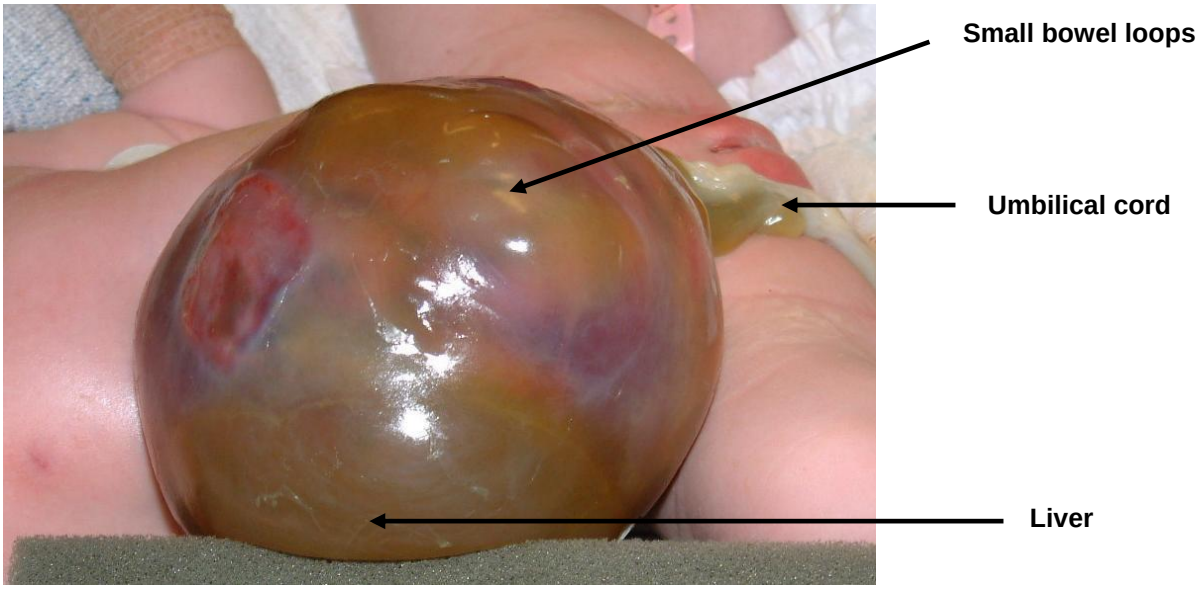
EXOMPHALOS – INITIAL MANAGEMENT ● 1/3

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

- http://www.bch.nhs.uk/content/neonatal-surgery
- http://www.bch.nhs.uk/find-us/maps-directions
- Parent information: https://bwc.nhs.uk/download.cfm?doc=docm93jijm4n2517