GUIDELINE

Exomphalos (Omphalocele)

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

Child Safe Organisation Statement of Commitment

CAHS commits to being a child safe organisation by applying the National Principles for Child Safe Organisations. This is a commitment to a strong culture supported by robust policies and procedures to reduce the likelihood of harm to children and young people.

This document should be read in conjunction with this disclaimer

Aim

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

Risk

Inappropriate 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 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.

Types

1. Exomphalos minor where the defect is less than 5 cm and only contains the intestine.

2. Exomphalos major where the defect is greater than 5 cm and/or with the liver inside the cord.

Antenatal Diagnosis

In most cases, exomphalos is seen on pre-natal ultrasound. Need further assessment for associated chromosomal and other anomalies.

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, 14,15, 18 and 21
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

- 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 with pulmonary hypertension requiring early intubation and ventilation.
- Clamping of the umbilical cord should only be carried out after careful assessment of the umbilical defect because bowel may be present at the base of the umbilical cord.

- 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 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:
   - 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 a gastroschisis.
   - All infants should be commenced on 80-100 mL/kg/day of maintenance fluid (10% glucose).
   - 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.
4. Infants with ongoing ventilation may have associated pulmonary hypertension and need early cardiac review.
5. Monitor glucose 4-6 hourly.
6. Start antibiotics including Metronidazole.

Investigation for Chromosomal Anomalies or Syndromes

- A full genetic work up should be sent.
- Consider Beckwith Wiedemann 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

- Treatment depends on the size of the defect, gestational age and presence of associated anomalies.
- 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.
- 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.
  - 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 the surgeon). GORD is common.
- Recurrent lung infections can occur in the postoperative recovery period.
- 60% of infants with exomphalos major will have feeding problems and failure to thrive.
- Most infants without associated abnormalities survive with good long-term growth and neuro-development.

Related CAHS internal policies, procedures and guidelines

Neonatology Clinical Guidelines

- Gastroschisis
- Pre-Operative Care
- Post-Operative Care
- Post-Operative Analgesia

References

1. Adams, A. D., et al. (2021). "Omphalocele-What should we tell the prospective parents?" Prenat Diagn 41(4): 486-496.
2. Verla MA, Style CC, Olutoye OO. Prenatal diagnosis and management of omphalocele. Semin Pediatr Surg. 2019 Apr;28(2):84-88
3. Baerg JE, Munoz AN. Long term complications and outcomes in omphalocoele. Seminar Pediatr Surg 2019 Apr; 28(2):118-121.
4. Christison-Lagay ER, Kelleher CM, Langer JC. Neonatal abdominal wall defects. Semin Fetal Neonatal Med. 2011 Jun;16(3):164-72

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