Congenital Diaphragmatic Hernia (CDH)

This leaflet offers more information about congenital diaphragmatic hernia. If you have any further questions or concerns, please speak to the staff member in charge of your baby's care.

What is a congenital diaphragmatic hernia?

A congenital diaphragmatic hernia (CDH) is when the diaphragm (the muscle that separates the chest and the abdomen) has not formed completely, leaving an opening allowing abdominal contents (bowel, spleen, liver, and stomach) to enter the chest cavity impacting on the growth and development of the lungs.

This is a serious and potentially life-threatening diagnosis. The lungs will be smaller than expected (pulmonary hypoplasia) having less space to develop and the blood vessels can also be less developed causing a high blood pressure within the lungs (pulmonary hypertension).

CDH occurs in 1 in 2,500 babies. There is currently no known cause or risk factor. In over 80% of cases the defect is on the left.

Diagnosis and during your pregnancy

CDH is frequently identified during pregnancy, usually on your routine 20-week antenatal ultrasound scan. This may occur in a regional centre. You will be referred to a specialist foetal medicine unit (FMU) to have further scans – St. George's is one of these centres.

The scans will ensure there is evidence of a CDH and look for markers of severity such as:

1. Extent of pulmonary hypoplasia (small lungs)
2. The lung to head ratio (LHR) and/or observed / expected lung to head ratio
   (O/E LHR)
3. Whether there is liver in the chest.

You will be offered further screening, which may involve taking a sample of amniotic fluid to test the baby's chromosomes and a detailed scan of the baby's heart. This scan is called an ECHO.

For the remainder of your pregnancy, you will have four weekly scans which may become more regular closer to the delivery date. These will monitor the growth of your baby and the amount of amniotic fluid around the baby.

Foetal Intervention

Foetoscopic endoluminal tracheal occlusion (FETO) is a foetal surgery procedure that may improve outcomes in babies with severe cases of CDH.

In severe cases, CDH can lead to serious disease and death at birth. For these babies, treatment before birth may allow the lungs to grow enough before birth so these children are capable of surviving and thriving. Your consultant in the Foetal Medicine Unit (FMU) will advise you regarding pre-natal intervention and a paediatric surgeon will discuss this treatment option with you during your evaluation to determine if it may be appropriate for your baby.

Delivery

How and when your baby needs to be delivered will be discussed with you.

You should be able to deliver your baby normally unless there are other reasons for requiring a caesarean section. When considering

your delivery, we need to make sure the neonatal unit has a cot available to ensure your baby's safety.

Babies born with CDH can need very intensive care and their condition can change very quickly. It is critical that you deliver within a hospital which has all aspects of specialised postnatal care immediately available in one location. St. George's Hospital is one of these centres with surgical tertiary neonatal specialists.

Your baby will be transferred to a large cot in the delivery room (Resuscitaire) and they will immediately have a breathing tube placed, which will be connected to a ventilator (breathing machine). They will also have a tube placed into the nose (NG) or mouth (OG) that reaches the stomach - to keep it decompressed so the lungs have room to expand during breathing. The baby will be treated with "gentle ventilation" which is a specialised technique of providing breathing support to babies with CDH.

You may not be able to hold your baby after birth, but once they are stabilised, you will have the opportunity to see your baby briefly before transfer. It may be a few hours before you can be next to your baby as the team will be working to stabilise them. This will include careful management of ventilation, the placement of lines (often including umbilical lines), X-rays, blood tests and other monitoring procedures.

You will be welcome to visit the neonatal unit as soon as you are able to do so.

Taking care of your baby over the next few days

The first 24 hours in NNU can be a critical time for your baby. He or she will need constant monitoring by the doctors and nurses.
Your baby will remain on the ventilator until after surgery and may require additional drugs to support their blood pressure and to ensure they have enough oxygen reaching their tissues. This is

needed because their lungs have not developed properly (pulmonary hypoplasia) during the pregnancy, as the abdominal contents were in the chest cavity and occupying space. Your baby may also receive an additional gas through the ventilator called Nitric Oxide (INO), which can help relieve pulmonary hypertension, if it is present. It is likely that your baby will be on medication to keep them very still (a muscle relaxant). Your baby may need Echo (scan of their heart) within the first few days of life.

If your baby doesn't respond well to initial ventilation, the mode of ventilation can be changed to high frequency oscillatory ventilation (HFOV) which delivers breaths and pressure differently to a conventional ventilator, causing the chest of the baby to "wobble" rather than the "normal" chest movement.

Very occasionally, if your baby is very unstable, they may be considered for extracorporeal membrane oxygenation (ECMO). ECMO allows the lungs to rest while a machine takes over the function of the heart and the lungs, delivering oxygen to the baby while removing carbon dioxide, the waste product of breathing. If this is the case your baby would need transfer to an ECMO centre.

Feeding

Your baby will not be able to receive any milk until after the surgery to repair the hole in their diaphragm. If you wish to breastfeed, the nursing staff will teach you how to express and store your milk, as this will be used to feed your baby when they are ready.

Your baby will initially receive their nutrition (parental nutrition) from a drip through a long line (a line that is placed in a small vein until it reaches a larger vein).

Surgery

The timing of surgery will depend on how much ventilatory and blood pressure support your baby requires. The operation is not performed

until your baby is stable and some of the additional life support has been reduced. This period of stability may take up to five to seven days.

The surgeon will discuss with you the type of operation needed to return the bowel and any other abdominal contents back to the correct place and repair the hole in the diaphragm. Depending on the size of the hole, the surgeon will decide whether to suture (stitch) the hole or to use a patch to repair the hole. The patch is made of a special material called ‘Gore-Tex®’ and should not cause any problems in the long term. This will be determined at the time of surgery. Your baby will still require ventilation and intensive care support following surgery. This will be reduced as your baby’s condition improves.

What are the long-term effects and after care?

This depends on how early the CDH was diagnosed and how much of the lungs was affected before birth. There may be no long-term effects and although most babies born with CDH can seem behind children of their age with regards to growth and developmental milestone, they do catch up in the long term. There will be regular check-ups by the surgical, respiratory and neonatal teams at St. George’s Hospital following their discharge from the unit.

Many CDH babies will have gastro-oesophageal reflux. This is a condition where milk can flow back into the oesophagus (food pipe) causing irritation and pain. This will be treated with medicine. This medicine is usually started after the operation and your baby is discharged home while taking it and should be continued until s/he is six to twelve months old as advised by your surgeon.

Some babies will have chronic lung problems or chest infections and may need repeated hospital admissions. Your baby will be referred to the respiratory team before being discharged home. Sometimes

they will need either oxygen or medications at home to ensure their lung function is well supported.

Occasionally recurrence of the CDH can happen, particularly with large defects necessitating further surgery. Chest wall and spinal problems can arise including pectus excavatum / carinatum, winging of the scapula and / or scoliosis. All of these should be picked up on routine follow up.

If St George's Hospital is not your local hospital

Once your baby has had surgery and made a good recovery, i.e. when their specialist medical and nursing requirements are fewer, the baby will be transferred back to the care of your local hospital. This transfer is a sign of progress and will not occur until the baby is ready. It will allow you to be closer to home and to become familiar with your local healthcare professionals.

Useful sources of information

BLISS

Bliss is a support group which can offer support and advice to families with babies with a range of conditions.

1st Floor North
10-18 Union Street
London
SE1 1SZ

Tel. 020 7378 1122
Email: hello@bliss.org.uk
Website: www.bliss.org.uk
Use your smartphone to scan the QR code (may require downloading a QR scanner.)

CDH UK

CDH UK consists of families, friends and medical professionals affected by Congenital Diaphragmatic Hernia (CDH). “Our website aims to provide support, information and advice on CDH by sharing experiences, providing news and information, raising awareness and working together with families and medical professionals to improve treatments and to further research.” Website: www.cdhuk.org.uk

For more information leaflets on conditions, procedures, treatments and services offered at our hospitals, please visit www.stgeorges.nhs.uk

Additional services

Patient Advice and Liaison Service (PALS)

PALS can offer you on-the-spot advice and information when you have comments or concerns about our services or the care you have received. You can visit the PALS office between 9.30am and 4.30pm, Monday to Friday in the main corridor between Grosvenor and Lanesborough wings (near the lift foyer).
Tel: 020 8725 2453 **Email:** pals@stgeorges.nhs.uk

NHS Choices

NHS Choices provides online information and guidance on all aspects of health and healthcare, to help you make decisions about your health.
Web: www.nhs.uk

NHS 111

You can call 111 when you need medical help fast but it’s not a 999 emergency. NHS 111 is available 24 hours a day, 365 days a year. Calls are free from landlines and mobile phones.
Tel: 111

AccessAble

You can download accessibility guides for all our services by searching ‘St George’s Hospital’ on the AccessAble website (www.accessable.co.uk). The guides are designed to ensure everyone – including those with accessibility needs – can access our hospital and community sites with confidence.

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