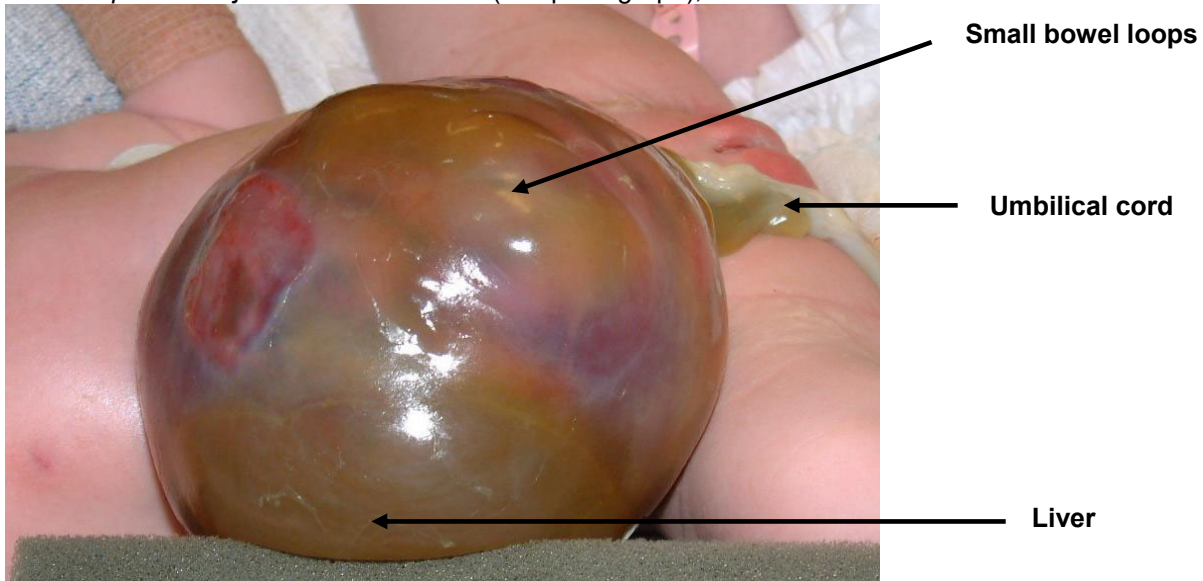


# EXOMPHALOS – INITIAL MANAGEMENT • 1/3

## DEFINITION

Congenital anterior abdominal wall defect, resulting in herniation of the abdominal contents through the umbilicus. Herniated viscera are covered by a sac

- *Exomphalos minor*: no liver in sac, defect diameter <5 cm
- *Exomphalos major*: sac contains liver (see photograph), defect diameter ≥5 cm



- Key issues to be aware of:
  - rupture or damage to protective sac
  - association with other major abnormalities (cardiac or chromosomal)
- Depending on individual patient factors, an exomphalos can be managed either by:
  - early surgical closure of the defect (as a neonate)
  - delayed surgical closure, after epithelisation of the sac using dressings

## Diagnosis and antenatal care

- Majority diagnosed antenatally
- Often associated with chromosomal and other abnormalities
- Multi-professional discussions needed to carefully plan antenatal and postnatal care
- If suspected antenatally
  - refer to **fetal medicine department** for further assessment
  - refer to **paediatric surgery** for antenatal counselling
- Give parents information leaflet
- Aim to deliver in hospital with appropriate **NNU** with either postnatal transfer to **paediatric surgical unit** or management by **paediatric surgical outreach team** at the **NNU**

## Pre-delivery

- Liaise with **on-call team** at **paediatric surgical centre** before making arrangements for elective delivery

## Delivery

- Experienced paediatrician/ANNP to attend delivery
- Clamp umbilical cord only after careful assessment of the umbilical defect (to avoid any bowel present at base of cord)
- Use plastic cord clamp (not artery forceps) on umbilical cord ≥10 cm away from where normal umbilical cord starts to avoid bowel injury
- Dry baby
- Provide resuscitation as required. Avoid prolonged mask ventilation
- Nurse in supine position
- Pass a size 8 Fr nasogastric tube (NGT) and fix securely with tape (see **Nasogastric tube insertion guideline**)
- Empty stomach by aspirating NGT with 10–20 mL syringe. If <20 mL fluid aspirated, check position of tube. Place tube on free drainage by connecting to a bile bag
- Put nappy on baby, taking care to fold it down under the defect

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- Place baby's legs and trunk, feet first, into a sterile plastic bag, to protect the defect and reduce fluid loss. Pull the draw-string under the arms, so that both arms are outside the top of the bag
- Show baby to parents and transfer to **NNU**

## In **NNU**

- Careful physical examination by experienced neonatal practitioner. If baby has a major lethal congenital abnormality, local consultant to decide whether referral for management is appropriate. May require discussion with **on-call consultant surgeon**. If the decision is not to transfer, inform **surgical unit**
- Nurse in supine position
- Insert IV cannula. Avoid vein which could be used for long line e.g. antecubital fossa, long saphenous or scalp
- Avoid umbilical lines
- Take blood for:
  - culture
  - FBC, CRP and clotting screen, including fibrinogen
  - U&E
  - blood glucose and venous blood gas
- Crossmatch sample will be taken at **surgical centre**
- Send 1 bloodspot on neonatal screening card marked as 'pre-transfusion' (for sickle cell screening) with baby to **surgical centre**
- Administer fluid boluses as indicated by baby's condition
- Start maintenance IV fluids (see **Intravenous fluid therapy** guideline)
- Give vitamin K (see **Vitamin K** guideline)
- Leave NGT on free drainage and aspirate NGT 4-hrly with a 20 mL enteral syringe
- Replace nasogastric losses mL-for-mL using sodium chloride 0.9% IV with potassium chloride 10 mmol in 500 mL bag
- Start broad spectrum antibiotics (see **Neonatal Formulary**) including metronidazole IV
- Monitor blood glucose 4–6 hrly
- Swab sac and send for culture and sensitivity
- Take a photograph of the exomphalos, with parent's consent
- Remove bowel bag and protect the sac by covering with a non-adhesive dressing (Jelonet) and sterile gauze, until assessed by **paediatric surgical outreach team**
- Discuss baby's condition and treatment plan with parents and ensure they have seen the baby before transfer. Take photographs for parents

## Referral

- Refer baby to planned **paediatric surgical unit e.g. BCH**. This may require a conference call with **on-call surgeon** to discuss urgency of transfer; an emergency surgical procedure is normally not indicated
- Some babies may not require transfer to **paediatric surgical unit** and can sometimes be managed on **NNU**
  - **for BCH this may include transfer to BWH for neonatal surgical outreach service**
- Obtain sample of mother's blood for crossmatch
  - sample tube must be clearly hand written and labelled with mother's name, date of birth, NHS number and date and time of collection
  - complete form
    - add baby's details to ensure it is clear sample relates to mother of baby being transferred (this information is required by **surgical unit** blood bank)
- Complete nursing and medical documentation for transfer. Electronically transfer any X-rays to **surgical unit** (or obtain copies of X-rays)
- Ensure mother's details included (including ward phone number if an inpatient and own number if discharged) as if operation necessary and an individual with parental responsibility unable to attend **surgical unit**, surgeon will require verbal telephone consent
- Ensure baby's documentation includes:
  - whether vitamin K has been given
  - name of referring consultant
  - whether parents received antenatal counselling
  - mother's name, ward (if admitted) and her contact details
- If the neonatal surgical decision is to perform a delayed closure of the exomphalos, the recommended dressing is manuka honey gel covered with a honey net dressing, sterile gauze and crepe bandage
- If exomphalos is to be managed with dressings on **NNU** this will be supported by the **surgical neonatal outreach service**

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## While awaiting transfer

- Reassess hourly for further fluid boluses and, if necessary, give either sodium chloride 0.9% or human albumin solution (HAS) 4.5% 10 mL/kg
- If evidence of a coagulopathy, treat appropriately (see **Coagulopathy** guideline)
- Aspirate NGT 4-hrly
- Replace nasogastric losses mL-for-mL with sodium chloride 0.9% IV with potassium chloride 10 mmol in 500 mL bag. Leave NGT on free drainage

## Transfer to **surgical unit**

- Place baby in transport incubator
- Take baby to parents (if not yet seen) in transport incubator, en-route to the ambulance
- Ensure mother's blood, baby's pre-transfusion bloodspots, letters for **surgical team** and all documentation accompany baby
- Make and document all usual observations during transport and on arrival at **surgical unit**

## Useful Information

- <https://bwc.nhs.uk/download.cfm?ver=3512>