

Cardiac Emergencies

1. Congenital Heart Disease (CHD)

Background:

- Congenital heart disease (CHD): Prevalence ranges from 6 to 13 per 1,000 live births.
- Cyanotic heart lesions account for approximately 15% of all cases of CHD.
- Cyanotic heart lesions can be categorized physiologically:
 - Decreased pulmonary blood flow:
 - Tetralogy of Fallot, tricuspid valve anomalies, pulmonary valve atresia.
 - Increased pulmonary blood flow:
 - D-transposition of the great arteries, truncus arteriosus, total anomalous pulmonary venous connection.
 - Heart failure:
 - Left-sided obstructive lesions including hypoplastic left heart syndrome, severe coarctation of the aorta, interrupted aortic arch, critical valvar aortic stenosis.
- Most newborns with cyanotic heart disease are discovered while in utero or in the newborn nursery, but some infants may present to the Emergency Department or Primary Care office.
- Infants with congenital heart lesions dependent on a patent ductus arteriosus for pulmonary blood flow may present in the first two weeks of life with profound cyanosis when the ductus closes.
- Early recognition, stabilization, and transport to a pediatric congenital heart center are essential to optimize patient outcomes.

Signs and Symptoms:

- History may include tachypnea, respiratory distress, poor feeding, decreased urine output, cyanosis.
- Cyanosis results when the absolute level of reduced hemoglobin in the capillary bed exceeds 3 gm/dL, which corresponds to an oxygen saturation level less than 85% in a patient with a hemoglobin of 15 gm/dL.
 - Peripheral cyanosis: Secondary to increased oxygen extraction resulting in an increased concentration of reduced hemoglobin on the venous side of the capillary bed.
 - Acrocyanosis of the newborn results from benign vasomotor changes and may cause cyanosis of the hands, feet, and mouth.
 - Central cyanosis: Pathologic condition secondary to systemic arterial oxygen desaturation.
- Fever or hypothermia may indicate an infectious etiology.

Evaluation:

- Physical exam: General appearance, vital signs, cardiovascular status (murmur, gallop, central and distal pulses in all extremities), respiratory status (tachypnea, increased work of breathing), neurologic status, hepatomegaly (heart failure).
- Pulse oximetry should be measured from a preductal (right hand) and postductal site (right or left foot).
- Blood pressure gradient between the upper and lower extremities may indicate severe coarctation of the aorta or interrupted aortic arch and a closed ductus arteriosus.
- Severe respiratory distress may indicate a congenital heart lesion with increased pulmonary blood flow or left-sided obstructive heart lesion resulting in heart failure.
- Hyperoxia test and arterial blood gas:
 - Utilize pulse-oximetry on the right hand unless a skilled proceduralist is available to perform arterial punctures.
 - Obtain arterial blood gas from right radial artery.
 - Provide 100% oxygen via non-rebreather for 10 minutes then obtain a second arterial blood gas from the right radial artery.
 - A partial pressure of arterial oxygen above 150 mmHg indicates probable pulmonary disease, while a value less than 150 mmHg makes cyanotic CHD more likely.

- An increase in oxygen saturations indicates probably pulmonary disease, while no or minimal increase in oxygen saturations makes cyanotic CHD more likely.
- CXR (cardiomegaly in heart failure, increased or decreased pulmonary vascular markings).
- ECG, ECHO.
- CBC, CRP, electrolytes, urinalysis, blood and urine cultures.

Management:

- Consult Pediatric Cardiology and Pediatric Critical Care.
- Utilize Pediatric Advanced Life Support (PALS) guidelines.
- See Supplemental Oxygen & Airway Management section in Preparing to Transport.
- Obtain vascular access: See Vascular Access section in Preparing to Transport.
- For shock: See Sepsis and Septic Shock section in Infectious Emergencies.
- For suspected infection: See Sepsis and Septic Shock section in Infectious Emergencies.
- For suspected ductal-dependent CHD:
 - Prostaglandin E1 infusion (0.05 mcg/kg/min; discuss dose with Pediatric Cardiology).
 - Apnea occurs in 10% of patients. Be prepared for endotracheal intubation.

2. Systemic-to-Pulmonary Artery Shunt Obstruction

Background:

- A number of cyanotic heart lesions (hypoplastic left heart syndrome, pulmonary atresia) are palliated through the use of a systemic-to-pulmonary artery shunt to improve pulmonary blood flow.
- The systemic-to-pulmonary artery shunt (modified Blalock-Taussig shunt, central shunt) connects the aorta or its branches to the pulmonary artery and is noted on auscultation as a continuous murmur.
- Occlusion of a systemic-to-pulmonary artery shunt is a life-threatening event and requires emergent medical care.

Signs and Symptoms:

- Acute onset of cyanosis, respiratory distress, shock.

Evaluation:

- Physical exam: General appearance (cyanosis), vital signs, cardiovascular status (murmur, gallop, central and distal pulses in all extremities), respiratory status (tachypnea, increased work of breathing), neurologic status, hepatomegaly (heart failure).
- Acute onset of cyanosis and absence of a continuous murmur noted on auscultation should trigger concern for a shunt obstruction.

Management:

- Consult Pediatric Cardiology and Pediatric Critical Care and coordinate emergent transfer.
 - Discuss use of systemic heparin and epinephrine with Pediatric Cardiology.
- Utilize Pediatric Advanced Life Support (PALS) guidelines.
- See Supplemental Oxygen & Airway Management section in Preparing to Transport.
- Obtain vascular access: See Vascular Access section in Preparing to Transport.
- For shock: See Sepsis and Septic Shock section in Infectious Emergencies.

3. Hypercyanotic (“Tet”) Spells

Background:

- Tetralogy of fallot accounts for approximately 10% of all cases of CHD and is one of the most common cyanotic heart lesions.

Signs and Symptoms:

- Presentation to medical care depends on degree of right ventricular outflow tract (RVOT) obstruction:
 - Severe RVOT obstruction with inadequate pulmonary blood flow: Present in the immediate newborn period with severe cyanosis.
 - Moderate RVOT obstruction with balanced systemic and pulmonary blood flow: Most often present for elective evaluation of a murmur, but may also present with hypercyanotic (“tet”) spells when the RVOT is obstructed during periods of agitation.
 - Mild RVOT obstruction: Present with increased pulmonary blood flow and heart failure.

Evaluation:

- Physical exam: General appearance (cyanosis), vital signs, cardiovascular status (murmur, gallop, central and distal pulses in all extremities), respiratory status (tachypnea, increased work of breathing), neurologic status, hepatomegaly (heart failure).

Management:

- Consult Pediatric Cardiology.
- For a hypercyanotic (“tet”) spell:
 - Place patient in knee-to-chest position to increase systemic vascular resistance.
 - Provide supplemental oxygen (pulmonary vasodilator).
 - If cyanosis persists:
 - Morphine (0.1 mg/kg/dose; max 2 mg/dose) IV once and 0.9% NS (20 mL/kg) IV bolus once over 20 minutes.
 - Propranolol (0.1 mg/kg/dose; max 5mg/dose) IV once over 10 minutes.
 - Phenylephrine (0.01 mg/kg/dose) IV once.

4. Postoperative Congenital Heart Disease

- Consult Pediatric Cardiology.
- Verify type of congenital heart disease, prior surgical procedures, “normal baseline” oxygen saturations, and recent ECHO findings including cardiac function.
- Children with CHD may have variable “normal baseline” oxygen saturations following surgical repair:
 - Parents often know their child’s “normal baseline” oxygen saturation level.
 - Expected oxygen saturations for patients with hypoplastic left heart syndrome:
 - Status post Norwood repair= 75-80%
 - Status post Glenn repair= 80-85%
 - Status post Fontan repair= 90-95%
- Patients may present for medical care with infection (pneumonia, sepsis), acute heart failure and/or volume overload, pericardial effusion (post-pericardiotomy syndrome), pleural effusion, wound infection, dysrhythmias, dehydration, electrolyte abnormalities, shock.
- If febrile or ill appearing: See Sepsis and Septic Shock section in Infectious Emergencies.
- Obtain ECG in any patient with history of cardiac disease or intervention.