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PEDIATRIC CARDIAC EMERGENCIES

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True cardiac emergencies in children are rare compared with the adult population. Cardiac diseases in infancy and childhood may be acquired or congenital. Acquired cardiac disorders, such as cardiac tamponade, usually evolve acutely, and result in sudden alterations in cardiac physiology that may lead to rapid deterioration. Although 0.8% to 1% of infants have congenital heart disease, most are stable and do not present with acute decompensation in the perinatal period.[26] [60] Infants with obstructive cardiac lesions are exceptions, usually requiring emergency medical or surgical intervention. Arrhythmias and congestive heart failure, which can be associated with congenital or acquired diseases, also require immediate attention. Timely identification, management, and stabilization of these patients are important goals. The following is a review of the more common pediatric cardiac emergencies and their management.

CONGENITAL CARDIAC LESIONS

Congenital forms of heart disease result from structural abnormalities of embryologic development that are present at birth, but the clinical manifestations of such disorders may not become evident for days, months, or years.[26] The age at presentation of congenital heart disease depends on the type of cardiac lesion and its effect on cardiac function. Although congenital heart disease generally is well tolerated during fetal life, postnatal circulatory changes have marked effects on the clinical presentation and course of the heart disease in the neonatal period. Severe obstruction to pulmonary or systemic blood flow may be masked in the first few days of life by persistence of a patent ductus arteriosus. Detection of a significant ventricular septal defect or atrial septal defect may be delayed for weeks. The wide variability in the age at presentation is related to the physiologic interactions of the systemic and pulmonary circulations after birth and to the size and location of the cardiac lesion.[26] Nonetheless, specific congenital cardiac defects do present
with acute illness, but they can produce nonspecific symptoms that may be confused with more common disorders, such as neonatal sepsis or overwhelming pneumonia. These disorders may require acute intervention in the newborn period that may include emergency cardiac catheterization or surgery.

**Indications for Emergency Cardiac Catheterization**

**Obstructive Left-Sided Cardiac Lesions**

Obstructive left-sided cardiac lesions typically present with acute illness in the newborn period. These lesions depend on a patent ductus arteriosus for systemic blood flow. Critical coarctation of the aorta, critical aortic stenosis, and hypoplastic left heart syndrome present with rapid onset of symptoms of congestive heart failure. Shock secondary to neonatal sepsis is often the presumed initial diagnosis.

Critical Aortic Stenosis and Critical Coarctation of the Aorta.

These lesions present with obstruction to left ventricular outflow and subsequent left atrial dilatation and pulmonary venous congestion. Increased left atrial pressure leads to left-to-right shunting by way of the foramen ovale, increasing pulmonary blood flow. Because systemic perfusion is maintained by way of the ductus arteriosus, when the ductus closes, congestive heart failure, shock, oliguria, and renal failure promptly ensue. Cardiac function is impaired further by the metabolic acidosis that develops in response to systemic hypoperfusion.

Hypoplastic Left Heart Syndrome.

This defect presents similarly to the previously described lesions. In general, left-sided structures are hypoplastic, whereas right-sided structures are hypertrophied. Left-to-right shunting occurs at the atrial level by way of a patent foramen ovale. Systemic perfusion depends on a patent ductus arteriosus. With preductal coarctation of the aorta and critical aortic stenosis, ductal closure causes cardiogenic shock and circulatory collapse.

The immediate management of patients with left-sided cardiac obstruction involves reopening and maintaining a patent ductus arteriosus. Intravenous infusion of prostaglandin E$_1$ (0.05 μg/kg/min) is the mainstay of therapy. With an open ductus, the relative flows into the systemic and pulmonary circuits depend, to a great degree, on pulmonary vascular resistance. Factors that tend to decrease pulmonary vascular resistance should be avoided because this results in systemic hypoperfusion. Systemic perfusion may be improved by modestly increasing pulmonary vascular resistance through controlled hypoventilation and positive end-expiratory pressure. Pulmonary vascular resistance also can be elevated by reducing the fractional inspired oxygen concentration to room air (0.21) or lower to maintain a Pa$_O_2$ of 30 mmHg. Inotropic support, sodium bicarbonate, and sodium nitroprusside (to decrease systemic vascular resistance) also may be required.

After the initial medical management, these defects require staged or definitive surgery or interventional cardiac catheterization. Balloon angioplasty of coarctation of the aorta has been effective in patients ranging from 3 days of age to adulthood. Fletcher et al observed that infants younger than 7 months of age usually require reintervention within a short time period. Aortic stenosis also may be amenable to interventional catheterization if there is no significant aortic regurgitation. Balloon aortic valvuloplasty can reduce transaortic pressure gradients and left ventricular peak systolic pressures successfully. These improvements seem to persist beyond infancy. As with surgery, balloon dilation can cause or worsen aortic regurgitation, but it now represents an accepted alternative to open heart surgery and aortic valvotomy. If the interatrial septum is markedly obstructive in hypoplastic left heart syndrome, causing severe pulmonary edema, relief of the obstruction may be accomplished with balloon atrial septostomy. Ultimately, survival of patients with hypoplastic left heart syndrome depends on heart transplantation or the palliative Norwood procedure.

**Obstructive Right-Sided Cardiac Lesions**

Pulmonary Valve Stenosis with an Intact Ventricular Septum.

This defect occurs in 8% to 10% of infants with congenital heart defects. Right ventricular hypertrophy is a characteristic anatomic feature. In its classic form, pulmonary valve stenosis with an intact ventricular septum is a conical lesion that projects into the main
pulmonary artery, frequently with complete fusion of the valve leaflets. The mortality from this lesion may be as high as 42% without intervention.\(^\text{[12]}\)

The neonate with unrelied valve obstruction shows signs and symptoms of severe right heart failure, including cyanosis and tachypnea. Resting cardiac output is usually normal, but it may be reduced by tachycardia, which shortens the diastolic filling time. Because there is no route for decompression of the right ventricle, pulmonary blood flow depends on a patent ductus arteriosus. Prostaglandin \(E_1\) therapy is essential even in the postoperative period until pulmonary vascular resistance decreases and right ventricular function gradually improves.\(^\text{[7]}\)

In the past, surgical intervention with an open valvotomy with inflow occlusion was the preferred approach in extremely ill neonates. Balloon pulmonary valvulotomy recently has become the preferred treatment for older children with isolated pulmonary valve stenosis. With the advent of smaller balloon catheters that do not obstruct the right ventricular outflow tract completely, balloon pulmonary valvulotomy also is now possible in the hypoxic neonate.\(^\text{[7]}\) Complications include balloon rupture, bradyarrhythmias and tachyarrhythmias, and bleeding from the catheter site. Balloon valvuloplasty in these patients is performed under general anesthesia, and should be limited to patients with a pulmonary systolic pressure gradient greater than 50 mm Hg.\(^\text{[56]}\)

### Inadequate Mixing of Pulmonary and Systemic Circulations

**Pulmonary Atresia with Intact Ventricular Septum and Restrictive Atrial Septal Defect.**

This abnormality affects approximately 1% to 3% of patients with congenital heart disease.\(^\text{[7]}\)\(^\text{[67]}\) It is a frequent cause of cyanosis and death in the neonatal period, affecting 30% of patients with cyanotic heart disease.\(^\text{[67]}\) Pulmonary blood flow depends on patency of the ductus arteriosus. Because death commonly follows ductal closure, early intervention is mandatory. Prostaglandin \(E_1\) therapy is essential to improve oxygenation and acidosis until surgery can be performed. The goal of surgery is to decompress the right ventricle to allow growth of the ventricular cavity and to improve pulmonary blood flow. Patients with severely restrictive atrial septal defects and a small ventricle may require balloon atrial septostomy (infants younger than 6 weeks of age) or blade atrial septostomy (infants older than 6 weeks of age) to enhance mixing of the pulmonary and systemic venous blood, as the initial procedure. Blade septostomy is the appropriate choice in infants older than 1 to 2 months of age, in whom the atrial septum is usually too thick to be torn adequately by balloon septostomy.\(^\text{[2]}\)\(^\text{[67]}\)

**D-Transposition of the Great Vessels.**

This is a common congenital cardiac abnormality, second in frequency only to isolated ventricular septal defect.\(^\text{[33]}\) The incidence of D-transposition of the great vessels in infants with congenital heart disease is reported to be approximately 5% to 7%.\(^\text{[17]}\) In the heart with D-transposition of the great vessels, the pulmonary and systemic circulations are isolated, with hypoxic blood circulating in the body and oxygenated blood circulating in the pulmonary circuit. Adequate mixing between the two circulations must be present for survival. Mixing can occur at the atrial level by way of an open foramen ovale or an atrial septal defect, at the ventricular level by way of a ventricular septal defect, or at the great arterial level by way of a patent ductus arteriosus. Systemic oxygenation depends on the left-to-right shunt (as the aorta arises from the right ventricle), and the return of deoxygenated blood to the pulmonary circulation depends on the right-to-left shunt. Although bidirectional shunting occurs through the atrial or ventricular septal defects, it is not possible at the level of the patent ductus arteriosus. Therefore, the presence of a patent ductus arteriosus without atrial or ventricular septal defects is not compatible with life. Prostaglandin \(E_1\) infusion dramatically improves oxygenation in the newborn with D-transposition of the great vessels until palliative or definitive surgery can be carried out. The Jatene arterial switch procedure is currently the preferred surgical correction. If mixing is inadequate, however, a Rashkind balloon atrial septostomy or blade atrial septostomy is performed emergently in the neonatal period. If the infant is hemodynamically stable with adequate oxygenation and the arterial switch procedure is to be performed within 24 hours, atrial septostomy may be of no added benefit.\(^\text{[2]}\)\(^\text{[17]}\)\(^\text{[33]}\)

**Tricuspid Atresia with Restrictive Atrial Septal Defect.**

This is a serious lesion that has a 50% mortality by 6 months of age without surgical intervention.\(^\text{[66]}\) Tricuspid atresia is the third most common cause of cyanotic congenital heart disease. The initial surgical intervention is usually a shunt procedure aimed at improving...
pulmonary blood flow so that physiologic correction by means of a Fontan-Kreutzer procedure may be performed later. All patients with tricuspid atresia demonstrate some degree of arterial hypoxemia. Because there is complete separation of the right atrium and ventricle, an atrial shunt with right-to-left flow is needed for survival. If the atrial septal defect or patent foramen ovale is restrictive, a large pressure gradient may exist between the right and left atria with subsequent congestive heart failure, necessitating urgent balloon atrial septostomy or atrial septectomy.

Total Anomalous Pulmonary Venous Connection.

This is a cyanotic congenital defect in which all the pulmonary veins drain to the right atrium, directly or by way of systemic veins. This lesion is believed to account for approximately 2% to 3% of congenital heart disease presenting in infancy. Because there is no direct connection between the pulmonary veins and the left atrium, an interatrial communication (usually a patent foramen ovale) is required for survival. If the interatrial communication is restrictive, there is a limitation of flow into the left ventricle with a resultant decrease in left ventricular output. Systemic hypoperfusion, hypotension, shock, or acidosis may ensue unless balloon or blade atrial septostomy is performed. Prostaglandin E infusion also may help stabilize the patient before surgical repair by permitting right ventricular blood to be shunted by way of the patent ductus arteriosus to the systemic circulation with improved mixed venous blood distribution.

Anesthetic Management

Neonates that present for emergency diagnostic or interventional cardiac catheterization are an anesthetic challenge because of the severity of their illness. In many patients, sedation and local anesthesia are used, with general anesthesia reserved for interventional procedures. Stable neonates may be premedicated with chloral hydrate, 50 to 70 mg/kg orally, before transfer to the cardiac catheterization laboratory. Premedication, however, may not be indicated in hemodynamically unstable newborns, and 0.05 to 0.08 mg/kg of midazolam may be administered intravenously during the procedure. Sedation may be supplemented with 1% lidocaine, 1 to 2 mL (maximum, 5 mg/kg) infiltrated in the groin to provide local anesthesia. Another drug that frequently is administered during procedures is intravenous ketamine, 1 mg/kg. Ketamine may provide perioperative analgesia and sedation with minimal depression of airway reflexes or respiratory drive. Propofol sedation may not be a good choice for critically ill neonates because of significant decreases in mean arterial pressure (greater than 20% of baseline) and arterial desaturation. These sedative agents must be used judiciously to prevent or minimize hemodynamic disturbances caused by respiratory and cardiovascular depression (e.g., alveolar hypoventilation, increased pulmonary vascular resistance, depression of cardiac output, and changes in systemic vascular resistance). Inadequate sedation causing patient agitation, however, also can produce inaccurate hemodynamic data.

If general anesthesia is chosen, it is also important to avoid changes in the heart rate, cardiac contractility, pulmonary or systemic vascular resistance, and venous return, which may alter hemodynamic measurements. In these patients, airway management usually is achieved with tracheal intubation. In certain patients, invasive hemodynamic monitoring, fluid administration, and inotropes or vasodilators may be indicated, especially in the patients with underlying congestive heart failure. Complications of cardiac catheterization include blood loss, arrhythmias, cardiac or vessel perforation, broken wires or catheters (that may need retrieval), anaphylactic reactions to contrast media, seizures, and embolic or thrombotic events. Blood loss should be monitored closely and treated aggressively with volume expansion when it exceeds 5 mL/kg or if the hemoglobin is less than 10 g/dL. Although an optimal hematocrit is controversial, it is a good rule-of-thumb to maintain hematocrits above 30% in patients with acyanotic lesions and above 40% in patients with cyanotic lesions.

ARRHYTHMIAS

Cardiac arrhythmias in children often are caused by an underlying congenital heart defect, especially after heart surgery. The arrhythmia may result from disturbances in impulse formation, conduction, or both. In many children, however, the cause of the arrhythmia is unknown. Although all patients who undergo cardiac surgery are at risk for developing cardiac arrhythmias, certain types of congenital heart disease are associated with a higher incidence of postsurgical rhythm disturbances. These conditions include corrected D-transposition of the great arteries, tetralogy of Fallot, Fontan repair, aortic and subaortic stenosis, ventricular septal defects, endocardial
cushion defects, large atrial septal defects, total anomalous pulmonary venous return, and congenital mitral stenosis.\textsuperscript{[26]}\textsuperscript{[60]}\textsuperscript{[76]}

The development of postsurgical arrhythmias is related to several factors, including the natural history of the specific defect, pre-existing arrhythmias, pre-existing hemodynamic abnormalities, type of surgical repair, age at repair, and the patient's postoperative hemodynamic status.\textsuperscript{[76]} The most common postoperative arrhythmias include supraventricular tachycardia, ventricular tachycardia, sick sinus syndrome, and complete heart block. Supraventricular arrhythmias are more common in congenital heart lesions requiring extensive atrial surgery or after repairs associated with elevated preoperative or postoperative atrial pressure.\textsuperscript{[76]} Postoperative arrhythmias associated with repair of specific congenital heart defects are listed in Table 1.

**TABLE 1 -- POSTOPERATIVE ARRHYTHMIAS ASSOCIATED WITH REPAIR OF SPECIFIC CONGENITAL HEART DEFECTS**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Associated Defects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sick sinus syndrome (seen after surgery near sinus node)</td>
<td>d-TGA, ASD, complex single ventricle lesions (after Fontan repair)</td>
</tr>
<tr>
<td>Supraventricular arrhythmias (seen after intra-atrial repairs)</td>
<td>d-TGA, single ventricle complexes (after Fontan repair)</td>
</tr>
<tr>
<td>Ventricular tachycardia</td>
<td>Tetralogy of Fallot, VSD, AV canal defects, aortic stenosis (especially subvalvular forms, such as idiopathic hypertrophic subaortic stenosis [IHSS]), Ebstein's anomaly, coronary artery anomalies, single ventricle complexes (after Fontan repair), d-TGA</td>
</tr>
<tr>
<td>Complete heart block</td>
<td>VSD</td>
</tr>
</tbody>
</table>

\(d\)-TGA = d-transposition of the great arteries, ASD = atrial septal defect, VSD = ventricular septal defect, AV = atrioventricular.

Other causes of arrhythmias in children include congenital complete heart block, Wolff-Parkinson-White syndrome, and long Q-T syndrome. Acquired heart diseases with associated arrhythmias include cardiomyopathies, viral myocarditis, rheumatic carditis, Kawasaki disease, and cardiac tumors. Arrhythmias also may be associated with myriad systemic disorders, including electrolyte derangements, neuromuscular disorders, endocrine disorders, inherited disorders of metabolism, and central nervous system diseases. When children present with new-onset arrhythmias, the possibility of drug and toxic substance ingestion also must be considered.\textsuperscript{[26]}

Young children and infants may not be aware of or be able to express awareness of an abnormal cardiac rhythm. In these instances, the diagnosis may be established from the patients' symptoms. Arrhythmias may manifest in several ways, including symptoms of congestive heart failure, decreased cerebral perfusion (e.g., syncope, dizziness, irritability, inappropriate behavior), decreased coronary perfusion (e.g., anginal chest pain), or perception of the arrhythmia (i.e., palpitations).\textsuperscript{[26]}

**Supraventricular Tachycardia**

**Cause**

Paroxysmal supraventricular tachycardia, previously called paroxysmal atrial tachycardia, is the most common arrhythmia presenting as a pediatric emergency.\textsuperscript{[21]}\textsuperscript{[24]}\textsuperscript{[25]}\textsuperscript{[26]}\textsuperscript{[42]}\textsuperscript{[60]}\textsuperscript{[76]} Because of improved methods of detection and awareness of supraventricular tachycardia, recent estimates suggest that supraventricular tachycardia occurs in 1 in 250 to 1 in 1000 children.\textsuperscript{[42]} This incidence is much higher than the previously reported incidence of 1 in 25,000.\textsuperscript{[25]} Although supraventricular tachycardia may be caused by enhanced automaticity of specialized atrial fibers, it is usually caused by an abnormal re-entrant mechanism. Electrophysiologic studies have shown that supraventricular tachycardia is actually several arrhythmias. In addition to being caused by abnormal impulse formation, supraventricular tachycardia also may be caused by re-entry within the sinoatrial node, atrium, atrioventricular nodal approaches, and accessory pathways, including the Wolff-Parkinson-White syndrome.\textsuperscript{[24]}\textsuperscript{[26]}\textsuperscript{[65]}
Fifty percent of cases of supraventricular tachycardia in infants are idiopathic (most likely involving concealed accessory pathways or other re-entrant circuits). Twenty-four percent have associated conditions, such as infection, fever, or drug exposure (most commonly cold medications containing sympathomimetic amines), 23% have congenital heart disease (most commonly, corrected transposition of the great vessels and Ebstein’s anomaly), and 22% have Wolff-Parkinson-White syndrome. The cause of supraventricular tachycardia in older children is more likely to be Wolff-Parkinson-White syndrome, concealed bypass tracts, or congenital heart disease. In a retrospective review of 217 pediatric patients with documented episodes of supraventricular tachycardia, Garson et al found that patients who were 4 months of age or younger at time of onset were more likely to present in congestive heart failure, but less likely to experience a recurrence of supraventricular tachycardia later in life.

**Clinical Presentation**

A typical ECG seen in a pediatric patient with supraventricular tachycardia shows a rapid rate and a narrow QRS complex. P waves usually can be seen, but may be obscured by the ST segment. If P waves are absent, a narrow and regular QRS complex is more likely to be of supraventricular rather than of ventricular origin. Supraventricular tachycardia is characterized by a regular heart rate that does not fluctuate with agitation, in contrast to sinus tachycardia. The rate of tachycardia in infants ranges from 220 to 320 beats per minute, whereas the rate in older children typically ranges from 150 to 250 beats per minute. The heart rate usually is not predictive of blood pressure.

The patient may present with a history of irritability, lethargy, or dyspnea with decreased appetite and vomiting. In more severe cases, mottling and cyanosis may be present with varying degrees of congestive heart failure. Infants with congestive heart failure have enlarged livers, but peripheral edema is rare. Older children may report a history of sudden onset of chest pain or palpitations, dizziness, restlessness, syncope, fatigue, and dyspnea. Occasionally, a relationship may be drawn to exercise or caffeine-containing foods.

**Medical Management**

Most infants and children are able to tolerate supraventricular tachycardia for several hours or days without developing clinical symptoms. In these patients, vagal maneuvers (e.g., carotid massage, Valsalva maneuver, gag reflex, abdominal compression, rectal stimulation in infants) and pharmacologic cardioversion are indicated as the first line of treatment. In hemodynamically unstable patients, however, synchronized cardioversion promptly should be used. The history and clinical findings should be assessed quickly to determine the most appropriate course of action.

In the asymptomatic child, direct current cardioversion is not required immediately. The therapeutic goal is to increase vagal tone, slowing conduction and prolonging refractoriness in the atrioventricular node. The success of vagal maneuvers is related to the age of the patient. A proper Valsalva maneuver also requires patient cooperation and 10 to 15 seconds of sustained effort. Vagal maneuvers are usually ineffective in infants and young children. Garson et al found the mean age of successful cardioversion through vagal maneuvers (carotid sinus massage, Valsalva) to be 8 years (with the youngest being 4 years). Although vagal maneuvers are often ineffective in younger children, the occasional successful attempt justifies their initial use. The application of crushed ice to the face or facial immersion in cold water for up to 30 seconds is an effective vagal stimulus that depends less on patient cooperation. This maneuver may result in intense peripheral vasoconstriction and subsequent increase in vagal tone (diving reflex). Although ocular compression may produce the most intense degree of vagal stimulation, it generally is not recommended because of the risk of retinal detachment.

Adenosine has become the first-line pharmacologic therapy for supraventricular tachycardia in pediatric patients who present to the emergency department. It has a rapid onset of action, usually within 10 seconds, and a half-life of only several seconds. Because of its short half-life, it is ineffective if administered too slowly. Clinically significant adverse effects of adenosine are rare in children. Transient bradycardia and asystole and reports of ventricular fibrillation, atrial fibrillation, and polymorphic ventricular tachycardia have been reported. Reported noncardiac adverse effects include apnea in a premature infant and bronchospasm in patients with asthma. In young patients, adenosine initially should be administered as a rapid intravenous bolus dose of 50 or 100 μg/kg, with each subsequent dose increased by 50 μg/kg until the desired effect is obtained or a maximal dose of 250 μg/kg is reached. Patients weighing more than 50 kg may be given an initial dose of 6 mg, with a subsequent dose of 12 mg given 1 to 2 minutes later, if required. Adenosine may be used in acutely ill patients, but should not delay immediate direct current cardioversion in severely compromised patients.
If intravenous access is delayed or if adenosine is not readily available or is unsuccessful in cardioverting a hemodynamically unstable patient, immediate synchronized cardioversion with 0.5 to 1 J/kg should be instituted. If the initial attempt is unsuccessful, the current should be doubled successively until effective or until a dose of 10 J/kg is reached. Because cardioversion may not be successful in the presence of hypoxia or acid-base imbalance, airway support and ventilation may be required to help correct an underlying acidosis. Once the patient's rhythm is converted to sinus rhythm, a pediatric cardiologist should be consulted for further management and evaluation.

In severely resistant cases, the supraventricular tachycardia can be terminated in the catheterization laboratory by overdrive atrial pacing. With an electrode catheter placed in the high right atrium, the atrium can be paced at a rate faster than the supraventricular tachycardia rate. This often interrupts the re-entrant cycle, and results in normal sinus rhythm. A less invasive method in infants and children is to use esophageal overdrive pacing, which requires the passage of a small bipolar electrode catheter through the nasogastric route. The catheter is positioned in the esophagus behind the left atrium, and again, atrial pacing is initiated at a rate faster than the supraventricular tachycardia to interrupt the re-entrant circuit.

Anesthetic Management

Deep sedation for cardioversion may be provided by intravenous midazolam, 0.1 mg/kg, and propofol, 1 to 2 mg/kg bolus. Patients presenting to the catheterization laboratory for treatment of supraventricular tachycardia, however, may require general anesthesia, especially if an ablation procedure is considered. Adequate premedication (midazolam, 0.05–0.08 mg/kg given intravenously) helps alleviate separation anxiety, reducing the chance of increased sympathetic activity at the time of induction. Premedication with atropine and glycopyrrolate should be avoided because of their vagolytic effects. Inhalational agents (e.g., isoflurane) that do not predispose to supraventricular excitement should be considered. Induction of anesthesia can be accomplished with thiopental, 1 to 4 mg/kg, or propofol, 2 mg/kg. Maintenance of anesthesia with propofol, 100 to 150 μg/kg/min, or isoflurane, 0.8% to 1.2%, can be supplemented with narcotics, such as fentanyl or alfentanil.

Complete Heart Block

Cause

Complete atrioventricular heart block may be congenital or acquired. Congenital complete atrioventricular heart block is seen in 1 in 22,000 live births. Congenital heart block may be associated with structural heart defects or with maternal collagen-vascular disease. It is speculated that maternal immunoglobins cross the placenta and damage the fetal cardiac conduction system. Acquired complete atrioventricular heart block may be secondary to nonsurgical causes, such as inflammatory or infectious diseases, or iatrogenic after cardiac surgery (especially after ventricular septal defect repair). Postsurgical complete atrioventricular heart block may be transient or permanent. Improved knowledge of the location of the conduction system has helped decrease the incidence of postsurgical heart block to less than 1%.

Clinical Presentation

All infants with congenital heart block present with bradycardia. The ventricular rate is usually less than 75 beats per minute and rarely more than 100 beats per minute while at rest. Although some patients may remain asymptomatic, other infants may present with signs and symptoms of congestive heart failure. Older children with congenital complete atrioventricular heart block tend to present with manifestations of ventricular arrhythmias and decreased cardiac output, such as syncope, diminished exercise tolerance, and fatigue. Patients may be evaluated with 24-hour Holter monitoring or echocardiography to assess the possibility of an associated congenital heart lesion or inadequate surgical palliation.

Anesthetic Management

Infants with complete atrioventricular heart block who present with severe congestive heart failure or shock should be intubated to optimize ventilation and oxygenation, and acidosis should be corrected. Patients who do not respond to this initial therapy with increased cardiac output need placement of a temporary pacemaker. Temporary pacing can be accomplished through transcutaneous, transesophageal, or transvenous routes. The subsequent implantation of a permanent pacemaker should be based on the severity of symptoms.

An infusion of isoproterenol, 0.02 to 0.5 μg/kg/min, or epinephrine, 0.05 to 0.5 μg/kg/min, may be necessary to provide pharmacologic
support while awaiting pacemaker placement. When providing general anesthesia for fluoroscopic-guided placement of transvenous pacemakers, the use of drugs (e.g., halothane, synthetic narcotics, vecuronium in combination with fentanyl or etomidate) that slow nodal pacemakers or myocardial conduction (directly by decreasing sympathetic tone or indirectly by increasing vagal tone) should be minimized or avoided.\textsuperscript{16} Temporary transvenous pacemakers should be left in place during the induction of anesthesia for permanent pacemaker implantation, because life-threatening arrhythmias can occur during this period.\textsuperscript{22} \textsuperscript{26}

Although isoproterenol and atropine increase heart rate by increasing the automaticity of the nodal pacemaker, they do not increase atrioventricular conduction. Atropine, however, may mitigate the effects of intraoperative vagal stimulation, and also should precede succinylcholine or neostigmine administration. Because cardiac output entirely depends on stroke volume in children with complete atrioventricular heart block, attention also must be given to maintaining intravascular volume.\textsuperscript{22}

CARDiac TAMponade

\textit{Cause}

Pericardial tamponade has been associated with various acquired disorders. It has been reported as a complication of cardiac tumors, hematologic malignancies, uremia, rheumatologic conditions, viral and bacterial infections, and autoimmune diseases. \textsuperscript{19} It also has been described as a complication of extracorporeal membrane oxygenation\textsuperscript{35} and positive pressure ventilation. \textsuperscript{18} As a complication of positive pressure ventilation, the tamponade was caused by pneumopericardium rather than by pericardial effusion. The anesthesiologist, however, is most likely to encounter patients presenting with cardiac tamponade from a few distinct causes, including blunt or penetrating chest trauma, acute myocardial perforation after cardiac catheterization procedures, myocardial perforation after central venous catheterization, and after cardiac surgery.

Myocardial perforation with acute tamponade occasionally occurs during interventional cardiac catheterization procedures.\textsuperscript{27} \textsuperscript{77} \textsuperscript{79} Hemopericardium resulting from ventricular puncture is usually self-limited, as the muscular ventricle seals the perforation after removal of the offending wire or catheter. Laceration of the thin-walled atrium, on the other hand, is more problematic, and may require surgical repair in the operating room.\textsuperscript{79}

Cardiac tamponade is a potential complication in the postoperative cardiac surgical patient. Tamponade occurring early in the postoperative course is usually secondary to continued bleeding, whereas tamponade occurring late in the postoperative course may be caused by postcardiotomy syndrome. The sudden cessation of chest tube drainage, with increased filling pressures and hypotension, may herald tamponade. Pericardial evacuation by facilitating chest tube drainage or reopening the sternotomy is required when cardiovascular stability is compromised. Aggressive circulatory support should be provided while awaiting definitive intervention.\textsuperscript{43}

Cardiac tamponade is also a rare but potentially fatal complication of central venous cannulation. The overall mortality rate is reported to be as high as 64\%.\textsuperscript{19} \textsuperscript{55} The diagnosis often is considered too late or not at all. Infants (especially younger than 6 months of age) seem to have a higher incidence of cardiac tamponade associated with central venous cannulation than adults, because the infant's thin-walled right atrium and right ventricle may be more susceptible to trauma.\textsuperscript{40} \textsuperscript{55} Premature infants seem to be predisposed particularly to this complication.\textsuperscript{19}

The primary cause of tamponade associated with central venous catheter use is related to the inappropriate placement of the catheter tip within the heart. \textsuperscript{4} In virtually all reported cases, the catheter tip was located in the right atrium or right ventricle. Perforation of the heart can occur during catheter insertion. Cardiac tamponade, however, typically is delayed, occurring more than 24 hours after central venous catheter insertion. It is speculated that the central venous catheter tip migrates into the right atrium or ventricle and lodges against the atrial or ventricular walls. Head and neck movements (especially flexion) or cephalad movements of the diaphragm and heart may cause the catheter tip to migrate into the right ventricle.\textsuperscript{40}

It is postulated that catheter-induced endocardial injury precipitates thrombus formation and adherence of the catheter to the endocardium. Progressive mural erosion then proceeds to perforation, perhaps enhanced by cardiac contractions.\textsuperscript{14} In cases in which perforation was not visualized on autopsy, it was hypothesized that tissue damage was caused by high-pressure infusions and hyperosmolar solutions at the site where the catheter became lodged against the heart wall. The subsequent transmural myocardial
necrosis resulted in the transudation of total parenteral nutrition, lipids, or other hypertonic solutions into the pericardial space.[9] [10] [19] [55] [69] [74]

Myocardial perforation and accumulation of pericardial effusions may be prevented by proper positioning of central venous catheters. Most authors agree that the tip of the catheter should lie in the superior vena cava, proximal to the pericardial reflection or outside the cardiac silhouette on the chest radiograph.[10] Leech et al.[40] recommend initially and intermittently transducing the distal lumen of the central venous catheter to identify catheter position and migration.

Other factors that may play a role in central venous catheter–induced cardiac tamponade include the stiffness of the catheter and the angle between the catheter tip and the vessel or myocardial wall.[10] [19] [40] [55] [74] Tamponade occurs more commonly with polyurethane than with silastic catheters.[10] [40] In vitro studies have demonstrated a higher incidence of tissue injury if the angle between the catheter tip and the vessel wall or myocardium approaches the perpendicular. Ideally, the catheter should be located in the long axis of the vein, which reduces the risk of lodging the tip in the vessel wall.[10]

Clinical Presentation

The clinical picture of cardiac tamponade results from an accumulation of fluid in the pericardial space that elevates intrapericardial pressure with subsequent impairment of diastolic filling of the heart, decreased stroke volume, and hypotension. If pressure in central veins exceeds right ventricular end-diastolic pressure, cardiac output and blood pressure are maintained. As the intrapericardial fluid pressure progressively increases and exceeds atrial pressure, equalization (at ~ 20 mm Hg) of left atrial and right ventricular end-diastolic filling pressures occurs. Without treatment, compensatory mechanisms ultimately may fail, causing profound hypotension and cardiovascular collapse.[66]

The ECG of a patient with cardiac tamponade may show electrical alternans (10%–15% of patients) or decreased voltage, caused by a short-circuiting effect of pericardial fluid. Evidence of myocardial ischemia also may be present if ventricular transmural pressure is elevated significantly to interfere with coronary perfusion. In addition to ECG changes, pulsus paradoxus also may be present. Pulsus paradoxus may reflect selective impairment of left ventricular filling during inspiration, but the exact mechanism is unclear.[43] [62] [66] [79]

A paradoxic pulse, however, is rarely seen in neonates because it is difficult to detect with rapid respiratory rates. Beck's triad, which consists of three classic findings in tamponade, hypotension, diminished heart sounds, and venous hypertension, also may not be evident until late in the course, if at all. The thin chest wall of the neonate allows heart sounds to be transmitted well even in the presence of a large pericardial effusion. The neonatal heart also can generate high heart rates, and can function normally at low filling pressures, which helps the neonatal circulation to compensate for advanced degrees of tamponade. Arterial pressure may be maintained until compensatory mechanisms are overwhelmed, resulting in sudden cardiovascular collapse. Straightening of the left heart border on chest radiograph is a useful finding. A progressively enlarging globular heart also is a reliable indicator of pericardial effusion, but this is characteristically a late finding. Echocardiography remains the definitive method for diagnosing and evaluating a pericardial effusion. Echocardiographic signs of diastolic collapse of atria and ventricles are considered pathognomonic for cardiac tamponade.[50] [64]

Anesthetic Management

When tamponade compromises cardiovascular stability, emergent pericardiocentesis is imperative. Percutaneous subxiphoid pericardiocentesis performed with local anesthesia is the most commonly used surgical method of treatment for cardiac tamponade. Echocardiography is useful in guiding the needle into the pericardial space. Drainage of small amounts of pericardial fluid often results in dramatic hemodynamic improvement. When cardiac tamponade results from trauma or develops after cardiac surgery, the recommended treatment is a pericardiotomy performed in the operating room under local or general anesthesia.[66]

Attempting percutaneous pericardiocentesis in a hemodynamically unstable, frightened, and combative child presents an anesthetic challenge. Anesthetic management should be tailored to maintain or to improve ventricular filling pressures, contractility, and cardiac output. Maintaining adequate preload with expansion of intravascular fluid volume; administering a β-agonist, such as isoproterenol, to maintain heart rate and contractility; and correcting any underlying metabolic acidosis are helpful temporizing measures. Metabolic acidosis resulting from low cardiac output should be treated with sodium bicarbonate, because an increased hydrogen ion concentration can depress myocardial contractility and attenuate the positive inotropic effects of catecholamines. Bradycardia resulting from vagal reflexes as intrapericardial pressures increase should be treated with atropine, because a slow heart rate is poorly tolerated in...
tamponade. Extreme tachycardia, however, should be avoided because it may not allow adequate time for diastolic filling and systolic ejection. Alpha-Agonists, such as phenylephrine and norepinephrine, should be used with caution in cardiac tamponade because of the depressant effects of increased afterload on myocardial contractility. Anesthetic agents used for sedation that excessively decrease preload or afterload and depress myocardial function can lead to life-threatening hypotension when combined with muscle paralysis and positive pressure ventilation, which further impairs ventricular filling. Rapid-sequence induction of anesthesia with high-dose pentothal, succinylcholine, and intubation of the trachea followed by positive pressure ventilation may be undesirable. Although not always possible, the preferred initial approach to the hemodynamically compromised patient is to perform pericardiocentesis under local anesthesia. Low doses of a narcotic, benzodiazepine sedation, and soft restraints of the child's upper extremities may be used while allowing the patient to breathe oxygen spontaneously during the procedure. Intravenously administered ketamine may be useful if adequate sedation cannot be achieved by using narcotics or benzodiazepines without significant respiratory depression. When general anesthesia is unavoidable, the goal of management is to maintain cardiac output. Bradycardia must be avoided, especially in infants who naturally have a high degree of vagal tone, in addition to the depressor vagal reflex present in tamponade.

CONGESTIVE HEART FAILURE

Cause

Heart failure in adults is usually secondary to rheumatic, arteriosclerotic, or hypertensive disease. In contrast, heart failure in the pediatric population is usually the result of decompensated congenital heart disease. Less frequently, it is secondary to myocarditis, cardiomyopathy, arrhythmia, respiratory disease, severe anemia, systemic or pulmonary hypertension, septicemia, cardiac tumors, central nervous system disease, storage diseases, and various metabolic disorders. In the postsurgical patient, heart failure may be caused by myocardial injury and subsequent ventricular dysfunction, arrhythmia, tamponade, excessive transfusion of fluids (resulting in failure of mechanisms regulating intravascular volume rather than failure of the heart as a pump), pulmonary hypertension, or malfunctioning prosthetic valves. Appropriate treatment relies on determining whether heart failure is secondary to a primary structural cardiac defect or to some other cause. The congenital cardiac defects that most commonly are associated with congestive heart failure are listed in order of frequency as follows:

- Transposition of the great arteries
- Coarctation of the aorta
- Ventricular septal defect
- Aortic atresia
- Common atrioventricular canal
- Transposition of pulmonary veins
- Single ventricle
- Patent ductus arteriosus

Ninety percent of the overall cases of congestive heart failure in children occur before the end of the first year of life. The time of onset of symptoms of congestive heart failure may help delineate the cause. Congestive heart failure presenting within the first few hours of life may be caused by volume overloading of the right heart (secondary to tricuspid or pulmonary regurgitation, a large systemic arteriovenous fistula, a large placental transfusion, or anemia), neonatal asphyxia (with sequelaes of acidosis and hypoxemia), or airway obstruction. Infants presenting with congestive heart failure in the first week of life are likely to have
lesions with obstruction of the left ventricular outflow tract. Infants presenting within the first several months of life may have lesions with left-to-right shunts. The onset of symptoms depends on the decrease in pulmonary vascular resistance that occurs gradually after birth. Children presenting later in life should be suspected of having acquired rather than congenital heart disease.\textsuperscript{21, 24}

**Clinical Presentation**

Common manifestations of congestive heart failure in the neonate in the approximate order of frequency include tachypnea, tachycardia, liver enlargement, cardiomegaly, pulmonary rales and rhonchi, and feeding difficulties. Less common signs and symptoms include peripheral edema, measurable elevated systemic venous pressure, inappropriate diaphoresis at normal room temperature, gallop rhythm, pulsus alternans, and ascites. Pleural and pericardial effusions caused by congestive heart failure are rare in infants.\textsuperscript{8, 41, 61} \([70, 71]\)

Because manifestations of heart failure and primary pulmonary diseases may be similar in infants, recognizing congestive heart failure can be difficult. Frequently, classic findings such as murmur are absent in infants with congestive heart failure. A murmur, when present, may be obscured by noisy respirations. Cyanosis or auscultation of rales and rhonchi may not be especially helpful because they may be present with cardiac or pulmonary disease. The administration of oxygen to ascertain improvement in arterial oxygen saturation does not always allow a differentiation between cardiac and pulmonary disease. Although rises in arterial oxygen tension in excess of 25 to 50 mmHg generally indicate a pulmonary contribution to hypoxemia, infants with severe pulmonary disease may not show a significant improvement in color or oxygen saturation. The presence of hepatomegaly and gross cardiomegaly, however, usually indicates heart failure.\textsuperscript{41} Echocardiography is the most accurate way to confirm the diagnosis of congestive heart failure in infants.\textsuperscript{61}

**Medical Management**

The therapeutic principles of treating congestive heart failure in adults also apply to treating infants and children.\textsuperscript{51, 61} Medical management of congestive heart failure is directed toward improving cardiac performance, augmenting peripheral perfusion, and decreasing systemic and pulmonary venous congestion. Aggressive measures should be taken to minimize the workload on the heart and to avoid unnecessary demands for increased cardiac output.\textsuperscript{23, 41, 70, 71}

Inotropic support frequently is required. Although digoxin is the mainstay of medical management in children, it requires some time to reach therapeutic serum levels and is not useful in an acute setting. Dopamine and dobutamine are more appropriate inotropes in the immediate management of congestive heart failure.\textsuperscript{60} Both drugs should be administered as continuous intravenous infusions at 5 to 20 \(\mu\)g/kg/min. Dobutamine may offer an advantage over dopamine because it has less arrhythmogenic potential and it reduces afterload because of its vasodilatory effect.\textsuperscript{26, 60} There is some concern about the efficacy of dobutamine in infants less than 1 year of age. In this population, dobutamine may improve cardiac output without a concomitant rise in blood pressure. If severe hypotension is present in a young infant, dobutamine may be more appropriate as an adjunct rather than as the primary inotrope.\textsuperscript{26}

The nonsympathomimetic bipyridine derivatives, amrinone and milrinone, also may be useful inotropic adjuncts in the treatment of congestive heart failure. These drugs have inotropic and vasodilator actions without increasing heart rate. They inhibit cyclic adenosine monophosphate (cAMP) breakdown by phosphodiesterase III in cardiac and vascular smooth-muscle cells. The elevated levels of cAMP result in positive inotropy in cardiac muscle and vasodilation in vascular smooth muscle. The pulmonary vasodilating effects of these drugs may be particularly useful in the setting of pulmonary hypertension or right heart failure.\textsuperscript{26, 37}

In patients who do not present with a rapidly deteriorating course, digitalis and diuretics remain the primary mode of therapy.\textsuperscript{21, 24, 26, 61, 71} The two primary indications for use of digitalis in infants and children are congestive heart failure and paroxysmal tachycardia of supraventricular origin.\textsuperscript{24, 26, 52} Digoxin is the most widely used digitalis glycoside. It enhances ventricular ejection by increasing the force of cardiac contractions, which results in increased cardiac output and decreased intracardiac filling pressures, cardiac size, and heart rate.\textsuperscript{26} Diuretic therapy also plays an important role in the management of congestive heart failure by promoting diuresis and natriuresis. Furosemide may be given intravenously as a bolus dose of 1 mg/kg initially, with onset in 5 minutes and a peak at 30 minutes.\textsuperscript{24}

**Anesthetic Management**

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Medical management alone is frequently insufficient to permanently control the deleterious effects of the abnormal loads imposed on the circulation by certain congenital cardiac defects permanently, particularly when severe obstruction to left or right ventricular outflow is present.\(^23\)\(^41\)\(^70\) When clinical evidence suggests that a structural cardiac defect is the cause of heart failure in an infant, diagnostic cardiac catheterization and angiocardiography are indicated. Continuing deterioration during the first 12 to 24 hours of medical management usually indicates immediate cardiac catheterization. A definitive anatomic and functional diagnosis helps guide further therapy towards surgical intervention versus continued or modified medical management.\(^23\)\(^41\)

Anesthetic management should be directed toward continued inotropic support with avoidance of anesthetic agents that cause direct myocardial depression. Newborn infants should be maintained in a neutral thermal environment to minimize their overall metabolic and circulatory requirements. Endotracheal intubation and ventilatory support with positive end-expiratory pressure are indicated in cases of severe pulmonary edema and respiratory failure.\(^24\)\(^41\)\(^61\)\(^70\) Pharmacologic agents that decrease pulmonary vascular resistance and enhance inotropy may be particularly useful (e.g., amrinone, 0.75 mg/kg loading dose followed by an infusion of 5 to 20 μg/kg/min, or milrinone, 37.5 to 50.0 μg/kg loading dose followed by an infusion of 0.375–0.75 μg/kg/min).\(^37\)

SUMMARY

Successful management of pediatric cardiac emergencies requires an accurate diagnosis to institute an appropriate plan of therapy. The diagnosis, however, is not always straightforward, as evidenced by the nonspecific clinical picture that can be presented by congenital heart defects. Entertaining the possibility of a cardiac problem in neonates with pulmonary symptoms unresponsive to standard therapies is crucial for successful management of patients with congenital heart disease. In addition to ventilatory support, prostaglandin \(E_1\) infusions or emergency interventional cardiac catheterization is often a life-saving initial measure in patients with acutely decompensated congenital cardiac lesions that require a patent ductus arteriosus for survival.

Pericardial tamponade is associated with various acquired and iatrogenic causes. Emergent pericardiocentesis is mandatory when cardiovascular compromise occurs. The goal of anesthetic management is to maintain cardiac output. With the increasing use of central venous catheters in neonatal ICUs and the high mortality rate for central venous catheter–related cardiac tamponade, the diagnosis must be considered in any patient with a central venous catheter in situ who acutely develops unexplained hypotension, bradycardia, and diminished pulses.

Arrhythmias also can cause hemodynamic instability in infants and children. Supraventricular tachycardia is by far the most common emergently presenting arrhythmia in the pediatric population. Unstable patients require immediate intravenous adenosine or synchronized cardioversion. Complete heart block is rare, but it can lead to congestive heart failure and occasionally to cardiovascular collapse and sudden death. Emergency treatment of complete heart block includes pharmacologic support and temporary or permanent pacemaker placement as indicated. In infants, congestive heart failure usually is related to congenital heart disease, whereas in older children, it tends to be secondary to an acquired cause. Supportive measures, fluid restriction, and inotropic support are the principles of initial treatment. Prompt recognition and initiation of appropriate therapy in pediatric cardiac emergencies are essential for favorable outcomes.

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