Will this happen again?

The chance of this happening again next pregnancy is low, in most cases. Occasionally, where there is an underlying genetic cause found, the risk could be higher, up to 25-50%. Your team will discuss options for testing in order to provide the best information about recurrence risk.

Your options

We acknowledge that a CDH is a very significant finding with considerable impacts for your baby and your family as a whole. This may leave you and your whānau in the very difficult situation of deciding whether or not to continue on with the pregnancy. We aim to provide you with as much information and support in order to make the best decision you can at this difficult time. If you decide not to continue with the pregnancy, your medical team will guide you through the next steps.

Supports available

Talk with your MFM service, LMC or health provider about supports that may be available to you such as:

- Access to counselling, cultural and spiritual supports for you and your whānau;
- Financial help with travel and accommodation when attending MFM specialist appointments away from where you live.
- Parent to Parent New Zealand: www.parent2parent.org.nz
- Facebook group: https://www.facebook.com/CDHNZ/
- SANDS: www.sands.org.nz
  Sands New Zealand is a network of parent-run, non-profit groups supporting families who have experienced the loss of a baby.
- Whetūrangitia: https://wheturangitia.services.govt.nz/
  Information for family and whānau experiencing the death of a baby or child.
- Baby loss NZ: http://www.babyloss.co.nz/practical-info/

Support groups may change over time - check with your MFM team

References

https://www.nationwidechildrens.org/-/media/nch/specialties/fetal-center/congenital_diaphragmatic_hernia_1.ashx

For more information please contact your nearest NZ MFM unit

Auckland
Auckland Hospital
2 Park Road
Grafton, Auckland 1023
Phone: 09 367 0000 extn 24951

Wellington
Wellington Hospital
Riddiford Street
Newtown, Wellington 6021
Phone: 04 806 0774

Christchurch
Christchurch Women's Hospital
2 Riccarton Ave, Christchurch 4711
Phone: 03 364 4557

Health New Zealand
Te Whatu Ora

About Wāhi Rua:

https://www.healthpoint.co.nz/public/wahi-rua-new-zealand-maternal-fetal-medicine/

Updated June 2024

Wāhi Rua
New Zealand
Maternal
Fetal Medicine
Network

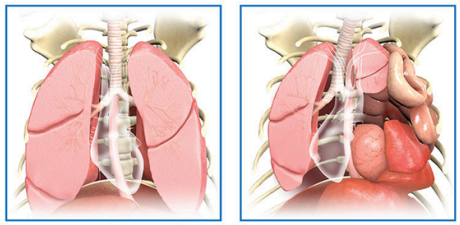
Congenital Diaphragmatic Hernia

Congenital diaphragmatic hernia

A **congenital diaphragmatic hernia** (CDH) is a hole in the sheet of muscle (the **diaphragm**) which separates the chest (which contains the heart and lungs) from the abdomen (which contains the liver, kidneys and bowel).

Usually, during very early development of the baby/pēpi in the womb, the sheets of muscle which form the diaphragm grow in from the chest wall and meet in the middle. In CDH, this process is disturbed and the muscles do not form as they should, leaving a gap in the muscle layer. This gap allows the contents of the abdomen to protrude through into the chest – this is called a hernia. This can include stomach, bowel, liver, and sometimes spleen and kidney. The exact reason a CDH occurs in some babies as they develop is not known.

CDH affects about one baby in 2,500 (15-20 babies per year) in Aotearoa New Zealand.



Normal Anatomy

Congenital Diaphragmatic Hernia

Diagnosis of CDH

Many cases of CDH are detected before birth on routine ultrasound scans. However, despite careful scanning, some CDH are not picked up until late in the pregnancy or even after birth.

Common signs of CDH on ultrasound include a finding of a fluid filled structure within the fetal chest, or abnormal appearance of the tissue within the chest. In some cases, the heart may be pushed to one side. On more detailed scanning it is then identified that these structures represent the stomach and bowel abnormally positioned within the chest, and a defect in the diaphragm is able to be identified. The majority of CDH occur on the left side but can sometimes be right sided or central.

If a CDH is suspected, you will be referred to your regional maternal fetal medicine service. You will be offered an appointment in the fetal medicine clinic.

What will happen at your fetal medicine appointment?

At your fetal medicine appointment, a detailed scan will be performed. You will have the opportunity to discuss with your specialist what this condition is and what having a baby with CDH might mean for you and your whānau. A detailed ultrasound scan is able to provide more information on the likely effect on your baby. It will assess the contents of the hernia itself, the size of the lungs, and any effect on heart function. It will also look in a lot of detail for whether there might be any other differences in baby's anatomy, especially heart differences, which can sometimes be associated with CDH. Your specialist will discuss the option of additional testing for genetic/chromosomal conditions which can be associated with CDH in about 1 in 10 cases. You may also be offered an MRI scan to try to see more detail.

How will this affect my baby and the rest of my pregnancy?

Having CDH can affect your baby's chance of life. The overall survival rate for babies with a CDH is up to 70%. Many babies born with CDH have good outcomes after the CDH has been surgically repaired. At present there is no 100% reliable test which will predict which babies with a CDH will survive and which babies will not.

It is important to know however, that having additional severe lung, heart or chromosome concerns are almost always associated with a very poor outcome, with over 90% of these babies passing away in the womb or soon after birth.

Are there any treatment options in pregnancy?

In cases diagnosed with severe CDH (a large amount of abdominal contents in the chest, leaving only a small amount of normal appearing lung), the overall survival may be predicted to be quite low. In very selected cases, it may be appropriate to consider a treatment called FETO, where a balloon is placed inside baby's windpipe by "fetoscopic" surgery (with baby still in the womb). This treatment is not performed in New Zealand, but may be arranged in Australia. If your fetal medicine team finds that you and your baby meet the criteria for this treatment to be considered, they will discuss with you in great detail all the pros and cons of this option. It is important to understand that this can be quite complex and should not be considered a "cure" for the condition.

How will my baby be managed during pregnancy?

We will monitor your baby's growth and wellbeing with regular ultrasound scans. Normally, we expect those carrying babies with a CDH to go into labour at around the time the baby is due.

However, sometimes a build-up of amniotic fluid (polyhydramnios) occurs which could put you at risk of premature delivery.Your medical team will discuss birth plans with you. Most often a normal vaginal delivery is possible, however in some cases a caesarean section may be recommended. While you are still pregnant, your maternal fetal medicine service will make an appointment for you to meet one of the paediatric surgeons and/or a neonatologist to discuss your baby's treatment after their birth.

What will happen to my baby after birth?

Your baby will need to be born at a hospital with a neonatal intensive care unit (NICU) so that the medical team are there to help with your baby's breathing at birth. Along with the main team looking after you during pregnancy, a paediatrician or neonatologist and a neonatal nurse will need to be present at the birth of your baby so that they can give specialist care to your baby as soon as they are born. As soon as your baby is born, they will be handed to the paediatrician who will start treatment. This involves placing a tube into the trachea (windpipe) to help with breathing and giving oxygen. Sadly, you will not be able to hold your baby until they are stabilised.

Your baby will be transferred quickly from the birthing suite to the NICU 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 48-72 hours of baby's life are critical. During this time we establish how well their lungs have developed. If their lungs are well developed then ventilation is easy and it is easy to maintain a normal level of oxygen in the blood.

We encourage you to express your breast milk after you have recovered from the delivery and freeze it so that we can use this to start feeding your baby.

Surgery to repair CDH

Once stable, your baby can have an operation to repair the diaphragmatic hernia. This is usually performed at 2-4 days of age. Surgery to repair a diaphragmatic hernia is performed under a general anaesthetic in the operating theatre. A surgical cut is made, the hole in the diaphragm is identified and the bowel is drawn out of the chest. The structural difference in the diaphragm is closed either with sutures or sometimes with a patch.

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.