Management of Gastroschisis and Exomphalos

**Introduction**

Management of a baby born with Gastroschisis /Ruptured Exomphalos is a surgical emergency. The majority of cases will be diagnosed on antenatal ultrasound scans. Ideally the babies known to have these conditions should be delivered in a Neonatal surgical unit or to be delivered in a level 3 Neonatal unit and transferred to a neonatal surgical unit as soon as possible.

**Definition**

Gastroschisis and Exomphalos are congenital abdominal wall defects that result in evisceration of abdominal contents at birth. The insertion of the umbilical cord into the abdominal wall distinguishes Gastroschisis from Exomphalos.

Gastroschisis occurs as a defect in the abdominal wall usually to the right of the umbilicus with herniation of bowel. The viscera are not covered by a peritoneal sac and so are exposed to amniotic fluid during pregnancy and to the air following delivery.
Exomphalos is a defect in the umbilicus, with herniation of intestine as well as other viscera into the umbilicus. It can be less than 5cm in size (minor) or greater than 5cm (major Exomphalos). The viscera are covered by a thin membrane of amnion and peritoneum if the sac ruptures, this is an emergency and should be managed as in gastroschisis. Associated congenital anomalies are more common in exomphalos.

Initial Management of an Infant born with Gastroschisis / Exomphalos

1. Management in delivery room:

   1. Delivery must be attended by the Neonatal team.
   2. Ensure the person delivering the baby uses a plastic cord clamp, rather than artery forceps on the umbilical cord and to cut the cord as long as possible.
   3. ABC resuscitation, as required. **Try to avoid mask ventilation** and have low threshold for intubation if baby shows signs of respiratory distress. Intubation and ventilation are preferable to V/T/ CPAP. This is to minimize the gaseous distension of the gut.
   4. Management of exposed bowel
      - Put on a pair of gloves and carefully handle the bowel to ensure the bowel is not twisted or kinked and there is no traction on the mesentery. Assess the colour and the alignment of the bowel. Avoid excess bowel handling.
Lay baby on top of a long length of precut cling film and wrap right around the abdomen aiming to keep the prolapsed gut well supported and to protect from injury and fluid loss.

Stabilize the bowel in the midline Fig 2
Pass a wide bore (8 FR) nasogastric tube and fix securely with tape. Place the NG on free drainage and aspirate every 20-30 minutes (to decompress and aspirate the gastric contents)

Place the baby on to the right side and support the bowel with towel rolls.
Insert venous access and consider fluid resuscitation if as required. Rapid fluid loss can occur in infants with exposed bowel.

When the baby is stabilized, allow parents to see them briefly before transferring to the neonatal unit.

**Management in NICU**

- Nurse the baby in an incubator with high humidity to reduce heat loss and to prevent hypothermia. Where possible, nurse the baby on its right side so that the eviscerated bowel is supported and does not flop over the abdomen potentially occluding the blood supply.
- Always review ABC on a regular basis. Ensure the baby is stable and has appropriate management for airway and breathing. Observe for tachycardia and monitor CRT frequently.

It is important to perform these procedures, as rapidly as possible, as these babies become cold very quickly and lose a lot of fluid from the exposed bowel.
• Insert I V cannula (x2) and take blood for Blood culture, Blood gas, FBC, Clotting studies, Urea and Electrolytes
• Check blood glucose immediately and monitor closely because of the association of Beckwith-Wiedemann syndrome with Exomphalos.
• Give Vitamin K IM according to guideline.

Fluid resuscitation:

- 10-20mls/kg of 0.9% normal saline bolus (in the delivery suite + in the unit as required). Repeat if required. If boluses exceed 20-30 ml/kg/day consider giving Human Albumin (4.5% HAS) as the babies lose large amounts of protein from the exposed bowel.
- Maintenance fluid 60mls/kg/day of 10% dextrose or PN
- Keep Nil by mouth.

• Start broad spectrum antibiotics according to unit policy.
• Leave the NGT on free drainage.
• Aspirate nasogastric tube hourly and replace the aspirate volume, ml for ml with intravenous 0.9% saline with 10mmol potassium chloride in each 500ml bag. The baby will need an accurate fluid balance record.
• Once stabilised, Refer the baby urgently to the Paediatric Surgeons and arrange emergency transfer to the surgical unit.
• While awaiting transfer to surgical unit continue to assess bowel perfusion every 15 minutes. Inspect gut for any discoloration, dilatation, obvious perforation, and stricture.

REMEMBER:
Exomphalos - has normal GI function, but has significant associated anomalies/defects. Survival about 60%
Gastroschisis - has horrible GI function but few associated anomalies/defects. Survival about 95-90%
Reference

1. Anthony D Owen, Sean Marven, Julie Bell, Gastroschisis: putting the bowel back safely (2009), Available at: http://www.infantgrapevine.co.uk/pdf/inf_026_gpd.pdf
2. Guy’s and St Thomas ’NHS Foundation Trust: Clinical guidance, Neonatal Manual Chapter 4: Neonatal Surgical Problems (August 2014)

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