EVALUATION OF THE NEONATE WITH SUSPECTED CRITICAL CONGENITAL HEART DISEASE AND REVIEW OF BASIC PRINCIPLES OF OXYGEN PHYSIOLOGY AND TRANSPORT

Department of Pediatrics
Resident Curriculum

Evaluation of the Neonate with Suspected Critical Congenital Heart Disease and Review of Basic Principles of Oxygen Physiology and Transport

The newborn with serious structural congenital heart disease may present with cyanosis, respiratory distress, heart murmur or subtly with decreased systemic oxygenation that may not be recognized on clinical examination. The specifics of this structural defect can be made accurately by echocardiography, but it is important that the pediatric resident understand the possible physiological derangements and be able to initiate appropriate initial management even before a specific anatomic diagnosis is made.

Critical congenital heart disease may present with:

- Decreased pulmonary blood flow
- Decreased systemic blood flow
- Abnormal mixing
- Increased pulmonary blood flow

Decreased Pulmonary Blood Flow

The infant with tetralogy of Fallot is a classical example. These infants present with cyanosis and a heart murmur. They have a ventricular septal defect. The severity of cyanosis or systemic oxygen desaturation depends on the severity of the obstruction to pulmonary blood flow from pulmonic subvalvar or valvar stenosis. In addition to the murmur from obstruction to pulmonary blood flow there may be a continuous murmur from persistent patency of the ductus arteriosus. The electrocardiogram is usually not diagnostic in these patients although a chest x-ray will often show an upturned cardiac apex and an absent main pulmonary artery segment. Management would include the administration of prostaglandin E_1 to keep patency of the ductus arteriosus.

Although not a classical structural congenital heart defect, infants presenting with persistent pulmonary hypertension have obstruction of pulmonary blood flow at the pulmonary arterial level from pulmonary vasoconstriction. These patients usually present as critically ill infants with severe cyanosis related to right-to-left intracardiac shunt through an atrial septal defect or reverse of flow through a patent ductus arteriosus. In many instances they may have intrapulmonary shunting from primary lung disease, prematurity or meconium aspiration.

Decreased Systemic Blood Flow

Newborns may present with inadequate systemic blood flow. The classical example of this physiology is the infant with aortic atresia and patent ductus arteriosus dependent systemic blood flow. Before the patent ductus constricts in these infants their presentation may be subtle. The only clinical finding may be an accentuated precordial activity with mild decrease in systemic oxygenation that is not clinically recognizable. It would be easy for the unsuspecting pediatrician to send an infant with aortic atresia home only to have this infant re-present two to three days later in *extremis* as the patent ductus arteriosus closes. The chest x-ray may be unrevealing and an electrocardiogram would only demonstrate an accentuation of the usual right ventricular hypertrophy and right axis deviation noted in the newborn. Management is to maintain patency of the ductus arteriosus.

Inadequate mixing

A classical example of this defect is transposition of the great arteries. The presentation of infants with transposition of the great arteries is not subtle as they have severe cyanosis with arterial PO_2 in the range of 30 mmHg or less. These infants present with severe cyanosis and in no respiratory distress. Chest x-ray and electrocardiogram can be normal although the chest x-ray can often show an absent pulmonary artery segment because of the anterior/posterior relationship of the great arteries. Management is prostaglandin and transfer to a center where balloon atrial septostomy can be performed.

A comment regarding total anomalous pulmonary venous return should also be made because these infants may present critically ill with what appears to be primary lung disease. This is particularly true if the exit from the pulmonary venous confluence is obstructed by exiting by way of portal venous system. These infants require prompt diagnosis and urgent surgical or supportive management with extracorporeal membrane oxygenation (ECMO).

Excessive Pulmonary Blood Flow

Infants with excessive pulmonary blood flow are asymptomatic as newborns. They can have simple or complex structural congenital heart disease that would be classified as cyanotic or non-cyanotic. Common examples are ventricular septal defect, transposition of the great arteries, or truncus arteriosus. After birth, there is a drop in pulmonary vascular resistance related to mechanical lung expansion and delivery of oxygen to the alveoli. Subsequent decrease in pulmonary vascular resistance occurs as there is regression of pulmonary arterial smooth muscle. It is during this further drop in pulmonary vascular resistance that infants become symptomatic with increased pulmonary blood flow. The increase in pulmonary blood flow causes changes in lung compliance and results in small and large airway obstruction. Presenting symptoms are tachypnea, increased work of breathing, although they can have poor growth parameters. Management consists of digoxin, Lasix, afterload therapy and early surgical intervention.

In summary, the pediatric residents need to understand the physiologic derangements that may occur in the newborn with critical structural congenital heart disease. It is not necessary to determine a specific structural diagnosis in order to institute early, appropriate, life-saving management.

References:

Hoke, T.R., Donohue PK, Bawa PK, Mitchell RD, Pathak A, Rowe PC, and Byrne BJ: Oxygen Saturation as a Screening Test for Critical Congenital Heart Disease: A Preliminary Study. Pediatric Cardiology 23:403-409, 2002.