Guideline

Stabilisation and transfer of an infant with exomphalos

1 Scope

For use within the Paediatric and Neonatal Decision Support and Retrieval Service (PaNDR) for the East of England.

2 Purpose

To provide safe, efficient and practical guidance for stabilisation and transfer of infants with Exomphalos.

3 Definitions and abbreviations

PaNDR: Paediatric and Neonatal Decision Support and Retrieval Service

Beckwith-Wiedemann syndrome:

An overgrowth disorder, usually present at birth. Common features include macroglossia (large tongue), macrosomia ('big body'), exomphalos and neonatal hypoglycaemia.

NBM: nil by mouth

NLS: newborn life support

Pentalogy of Cantrell:

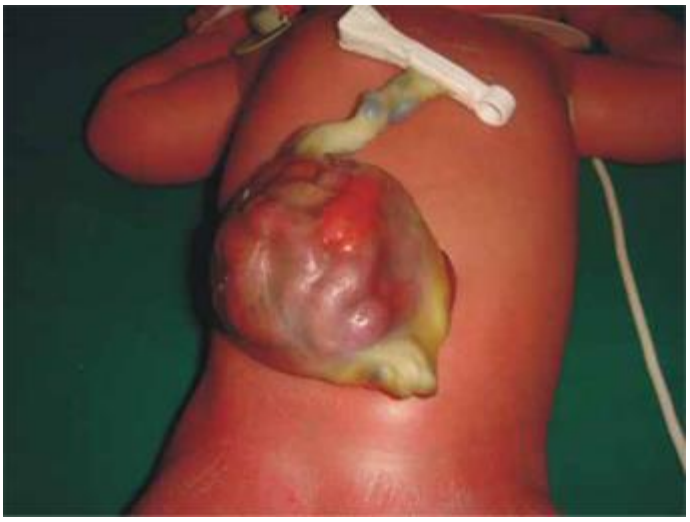
A rare syndrome characterized by the following five features: exomphalos, diaphragmatic hernia, sternal cleft, pericardial defect, cardiac defect. Severity varies from mild defects with incomplete expression of the disorder to complete ectopia cordis (heart displaced outside the thoracic cavity so not protected by the chest wall) with large exomphalos.

Trisomies: Genetic abnormalities involving three instances of a particular chromosome instead of the normal two (eg trisomy 18, trisomy 21).

4 Introduction

- Exomphalos is a congenital abdominal wall defect. Abdominal wall defects occur when a fetus's abdominal wall does not develop fully while in utero, resulting in the intestine developing outside the abdomen. The intestines develop inside the umbilical cord in early pregnancy, and then move inside the abdomen by 12 weeks of pregnancy. An exomphalos occurs if the abdominal contents protrude into the base of the umbilical cord and are covered by a peritoneal membrane. Non-rotation of the intestines is commonly seen.
- If the abdominal defect at the base of the exomphalos is less than 5cm it is called exomphalos minor and may contain only tissue left over from structures in the unborn fetus's digestive tract. If it is >5cm it is called exomphalos major. Part of the liver may also be present in the sac along with bowel.
- The cause of exomphalos is not always known. Familial occurrence has been described. Up to 80% of babies with exomphalos have other serious abnormalities such as heart defects, gastro-intestinal and renal anomalies and chromosomal abnormalities (eg trisomies). It can also occur as part of a syndrome such as Beckwith-Weidemann syndrome or Pentalogy of Cantrell.

Figure 1: Exomphalos minor



5 Management

- **Resuscitation**
  - Resuscitate as required according to NLS guidelines
  - Aim to **avoid** prolonged mask ventilation, so as not to distend the bowel, which may make surgical repair more difficult.
- **Cord clamping**
  - Ensure careful clamping of the cord **distal** to normal practice so as not to damage any bowel that may be present within it.
- **Care of membranous sac**
  - Wrap the abdomen securely with cling film and ensure the gut is well supported to avoid compromising its blood supply.
  - Handle the baby carefully to avoid tearing the sac.
  - If the sac is torn (at delivery or postnatally) urgent surgical intervention is required.
- **Nursing position**
  - Nurse supine in the transport incubator so that the gut can be visualised within the sac during transfer.
  - Before moving into the transport incubator the infant can also be nursed in the left lateral position, ensuring the abdominal contents are lying supported straight in front of the baby.
- **NG tube**
  - Ensure early introduction of a large bore NG tube (size 8 or 10F) on free drainage with at least hourly aspiration to avoid the bowel becoming distended with air.
- **Access**
  - Do **not** attempt umbilical lines
  - Transfer with 2 x peripheral venous lines
- **Fluids**
  - NBM
  - Start maintenance IV fluids (10% dextrose) and replace NG losses ml/ml with 0.9% Sodium Chloride with 10mmol/500ml Potassium Chloride.
  - Infants with abdominal wall defects are at risk of dehydration from excessive fluid losses, though the peritoneal membrane

covering the intestines in exomphalos offers some protection from this (unlike in gastroschisis where the gut is completely exposed). Observe closely for the need for additional fluid boluses.

- **Monitor temperature**
- **Glucose**
    - It is important to monitor blood glucose levels regularly and correct any hypoglycaemia with IV dextrose (in view of association with Beckwith-Weidemann Syndrome)
- **Sepsis**
    - Consider antibiotics if risk factors for sepsis present or there are additional clinical concerns.
- **Associated anomalies**
    - Examine baby for other anomalies

6 Monitoring compliance with and the effectiveness of this document

The transport team will monitor that the guideline is being adhered to on a case by case basis (very rare condition). This will be fed back to the senior team at a governance meeting.

7 References

London Newborn Transport Service guideline - https://london-nts.nhs.uk/wp-content/uploads/2015/01/Exomphalos-NTS-Guideline.pdf

Southern West Midlands Newborn Network guideline - https://www.networks.nhs.uk/nhs-networks/southern-west-midlands-newborn-network/documents/Attachment%206%20Exomphalos%20Final%20jan%202012.pdf

National Organisation for Rare Disorders (NORD) website – www.rarediseases.org

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Document management

Approval:	7 February 2020		
Owning department:	PaNDR		
Author(s):	Dr Samantha O'Hare		
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File name:	Stabilisation and transfer of an infant with exomphalos Version2 February 2020.doc		
Supersedes:	Version 1, May 2016		
Version number:	2	Review date:	February 2023
Local reference:		Document ID:	37712