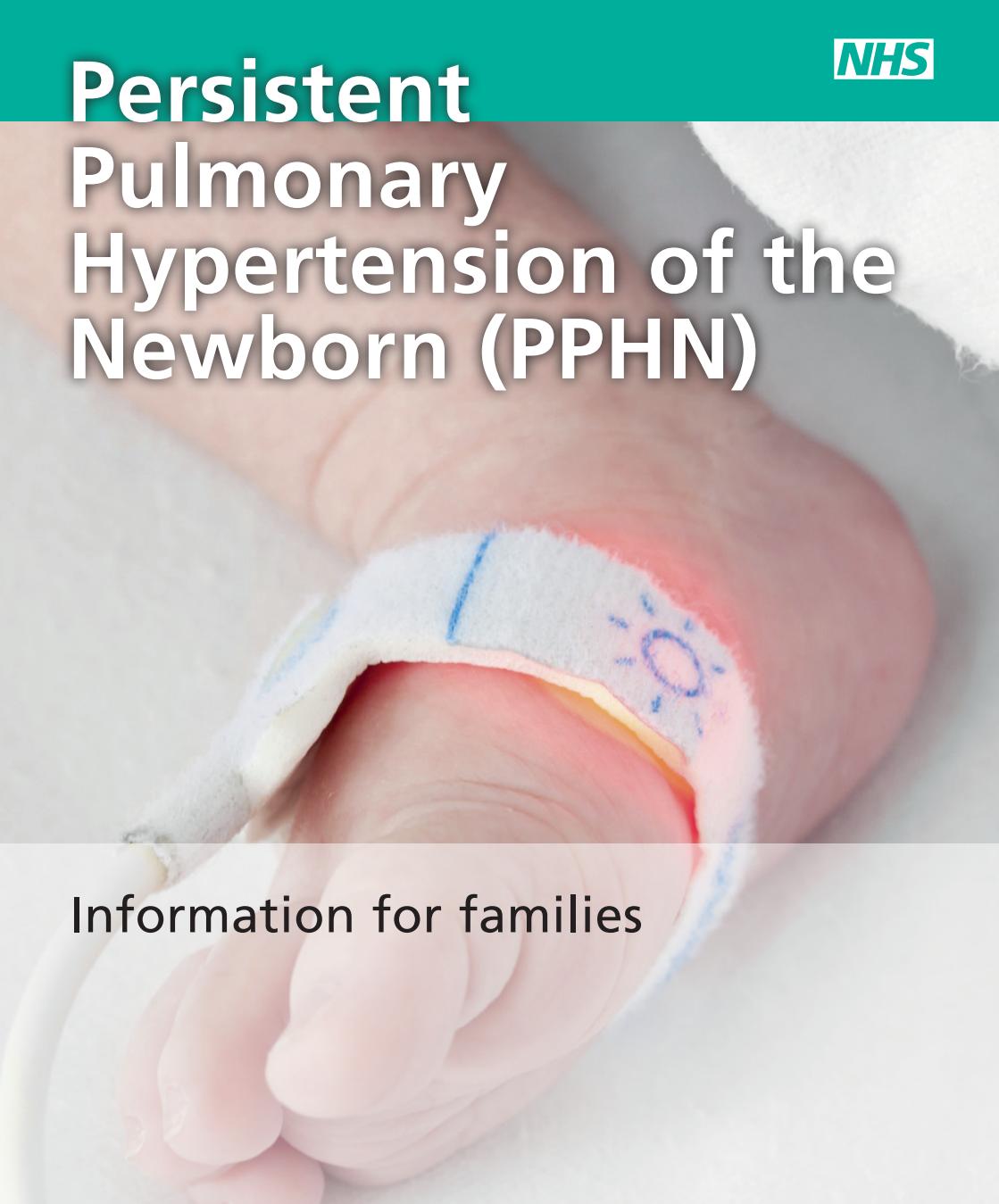


Persistent Pulmonary Hypertension of the Newborn (PPHN)



Information for families

Great Ormond Street Hospital
for Children NHS Foundation Trust

This information sheet from Great Ormond Street Hospital (GOSH) explains the causes, symptoms and treatment of persistent pulmonary hypertension of the newborn (PPHN). It also describes what to expect when your child is being treated at GOSH.

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

Before a baby is born, they do not need to use their lungs to breathe, because they receive oxygen via the umbilical cord and placenta from their mother. In the womb, the lungs are filled with amniotic fluid, and the blood vessels (arteries and veins) which take blood from the heart to the lungs are constricted, or closed. This means that the pressure inside the blood vessels of the lungs is high – you will often hear doctors talk about ‘high pulmonary pressures’.

When a baby is born, they take a big breath or cry, and their lungs fill with air instead of fluid. When the lungs fill with air, the blood vessels which take blood from the heart to the lungs open up (dilate), and this means that oxygen can be carried from the lungs, back to the heart, and pumped to the brain and the rest of the body once the umbilical cord is cut. The pressure inside the lungs and the blood vessels is now low.

If there is a problem around the time of birth which interferes with this process, the blood vessels may not open up properly so the pressure inside them remains high. This is called persistent pulmonary hypertension of the newborn (PPHN). As a result of the blood vessels not opening up, blood cannot get into the lungs to pick up oxygen and then the body will not have enough oxygen for the brain and other organs, and this can make your baby unwell.

Who does PPHN affect?

PPHN occurs in about two in every 1000 births. It usually occurs in babies that are born at term (at nine months) but can occasionally occur in premature (born too early) babies as well.

What causes PPHN?

Doctors do not always know the cause of PPHN, but we do know that the following may be factors:

- **Meconium aspiration** – this is when a baby has passed faeces (poo) while still in the womb, and because the poo becomes mixed with the amniotic fluid (the fluid inside the womb), the baby can breathe it into their lungs.
- **Infection** – infections such as pneumonia (lung or chest infection) and bloodstream infections can make PPHN more likely, and there may be an increased risk of these conditions if the waters broke a long time before the baby was delivered, or if there was a group B strep infection present.
- **Congenital abnormalities of the heart and lungs** – a small number of babies who get PPHN will have it because they have been born with an abnormality of their heart or lungs such as a diaphragmatic hernia, or a blocked heart valve, or lungs that are smaller than they should be.

How do doctors diagnose PPHN?

The main feature of PPHN is that not enough oxygen is getting to the heart, brain and other organs. This causes babies to look blue or pale and to have difficulty in breathing.

Doctors, nurses and midwives will use oxygen saturation monitoring to measure the amount of oxygen in the blood, expressed as a percentage.

Monitoring involves putting a small probe which looks like a sticky plaster around the baby's hand or foot, then displaying a number on a screen like the image bottom. If this number is low (below 92 per cent), and does not come up to 100 per cent easily when doctors treat problems with breathing using oxygen or breathing machines, they may diagnose PPHN.

In some hospitals, it may be possible to do an ultrasound of the baby's heart called an echocardiogram, which can look to see if the blood vessels in the lungs are closed or constricted (pulmonary hypertension, sometimes called 'high pulmonary pressures') and also rule out any abnormality of the heart which may be causing the low oxygen levels.

It is likely that doctors will also do a chest X-ray to look at the baby's lungs, and some blood tests to look for signs of infection.

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How is PPHN treated?

Initial treatment of PPHN will consist of simple measures such as keeping your baby warm (but not too hot) and giving oxygen, usually through small prongs (short plastic tubes) in the nostrils, or in an incubator.

Doctors will usually insert a cannula or drip into your baby's hand or foot, and use this to give some antibiotics.

As your baby is not likely to feed well while they have PPHN, they will receive fluids containing sugar for energy through a drip.

If these simple measures do not bring the oxygen levels up easily, your baby is likely to need to be moved to a neonatal intensive care unit or NICU.

Treatment of PPHN in the intensive care unit (PICU or NICU)

■ Ventilation (breathing machine)

– it is likely that doctors will pass a breathing tube into your baby's airway through their mouth or nose, which is connected to a breathing machine or ventilator. This will breathe for your baby while they are unwell. The machine reduces the amount of effort and energy needed to breathe, which in turn reduces the amount of oxygen that your baby needs and it also delivers the maximum possible amount of oxygen into the lungs. When your baby is connected to the breathing machine, doctors will give them some medicines to make them sleepy so that they do not feel any discomfort as well as some medicines to stop them from moving. As the breathing tube passes into their airway, your baby will not be able to cry or make noises while they are on the machine.

■ High Frequency Oscillation (oscillator)

– this is another type of breathing machine that doctors use to help deliver oxygen into the lungs of babies with PPHN. This machine pushes air in and out of the lungs very quickly, so it is very noisy.

Extra-corporeal membrane oxygenation (ECMO)

- **Nitric oxide** – this gas is a combination of nitrogen and oxygen which is given to your baby through the breathing machine, straight into the lungs. It works by opening up the closed blood vessels so that more blood flows into the lungs, and the high pressures are reduced.
- **Inotropes** – these are medicines that are given directly into the bloodstream via a drip to help keep your baby's blood pressure high, as this helps the heart to pump blood into the lungs.

If doctors have tried all of the treatments described above, and your baby's oxygen saturations are still low, they may consider a treatment called extra-corporeal membrane oxygenation (ECMO), which is like a heart bypass machine during heart surgery. It takes over the work of the heart and lungs, putting oxygen directly into a baby's blood rather than relying on the lungs to add oxygen. For this treatment your baby will be moved to the Cardiac Intensive Care Unit – we will give you further written information about this if it becomes likely.



Moving forward and discharge from intensive care

Once your baby's oxygen saturation levels are normal, the doctors and nurses will start to reduce the amount of nitric oxide delivered through the breathing machine, until it is turned off altogether. They will then gradually reduce the amount of oxygen used, and the amount of breathing work being done by the ventilator. You may hear them referring to this as 'weaning'. During this time the doctors will reduce the medicine that keeps your baby still and sleepy so that they will become more awake and aware of their surroundings.

At the same time, the doctors will gradually reduce the inotropes (medicines being given to raise blood pressure) and might consider stopping antibiotics if a full course has been given. At this time the nurses may use a feeding tube, passed into one of your baby's nostrils and down the throat to the stomach, to start some milk feeds. These can be expressed breast milk or formula, depending on your preference.

Once your baby is doing most of the work of breathing themselves, with only a small amount of help from the breathing machine, doctors will try to take your baby off the breathing machine altogether.

Once this has happened, it is likely that you will be ready to go back to your local hospital, where your baby will finish any antibiotics that are still needed, continue oxygen treatment until it is no longer needed, and help you to establish feeding before you are discharged home.

What is the outlook for babies who have had PPHN?

PPHN is a serious condition, and if you have been moved to the intensive care units at GOSH, that is because your baby is very poorly. The mortality is thought to be under 10 per cent (fewer than 1 in 10 babies affected will die).

There are undoubtedly some after-effects from the lack of oxygen to the brain during the illness, and up to a quarter of babies affected will have some impairment because of their illness as they grow older. This includes difficulties such as learning problems and deafness.

Further information and support

BLISS is the main UK organisation offering support and advice to anyone with a premature or sick baby. Call their helpline on 0500 618 140 or visit their website at www.bliss.org.uk





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Ref: 2016F1863

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