfusion of end organs and acidosis. As previously noted, hypercapnia and low inspired oxygen concentration promote increases in PVR; these factors can be regulated by altering inspired gas mixtures, with the addition of CO_2 or N_2. In the operating room, CO_2 is added to achieve an Spo_2 near 85%. Metabolic acidosis is treated by attempting to improve systemic flow and, on occasion, IV sodium bicarbonate.

The infant with univentricular heart undergoing initial palliation (the first stage in the series of operations leading to Fontan physiology) to provide pulmonary blood flow requires special considerations. The use of an arterial to pulmonary artery shunt impacts on the site of arterial cannulation; for a planned right modified Blalock-Taussig shunt, the right upper extremity is avoided. The diminutive size of the ascending aorta in hypoplastic left heart syndrome often preclude its cannulation and use for initiating bypass; an alternative strategy uses pulmonary artery cannulation, with subsequent snaring of the branch pulmonary arteries to allow systemic flow through the ductus arteriosus. Surgical repair reconfigures the pulmonary trunk into the neoascending aorta, such that this site can be recannulated after DHCA for reinitiating CPB. Depending on the size of the native aorta, homograft or prosthetic material is used for augmentation. An arterial to pulmonary artery shunt, either from the right subclavian or the right carotid artery, will supply the pulmonary circulation. Occasionally, a centrally located shunt between the ascending aorta and the pulmonary artery may be used.

Separation from bypass is achieved once inotropic support is started, ventilation is initiated, and shunt flow is established. Issues that complicate postcorrection management in the operating room and the intensive care unit include hypoxemia and metabolic acidosis. Hypoxemia after a Stage I palliation results from several

factors including inadequate ventilation, impaired shunt flow, and high pulmonary vascular resistance. The placement of a fresh endotracheal tube with a large internal diameter, the process of suctioning the lungs before resuming ventilation after DHCA, and the use of a neonatal ventilator minimize the opportunity for inadequate ventilation. Impaired shunt flow can result from thrombus or kinking of the shunt, excessive length, or inadequate diameter. Manipulation of pulmonary vascular resistance, using ventilation, inspired oxygen concentrations, and pulmonary vasodilators may provide some improvement in oxygen saturation. Compression of vascular structures after chest closure occasionally diminishes pulmonary blood flow and may require leaving the sternum open for 24–48 h. Although some metabolic acidosis is anticipated after separation from CPB, its persistence suggests impaired systemic perfusion, either from aortic arch obstruction, ventricular dysfunction, or severe atrioventricular valve regurgitation. Echocardiography assists in diagnosing these problems. Inotropic support and afterload reduction may improve systemic flow. Expectant management of mild arch obstruction may allow balloon dilation in the cardiac catheterization laboratory at a later date; severe arch obstruction necessitates surgical revision.

Anesthesia for Closed Heart Operations

Early corrective repair in infancy has significantly reduced the number of noncorrective palliative closed heart operations. Corrective closed heart procedures include PDA ligation and repair of coarctation of the aorta. Noncorrective closed heart operations include pulmonary artery banding, extracardiac shunts such as the Blalock-Taussig shunt. All these procedures are performed without cardiopulmonary bypass. Therefore, venous access and intraarterial monitoring are important in evaluating and supporting these patients. A pulse oximeter remains an invaluable monitor during intraoperative management.

PDA ligation is typically performed through a left thoracotomy, although video-assisted thoracoscopic techniques are increasingly prevalent. Physiologic management is that of a left-to-right shunt producing volume overload. Patients with a large PDA and low PVR generally present with excessive pulmonary blood flow and congestive heart failure. Neonates and premature infants also run the risk of having substantial diastolic runoff to the pulmonary artery, potentially impairing coronary perfusion. Thus patients range from an asymptomatic healthy young child to the sick ventilator-dependent premature infant receiving inotropic support. The former patient allows for a wide variety of anesthetic techniques culminating in extubation in the operating room. The latter patient requires a carefully controlled anesthetic and fluid management plan. Generally, a trial of medical management with indomethacin and fluid restriction is attempted in the premature infant before surgical correction. In the premature infant, transport to the operating room can be especially difficult and potentially hazardous, requiring great vigilance to avoid extubation, excessive patient cooling, or venous access disruption. For these reasons, many centers are now performing ligation in the neonatal intensive care unit. Intraoperatively, retractors may interfere with cardiac filling and ventilatory management so that hypotension, hypoxemia, and hypercarbia occur. Complications include inadvertent ligation of the left pulmonary artery or descending aorta, recurrent laryngeal nerve damage, and excessive bleeding resulting from inadvertent PDA disruption. After ductal ligation in premature infants, worsening pulmonary compliance can precipitate a need for increased ventilatory support, and manifestations of an acute increase in left ventricular (LV) afterload should be anticipated, especially if LV dysfunction has developed preoperatively. More recently, PDA ligation has been performed in infants and children using thoracoscopic surgical techniques. This approach has the advantages of limited incisions at thoracoscopic sites, promoting less postoperative pain and discharge from the hospital the same day of surgery.

Coarctation of the aorta is a narrowing of the descending aorta near the insertion of the ductus arteriosus. Obstruction to aortic flow is the result, and this may range from severe obstruction with compromised distal systemic perfusion to mild upper extremity hypertension as the only manifestation. Associated anomalies of both the mitral and aortic valves can occur. In the neonate with severe coarctation, systemic perfusion is dependent on right-to-left shunting across the PDA. In these circumstances, LV dysfunction is very common and PGE 1 is necessary to preserve sufficient systemic perfusion. Generally, a peripheral IV line and an indwelling arterial catheter, preferably in the right arm, are recommended for intraoperative and postoperative management. In patients with LV dysfunction, a central venous catheter may be desirable for pressure monitoring and inotropic support. The surgical approach is through a left thoracotomy, whereby the aorta is cross-clamped and the coarctation repaired with an onlay prosthetic patch, a subclavian artery flap, or resection of the coarctation with an end-to-end anastomosis. During cross-clamping, we usually allow significant proximal hypertension (20–25% increase over baseline) because vasodilator therapy may jeopardize distal perfusion and promote spinal cord ischemia. Intravascular volume loading with 10–20 mL/kg crystalloid is given just before removal of the clamp. The anesthetic concentration is decreased, and additional blood volume support is given until the blood pressure rises. Postrepair rebound

hypertension resulting from heightened baroreceptor reactivity is common and often requires medical therapy. After cross-clamping, aortic wall stress attributable to systemic hypertension is most effectively lowered by institution of β-blockade with esmolol or α/β-blockade with labetalol. Propranolol is useful in older patients but can cause severe bradycardia in infants and young children. Although it actually increases calculated aortic wall stress in the absence of β blockade by accelerating dP/dT, the addition of sodium nitroprusside may become necessary to control refractory hypertension. Captopril or an alternative antihypertensive regimen is begun in the convalescent stage of recovery in those patients with persistent hypertension.

The management of infants undergoing placement of extracardiac shunts without cardiopulmonary bypass centers around similar goals as other shunt lesions: balancing pulmonary and systemic blood flow by altering $Paco_2$, Pao_2, and ventilatory dynamics. Central shunts are usually performed through a median sternotomy, whereas Blalock-Taussig shunts may be performed through a thoracotomy or sternotomy. In patients, in whom pulmonary blood flow is critically low, partial cross-clamping of the pulmonary artery required for the distal anastomosis causes further reduction of pulmonary blood flow and desaturation, necessitating meticulous monitoring of pulse oximetry. Careful application of the cross-clamp to avoid pulmonary artery distortion will help to maintain pulmonary blood flow. Under circumstances in which severe desaturation and bradycardia occur with cross-clamping, CPB will be required for repair. Intraoperative complications include bleeding and severe systemic oxygen desaturation during chest closure, usually indicating a change in the relationship of the intrathoracic contents that results in distortion of the pulmonary arteries or kink in the shunt. Pulmonary edema may develop in the early postoperative period in response to the acute volume overload that accompanies the creation of a large surgical shunt. Measures directed at increasing PVR, such as lowering inspired oxygen to room air, allowing the $Paco_2$ to rise, and adding positive end-expiratory pressure, are helpful maneuvers to decrease pulmonary blood flow until the pulmonary circulation can adjust. Decongestive therapy such as diuretics and digoxin may alleviate the manifestations of congestive heart failure. Under such circumstances early extubation is inadvisable.

Pulmonary artery banding is used to restrict pulmonary blood flow in infants who are deemed uncorrectable for either anatomic or physiologic reasons. These patients are generally in congestive heart failure with reduced systemic perfusion and excessive pulmonary blood flow. The surgeon places a restrictive band around the main pulmonary artery to reduce pulmonary blood flow. Band placement is quite imprecise and requires careful assistance from the anesthesia team for successful placement. Many approaches have been suggested. We allow the patient to breathe 21% inspired oxygen concentration and maintain the $Paco_2$ at 40 mm Hg to simulate the postoperative state. Depending on the malformation, a pulmonary artery band is tightened to achieve hemodynamic (e.g., distal pulmonary artery pressure 50–25% systemic pressure) and/or physiologic goals (e.g., Qp:Qs approaching 1). Should the attainment of these objectives produce unacceptable hypoxemia, the band is loosened.

Blalock-Hanlon atrial septectomy is now an uncommon procedure for enlarging an intra-atrial connection. This procedure is done by occluding caval flow and creating an intra-atrial communication through the atrial septum. In patients with hypoplastic left heart syndrome with an intact atrial septum, this procedure is lifesaving and must be performed within hours of birth. Balloon atrial septostomies (Rashkind procedure) and blade septectomies performed in the cardiac catheterization laboratory have replaced surgical intervention, except when left atrial size is very small or the atrial septum is thickened. Improved safety of CPB has led to the virtual elimination of such intracardiac procedures using inflow occlusion. Surgical septectomies, because they are currently confined to the most difficult subset, are rarely performed without benefit of CPB.