In a child with Tetralogy of Fallot, there is obstruction to right ventricular outflow, and anything that causes a decrease in a systemic vascular resistance may precipitate a hypercyanotic spell. The hypercyanotic spell is caused by right to left shunting of blood across the ventricular septal defect (VSD) when systemic vascular resistance is decreased or pulmonary resistance is increased with deoxygenated blood that has not passed through the pulmonary circulation being pumped into the systemic circulation. This results in profound hypoxemia, which manifests as cyanosis, irritability, increased respiratory rate without respiratory distress, a decrease in the intensity of the child's heart murmur, increased tissue oxygen consumption, and acidemia. Increased systemic venous return further increases right-sided heart pressure and right to left shunting of blood, and the acidemia results in reflex tachypnea, thereby creating a vicious cycle.

A decrease in systemic vascular resistance resulting in a "Tet" spell often occurs during or after feeding or defecation, when the infant begins crying, or early in the morning after awakening. Management of a hypercyanotic spell involves efforts to break the vicious cycle.

FACULTY NOTES: Read Out Loud

Question #5: What are further management priorities?

Answers:

Management of a Hypercyanotic Spell

BLS treatment priorities

- Avoid further agitation.
- Provide oxygen, although this is unlikely to improve oxygen saturation in this setting.
- Have the caregiver pick the baby up and hold him in a knee-chest position on her shoulder or place the infant on his abdomen in the knee-chest position. This simple maneuver will sometimes terminate a hypercyanotic spell.
- Transport with frequent reassessments.

ALS

ALS treatment priorities

- The risks of prolonged scene time and worsening the child's agitation and hypoxia must be weighed against potential benefits of ALS interventions.
- Attempt vascular access and give a cautious 10-mL/kg bolus of normal saline. Isotonic fluids are given to increase systemic vascular resistance.
- With long transport times, morphine may be considered to reduce agitation, decrease tachypnea, and improve hemodynamics; 0.1 mg/kg can be administered IV/intramuscularly/subcutaneously.

FACULTY NOTES: Read Out Loud

Question #6: Does this case raise any special issues?

Answers:

1. How can you separate cardiac from pulmonary disease in an out-of-hospital setting?
   This may be difficult to do without blood gas analysis. A rough "hyperoxia test" can be performed by giving 100% oxygen and watching for a significant improvement in oxygen saturation. If oxygen saturation does not improve significantly with the administration of supplemental O_2, right to left intracardiac shunting is likely.

   Cyanosis without respiratory distress should always suggest a cardiac etiology. If an infant becomes dusky or cyanotic or has increased work of breathing with feeding, a cardiac cause should be considered. The absence of a heart murmur does not mean the absence of heart disease.
2. How common is Tetralogy of Fallot, and at what age is it diagnosed?
   Recall that the four anatomic abnormalities that make up the tetralogy are ventricular septal defect, pulmonic stenosis, over riding of the aorta, and right ventricular hypertrophy.

   Tetralogy of Fallot is the most common cyanotic congenital heart disease in children beyond infancy. Although many present with cyanosis in the newborn period, the severity depends on the degree of right ventricular outflow obstruction, and many patients may go undiagnosed for several months.
3. What is a "duct-dependent" (or ductal-dependent) lesion?
   There are a number of causes of congenital heart disease in infants where mixing of blood between the pulmonary and systemic circulation is critically dependent on a patent ductus arteriosis, an embryonic