

connection that usually closes at the time of birth. If the duct closes in a child with a duct-dependent cardiac lesion, there is no mixing of blood between the right and left heart, and the systemic and pulmonary circulations function independently of one another. This means that the left ventricle is pumping deoxygenated blood, which will rapidly lead to tissue ischemia and shock.

Lesions in this category include the 5 Ts: Tetralogy of Fallot with pulmonic stenosis, transposition of the great vessels (where aorta and pulmonary artery are attached to the wrong ventricles), tricuspid atresia, total anomalous pulmonary venous return (the pulmonary veins drain into the right atrium, coronary sinus, the superior vena cava, or inferior vena cava rather than the left atrium), truncus arteriosus (a single arterial trunk leaves the heart and forms the pulmonary, systemic, and coronary circulation), and hypoplastic left heart, severe aortic stenosis, and some cases of coarctation of the aorta. If the ductus arteriosus closes, there is immediate intense cyanosis and circulatory collapse.

Closure of the ductus arteriosus in an infant with a duct-dependent lesion is a true medical emergency. Treatment involves administration of a continuous infusion of the hormone prostaglandin E₁ (PGE₁). PGE₁ dilates the ductus arteriosus and allows temporary stabilization pending surgical correction. Because these situations are rare and PGE₁ infusion is commonly associated with apnea, hyperthermia, and seizures, this is not a medication that is carried by EMS units. These patients need rapid transfer to an emergency department that is capable of providing PGE₁ and preferably a center that has pediatric cardiac surgery capability.

4. How do we separate a hypercyanotic Tet spell from cyanosis associated with closure of a patent ductus arteriosus?

Hypercyanotic spells are rarely associated with circulatory collapse and loss of peripheral pulses. They may occur in a child with unrepaired Tetralogy of Fallot of any age.

Circulatory collapse of a child with a ductal-dependent lesion usually occurs in a neonate who has not yet been diagnosed with congenital heart disease. When the ductus arteriosus closes in such infants, the child is not only cyanotic, but also in shock.

5. How can you recognize cyanosis in dark-skinned children?

Observe for central cyanosis by looking at the lips and oral mucous membranes.

6. What is acrocyanosis?

Newborn infants and infants in the first few months of life may have vasomotor instability that results in acrocyanosis (cyanosis of the hands and feet only) when the child is cold. This is a normal phenomenon, not to be confused with central cyanosis that always reflects hypoxemia.

7. Does the hemoglobin concentration of the blood affect the presence or absence of cyanosis?

It is necessary to have at least 5 g/dL of desaturated hemoglobin in order for cyanosis to be noted in the skin. The presence of anemia reduces apparent cyanosis, and children with severe anemia (less than 5 g/dL) will have no cyanosis evident. Conversely, polycythemia (abnormally high serum hemoglobin levels) increases apparent cyanosis. Children with cyanotic congenital heart disease are often polycythemic and may appear blue even when at their baseline.

8. When is oxygen risky to give patients with heart disease?

Excessive amounts of oxygen have the potential for dilating pulmonary vessels and decreasing shunting. If there is left heart obstruction, oxygen should be administered very cautiously. However, in the prehospital setting with little diagnostic information and relatively short transport times, err on the side of oxygen administration if the child's oxygen saturation is below their baseline.

9. If this patient had history of repaired Tetralogy, as evidenced by a sternal scar, what kind of complications might occur?

Any child with congenital heart disease who has had a repair performed can be divided into two categories:

- a. Corrections with ventriculotomy: If the ventricular myocardium has been opened with a surgeon's knife, there is always a risk of dysrhythmias because of interruption of the normal conduction pathways. Right bundle branch block rhythms are common, and PVCs and A-V blocks can occur. Sudden death from ventricular dysrhythmias (ventricular tachycardia or ventricular fibrillation) can occur.
- b. Complications of temporizing shunts (artificial conduits that are placed to improve blood flow between the right and left heart circulation) can be
 - Too large, resulting in increased pulmonary blood flow and congestive heart failure
 - Too small, with inadequate pulmonary blood flow and cyanosis
 - Result in clot formation, with embolism and stroke

Children with cyanotic heart disease are at risk for embolic phenomenon. Whenever there is acute alteration in neurologic status or focal neurologic findings (hemiparesis, hemiplegia), a stroke should be considered. They may also be at risk for congestive heart failure, presenting with increased work of breathing, inspiratory crackles on auscultation, and hepatomegaly.