Persistent Pulmonary Hypertension in the Neonate

Persistent pulmonary hypertension in the neonate (PPHN) is a serious disorder of impaired transition to extrauterine circulation that primarily affects infants who are term or near term. PPHN is frequently associated with respiratory disease or failure from other causes such as meconium aspiration, sepsis, or congenital diaphragmatic hernia. Occasionally, PPHN occurs without any other conditions that can be identified.

During pregnancy, the infant's mother and the placenta provide the infant with oxygen, and very little of the infant's blood goes to the lungs. The blood vessels in the lung constrict (high pulmonary vascular pressure) to decrease blood flow to the lungs during this time. As a fetus, blood can bypass the lungs using the patent ductus arteriosis (PDA) and the foramen ovale. When an infant is born and takes his or her first breaths, the blood vessels in the lungs expand and allow blood to travel to the lungs to pick up oxygen and eliminate carbon dioxide. When these blood vessels fail to fully relax or fail to allow adequate blood to reach the lungs, it is called persistent pulmonary hypertension. The failure of these vessels to dilate may be related to underdevelopment of the vessels, hypertrophy of the muscle layer in the blood vessels, or a situation such as a difficult birth that interferes with the transition to extrauterine life. Because newborns still have the PDA and a foramen ovale, blood can bypass the lungs. This process is called shunting and can cause cyanosis. Oxygen is one of the molecules that helps vasodilate the newborn's pulmonary blood vessels. When an infant develops hypoxia or cyanosis, it acts as a pulmonary vasoconstrictor and tends to worsen the pulmonary hypertension. This cycle can be difficult to break.

The incidence of PPHN may be increased if the mother took certain medications during the last months of pregnancy, including nonsteroidal anti-inflammatory agents and serotonin reuptake inhibitors.

The diagnosis of PPHN should be considered in any infant with severe cyanosis or high oxygen requirements. PPHN



may be suspected due to an infant's clinical picture, but diagnosis should be confirmed with echocardiography (showing increased right atrial pressures, increased right ventricular pressures, or tricuspid insufficiency).

Treatment of PPHN includes support of oxygenation and ventilation, fluids and medications to maintain good cardiac output, and sedation. Surfactant may be given to improve lung function. Infants may require inhaled nitric oxide, a clear, odorless gas that when inhaled acts as a pulmonary vasodilator. In extreme cases, the infant may require extracorporeal membrane oxygenation, which is a form of heart lung bypass to avoid lung injury and allow the PPHN to resolve.

Persistent pulmonary hypertension puts the infant at risk for low blood oxygen levels, increased requirements for support, and long-term neurodevelopmental impairment such as developmental delays, motor delays, and hearing loss.

Bibliography

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Persistent Pulmonary Hypertension in the Neonate: Information for Parents

Persistent pulmonary hypertension in the neonate (PPHN) is high blood pressure in the lungs and is a serious respiratory disorder that primarily affects infants who are born full term or near term (usually 34 weeks or more). PPHN is frequently associated with respiratory problems.

During pregnancy, the baby's mother and the placenta provide the baby with oxygen, and very little of the baby's blood goes to the lungs. The blood vessels in the lungs are mostly closed, because the lungs are not being used. When an infant is born and takes his or her first breaths, these blood vessels expand and allow blood to go to the lungs to pick up oxygen. When these blood vessels do not fully relax or do not allow enough blood to reach the lungs, it is called PPHN. PPHN is very dangerous because it can limit how much oxygen the baby can deliver to his or her brain and organs.

The treatment of PPHN may include the use of oxygen, special ventilators that breathe for the baby at a very fast rate, a gas called nitric oxide, or even temporary heart lung bypass.



After treatment for pulmonary hypertension, your baby's lungs will take weeks or even months to recover fully. It will be important to help protect your baby from catching colds or flus. Good handwashing and keeping your baby away from sick people and large crowds will help. It also will be especially important for your baby to see his or her pediatric provider and other specialists regularly to be screened for normal developmental milestones.