GUIDELINE

Exomphalos (Omphalocele)

Scope (Staff):	Nursing and Medical Staff
Scope (Area):	NICU KEMH, NICU PCH, NETS WA

CAHS commits to being a child safe organisation by applying the National Principles for Child Safe Organisations.

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CAHS Child Safe Organisation Commitment Statement

This document should be read in conjunction with this disclaimer

Aim

The purpose of this guideline is to outline the antenatal, delivery room, general pre and postoperative management of neonates with exomphalos.

Risk

Inappropriate or delayed management could compromise the safety of the neonate, increasing risk of infection and postoperative complications.

Definition

- An exomphalos is herniation of abdominal viscera through a central abdominal wall defect.
- The herniated viscera are 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, usually para-umbilical and is more likely to have associated anomalies or be part of a syndrome.
- **Embryology**: During the 6^{th}-10^{th} 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.

Classification

1. Exomphalos minor- Defect < 5 cm, contains the intestine only.
2. Exomphalos major- Defect ≥5 cm and/or contains liver.

Antenatal Diagnosis

In most cases, exomphalos is seen on routine pre-natal ultrasound. Need further assessment for associated chromosomal and other anomalies. Delivery should occur in a tertiary care centre.

Associated Anomalies

- Associated anomalies are observed in up to 70% of infants.
- Most commonly associated with chromosomal defects or part of a syndrome.
- Structural anomalies could be present in 50% of infants even with a normal karyotype.
- The incidence of Beckwith Wiedemann is high with isolated omphalocele.

Associations	Notes
Chromosomal abnormalities	Trisomy 13, 18 and 21(and occasionally 14,15)
Syndromes	Beckwith-Wiedemann, Pentalogy of Cantrell, lower midline syndrome (bladder/cloacal exstrophy, imperforate anus, meningomyelocele)
Cardiac anomalies	Cardiac anomalies seen in up to 20% of cases
Pulmonary hypoplasia	Commonly associated with exomphalos major (20%)
VACTERL anomalies	Vertebral defects, Anal atresia, Cardiac defects, Tracheo-Esophageal fistula, Renal anomalies, and Limb abnormalities
Nervous system	Holoprosencephaly and anencephaly

At Birth

- **Mode of delivery:** Vaginal delivery is acceptable; caesarean section may be considered for large liver-containing sacs to prevent rupture or hepatic injury.
- **Resuscitation:**
    - Experienced neonatal staff should be present. A consultant should be present where possible if an exomphalos major is expected.
    - Be prepared for pulmonary hypoplasia and pulmonary hypertension: early intubation and ventilation may be required.
    - Clamping of the umbilical cord should only be carried out after careful assessment of the umbilical defect because bowel may extend into the cord base.
    - Commence early intravenous access and thermoregulation processes.

- 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.

Immediate Post-Resuscitation Care:

1. **Care of sac and Viscera**:
   - Handle sac gently by the membrane edges.
   - A rupture in the sac should be managed as a gastroschisis with urgent surgical referral and transfer to Ward 3B PCH.
   - The bowel should be decompressed with a large bore gastric tube and then left on free drainage.
   - 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**:
   - Commence 10% dextrose at 80-100 mL/kg/day of maintenance fluid; adjust for gestation
   - If there has been a rupture in the sac then fluid management is the same as a gastroschisis
   - Normal saline boluses(10-20ml/kg) for poor perfusion or metabolic acidosis.
   - Monitor electrolytes and glucose 4-6 hourly
3. **Temperature regulation**:
   - High risk of heat loss- use servocontrolled warmer and occlusive covering
4. **Respiratory and Cardiovascular management**:
   - Anticipate pulmonary hypoplasia and PPHN- ventilate early if oxygen requirement increases or respiratory distress worsens.
   - Obtain early cardiac echocardiogram.
5. **Antibiotics**
   - Commence antibiotics including Metronidazole.

Investigation for Associated anomalies

- Chromosomal microarray and methylation analysis (Beckwith-Wiedemann)
- Chest X-ray and spinal X-ray as indicated
- Abdominal & renal ultrasound

- Head ultrasound
- Echocardiography
- Limb X-rays if structural abnormalities suspected.

Surgical Management

Principles

- Individualise based on defect size, sac integrity, gestational age and presence of associated anomalies.
- Discuss with paediatric surgeons.
- Intact sac: Usually repaired electively once stabilised.
- Ruptured sac: Urgent surgical referral.

Options

- Small defects may be repaired early 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.
  - Conservative (“paint and wait”) management allowing the sac to epithelialise using topical agents (povidone-iodine or silver sulfadiazine) and delaying definitive ventral hernia repair to 6-12months.
  - Other adjunctive techniques include use of negative pressure dressing, flaps, patches 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)
- Liver capsule tear (Glisson’s capsule)
- Inadvertent damage of the bladder can occur if it is within the exomphalos
- Infection in infants with a patch or mesh
- Bowel adhesions may develop post-operatively
- Prolonged parenteral nutrition and cholestasis
- Large residual ventral hernia after delayed repair.

Post-Operative Course

- Most infants require mechanical ventilation for several days.
- A nasogastric tube should be used for gastric decompression.
- Commence trophic feeds when the nasogastric output is decreasing (this will be guided by the surgeon).
- Anticipate GORD.
- Recurrent lung infections can occur in the postoperative recovery period.
- 60% of infants with exomphalos major will have feeding problems and failure to thrive- multidisciplinary follow-up (surgery, feeding team, developmental).
- Prognosis is excellent in isolated cases without associated anomalies.

Related CAHS internal policies, procedures and guidelines

Neonatology Clinical Guidelines

- Gastroschisis
- Pre-Operative Care
- Post-Operative Care
- Post Operative Pain and Sedation
- Sepsis: Neonatal

WNHS Neonatal Medication Monograph

- Metronidazole Neonatal

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This document can be made available in alternative formats on request.

Document Owner:	Neonatology		
Reviewer / Team:	Neonatology Coordinating Group		
Date First Issued:	July 2006	Last Reviewed:	February 2026
Amendment Dates:		Next Review Date:	24th February 2029
Approved by:	Neonatology Coordinating Group	Date:	24th February 2026
Endorsed by:	Neonatology Coordinating Group	Date:	
Aboriginal Impact Statement and Declaration (ISD)		Date ISD approved:	24th February 2026
Standards Applicable:	NSQHS Standards: Child Safe Standards: 1, 2, 3, 4, 5, 6, 7, 8, 9, 10		

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