Pediatric Update

The case of the blue baby: ED management of tetralogy of Fallot

Author: Scott DeBoer, RN, BSN, CEN, CCRN, CFRN, Dyer, Indiana

Nothing in the ED setting seems to cause as much anxiety among even the most experienced nurses as a critically ill child. We are now accustomed tc "common" conditions such as asthma, dehydration, and trauma, but times are changing. We are seeing more infants and children with chronic diseases, children whose health is maintained by technologic assistance, and infants with severe birth defects, some of which may be unrecognized. In the past the expected length of hospital stay after a normal vaginal delivery was up to 1 week. Now some mothers and babies are discharged only 6 hours after delivery.

It is amazing that most children progress through the complex pulmonary and hemodynamic changes that occur after delivery so smoothly and uneventfully,[1] and that most infants make the transition from a liquid environment to air without difficulty. Congenital heart disease, which affects approximately 1 in every 100 infants,[2] may not manifest itself until after the first few hours or even months of life. Even when congenital heart disease is recognized, it may not be treated immediately. Children with an initial diagnosis of tetralogy of Fallot at birth do not always undergo surgery immediately; because of their small size, waiting may provide a better chance for survival. In addition, other factors include the physiologic stability of the child's condition, size of the pulmonary arteries, and the possible presence of other complicating defects. The optimal age for elective repair is 3 months to 3 years because of the maturation and development of the pulmonary arteries and associated structures. Children with tetralogy, even if their condition is diagnosed in infancy, are occasionally sent home (with detailed parental instructions) to grow before definitive repair of their defect is performed; therefore they may be brought to the emergency department.

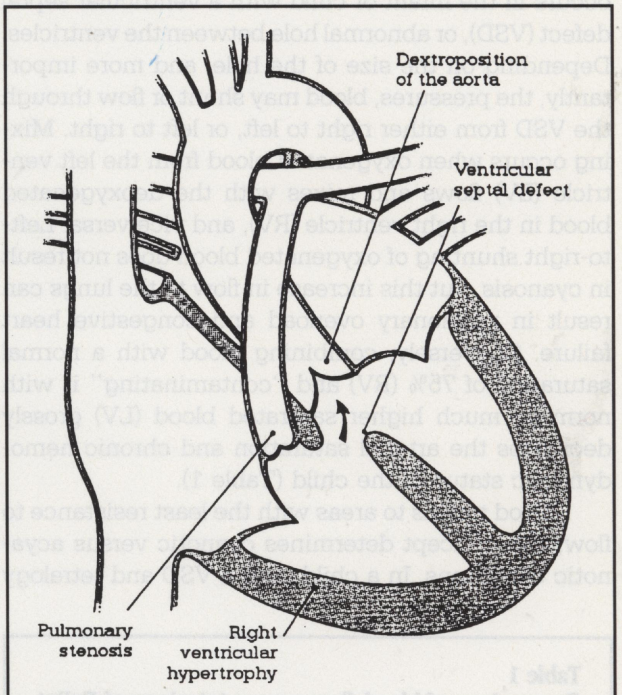

Figure 1
Tetralogy of Fallot. From Hazinski MF. The cardiovascular system. In: Howe J, et al., eds. The handbook of nursing, Copyright © 1984, John Wiley & Sons, Inc. Reprinted by permission of John Wiley & Sons, Inc.

Pathophysiology of tetralogy of Fallot

A review of normal adult circulation and assimilation of two key concepts are needed to understand the pathophysiology and treatment of tetralogy of Fallot, and associated "tet spells." In the adult circulation, deoxygenated blood flows from the right side of the heart to the lungs, where blood is oxygenated, and waste products are released. Oxygenated blood then returns to the left structures of the heart through the

Mr. DeBoer is a flight nurse, UCAN/University of Chicago Hospitals, Chicago, Illinois.

For reprints, write Scott DeBoer, RN, BSN, CEN, CCRN, CFRN, 2301 Flint Court, Dyer, IN 46311-1913.

J EMERG NURS 1996;22:73-6

0099-1767/96 $5.00 + 0 **18/9/71105**

pulmonary veins, and is then ejected to perfuse the body through the aorta. When defects or problems with either the "pump or the plumbing" occur, the concepts of pressure and mixing are involved.

Simply stated, blood and other fluids follow the path of least resistance. Increased pressure or resistance to flow results in reduced flow, just as less water can flow through a garden hose than a drain pipe. In addition, if increased pressure is encountered, and a hole or other pathway is present between two areas, fluids again flow through the area with the least pressure and resistance.

Mixing of oxygenated and deoxygenated blood occurs in the infant or child with a ventricular septal defect (VSD), or abnormal hole between the ventricles. Depending on the size of the hole, and more importantly, the pressures, blood may shunt or flow through the VSD from either right to left, or left to right. Mixing occurs when oxygenated blood from the left ventricle (LV) flows and mixes with the deoxygenated blood in the right ventricle (RV), and vice versa. Left-to-right shunting of oxygenated blood does not result in cyanosis, but this increase in flow to the lungs can result in pulmonary overload and congestive heart failure. Conversely, combining blood with a normal saturation of 75% (RV) and "contaminating" it with normally much higher saturated blood (LV) grossly decreases the arterial saturation and chronic hemodynamic status of the child (Table 1).

Blood travels to areas with the least resistance to flow; this concept determines cyanotic versus acyanotic conditions. In a child with a VSD and tetralogy of Fallot, the most common type of cyanotic congenital heart disease (2 occurrences per 10,000 live births), both abnormal holes and pressures occur. If the left (aortic) pressures are higher than the pulmonary arterial pressure (this is normal), left-to-right shunting occurs. If the right ventricular pressures are higher than the left, as with pulmonary hypertension or stenosis, blood shunts in the reverse direction and cyanosis/systemic desaturation occurs.

Table 1
Comparison of blood flow versus tetralogy of Fallot

Cardiac shunting	Tetralogy of Fallot
Right to left	VSD
Increased pulmonary resistance	"Hole" in ventricular septum
Desaturated blood from RV to LV	Pulmonary stenosis
Systemic desaturation and acute cyanosis	Tight area of pulmonary valve or tissue
Left to right	Right ventricular hypertrophy
Normal with VSD	Ventricular enlargement as result of stenosis
Saturated blood from LV to RV	Overriding aorta
Increased pulmonary flow	Aorta sits on top of VSD and receives mixed blood from both ventricles
Possible CHF	
Acyanotic unless severe CHF	

CHF, Congestive heart failure; *LV*, left ventricle; *RV*, right ventricle; *VSD*, ventricular septal defect.

Tetralogy of Fallot components (Figure 1)

Tetralogy of Fallot involves four distinct defects: (1) VSD, (2) pulmonary stenosis (valve or arterial tissue) (3) RV hypertrophy as a result of pulmonary stenosis, and (4) an overriding aorta (directly on top of the VSD).

Ventricular septal defect

An opening in the ventricular septum allows communication between the ventricles and mixing of oxygen-enriched and desaturated venous blood. Left-to-right shunting results in increased pulmonary flow, but no decrease in systemic saturation unless CHF or symptoms of cor pulmonale are present. Right-to-left shunting, as is most common with tetralogy, results in right ventricular blood flowing to the left ventricle, and systemic desaturation and acute/chronic hypoxemia.

Pulmonary stenosis

The stenosis or "tight area" in the right ventricular outflow tract (pulmonary artery or valve) results in decreased blood flow to the lungs. This results in right ventricular hypertrophy and contributes to cyanosis and right-to-left shunting.

Right ventricular hypertrophy (RVH)

RVH occurs as a result of increased pulmonary vascular resistance (PVR) and obstruction to flow from the ventricle as a result of pulmonary stenosis. The key

Table 2
Management of tetralogy of Fallot

Tet spell signs and symptoms	Management
Hypercyanosis	Decrease pulmonary resistance
Irritability	Supplemental O_2
Tachypnea	Calming measures
Metabolic acidosis	IV morphine
Diaphoresis	IV propranolol
Poor peripheral perfusion	Increase systemic resistance
	Isotonic fluid boluses (20 ml/kg) LR or 0.9NS
	Dopamine or Neo-Synephrine
	Knee-chest position

concept to remember is Starling's law, in which the heart muscle stretches much like a rubber band. The band stretches and results in a higher length of "flight," but only to a point, when the rubber band, like the ventricle, breaks and hemodynamic instability may occur.

Overriding aorta

The aorta sits directly on the VSD, instead of the left ventricle alone, so that mixed right and left ventricular blood (with low oxygen saturation), flows into the systemic circulation. This defect, combined with the VSD and pulmonary stenosis, contributes to systemic desaturation and hypoxemia.

Initial signs and symptoms

When children with tetralogy come to the emergency department, the initial signs and symptoms can be vague and easily missed (Tables 2 and 3). At birth there may only be a mild heart murmur and no other dramatic signs. Typically, if a heart murmur persists after birth, an echocardiogram is performed before discharge, and the majority of patients with tetralogy have a diagnosis made and their condition is treated. However, manifestations depend on the size of the VSD (mixing ability) and the degree of stenosis (how much blood reaches the lungs). The classic sign of a "tet spell" is hypercyanosis caused by an acute increase in PVR, or a decrease in oxygen availability versus demand. If the PVR is greater than the systemic vascular resistance (SVR), or arterial blood pressure, less blood reaches the lungs, and more deoxygenated blood flows through the VSD from the right ventricle to the left. This increase in PVR may be caused by crying, agitation, feeding, or bowel movements. All of these activities increase the PVR and oxygen demand. The concept of supply and demand is relevant to this crisis. Children with unrepaired tetralogy defects have a relatively fixed degree of obstruction; the pulmonary obstruction does not grow proportionately as the child does. As the child requires more oxygen, and less and less is available, cyanosis is exhibited. When the activities that increase pulmonary resistance or oxygen demand decrease, the systemic pressure becomes higher than the pulmonary resistance, and blood flow to the lungs is increased with less detrimental shunting. Undiagnosed or unrepaired tetralogy of Fallot, as described earlier, is rare. Other more common reasons for cyanosis and distress, such as pneumonia and sepsis, should *always* be considered much higher in the differential diagnosis of cyanosis.

When too little blood reaches the lungs for oxygenation and the mixing from shunting results in hypoxemia (acute superimposed on chronic), the degree of respiratory distress exhibited ranges from mild to (seldom) fulminant respiratory failure necessitating intubation and ventilatory support. Continued hypoxia from poor pulmonary flow and increased mixing can result in loss of consciousness, cerebral infarctions, seizures, and death. Classic signs of hypoxia, including irritability, tachypnea, acidosis, and diaphoresis, can also be found.[3] This occurs because of the discrepancy in the amount of oxygen required versus what is available. Perfusion will be poor during the tet spell. The decrease in systemic flow may result in additional metabolic acidosis, and clinical signs such as diminished peripheral pulses, delayed capillary refill, and poor urine output may be noted.

ED management

Treatment options during an acute episode focus on three areas of intervention: (1) increasing the arterial oxygen saturation, (2) lowering the pulmonary resistance, and (3) increasing the systemic pressures.

Alveolar hypoxia is the most potent stimulus for pulmonary vasoconstriction.[4] The lowered oxygen levels may cause the active constriction of the muscular resistance vessels in the lungs, which in turn permit only a very small amount of flow through the pulmonary circulation.[1] Supplemental oxygen is essential in all patients with hypoxia and respiratory distress to enhance arterial saturation and prevent the sequelae of hypoxemia and acidosis. Administration options may range from delivery of 100% oxygen by nonrebreather mask to intubation (rare).

Lowering the pulmonary resistance is the key to resolving a tet spell. Increased PVR and pulmonary stenosis impede pulmonary blood flow. This leads to hypoxemia, acidemia, and eventually metabolic acidosis. The pulmonary arterioles respond to this process with further constriction, promoting an additional decrease in blood flow, and thus a cyclic pattern is established.[2] As the situation worsens, the body attempts to correct the problem; however, during a tet spell, the child's condition and the situation can deteriorate rapidly. Hypoxia is the most potent initiator of pulmonary vasoconstriction. In such children, ventilation is not usually difficult; however, maintenance

Table 3
Diagnosis of tetralogy of Fallot

Echocardiography—tetralogy
EKG—right ventricular hypertrophy
Arterial blood gas studies—hypoxemia, acidosis
Heart murmur—systolic ejection murmur, thrill?
Chest x-ray—right ventricular hypertrophy, decreased pulmonary markings
Cardiac catheterization—tetralogy

of adequate pulmonary perfusion may be difficult. Oxygen can be delivered relatively easily to the alveoli; the difficulty lies in allowing blood to reach these alveoli to receive oxygen and release wastes. Options for decreasing PVR include the following:

- Calming measures. Decreasing anxiety, agitation, or crying through parent support, swaddling, and similar actions decreases the PVR and oxygen demand. Minimizing crying decreases oxygen requirements and subsequently assists in the resolution of this crisis.
- IV morphine sulfate. Administration of morphine, 0.1 to 0.2 mg/kg intravenously, may calm and sedate the child, thus decreasing crying and agitation, and lowering PVR and maximizing pulmonary flow.

Administration of morphine, 0.1 to 0.2 mg/kg intravenously, may calm and sedate the child, thus decreasing crying and agitation, and lowering PVR and maximizing pulmonary flow.

- IV propranolol (Inderal). Use of propranolol, a β-adrenergic blocking agent, 0.15 to 0.25 mg/kg intravenously, may relieve spasms of the right ventricular outflow tract and subsequently enhance oxygenation.[4] Most children with tetralogy who are at home before later surgical repair receive an oral form of propranolol to minimize these spasms. This intervention alone has greatly decreased the frequency of tet spells treated in the emergency department.

Attempts to decrease the PVR, thus amplifying pulmonary flow, increasing the SVR will result in a decreased shunt across the VSD and enhanced pulmonary flow. This may be accomplished by one of three interventions: (1) fluid challenges of 20 ml/kg of lactated Ringer's solution or 0.9% normal saline solution to raise systemic blood pressure. It is crucial to ensure that there is no air in any of the child's IV lines. Because of the VSD and associated shunting, any air can easily cross into the left ventricle, aorta, and systemic circulation, possibly causing a cerebral air embolus; (2) administration of dopamine or phenylephrine (Neo-Synephrine) infusions to increase SVR and cardiac output[5]; (3) knee-chest positioning. Older children will instinctively squat to attempt to increase SVR and diminish shunting. Knee-chest positioning increases the lower extremity resistance to flow and augments upper torso and pulmonary flow.[2]

The intensity and speed of a tet spell with its potentially fatal outcomes are truly frightening. However, remembering the principles of mixing and pressures to enhance pulmonary blood flow helps to effectively manage and stabilize the condition of affected children.

References

1. Klaus M, Fanaroff M. Care of the high-risk neonate. Philadelphia: WB Saunders, 1993:260.
2. Daberkow E, Washington R. Cardiovascular diseases and surgical interventions. In: Merenstein G, Gardner S, eds. Handbook of neonatal intensive care. St Louis: Mosby–Year Book, 1993:365.
3. Clark J, Starr-McNamara L. Heart disease: more than just blue babies. JEMS 1993;10:37-43.
4. Hazinski M. Nursing care of the critically ill child. Chicago: Mosby–Year Book, 1992:129.
5. Baker L, Alyn I. Pharmacologic manipulation of preload, afterload, and contractility in the young. J Cardiovasc Nurs 1992;6:14-8.

Contributions to this column can be sent to Deborah Parkman Henderson, RN, MA, CEN, CCRN, 1255 Linda Ridge Road, Pasadena, CA 91103, or Donna Ojanen Thomas, RN, MSN, CEN, 2822 E. Canyon View Dr., Salt Lake City, UT 84109; phone (801) 588-2240.