Exomphalos / Omphalocele

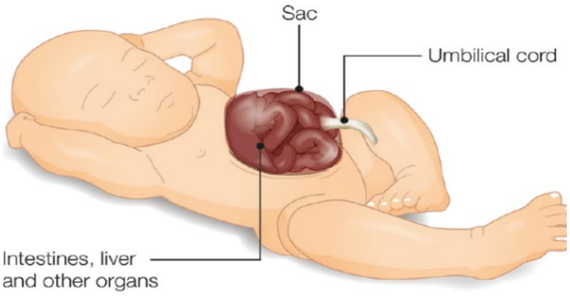
Whānau/Family Information – Neonatal Services

A scan has shown that your pēpi/baby has a condition called Exomphalos (also known as an omphalocele). This means that some, or a lot, of the contents of your pēpi tummy are lying in the umbilical cord.

This leaflet provides information to support the discussions you will have with health professionals about your care and the care of your pēpi both during pregnancy and after your pēpi birth.

What is an exomphalos?

Exomphalos occurs when a pēpi abdominal (tummy) wall does not fully close around the base of their umbilical cord (where the tummy button will form). This allows the organs that usually grow inside the abdomen to grow outside of their abdomen in a protective membrane (loose sac) that surrounds the umbilical cord.



Exomphalos usually affects the pēpi bowel but it may affect their liver and other organs. The size of pēpi exomphalos/omphalocele will depend on the number of organs growing outside of their abdomen.

What causes an exomphalos?

The cause is not always known. It occurs in about 4 babies out of every 10,000 (0.04%) births. Many pēpi born with an exomphalos also have other problems (eg. with chromosomes or the heart). Regular ultrasound scans, including a detailed look at the heart, will be done during your pregnancy to assess your pēpi wellbeing. You may have an amniocentesis to assess for a genetic problem.

Where should I have my pēpi?

You will get to meet several people who will become involved in the care of you and your pēpi before, during and after delivery. These include Neonatologists (specialised pēpi doctor) Paediatric Surgeons (specialised in children's surgery) Obstetricians (care for women and pēpi during pregnancy, labour and delivery). Progress is discussed at the Foetal Anomaly Advice Committee (deals with high risk pregnancies).

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 the neonatal unit is experienced in caring for pēpi with exomphalos and has paediatric surgical services available. 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 around the time of birth?

It is usually possible to give birth to your pēpi vaginally. Other health reasons may indicate the need for a Caesarean and your obstetrician will decide on this with you.

When your pēpi is born they will be cared for by the neonatal team who will assess his/her wellbeing. Your pēpi may need breathing support. The exomphalos will be wrapped in see through plastic to prevent heat loss and protect the membrane. A tube will be passed through your pēpi nose or mouth, down into their stomach to keep it empty of air and fluid. Your pēpi will then be transferred to the NICU and put in an incubator where he/she will be monitored. Intravenous lines (drip) will be placed in the pēpi hands or feet, so fluids can be given directly into their veins to make sure they are hydrated. Your pēpi may need an echocardiogram (ECHO) which checks the heart's structure and function.

The neonatal team will explain the reason for any treatment your is receiving and they will also be happy to answer any questions you may have.

When will my pēpi have surgery?

Your pēpi will need to have an operation to place the contents of the exomphalos inside their abdomen and repair their abdominal muscles and skin. If the exomphalos is small, it is possible to do this in one operation.

If the exomphalos is too big to close at birth, or if there is too much bowel or liver outside the tummy, more than one operation may be needed. The first operation sometimes involves placing the exomphalos within a temporary transparent envelope called a silo. The silo is made smaller over 5 to 7 days, so that the abdominal contents are gradually pushed back inside the abdomen. A full or partial repair of the hole then occurs. Sometimes surgery will be delayed longer than this.

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 (medicines used to send your pēpi asleep).

What happens after surgery?

After the operation, your pēpi will be returned to NICU where you will be able to see your pēpi. Your pēpi will be monitored closely and will initially be on a ventilator (breathing machine) until they are ready to breathe on their own.

Pain relief drugs will be given through an intravenous infusion. A small catheter (tube) may be inserted into your pēpi bladder to drain urine for a while.

Feeding: Your pēpi will continue to be fed intravenously. 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 a tube going through your pēpi mouth or nose into their stomach. As the milk feeds increase the TPN will decrease until your pēpi is fully milk fed.

Once the bowel has fully recovered your pēpi should be ready to breast or bottle feed. You will receive lots of help and support with feeding.

How long will my pēpi stay in hospital?

It is difficult to say how long your pēpi will be in hospital, the length of stay will depend on how big the exomphalos was, and whether there is additional organ, eg. heart or chromosome problems.

It is perfectly normal for you to feel anxious and worry. All the monitoring, intravenous infusions, tubes, the length of time in hospital 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, doctors and NICU social worker if you have any questions or concerns and what you can do to care for and bond with your pēpi.

Follow up

If at any time following discharge from NICU you have concerns about your pēpi it is important you contact your GP or hospital immediately if your pēpi is not well and vomits especially with bile (green) as sometimes adhesions, a kink or obstruction in the bowel can occur.

To monitor your pēpi progress regular outpatient check-ups will be arranged with a paediatrician close to where you live. The paediatric surgical team will also see your pēpi to check for wound healing and the appearance of the area where the operation took place. This can usually be done at your local hospital.

At these check-ups, please ask questions and express any concerns you may have.