Gastroschisis is a congenital anomaly characterised by a full thickness abdominal wall defect through which abdominal viscera (not covered by a protective sac) usually the intestines and occasionally other organs such as the liver or spleen etc may also protrude. Oligohydramnios is commonly associated with gastroschisis. If polyhydramnios is seen antenatally, this may indicate an associated intestinal atresia which occurs in approximately 10% to 20% of cases. The defect is usually located to the right of the umbilicus. The incidence of gastroschisis has increased from 2.5 per 10000 total births in 1994 to 4.4 per 10000 in 2004 and the incidence in Wales is generally higher at 6.2 per 10000 total births. Young maternal age and a history of drug abuse are risk factors for gastroschisis.

Immediate Management in labour ward:
1. Note the antenatal history and presence of any bowel dilatation. Standard neonatal resuscitation as per the NLS guidelines. If respiratory support is required, intubation and ventilation are preferable to NCPAP in order to minimise gaseous distension of stomach and intestines.
2. Insert a wide bore nasogastric tube (NGT), aspirate the stomach contents and leave on free drainage.
3. The mid gut is supplied by a narrow vascular pedicle: kinking or torsion of the vascular pedicle must be avoided as this may lead to ischemia or infarction of gut. Stabilise the bowel loops in the midline with a doughnut ring made of gauze and cotton. The exposed bowel should then be covered with transparent cling film to allow frequent assessment of bowel perfusion. Lay the cling film on the resuscitaire before birth and place the baby on top to facilitate easy covering of the loops.
4. If the bowel appears to have impaired blood supply or drainage (looks dusky, dark purple or suffused) try gentle manipulation of the bowel into other positions to see if the circulation can be improved. Contact the paediatric surgeon for urgent review.
5. Care must be taken during the initial stabilisation and transport to ensure that the bowel is well protected and supported. Nurse supine. Avoid unnecessary handling of the bowel.

On admission to Neonatal Unit:
1. Nurse in a warm environment in a supine position. Watch the bowel to note colour and position.
2. Examine carefully for any other anomalies and contact the surgical team.
3. Insert wide bore NGT if not already inserted at delivery to decompress the stomach contents and allow for free drainage. Aspirate hourly until in theatre. Replace NG losses ml/ml with normal saline 0.9% with KCl 20mmol/1Lbag.
4. As bowel is herniated external to the abdominal wall a significant amount of serous fluid is lost as ooze which contributes to radiant heat loss and can lead to hypothermia. Obtain IV access (do not use umbilical lines and avoid possible long lines sites) and start maintenance IV fluids.
5. Regularly assess perfusion, and maintain a low threshold for giving a normal saline fluid bolus as these babies have extra ongoing losses. Crystalloid can be used initially but consider colloid eg 4.5% Albumin early due to likely protein ooze from the gut. Consider early use of inotropes.
6. Obtain FBC, blood group & cross-match, blood gas, blood culture and biochemistry. Clotting profile only if there are clinical concerns. Ensure that cross match sample has been taken prior to surgery.
7. Start IV antibiotics after collecting blood cultures if there are any medical concerns regarding sepsis. Otherwise Augmentin is usually given at induction in theatre.
8. Administer 1mg IM Vitamin K.
9. Occasionally the surgeons will decide to place the bowel in a preformed silo, on the neonatal unit without anaesthesia, but most cases are taken to theatre as a category 1 (surgery within 2 hours).

Surgery:
The aim of surgery is primary closure however; this may not be possible in all babies as the abdominal cavity may be too small to accommodate the intestine. In some cases, closure is possible using a Permacol® patch which may be left exposed but covered with a saline soaked gauze. If there is severe
viscero-abdominal disproportion a silo repair is done which allows for gentle staged reduction of the bowel into the abdomen.

Following silo placement, the herniated gut is gradually reduced into the abdominal cavity over a period of time dictated by the surgical team. This may take a few days or longer than a week. Carefully observe the bowel in the silo for any discolouration. Healthy bowel in the silo should look pink, if the bowel is changing colour to grey or purple it may indicate vascular compromise. Alert the surgical team.

Some infants may not adapt quickly to the increased amount of gut in the abdominal cavity causing compression of the mesenteric blood vessels resulting in ischemia to the bowel loops, abdominal compartment syndrome. If this is not recognised and treated promptly, it can lead to gut necrosis and ultimately short gut syndrome. Compartment syndrome may also cause renal impairment and cool limbs.

One or more of the following clinical features should suggest the possibility of abdominal compartment syndrome:

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

Alert the surgeons immediately if one or more of these clinical features develop, in particular decreasing/absent urine output or progressive metabolic acidosis.

Ongoing management:

Fluid management can be complex in these babies. Fluid loss and resultant fluid requirements may be greatly increased in the peri-operative period due to evaporative loss from exposed viscera and third space loss into the abdominal cavity and surrounding tissues. Venous compression due to the increased intra-abdominal pressure can compromise renal blood flow and lead to decreasing urine output. A urinary catheter may be necessary to allow for an accurate measurement of urine production.

Blood pressure, capillary refill, urine output, blood gas and electrolyte measurements should all help guide fluid therapy.

Keep the large bore NGT (size 8 or 10) on free drainage with 1–2 hourly aspirations to prevent gastrointestinal distension caused by post-operative ileus. Monitoring and replacement of gastric aspirates is vital as some infants can have significant losses. NGT aspirate losses are replaced ml for ml, usually as 0.9% normal saline with added potassium chloride (1mmol/50ml).

Some babies may require paralysis in the post operative period, particularly if abdominal closure was difficult and the intra-abdominal pressure is considered elevated.

Insert a long line as soon as possible following return from theatre. Enteral feeds are started when the NGT aspirates are non bilious and minimal, the median duration to establishing feeds is around 3-4 weeks. Once enteral feeds are started advance slowly @1ml/day. Human milk or colostrum is the preferred milk but if not available use normal term or preterm formula.

Delayed gut motility can persist for weeks post op and prolonged parenteral nutrition is required. Moreover, if there is an associated atresia time to full feeds may be significantly delayed. If gut function is slow to improve, investigations such as contrast studies (both upper and lower) and rectal biopsies can aid further management.

Occasionally Erythromycin may be used empirically as a prokinetic agent. If enteral feeds are not established by 6 weeks post op, consider referring to Paediatric Gastroenterology.

References:

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2. CARIS Review 2015
3. J Waldhausen; Surgical Management of Gastrochisis; NeoReviewsVol.6 No.11 November 1, 2005, p500-507
4. James A, Drayton M Outcome of newborns with gastroschisis; Wales Paediatric Journal WPJ: Volume 33, 2010