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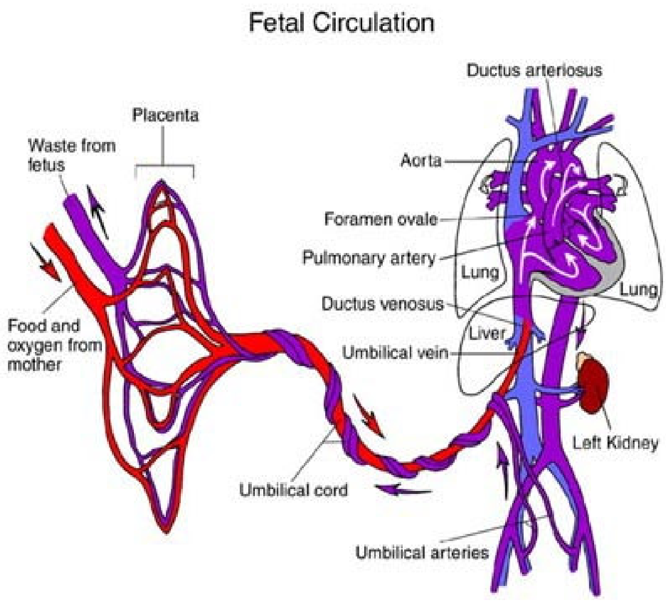
PERSISTENT PULMONARY HYPERTENSION

What is persistent pulmonary hypertension of the newborn (PPHN)?

When your baby is in the uterus most of the blood from the heart bypasses the lungs through a vessel called the ductus arteriosus. This is because the placenta supplies the baby's blood with oxygen. After a baby is born the ductus arteriosus should close and blood passes through the lungs. The lungs oxygenate the blood before it returns to the left side of the heart to be distributed around the body. Some babies after birth do not change to the newborn pattern of circulation which results in persistent pulmonary hypertension (PPHN) also known as persistent fetal circulation. In this situation the ductus arteriosus does not close and the blood continues to bypass the lungs. As the placenta is no longer available to supply the baby with oxygen the oxygen supply in the baby's blood becomes low.



http://www.rmgh.net/wiki/images/b/b4/Fetal_circulation_1.gif

Which babies are at risk of PPHN?

About 1 in 500-700 babies develop PPHN. Risks for PPHN include:

- premature birth
- meconium aspiration syndrome
- requiring resuscitation at birth
- severe infection

What causes PPHN?

When your baby is born the first few big breaths they take allows oxygenation of the blood which then helps the blood vessels in the lungs to dilate (get wider). This allows more blood to flow through the lungs and become oxygenated as the baby continues to breathe and also makes the ductus arteriosus close. If your baby does not take these first few big breaths due to the conditions listed above the blood vessels of the lungs do not dilate and the ductus arteriosus does not close. Very occasionally there does not appear to be a cause for the persistence of the fetal circulation pattern.

What symptoms will my baby have?
The signs and symptoms of PPHN include:

- rapid breathing (tachypnoea)
- rapid heart rate (tachycardia)
- increased work of breathing where the baby can be seen to use extra muscles to help him/her breathe
- grunting
- cyanosis – “blue baby” where the skin is blue due to inadequate oxygen supply

How do we diagnose PPHN?
PPHN is diagnosed by an ultrasound of the heart (echocardiogram) in combination with a chest x-ray and blood tests measuring the amount of oxygen in the baby’s blood (arterial blood gas).

How do we treat PPHN?
Babies with PPHN need intensive care and may become critically ill. The most important part of treatment is to provide the lungs with as much oxygen as possible as high oxygen levels will help the baby’s circulation to change over to the newborn pattern. This is done by intubating (placing a tube in the trachea – the windpipe) and providing 100% oxygen by a ventilator which also assists their breathing. To maximise their ventilation we will frequently use medication that relaxes his/her muscles. We will also use medications that support his/her blood pressure aiming to improve the circulation of blood to the lungs.

Many babies with PPHN will have low blood pressure. Medications called inotropes (for example dopamine and dobutamine) will be used to help increase the blood pressure. It is important to maintain a good blood pressure so that as much blood as possible can be sent to the lungs to be oxygenated.

Many babies with PPHN will be treated with a gas called nitric oxide. This gas, like oxygen, helps to dilate the blood vessels of the lungs. This assists in oxygenating the blood and closing the ductus arteriosus, allowing the adult circulation pattern to be maintained. This gas is considered a standard treatment for this condition but has currently not been passed for use in Australia, and requires consent by the parents to be used.

During the critical part of the illness your baby will not be given any milk feeds. He/she will be provided with nutrition via intravenous fluids. Once your baby has recovered, he/she will be commenced on small amounts of milk feeds via a nasogastric tube (tube passed through the nose into the stomach). As your baby improves these feeds will be increased.

Are there any long-term complications?
With improvement in ventilators and the use of nitric oxide the majority of babies with this condition now survive (more than 80%). Depending on the underlying reason for the PPHN there may be some long-term physical or developmental problems. Infants are at higher risk of hearing loss and will have their hearing checked prior to discharge and again at 3 months of age. If your baby required resuscitation at birth and PPHN occurred following this, the medical staff will discuss with you any concerns they may have about long-term developmental problems.

Some babies may continue to have an ongoing oxygen requirement and may be commenced on sildenafil which is an oral medication that acts as a pulmonary vasodilator. The adverse effects include a minor reduction in blood pressure, headache, flushing, nasal congestion and rash (all of these are unusual in children ranging from 2-10%). Your baby will be started on the medication at a low dose and the medication increased every 24-48 hours. They will be observed for any side effects and any improvement in oxygen and respiratory requirements.

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

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