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 5 mg/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.