



**CLINICAL PRACTICE GUIDELINE**

Guideline coverage includes NICU KEMH, NICU PCH and NETS WA

# Exomphalos (Omphalocele)

This document should be read in conjunction with the [Disclaimer](#)

## Definition

- An exomphalos is herniation of abdominal viscera through a central abdominal wall defect.
- The herniated viscera is covered by 3 layers: peritoneum, amnion and Wharton’s jelly.
- Exomphalos is different from a gastroschisis in that it has a membrane that covers the abdominal contents and is more likely to have associated anomalies or be part of a syndrome.
- **Embryology:** During the 6<sup>th</sup>-10<sup>th</sup> week the foetal intestine migrates through the umbilical ring into the cord, and then returns to the abdominal cavity. Failure of viscera to return results in an exomphalos.

## Types

1. **Exomphalos minor** where the opening is less than 4 cm and only contains the intestine.
2. **Exomphalos major** where the opening is greater than 4 cm and/or with the liver inside the cord.

## Antenatal Diagnosis

In most cases, exomphalos is seen on pre-natal ultrasound with associated anomalies investigated.

## Associated Anomalies

- Associated anomalies are observed in up to 72% of infants and it is commonly associated with chromosomal defects or part of a syndrome.
- Of infants with normal karyotypes, nearly 80% have multiple other anomalies.
- Multiple anomalies are more common with minor (< 4 cm) versus major exomphalos (55% vs 36%).

Associations	Notes
Chromosomal abnormalities	Trisomy 13, 14,15, 18 and 21
Syndromes	Beckwith-Wiedermann, Pentalogy of Cantrell, lower midline syndrome (bladder/cloacal extrophy, imperforate anus, meningomyelocele)
Cardiac anomalies	Cardiac anomalies seen in up to 20% of cases
Pulmonary hypoplasia	Commonly associated with exomphalos major (20%)
VACTERL anomalies	
Nervous system	Holoprosencephaly and anencephaly

## At Birth

- Neither vaginal delivery nor caesarean section has been shown to be superior.
- In exomphalos major, caesarean section may reduce liver injury or sac rupture during vaginal delivery.
- Medical staff should attend the resuscitation. A consultant should be present where possible if an exomphalos major is expected.
- These infants may have unsuspected pulmonary hypoplasia requiring early intubation and ventilation.
- Fluid resuscitation may be required early especially if there is rupture of the covering sac.
- Congenital heart disease should be suspected if the infant is cyanosed/not responding to resuscitation.

## Post-Resuscitation Care Should Focus On:

1. Care of the exomphalos, covering sac and blood supply:
  - A rupture in the sac should be managed as a gastroschisis with urgent surgical referral and transfer to 3B.
  - The bowel should be decompressed with a large bore gastric tube.
  - An intact sac can be dressed with saline-soaked gauze and impervious dressing to minimize fluid loss (**discuss with surgical team first**).
  - The underlying viscera should be inspected for perfusion and colour.
2. Fluid resuscitation:
  - Despite having a covering sac, these infants have higher fluid losses.
  - If there has been a rupture in the sac then fluid management is the same as gastroschisis.
  - All infants should be commenced on 80-100 mL/kg/day of maintenance fluid (10% glucose/0.22% saline).
  - Normal saline boluses may be required if perfusion worsens.
3. Temperature regulation:
  - These infants lose heat through the exomphalos so temperature control is very important.

## Investigation for Chromosomal Anomalies or Syndromes

- A full karyotype should be sent.
- Consider Beckwith Weidemann in infants with hypoglycaemia or are large for gestational age.
- The following investigations should be considered - chest X-ray, spinal series, abdominal & renal ultrasound, head ultrasound, echocardiography, limb X-rays (where appropriate).

## Surgical Management

- Surgery should be considered electively in all with an intact sac.
- The infant should have an urgent surgical referral if a rupture of the sac has occurred.
- Treatment depends on the size of the defect, gestational age and presence of associated anomalies.

- Small defects may be repaired with inversion of the sac and primary closure of the fascia and skin.
- In larger defects:
  - Primary closure may be difficult due to excessive increase in intra-abdominal pressure.
  - Options include use of flaps, patches, negative pressure dressing or use of a silastic pouch (silo) to allow gradual reduction of the defect and scarification.

### Surgical Complications

Complications are dependent on the size of the lesion, type of surgery, gestational age and associated anomalies. These include:

- Abdominal compartment syndrome (more common with primary closure of larger defects).
- A tear of Glissen's capsule of the liver may occur when removing the sac covering the liver.
- Inadvertent damage of the bladder can occur if it is within the exomphalos.
- Infection risk in infants with a patch or mesh.
- Bowel adhesions may develop post-operatively.

### Post-Operative Course

- The majority of infants will require mechanical ventilation for a few days postoperatively.
- A nasogastric tube should be utilized for gastric decompression.
- Feeding can begin when the nasogastric output is decreasing and no longer bilious (this will be guided by surgeon). GORD is common. 60% of infants with exomphalos major will have feeding problems.
- Survival ranges from 75 to 95% of cases. Asthma and recurrent lung infections can occur in the postoperative recovery period.
- Most infants without associated abnormalities survive with good long-term growth and neuro-development.

### References

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2. Poddar R, Hartley L. Exomphalos and gastroschisis. *Continuing Education in Anaesthesia, Critical Care and Pain.* 2009; 9(2):48-51

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