

The Specialist

Complications of Polycythemia in Patients Who Have Cyanotic Congenital Heart Disease

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A 13-mo-old child is brought to his pediatrician following sudden onset of left-sided hemiplegia. The pediatrician accompanies the child and family to the emergency room. The pediatrician has been following the child, who has a diagnosis of possible cyanotic congenital heart disease. The child has never been known to have had a hypercyanotic episode. Oxygen saturation determinations by pulsed oximeter had been in the range of 84% to 88%. On one occasion, when the child was very upset, a reading of 78% was observed. At his 1-year visit, his hemoglobin level was 13 g/dL.

On admission to the emergency department, a computed tomographic scan of the head was ordered. It demonstrated a right-sided nonhemorrhagic cerebrovascular lesion.

What is the likely pathophysiology of this event? What are the more common complications of hypoxemia in the older infant and young child? This clinical scenario is uncommon, but it represents one of the two feared central nervous system complications of cyanotic congenital heart disease, (ie, cerebrovascular accident and brain abscess).

A uniform response to hypoxemia of cardiac etiology is the production of erythropoietin to produce more red blood cells. This is a compensatory mechanism to maintain oxygen delivery to the peripheral tissues. Normally, hemoglobin is about 96% saturated with oxygen. Therefore, the oxygen-carrying capacity of blood with a normal hemoglobin concentration of 15 g/dL is approximately 20.3 mL of oxygen per 100 mL of blood (ie, 15 g of hemoglobin \times 1.35 mL of O₂ per g of hemoglobin = 20.3). The oxygen content of blood equals the oxygen-carrying capacity multiplied by the oxygen saturation. At a normal oxygen saturation of 96%, the O₂ content of arterial blood (Hgb 15 g/dL) equals 19.5

mL/dL (96% \times 20.3 mm³/dL) or 195 mL per liter of cardiac output.

The arterial O₂ content of this child, assuming an average arterial saturation of 85%, will be 11.1 mL/dL. Therefore, every liter (10 dL) of cardiac output will carry 111 mL of O₂ or 84 mL of O₂ less than the child with a 15 g/dL hemoglobin level. The normal compensation for this deficit is either to increase cardiac output (to deliver more blood with less O₂) or to increase the red blood cell mass so that every 100 mL of blood has more hemoglobin and is able to carry more O₂. A fairly typical hemoglobin level for a child with an O₂ saturation of 85% would be 17 g/dL. This would provide an O₂ content of 19.5 mL/dL (17 \times 1.35 \times .85), which is similar to the normal child's oxygen content.

So why was this patient's hemoglobin only 13 g/dL? Investigation of the child's hematologic status revealed a mean corpuscular volume of 66 fL, mean corpuscular hemoglobin concentration of 29.8 g, a zinc protoporphyrin level of 105 mg/dL, a transferrin saturation of 4%, and a serum ferritin of 2.8 ng/mL, indicating significant iron deficiency anemia.

Children who have cyanotic heart disease and iron deficiency anemia are especially susceptible to cerebrovascular accidents secondary to the decreased deformability of the red blood cells as they traverse the cerebral microcirculation. Therefore, even though this 13-mo-old's hemoglobin value would be normal for a normal child, it is insufficient for a hypoxic child.

Children who have cyanotic heart disease may have hemoglobin levels in the range of 16 to 17 g/dL and still be iron-deficient. Therefore, every hypoxic child should have his or her iron status evaluated routinely. Many of these infants and young children are poor eaters because of their disease and may require substantial iron and vitamin supplementation as well as caloric supplementation if they are not growing well. Therefore, infants and young children who have cyanotic congenital heart disease are at risk for polycythemia, iron deficiency anemia, and cerebrovascular accidents.

Interestingly, children who have pulmonary parenchymal disease, especially cystic fibrosis, seem to have an inadequate erythroid response to hypoxemia. It has been demonstrated that the inadequate erythroid response is secondary to a disturbance of erythropoietin regulation and iron availability.

Older children who have congenital heart disease and are cyanotic because of an inoperable defect or because of the development of pulmonary vascular obstructive disease (ie, Eisenmenger syndrome) may become

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excessively polycythemic. These adolescents become symptomatic because of increased blood viscosity. The symptoms most frequently described are decreased exercise tolerance, headache, and chest pain, which are observed most commonly when the hematocrit exceeds 65%. These youngsters frequently will benefit from an erythrophoresis, which removes erythrocytes and replaces the volume extracted with a similar volume of colloid (fresh frozen plasma, dextran, hetastarch). As with the younger child, iron deficiency may occur because of the removal of erythrocytes and their iron content.

It is important not to perform a simple phlebotomy, which would reduce intravascular volume, decrease systemic resistance, and promote a larger right-to-left shunt. This would increase hypoxemia further,

resulting in systemic vasodilatation that would increase the right-to-left shunt, furthering hypoxemia and resulting in acidosis and potential cardiovascular collapse.

The pathophysiology of the vascular system in cyanotic congenital heart disease is simple: flow goes where resistance is least. It is important for all pediatricians to have an understanding of this mechanism and potential complications. Any procedure that has the potential to reduce blood pressure should be avoided unless the child is in a well-supervised and monitored situation.

One final caution in managing children who have right-to-left intracardiac shunts is the potential for paradoxical systemic embolus. Any intravenous fluids or medications must be given with scrupulous attention to avoiding injection of any air

or foreign material. In the normal child, these minute particles would be filtered in the pulmonary circuit. The patient who has an intracardiac right-to-left shunt is at high risk for those particles entering the systemic circulation, especially the cerebral circulation. The risk can be reduced substantially by incorporating an air filter into all intravenous systems and giving medications distal to the filter.

In summary, attention to detail is critical in the care of children who have cyanotic congenital heart disease to avoid preventable complications. Oxygen saturation may be monitored transcutaneously with pulsed oximetry and iron status by blood count and indices. Intravenous fluids and medications should be administered through a system that contains an air filter.

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