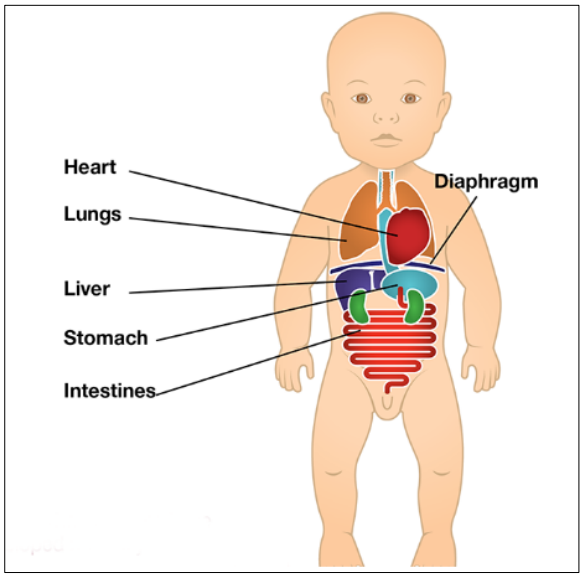
Congenital Diaphragmatic Hernia (CDH)

Whānau/Family Information – Neonatal Services

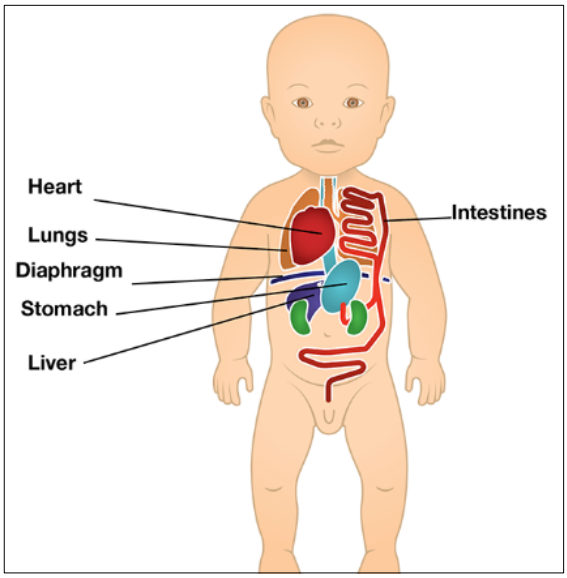
CDH is a is a life-threatening condition where the pēpi/baby has a hole in the layer of muscle that separates the chest and the abdomen (tummy). This muscle is called the diaphragm and helps us to breathe. It also keeps the heart and lungs separate from the abdominal organs, such as the stomach, liver and bowel.

If there is a hole in this muscle, the abdominal organs can move up (herniate) into the chest, putting pressure on and squashing the developing heart and lungs. Because of this, your pēpi could have problems with breathing at birth and it may take time to establish feeding. The lungs will be smaller (pulmonary hypoplasia) than expected and cause breathing problems after your pēpi is born. In the womb pēpi get oxygen from the māmā/mother's bloodstream through the placenta. After being born, the lungs need to provide the body with oxygen, so organs can work properly. If the lungs are small or do not have much space to expand, they may not work properly.

Sometimes the pēpi heart is also pushed over to the 'wrong' side of the chest (as shown below). This can affect the growth of the lung on the opposite side.



Pēpi whose organs have developed as expected



Pēpi with congenital diaphragmatic hernia

What causes CDH?

It is not caused by anything you have or have not done.

It is a rare serious condition that has several potential complications, and a significant risk of death. It affects approximately one in 2,500 births with 80% occurring in the left side of the chest.

CDH can be an isolated condition (which means no other condition occurs with it), but it is also more common in pēpi who may have a heart defect or those who have a gene or chromosome condition. You will be able to discuss your individual circumstances with a specialist team.

How will CDH be diagnosed?

This condition can often be diagnosed antenatally by an ultrasound scan, by the presence of abdominal organs in the chest or polyhydramnios (increased fluid around the pēpi). Sometimes though it is not diagnosed until after pēpi is born.

Once the findings have been found on ultrasound scanning, several people will review the ultrasound pictures and become involved in the care of you and your pēpi before, during and after delivery. The team of people will include Foetal Anomaly Advice Committee (deals with high-risk pregnancies). Neonatologists (specialised pēpi doctor) Paediatric Surgeons (specialised in children's surgery) Obstetricians (care for women and pēpi during pregnancy, labour and delivery) and Geneticist (studies genes). A social worker will offer support during your stay in the Neonatal Unit.

Where should I have my pēpi?

A meeting will be arranged for you to meet with the Paediatric Surgeon and Neonatologist to discuss the condition, possible complications and treatment. It is necessary for your pēpi to be born at Christchurch Women's Hospital where we are experienced in caring for pēpi with CDH. You will have the opportunity to look around the Neonatal Intensive Care Unit (NICU) with a member of the nursing staff before your pēpi is born.

What will happen after birth?

Your pēpi will require immediate specialised treatment after being born, including being placed on a machine to help with their breathing (ventilator). So, you may not hear your pēpi cry or hold your pēpi immediately after birth. They will also have a tube placed into the nose (NG) or mouth (OG) to the stomach to keep it decompressed (reduce pressure) so the lungs have room to expand during breathing.

They will be transferred to NICU where special lines will be placed into the blood vessels of the umbilical cord to monitor different blood levels and provide fluids and medications. Additional intravenous lines will be placed in the pēpi hands or feet if needed. Heart scans are done frequently to check on how well it is functioning, and which medications are needed to support the heart.

Your pēpi will require surgery to repair the hole in the diaphragm. The timing of surgery will depend on the amount of lung damage from this condition as well as the consequences of abnormal blood flow between the heart and lungs. The majority of pēpi will require several days of stabilisation before surgery. The circulation through the lungs can be a problem in some pēpi and they may require additional drugs (Nitric Oxide). The best outcome for these children will be achieved by improving their underlying heart and lung function before proceeding with surgery.

What happens before the operation?

The surgeon and anaesthetist will explain about the operation and anaesthetic and discuss any worries you have. They will ask you to sign consent forms giving permission for the operation and general anaesthetic.

You will be able to go the anaesthetic room with your pēpi and stay until they are asleep. Sometimes the surgery takes place in NICU. The surgeon will phone you once the surgery is finished to update you.

What will happen after surgery?

After the operation, your pēpi will return to NICU and be monitored closely and will receive support with their breathing until the lungs recover. This period of support depends on how the lungs and heart were affected. Pain relief drugs will be given through an intravenous infusion. A morphine infusion is given for pain relief. Midazolam infusion can help if they are distressed with their cares. A small catheter (tube) may be inserted into your pēpi bladder to drain urine for a while.

Before and after the operation, your pēpi will be fed intravenously (straight into a vein). This is called total parental nutrition (TPN) and will be continued until your pēpi is ready to have small amounts of either breastmilk or formula through the NG or OG tube. As the milk feeds increase the TPN will decrease until your pēpi is fully milk fed. Once your pēpi is ready to feed by mouth you will receive lots of help and support.

If your pēpi has CDH it is perfectly normal to worry. All the monitoring, intravenous infusions, tubes and the number of people involved in your pēpi care can be very daunting. You will receive lots of guidance and support but please do not hesitate to ask your nurses and doctors if you have any questions or concerns and what you can do to care for and bond with your pēpi.

CDH can be relatively straightforward to treat, but others are very complicated. Survival will depend on the side affected, and the severity of the heart and lung changes. They can also have several medical issues including gastro-oesophageal reflux (stomach contents are released back up towards the mouth causing pain and irritation), delayed oral feeding due to rapid breathing. Your pēpi development will be checked in paediatric follow-up. Yearly surgical follow-up will occur for a few years.