The Leeds Teaching Hospitals NHS Trust

Diaphragmatic Hernia

Information for patients

Leeds Maternity Care

This leaflet is aimed at pregnant women with a confirmed congenital diaphragmatic hernia in their baby. The leaflet should only be used to supplement a consultation with an experienced obstetrician.

What is a Diaphragmatic Hernia?

The diaphragm is a sheet of muscle which separates the chest (which contains the heart and lungs) from the abdomen (which contains the liver, kidneys and bowel). A congenital diaphragmatic hernia (CDH) is a defect or hole in this muscle. CDH affects about one baby in 2,500. Each year, in Leeds, we see 15-20 babies with a CDH.

During very early development, the sheets of muscle which form the diaphragm, grow in from the chest wall and meet in the middle. At the same time as the diaphragm is developing, the lungs are starting to form. The precise cause of a diaphragmatic hernia is not known. However, we do know that whatever causes the CDH also disturbs lung growth. The majority of diaphragmatic hernias occur on the left side. Rarely the CDH's are right sided or occasionally central. Bowel or liver herniates through the hole in the diaphragm from the abdomen into the chest.

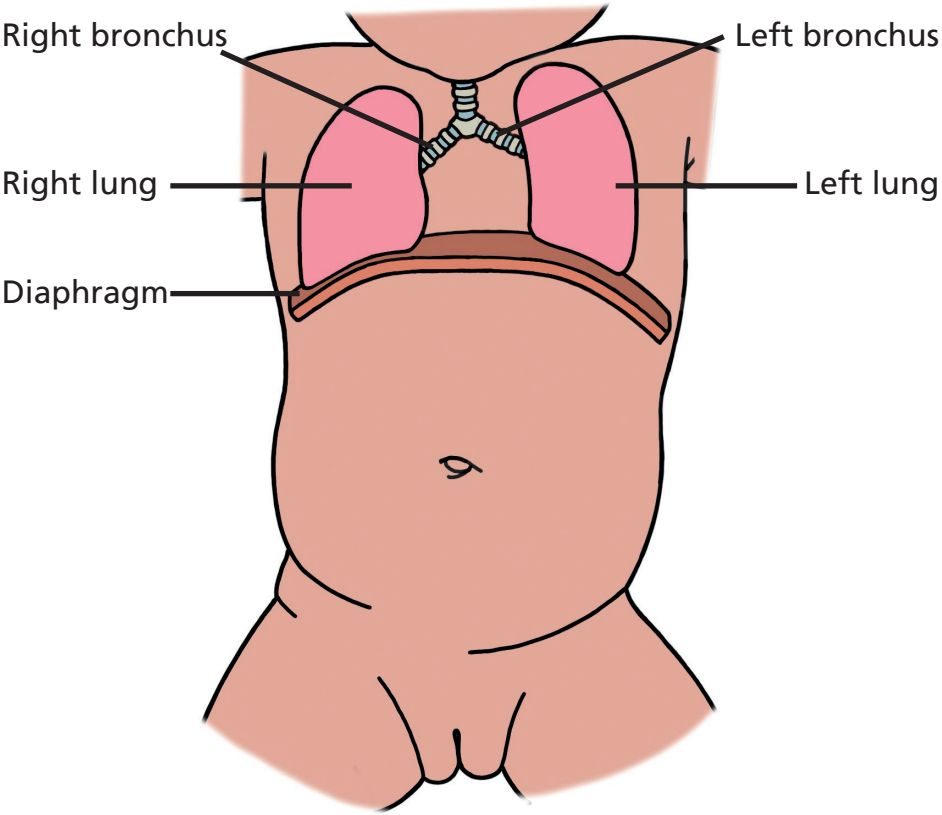
How is it diagnosed?

Most congenital diaphragmatic hernias can be diagnosed before birth using an ultrasound scan.

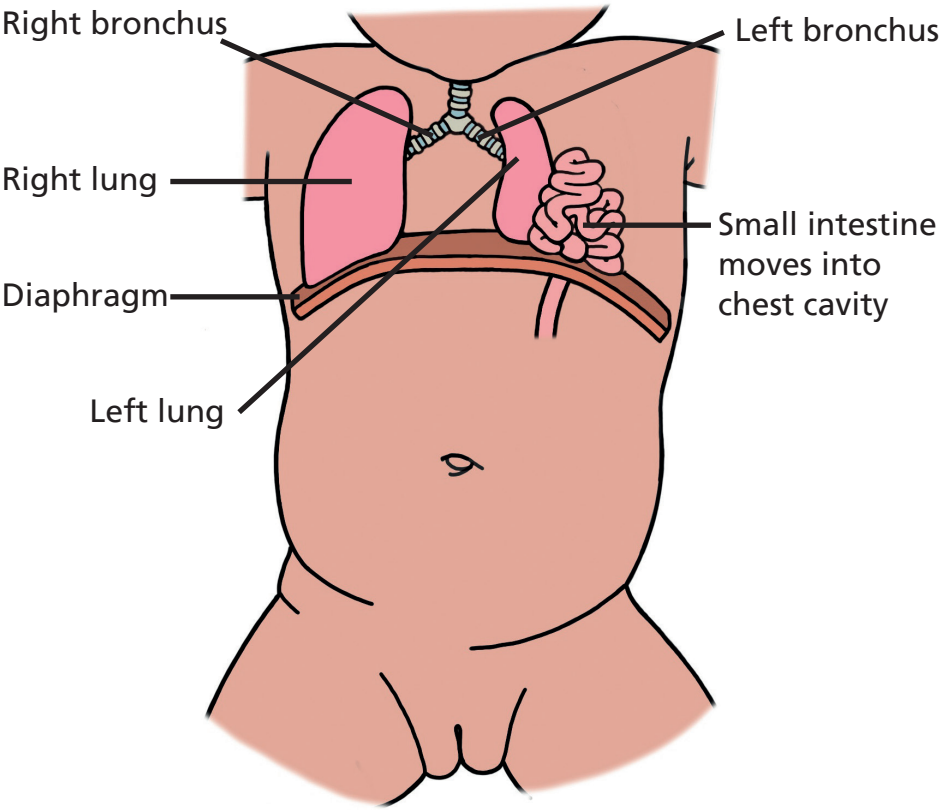
When there is a diaphragmatic hernia the fetal stomach is seen in the chest rather than in the abdomen. Despite careful scanning some diaphragmatic hernias are not picked up until late in the pregnancy or even after birth.

When the diagnosis is not made until the late stages of pregnancy, babies tend to do well.

Normal Chest Cavity



Congenital Diaphragmatic Hernia



What does it mean for your baby?

Associated problems

Congenital diaphragmatic hernia may be an isolated problem or it may occur with other abnormalities.

Cardiac (heart) abnormalities are among the commonest and are found in 25% (1 in 4) of babies with CDH.

In some babies, especially if we find other anomalies, the CDH is part of an underlying problem with the baby's chromosomes and we offer a diagnostic test to see if the chromosomes are normal. Chromosome anomalies affect about 3 in 10 babies with a CDH.

Lung development

Lung development is abnormal in babies with a congenital diaphragmatic hernia. This is called pulmonary hypoplasia.

This is a medical term for very small lungs. Because of pulmonary hypoplasia a substantial number of babies with CDH do not survive more than a few days after birth.

In general terms, the earlier in the pregnancy the CDH is identified, the more severe the pulmonary hypoplasia and the higher the chance of the baby dying

- If the CDH is found before 25 weeks gestation the chance of survival is about 50%, which means that only 5 out of 10 babies will survive
- If the CDH is on the right side of the chest the outlook for the baby is considerably worse, although this is rare
- The presence of other abnormalities in the baby, whether physical abnormalities such as a heart defect, or a chromosomal abnormality, is almost always associated with a very poor outcome with over 90% of these babies dying

At present there is no 100% reliable test which will predict which babies with a diaphragmatic hernia will survive and which babies will not.

We are working on methods to predict pulmonary hypoplasia at an early stage in the pregnancy.

Measuring the size of the baby's lungs in relation to the head (LHR) may be helpful in predicting outcome and this will routinely be measured as part of your baby's scans.

Further investigations

- A detailed anatomy scan will be arranged with one of the Fetal Medicine Consultants in Leeds
- All women whose baby had a CDH are offered a test (amniocentesis or chorionic villus sample) to examine the baby's chromosomes in detail
- If an amnio or CVS is carried out, a chromosome test called a microarray will be done. This test looks at the baby's genetic make-up in fine detail but it is not possible to exclude all genetic problems before birth
- A fetal echocardiogram (scan of the baby's heart) will also be offered since around 25% of babies with CDH also have a heart abnormality
- Regular ultrasound scans are recommended throughout the pregnancy to monitor the baby's growth and the amount of amniotic fluid around the baby

What does it mean for you?

- You will be offered the tests outlined above, all of which will take place at the Leeds General Infirmary

Sometimes a build-up of amniotic fluid (polyhydramnios) occurs which can put you a risk of premature delivery if severe

- Normally we expect mothers carrying babies with a CDH to go into labour at around the time the baby is due. A normal vaginal delivery is best and there is no advantage to either you or your baby in delivery by caesarean section
- We recommend that your baby is born at a hospital with a neonatal surgical unit, which usually means the Leeds General Infirmary. Your baby will need intensive care as soon as he / she is born and, if stable, an operation to repair the diaphragmatic hernia

What will happen to my baby after birth?

Immediate care

A team of neonatal doctors and nurses will be present at the birth of your baby. As soon as your baby is born he/she will be handed to the neonatologist who will start treatment.

This involves placing a tube into the windpipe to help with breathing and giving oxygen. Your baby will be transferred quickly from the Delivery Suite to the Neonatal Intensive Care Unit for stabilisation. We routinely sedate babies with a CDH very deeply because this helps us to take over their breathing with a machine called a ventilator and also because it means that the baby is in no distress.

The first few days

The first 48-72 hours of life are critical. During this period we establish how well the baby's lungs have developed (whether the pulmonary hypoplasia is severe or not). If the baby's lungs are well developed then ventilation is easy and it is easy to maintain a normal level of oxygen in the blood.

We usually wait for 2-4 days before operating to repair the diaphragmatic hernia even if the baby seems to be fine at birth because most babies deteriorate temporarily after surgery.

Surgical repair

Surgery to repair a diaphragmatic hernia is performed under a general anaesthetic in the operating theatre. We usually make an incision 2-3 inches long to the left of the umbilicus (belly button).

The hole in the diaphragm is identified and the bowel drawn out of the chest. The defect in the diaphragm is closed either with sutures or sometimes with a patch of surgical Gore-Tex.

Post-operative care

Most babies stay in hospital for about a month after repair of a CDH. After they come off the ventilator they are usually quite breathless for a couple of weeks and tend to need feeding through a tube.

We encourage mothers to express their milk after they have recovered from the delivery and freeze it so that we can use it to start feeding their baby.

Long-term outlook

Most babies who survive with a CD, grow up to be healthy normal children. In the early years they often suffer from chest infections during the winter but after that they rarely have problems.

Lung growth continues until 7 or 8 years of age and we know that catch up growth means that lung function is usually normal in CDH survivors.

A small number of babies who are very poorly in the early weeks of life will go on to develop long-term problems such as lung disease and/or developmental problems (physical or mental handicap). Until relatively recently these babies would not have survived.

Can anything be done before delivery?

Attempts have been made to repair the defect before delivery by performing a caesarean section, operating on the baby and then closing the uterus. Results from the USA have been disappointing and this procedure has been abandoned.

There is some evidence that blocking the windpipe of a baby before birth causes the lungs to grow at a faster rate than normal. Not all cases are suitable for this procedure however, the Fetal Medicine consultant will discuss this option with you and, if you wish, will refer you to a centre in London for assessment.

What about future pregnancies?

The risk of having further babies with a diaphragmatic hernia is very small for the majority of mothers. We usually quote a risk of recurrence of the order of 1-2% and offer ultrasound scans in any future pregnancies to look specifically for a CDH.

In a small number of families the CDH occurs as part of a genetic syndrome which means that the risk of it happening again may be as high as 1 in 4 (25%). If we suspect that this we will arrange for you to see one of the Consultant Geneticists to discuss things in more detail.

What happens next?

You will have been referred to the Fetal Medicine Unit at Leeds. The Fetal Medicine consultant will discuss and agree the best plan for you and your family, according to your wishes.

You will have the option to continue with the pregnancy and receive care as outlined above or, to end the pregnancy (have a termination) because of the severity of the baby's condition.

An appointment will be made for you to meet with one of the Paediatric Surgeons and for you to look around the Neonatal Intensive Care unit at the LGI.

Where can I get more information and support?

Be sure to ask questions to the doctor supplying you with this leaflet and make a note of any questions you would like to ask at your Fetal Medicine Unit appointment.

Your local hospital will also have a specialist midwife who you will be able to contact for further discussion.

There is a list of useful websites for organisations that provide support to families with a diagnosis of CDH either during a pregnancy or afterwards.

Antenatal Results and Choices

- **www.arc-uk.org**

Total trial

- **www.totaltrial.eu**

Questions / Notes

What did you think of your care?

Scan the QR code or visit **bit.ly/nhsleedsfft**

Your views matter

Developed by: The Fetal Medicine Team
Produced by: Medical Illustration Services • MID code: 20200428_018/MH

LN001586
Publication date
01/2021
Review date
01/2023