

PULMONARY HYPOPLASIA

What is Pulmonary Hypoplasia?

Pulmonary Hypoplasia (PH) is where the lungs do not develop normally while the baby is in the uterus (womb). This condition may affect one or both lungs. As a consequence there are not enough alveoli (air sac units for exchange of gases) or normal blood supply to the alveoli resulting in poor exchange of oxygen and carbon dioxide.

Who gets Pulmonary Hypoplasia?

Occasionally PH is a spontaneous unexpected condition detected after birth. However, most commonly PH is associated with other congenital conditions where the development of the lungs is compromised by other organs or growths within the chest cavity, or there is insufficient amniotic fluid around the baby. Congenital diaphragmatic hernia where the abdominal organs are in the chest may compromise development of the lungs. Other lung conditions such as congenital adenomatoid malformation of the lung and lung sequestrations may compromise lung development. Prolonged rupture of membranes with minimal amniotic fluid around the baby from early gestation i.e. 18-24 weeks may result in poor lung development. Conditions affecting normal kidney development such as bilateral renal agenesis, polycystic kidney disease or posterior urethral valves will result in decreased amniotic fluid and compromise lung development. Antenatal ultrasounds are not able to predict with certainty whether lungs will work normally when a baby is born.

What are the symptoms of Pulmonary Hypoplasia?

Babies with PH have signs of respiratory distress from birth, including cyanosis (blue skin and lips), grunting respirations and increased work of breathing with use of abdominal muscles. The carbon dioxide levels in the blood will be high and the oxygen levels will be low.

How do we diagnose and treat Pulmonary Hypoplasia?

The definitive diagnosis of PH can only be made at the time of an autopsy where the fine architecture of the lungs is examined under the microscope. However it is clinically suspected with the history of other known findings on antenatal scans and the symptoms of respiratory distress described above. Your baby will require assistance with breathing from a ventilator, and often this will involve a method of ventilation called high frequency oscillation where your baby's lungs are "wiggled" to enable diffusion of gases while causing minimal damage to the lung. Your baby will also frequently be treated with a gas called nitric oxide which helps dilate the blood vessels in the lungs to help diffusion of gases. Blood pressure medications may also be utilised to try and assist with blood flow to the lungs.

Are there any long term consequences of CLD?

Unfortunately many babies with PH do not survive. Survival frequently depends on the other conditions which may have caused the PH. Baby's that survive may have reduced lung function for the rest of their life and be at increased risk of respiratory compromise with colds and chest infections.

If you have any further questions please ask any of the nursing or medical staff.