GASTROSCHISIS (ALSO SEE THE NETS WA MANUAL)

Gastroschisis is a congenital anomaly characterised by a full thickness abdominal wall defect, usually just to the right of an intact umbilical cord, through which a variable amount of intestine protrudes without a covering membrane; other abdominal viscera like liver, spleen, gonads etc may also protrude. The size of the abdominal wall defect is variable; small defects are a concern and may cause significant bowel ischaemia.

The term ‘Vanishing Gastroschisis’ is used to describe the apparent foetal ultrasonic reduction and disappearance of exteriorised bowel due to the abdominal wall defect spontaneously closes, leading to significant bowel ischaemia and short bowel syndrome.

MANAGEMENT AT BIRTH

LIAISE WITH THE PAEDIATRIC SURGEONS AND THE CHILDREN’S HOSPITAL NICU
Alert the Children’s hospital NICU and the on call paediatric surgical team well before delivery. The Children’s hospital NICU may need to make room for the admission by moving existing patients and the paediatric surgeons may wish to be present soon after delivery in order to place the exposed bowel into a silo before the baby is transferred to Children's NICU.

MANAGEMENT AT BIRTH IN LABOUR WARD
1. Standard neonatal resuscitation as per NRP guidelines.
2. The infant’s legs and torso should be placed inside an impermeable bowel bag (cling film when no bowel bag available), ensuring it is secured under the infant’s axillae and carefully watching for bowel discolouration. Lay the infant on the right side for transport to NICU.
3. Insert size 8 Nasogastric tube (NGT) and fully aspirate the stomach contents and leave on free drainage.
ON ADMISSION TO SCN3 AT KEMH

1. Immediately upon admission to NICU, contact the Children’s Hospital NICU, NETS WA and the surgical team and confirm that delivery has occurred (see above). If the surgeon is unable to attend the infant at KEMH, arrange for transfer to the Children’s NICU as soon as the infant is stable for transport.

2. Nurse the infant under a radiant warmer. The infant’s legs and torso should remain inside the impermeable bowel bag in order to limit fluid and heat loss from the bowel and also to allow easy inspection of the bowel and help protect the bowel and limit the risk of sepsis.

3. Insert size 8 NGT if not inserted at delivery and fully aspirate the stomach contents and leave on free drainage.

4. Watch bowel colour and report any discoloration that develops; position the bowel and baby carefully, usually right side down. At any time, if bowel turns dusky, consider gently repositioning the bowel in relation to the abdominal opening and ask for an urgent surgical review.

5. Insert an IV and commence maintenance fluids (7.5 to 10% glucose): 80 mL/kg/day in neonates >37 weeks and 100 mL/kg/day in preterm infants. In addition, commence an infusion of Normal Saline at a rate of 10 mL/kg/hr. The Normal Saline infusion should continue until the surgeons apply a silo or until a formal reduction of gastroschisis is achieved (whichever happens first). Once the Silo is applied make sure to stop the Normal Saline infusion.

6. Give additional or subsequent boluses of Normal Saline if hypoperfusion, hypotension or metabolic acidosis is present. Consider early use of Dopamine or Dobutamine if no improvement in spite of the Normal saline infusion.

7. Give IV prophylactic antibiotics Amoxicillin/ Gentamicin/ Metronidazole after collecting blood cultures.

8. Examine the infant for associated anomalies.

9. FBC, blood group and hold or cross-match, blood gas, blood culture. Ensure that 10mls of clotted maternal blood for cross-matching is collected.

10. Once the baby is stable transfer to the Children’s Hospital NICU.

MANAGEMENT OF NEONATES WITH GASTROSCHISIS DURING TRANSPORT

(PLEASE REFER TO THE NETS WA MANUAL).

SPECIAL MENTION ABOUT GASTRIC DECOMPRESSION AND WATCHING FOR INTESTINAL VASCULAR COMPROMISE

Free drainage with wide bore NGT is essential to prevent gastric distension, vomiting and aspiration. Care must also be taken during initial stabilisation and transport to ensure that the intestinal contents are well protected and supported. Nurse the infant supine with boundaries positioned on either side to support the herniated bowel. Otherwise position the infant on the right side so that there is no tension or kinking or the mesenteric arteries leading to vascular compromise. If a length of bowel appears dusky in appearance (i.e. impaired blood supply) then reposition to see if this improves the colour.

PREOPERATIVE CARE ON ARRIVAL AT THE CHILDREN’S HOSPITAL NICU

1. Alert surgeon of infant’s arrival if the silo is not in place or if there are new concerns.

2. Continue with care as detailed above
3. Assess the infant’s hemodynamic status using skin perfusion, blood pressure and acid base status. If peripheral perfusion is poor and or metabolic acidosis develops, consider more Normal Saline boluses (10-20mls/kg over 30 minutes) and/or inotropes, e.g. Dopamine or Dobutamine.

4. Continue maintenance fluids of 80 mL/kg/24hrs in full term infants (>37 weeks) and 100mL/kg/day in preterm infants (7.5% or 10% Glucose).

5. If silo has already been applied, ensure that the Normal Saline infusion has been ceased.

6. If the silo has not yet been applied, prepare for reduction either in NICU or theatre depending on the surgeon’s preference.

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**PRIMARY REDUCTION OF THE BOWEL AND CLOSURE OF THE ABDOMINAL WALL DEFECT IS RARELY PERFORMED.**

**THE GENERAL MANAGEMENT OF A GASTROSCHISIS INFANT WITH A SILO IN SITU.**

1. Nil oral and keep the stomach on open drainage.

2. Maintenance parenteral nutrition to ensure adequate weight gain and metabolic balance.

3. Extra IV fluids will be required if gastric losses are greater than 10mls/kg/12hour period. The total fluid loss in the previous 12 hour period should be replaced, (half as TPN solution and half as Normal Saline via a side line). Please review the [Replacement of Gastrointestinal Fluid Losses in Surgical Neonates](#).

4. Surgeons will gradually reduce the intestinal contents as the baby’s condition permits; usually over 2-5 days.

5. Carefully observe the visible bowel for discolouration. Normal bowel in silo should look pink. Grey, purple or black looking bowel within the silo indicates vascular compromise. Areas of ‘peel’ may be present and appear discoloured. This situation should be reviewed by senior neonatal staff.
POST OPERATIVE MANAGEMENT FOLLOWING COMPLETE CLOSURE, EITHER AFTER SILO OR AS AFTER PRIMARY CLOSURE

Some infants may not adapt quickly to the extra amount of gut which has been reduced into the abdominal cavity. This could result in compression of the mesenteric blood vessels leading to ischemia of the bowel. This is called “abdominal compartment syndrome”. If this is not diagnosed and treated quickly, it can have catastrophic consequences, such as necrosis of the bowel and short gut syndrome. One or more of the following clinical features should suggest the possibility of abdominal compartment syndrome:

1. A significant increase in ventilator pressures compared to the pre-reduction pressures.
2. Tense and possibly tender abdomen
3. Discolouration of abdominal wall and lower limbs
4. Requirement of high dose of morphine for analgesia
5. Progressive worsening of metabolic acidosis and or respiratory acidosis, high lactate levels
6. Hypotension requiring inotropic support.
7. Decreased or absent urine output.

If one or more of these clinical features develop, you should seek an urgent senior neonatal review before contacting the surgical team.

PAIN MANAGEMENT

Analgesia should be provided as needed. Preferably with intravenous paracetamol or morphine. A balance should be maintained between effective analgesia and side effects of opiates on respiratory depression and gut motility. Normally infants with a silo do not require significant analgesia. If an infant needs high dose morphine infusions, it should alert the clinicians to the possibility of bowel ischemia.

FLUID AND ELECTROLYTE BALANCE

Frequent blood gas and electrolyte measurements (6-12 hourly) should be performed to guide fluid therapy. Blood pressure, capillary refill, pulse rate, colour, amount of gastric aspirates and urine output should be taken into consideration in adjusting fluid therapy.

GASTRIC LOSSES

Please review the Fluid replacement therapy for surgical neonates

A large bore gastric tube (8 or 10) on free drainage with 1–2 hourly gentle aspirations is needed to prevent gastrointestinal distension caused by post-operative ileus and the reduction process. Monitoring and replacement of losses is necessary as some infants can loose very large volumes of stomach fluids. Losses are generally replaced as half normal saline, but are tailored to the baby’s electrolyte status. In some cases, this may last for weeks until gut motility improves.

URINARY OUTPUT DUE TO DECREASED URINE PRODUCTION OR BLADDER RETENTION

A decrease in urine output may be due renal venous compression due to the raised intra-abdominal pressure or an inadequate intravascular volume (due inadequate fluid loss replacement or ‘third’ spacing. Bladder retention is a frequent problem, often as a side effect of morphine analgesia. A urinary catheter may be necessary to relieve bladder distension and allow a more accurate measurement of actual urine production.

NUTRITION

Consider early placement of a percutaneous long line. Parenteral nutrition should be started as soon the baby is stable after birth. Enteral feeding will not start until the abdominal wall defect has
been closed and gastric aspirated are reducing. Trophic feeds are often given even when there are relatively large gastric aspirates, decided on a case by case basis. Enteral feeds are increased as tolerated; as judged by the amount of gastric aspirates/vomits, abdominal distension and bowel actions. Gut dysmotility can persist for weeks after gastroschisis closure and longer term TPN may be needed to supplement inadequate enteral intake. Early initiation of enteral feeding has been shown to improve the gut function in babies with gastroschisis. Occasionally a prokinetic agent may be helpful in promoting gut motility.

**ANTIBIOTICS**
Antibiotics are routinely started immediately after birth and usually continued until the abdominal wall defect is closed. Routine antibiotics are Amoxicillin, Gentamicin and Metronidazole. Vancomycin is frequently added if there is significant abdominal wall redness. It is reasonable to monitor CRP levels whilst on antibiotics.

**REFERENCES**
Davies MW, Kimble RM, Woodgate PG. Ward reduction without general anaesthesia versus reduction and repair under general anaesthesia for gastroschisis in newborn infants. The Cochrane Database of Systematic Reviews 2002, Issue 3
