Section 13: Surgical conditions
Exomphalos (Omphalocele)
Date last reviewed: 2006
Date this revision: Sept 2014
Review date: Sept 2017

Neonatology Clinical Guidelines
King Edward Memorial/Princess Margaret Hospitals
Perth Western Australia
Authorised by Neonatal Coordinating group

EXOMPHALOS (OMPHALOCELE)

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^{th}–10^{th} week the fetal intestine migrates through the umbilical ring into the cord, 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 4cm and only contains the intestine,
2. ***Exomphalos major*** where the opening is greater than 4cm 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 a 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 6B
    - 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 80-100mL/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 Weidermann 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.

POSTOPERATIVE 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:

Christison-Lagay ER, Kelleher CM, Langer JC. Neonatal abdominal wall defects. Semin Fetal Neonatal Med. 2011 Jun;16(3):164-72

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