Blue Baby Syndrome (Cyanotic Newborn) and Hyperoxia Test

Blue babies lack sufficient hemoglobin, resulting in the bluish discoloration of tissues, a term referred to as cyanosis. There are three major types of cyanosis: central, peripheral and Acrocyanosis and each of them have its specific symptoms, even though the symptoms of central cyanosis overlaps with that of peripheral cyanosis. Cyanosis is diagnosed with auscultation, chest x-ray, electrocardiogram and recently hyperoxia test.

Definition

Cyanosis refers to the situation where the tissues present with a bluish discoloration resulting from reduced volume of hemoglobin in the capillary bed less than 3 g/dL [1-3]. Usually, the baby will be blue if the total volume of hemoglobin is decreased, in contrast to the reduction in the ratio of decreased oxygenated hemoglobin. Cyanosis is a very frequent outcome in newborn babies. Neonatal cyanosis, especially of the central type, can result in significant and possibly life-threatening conditions as a result of conditions related to the cardiopulmonary, metabolic, neurological conditions as well as infections.
Types of Cyanosis

Central cyanosis

Central cyanosis results from a reduction in the rate of arterial oxygen saturation. Newborn babies usually present with central cyanosis for at least 5 to 10 minutes after birth, because oxygen saturation gradually increases to 85 to 95% at 10 minutes of age. However, persistent central cyanosis is usually abnormal and must be assessed and treated immediately.

Peripheral cyanosis

Babies having peripheral cyanosis present with an optimal systemic arterial oxygen saturation as well as heightened uptake of oxygen from the tissues that results in an enlarged systemic arteriovenous oxygen difference of 60% (which is higher than the optimal 40%) leading to an increase in the level of decreased hemoglobin on the venous part of the capillary bed. Peripheral cyanosis is characteristically seen in the distal part of the extremities and less frequently around the circumoral or periorbital regions [1]. In this case, the extremities will feel cold or clammy. Peripheral cyanosis is likely to be associated with peripheral vasoconstriction or several etiology associated with central cyanosis. Neonates presenting with peripheral cyanosis, have a pinkish coloration of the mucous membranes, which makes it different from central cyanosis.

Acrocyanosis

Acrocyanosis is a benign condition, which is sometimes present in healthy newborns, refers to the peripheral cyanosis surrounding the mouth and extremities (i.e. hands and feet). The cause of acrocyanosis is a benign vasomotor alteration that leads to peripheral vasoconstriction and heightened tissue oxygen uptake. What makes acrocyanosis different from other types of cyanosis is that it results in some type of significant pathology (such as septic shock) because it happens just after a healthy baby is born. It is usually a common presentation and may likely to last for 24 to 48 hours.

Causes of Cyanosis

Central Cyanosis

In newborn babies, central cyanosis is observed in situations where there are critical issues with the respiratory system, breathing, heart, and blood circulation. If this is the case, such babies need quick evaluation so that the problem can be corrected as soon as possible. Even though cyanosis is present at birth, this should clear after some minutes, and if it does not, immediate action should be taken.

The causes of central cyanosis in infants arising from heart problems are referred to as cyanotic heart diseases or congenital heart diseases. Examples of these conditions include:

- Transposition of great arteries,
- Tetralogy of Fallot,
- Total anomalous pulmonary venous return,
- A left heart that is Small or hypoplastic,
- Truncus arteriosus,
- Persistent fetal circulation

The above conditions are likely to occur if the infant has a congenital problem such as Down syndrome, Turner syndrome, Marfan’s syndrome, Noonan syndrome, among others. It can also occur if the infant’s mother had infections such as German measles or rubella while she was pregnant.

Also, women who have diabetes during pregnancy as well as those on certain prescription and over-the-counter medications and illicit drugs during pregnancy present with a higher risk of delivering a child with a congenital heart defect.

Causes of central cyanosis that are caused by a defect in the respiratory system:

- Birth trauma or asphyxia,
- Transient tachypnoea of the newborn,
- Respiratory distress syndrome,
- Pneumothorax,
- Pulmonary or lung edema,
- Accidental aspiration or swallowing and choking on meconium,
- A diaphragmatic hernia,
- Pleural effusion,
- Trachea-oesophageal fistula
- Obstruction of the upper respiratory tract.

Other causes of central cyanosis in infants include:

- Low blood sugar
- Inadequate blood magnesium
- Infections
- Epilepsies

Peripheral cyanosis

The causes of peripheral cyanosis are:

- Central cyanosis and all its causes
- Reduction in the volume of blood pumped by the heart or decreased cardiac output, as observed in heart failure or circulatory shock
- Defects in circulation ob blood such as thrombosis or embolism
- Reduction in the size of blood vessels of the arms, legs, fingers, and toes due to:
  - Exposure to cold
  - Raynaud’s phenomenon
  - Spasm of the tiny skin capillaries or arteries referred to as acrocyanosis
  - Erythrocyanosis taking place in young women or as an unwanted effect of beta blocker drugs taken for hypertension

Clinical Features

The frequent symptoms of this condition are the bluish coloration of the lips, extremities, fingers, and toes. However, the symptoms of cyanosis can be divided into two separate groups; for central and peripheral
Symptoms of central cyanosis

It occurs in conditions affecting the heart and/or lungs as well as in hemoglobin defects such as methemoglobin and sulfhemoglobin etc. The cyanosis, in this case, results from desaturation of main arterial blood as a result of diseases of the heart and lungs.

Instead of the deoxygenated blood to be transported to the lungs for oxygenation, it is transported to the body’s overall blood circulation resulting in the manifestation of cyanosis.

This is seen as bluish or purple discoloration of tongue and lips and mucous membranes of the mouth. Most of the times, symptoms of central cyanosis is accompanied by those of peripheral cyanosis.

Other features such as increased rate of breathing, shortness of breath, bluish or purple discoloration of the mouth, fast and shallow breathing may be seen.

The hands and feet frequently feel like normal temperature or warm. However, if it is associated with peripheral cyanosis, they may be cold.

Symptoms of peripheral cyanosis

This results from decreased blood flow to the peripheral organs and limbs. It results from stasis or stagnation of blood as well as overutilization of oxygen from the blood. This makes blood deoxygenated and leads to cyanosis.

Any area that is affected becomes turn bluish or purple, and feel cold to touch. This is commonly observed in the nail beds.

It may resolve on its own after the initial warming, without causing bluish discoloration of the mucous membrane of the oral cavity.

Symptoms of cyanosis in Babies

Babies born with cyanosis are likely to get tired easily while eating or playing and will sweat profusely while eating. They are likely to present with low weight gain. They are likely to be born with a low birth weight. May probably be irritable and sometimes feel floppy or weak.

There is a flaring of the nose, revealing that the baby is breathing with stress (labored breathing). A few infants may be heard grunting or producing an abnormal noise as they breathe.

Diagnosis of a Blue baby

The assessment will systematically evaluate the baby for airway, pulmonary, and circulatory reasons for the cyanosis.

History

A detailed history that history of pregnancy, labor, and infant risk factors should be obtained. If the mother has a previous history of maternal diabetes, this heightens the risk of developing congenital heart disease.

Check for oligohydramnios, and if present may point to renal abnormalities related to
hypoplastic lungs, while polyhydramnios is likely to show airway, esophageal, or neurological abnormalities.

A history of prolonged labor may have led to intracranial hemorrhage or phrenic nerve paralysis. Ask for a history of birth trauma.

Clinical Examination

To carry out a physical examination on the infant, make sure he/she is quiet and relaxed. The pattern and features of growth should be noted because babies who are either small or large for their age are more likely to develop polycythemia. The main aim is to determine the extent of respiratory distress because it will point to a congenital heart disease or methemoglobinemia.

Lack of adequate respiration caused by the pulmonary disease is naturally revealed by fast respiration associated with chest retractions and nasal flaring. Neurological diseases may lead to cyanosis owing to hypoventilation and may cause reduced or irregular respirations. It is also imperative that the baby’s breathing tone and activity are evaluated to check for periodic breathing and/or presence of apnea.

The cardiac examination must include an evaluation of the baby’s heart rate, peripheral pulses, as well as perfusion. While auscultating the heart, concentrate the second heart sound, which is likely to be loud and heard once in pulmonary hypertension. Carrying out an auscultation for heart murmurs is usually not useful because serious conditions like transposition do not produce murmurs, and loud murmurs are often caused by a harmless lesion such as a small ventricular septal defect.

However, note that a serious ejection murmur points to pulmonary stenosis.

Investigations

Laboratory

Pulse oximetry should be carried out to determine the level of oxygen saturation. It is important that measurement should be obtained from both the right hand and a foot to ascertain the pattern of flow via the ductus arteriosus. It is not advisable to use the left hand for pulse oximetry because the left subclavian artery is likely to have a preductal or postductal root from the aorta.

While a venous blood gas is important for evaluating pH and PaCO2, it should not be utilized to ascertain oxygenation. However, if significant metabolic acidosis is revealed, it may point to cardiac failure, sepsis, asphyxia, or metabolic disorders.

Chest Radiography

A chest radiograph is a very vital part of evaluating a cyanotic baby. It will help to show the positions of stomach, liver, and heart, and this will help eliminate dextrocardia and situs inversus. Look at the lung fields for signs of parenchymal lung disease (note that newborns with pneumonia characteristically have a infiltrates spread out rather than focal), and lung abnormalities including cystic adenomatoid malformation.

Elevation of any of the hemidiaphragm by higher than two intercostal spaces in comparison to the other side points to diaphragmatic paralysis caused by phrenic nerve
injury. Sometimes, we can see hyperinflated lung fields if the baby has lobar emphysema or cystic lesions of lungs. Reduced pulmonary vascular outlines suggest pulmonary stenosis or pulmonary atresia with poor ductal shunting which is probably present in infants with idiopathic persistent pulmonary hypertension.

The size and shape of the heart are likely to help point to a diagnosis: for example, if cyanosis is caused by Tetralogy of Fallot, it will be “boot shaped”, transposition presents as an “egg on string” and a characteristic large cardiomegaly points to Ebstein’s anomaly.

If the baby has an idiopathic persistent pulmonary hypertension, the lung fields will be very clear with signs of reduced vascularity.

**Electrocardiogram (EKG)**

An electrocardiogram (EKG) is important to diagnose cardiac arrhythmias. That said, it should be noted that EKG not very useful in assessing a baby with congenital heart disease because most times it appears totally normal even when a baby has a life-threatening cardiac condition such as transposition. An exception though will be a baby with left axis deviation caused by left ventricular hypertrophy, pointing to tricuspid atresia.

**Hyperoxia Test**

The hyperoxia test is an important clinical tool for differentiating pulmonary conditions from cardiac diseases in cyanotic babies. It is based on the theory that if there is an absence of persistent cardiac shunts, then, 100% oxygen will enhance alveolar PO$_2$, resulting in a heightened level of pulmonary venous and systemic arterial PO$_2$. In babies with cyanotic congenital heart disease, we will see no observable difference in the amount of PaO$_2$ even after breathing 100% O$_2$.

However, a similar outcome is expected in babies with significant pulmonary hypertension due to persistent right-to-left shunting or extrapulmonary shunts (caused by ductus arteriosus and foramen ovale). That said, the hyperoxia test should only be carried out if an echocardiography is unavailable or after speaking to a cardiologist.

**References**


