NTS Clinical Guideline

Title:	Exomphalos initial stabilisation and transfer		
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Description

Congenital defect of the anterior abdominal wall where it joins with umbilicus, resulting in herniation of abdominal contents through the umbilicus. The abdominal contents are covered by a sac. If the defect is < 5 cm it is called Exomphalos Minor and if it is > 5 cm then it is called Exomphalos Major, which may contain liver within the sac. The abdominal cavity is small in size as the contents are lying outside.

Incidence 1 in 5000

Associated with other anomalies in about 40-50% cases (e.g. trisomies, cardiac defects, G.I. and renal anomalies, Beckwith Weidemann syndrome, Pentalogy of Cantrell)

Survival rates are dependent on whether other anomalies are present.

Stabilisation and management

- Careful clamping of cord, as some bowel may be present at base of the umbilical cord. The clamp should ideally be placed distal to where the normal cord starts to prevent any bowel injury.
- Initiate resuscitation as required. **Avoid prolonged mask ventilation**.
- Nurse in supine position. Wrap abdomen of baby in cling film with gut lying well supported.
- Pass large NG tube size 8 or 10F, place on continuous low suction (5-10 kPa) or leave on free-drainage and aspirate hourly. Distal end of the nasogastric tube should be lower than the baby.
- Nil by mouth
- Peripheral vascular access x 2. Do not try umbilical lines.
- Maintenance IV fluids. Replace NG losses ml/ml with 0.9% saline and Potassium Chloride (10 mmol / 500 ml)
- Monitor temperature
- Carefully monitor blood sugars for hypoglycaemia - association with Beckwith-Weidemann syndrome
- Consider antibiotics if risk factors for sepsis.
- Look for other anomalies.