1. Introduction / Background

Gastroschisis is one of the most common fetal surgical anomalies with an increase in reported incidence in recent years to as high as 5:10000 (Boyd et al, 2011). It is easily diagnosed antenatally with reported pick up rates of 90-100% in centres with dedicated second trimester screening.

The precise aetiology is unknown and thought to be due to a vascular accident of the fetal vessels. A recent hypothesis suggests that the vascular thrombosis is due to fetal thrombophilia caused by the maternal oestrogen surge (Lubinsky, 2012).

Gastroschisis is most commonly an isolated defect. However, 10-15% of cases appear to have associated intestinal atresia (Duke and Schwartz, 2009). It commonly occurs on the right side of the umbilical defect and it is characterised by of the evisceration of intra-abdominal content not covered by the amnion (in contrast with exomphalos). The overall survival is more than 90% (Duke and Schwartz, 2009).

2. Patient Group

All neonates with gastroschisis.
3. Management

3.1 Antenatal diagnosis and management

Antenatal diagnosis is possible in all cases where a second trimester scan is performed. As part of the surgical care pathway, counselling should be arranged by the fetal medicine team, with the surgical team. Early in counselling the importance of breast milk should be stressed and mothers should be encouraged to express breast milk (Kohler et al, 2013). Prospective parents should be offered a visit to neonatal unit with surgical care pathway nursing team or the family care sister. Parents should be provided with the ‘Gastroschisis: Information for parents’ booklet during the antenatal period.

Babies who initially book at the City campus continue antenatal care at the City campus, but all babies with Gastroschisis will have a planned delivery at the QMC campus. Fetal care team will facilitate this process. All other Trent Perinatal Network babies with gastroschisis are referred to QMC campus for antenatal care and delivery (See Trent Perinatal Network Referral for Surgical Assessment guideline http://www.centralandtrentneonatalnetwork.nhs.uk/index.php/health-professionals/guidelines)

Obstetric guidelines are available for the antenatal management of gastroschisis. Antenatally all mothers are given their own ‘Vi-Drape bag’ to use in case of unexpected delivery any where other than at QMC.

3.2 Presentation after birth

It is uncommon to have first presentation of this condition at birth as almost all of them get diagnosed on routine antenatal scans. However it would be obvious at the time of delivery. Absence of covering membranes that occurs most commonly in Gastroschisis differentiates it from another similar condition like Exomphalos. The deliveries are often associated with green staining liquor with green colour originating from bile. It is very unusual for babies to have meconium stained liquor.

3.3 Delivery room care

The primary aim is to stabilise the baby prior to urgent surgery. The exposed gut must be handled with care. Problems with thermoregulation and fluid loss from the exposed bowel need to be anticipated.

- Avoid drying and place the baby directly into a Vi-Drape bag. Secure it under the baby's arms.
- Keep the baby warm under radiated heat and resuscitate as required according to NLS guidelines (see Guideline A5).
- Place the bowel loops either in the midline on top of the baby, or next to the baby with baby lying on his / her side to avoid twisting of the mesentery and constriction at the edge of the defect with possible compromise of blood supply. It may be helpful to put towels at the side of the baby (in supine position) or underneath the baby (in lateral position) to maintain the bowel position.
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- Bowel loops should be observed and their appearance and colour documented every 15 minutes. Duskeness of bowel loops should warrant urgent discussion with surgical team.
- Most of babies with gastroschisis do not need respiratory support in the delivery room. However, if respiratory support is required, the infant should be intubated and ventilated. Avoid the use of CPAP which would cause bowel distension.
- Pass a wide bore nasogastric tube and aspirate. The stomach is often distended and containing bilious fluid following delivery. The nasogastric tube should be left on free drainage and aspirated regularly.

3.4 Care on admission to the neonatal intensive care unit

The goals of early care are maintenance of adequate bowel perfusion, thermoregulation and adequate fluid resuscitation. The Consultant Neonatologist and Consultant Paediatric Surgeon should be informed as soon as possible. The surgical team will contact the anaesthetic team. Surgery for gastroschisis is a surgical emergency.

The aim is for inborn babies to have a surgical intervention by 2 hours of age.

3.4.1 Early Care

All observations and management initiated in the delivery room should be continued.
- Access and monitoring - Peripheral venous access should be obtained. Peripheral arterial access may be warranted if there are concerns about blood pressure and fluid resuscitation. Full intensive care monitoring including blood pressure monitoring should be commenced.
- Blood tests – The usual admission bloods should include full blood count, CRP, blood cultures, group and save and request a unit of red cells in preparation for theatre.
- Fluids and perfusion – Large fluid and protein losses may occur from the exposed bowel. Increased fluid input may therefore be required. Give fluid boluses of 10ml/kg 0.9% sodium chloride if there are signs of circulatory impairment (raised lactate, low blood pressure, poor peripheral perfusion) or if exposure of the bowel is prolonged. Do not include the often large volume of the first nasogastric aspirate in the calculations for replacing nasogastric losses. Reassess frequently including capillary refill time, heart rate, blood pressure, urine output, fluid balance and blood gases for monitoring of lactic acidosis. Anticipate the need for additional fluids depending on the timing of surgery.
- Antibiotics – Benzyl penicillin and Gentamicin should be prescribed.
- If respiratory support becomes necessary intubate and ventilate. Do not use CPAP. Excessive air going past the stomach may distend the gut and lead to potential compromise of s blood supply. The nursing team should complete the pre-op checklist and prepare the transport incubator.
- Written parental consent should be obtained by the surgical team.

3.4.2 Transfer to theatre
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A nurse and doctor/ ANNP would both transfer the baby to theatre. Positioning of the baby and care of the bowel should be continued as in section 3.3 with the nasogastric tube on free drainage.

3.4.3 Postnatal transfer

This section applies to babies who deliver in any other hospital other than QMC. If born outside of Nottingham urgent transport of these babies after immediate stabilisation will be required. Trent Perinatal Network Surgical referral guideline should be followed and the referring team should be supported by the neonatal, surgical and CenTre team to facilitate the transfer of baby to QMC or an alternative surgical bed if it is not possible for the baby to be transferred to QMC. Transport management is a combination of delivery room and early care as detailed above, and meticulous transport stabilisation with particular attention to the positioning and perfusion of the bowel.

3.5 Surgery

Surgery for gastroschisis – either primary closure or silo formation with staged reduction – is a surgical emergency. The surgical team should be informed of the impending delivery where appropriate. The surgical team must be informed of the birth on arrival on the NICU who will review the baby on the NICU.

Mostly the surgery is performed in theatre. However, in some carefully evaluated circumstances, the surgeons may reduce the bowel on the Neonatal Unit. In these cases, careful attention should be paid to intubation and ventilation and to pain assessment and pain relief as in Neonatal Guideline G9.

3.6 Post-operative care

Postoperative care is multi-disciplinary with nursing, medical and surgical teams all providing input.

3.6.1 Immediate post-operative care

- Measure temperature and correct appropriately
- Ventilation - Stabilise on the ventilator and closely monitor respiratory status and blood gases. Babies with significant postoperative abdominal distension may be at risk of basal lung atelectasis. In these patients, higher PEEP of 6-7cm of H2O may be helpful. Aim for normal pH. Do not underestimate pain especially in cases where stretching of abdominal wall has been performed as a part of surgical procedure. See Guideline G9. Weaning and extubation should be guided by pain control, degree of opiod use for pain control, respiratory drive along with standard weaning parameters (Guideline B9)
- Cardiovascular system - Carefully assesses paying particular attention to urine output, heart rate, blood pressure, peripheral perfusion and lactate concentration.
- Fluids - Review fluid input and losses during surgery. Additional fluid may be required. Fluid balance must be measured 12 hourly for 72 hours. These babies should have daily weights as a part of their fluid assessments. This may be challenging for babies with silo. Discuss with the nursing team and service consultant.
Antibiotics – The duration of antibiotics should be decided in discussion with neonatal medical and surgical team

- Inert peripheral long line at earliest opportunity
- Close monitoring of PN profile should be necessary when the baby is started on parenteral nutrition.
- Signs of increased abdominal pressure – This may be indicated by increased lactate concentration, abdominal distension, upper leg oedema and decreased urine output. Increased abdominal pressure may require surgical intervention and the surgical team should be informed as soon as possible.
- Pain should be assessed, scored and treated as per Neonatal Guideline G9.

3.6.2 Nursing care of infants with Silos

These infants do not need to be muscle relaxed. Pain should be assessed and managed as per Neonatal Guideline G9.

- Colour of the bowel – the bowel must be observed for changes in colour (darker or duskier) which may suggest compromised blood supply and require a surgical opinion. Document this every 15 minutes for the first 4 hours and hourly thereafter.
- Fluid loss – There may be loss of fluid from the silo. The baby may require nursing on an incopad. If possible, gauze swabs should be used to allow a measurement of fluid loss. This should be accurately included in the fluid balance.
- Attachment of the silo to the incubator – The silo bag is usually attached to the top of the incubator. Care should be taken that there is not too much tension on the silo especially as the bowel is gradually allowed to return into the abdomen.
- Do not use the Giraffe OmniBed incubator due to the risk of inadvertently lifting canopy to which the silo bag is attached to.
- Skin care – and There remains a risk of developing pressure sores as the baby is nursed supine. Adequate attention for prevention of this should be included in nursing care. e.g. regular inspection and ure reduction mattress wheer necessary.
- Temperature control measures should be adopted

3.6.3 Abdominal assessment

Signs of increased abdominal pressure should be noted and acted on as in 3.6.1. After primary repair, there may be redness of the wound site. In discussion with the surgeons, Flucloxacillin may need to be started if there is increasing erythema or temperature of the affected skin in combination with associated signs of cellulitis/sepsis in the baby.

3.6.4 Ongoing post-operative care

- Nasogastric losses - Post-operatively, nasogastric losses greater than 20ml/kg/day should be replaced with 0.9% sodium chloride with 10mmol potassium chloride per 500 ml. The first nasogastric aspirate obtained after delivery, may be large and should not be included in the calculations for intravenous replacement.
• Parenteral Nutrition – This should be ordered to commence as soon as possible after surgery. Percutaneous central venous access will need to be obtained.

• Broviac line insertion – All patients with gastroshisis need parenteral nutrition. Some patients, especially those with complex gastroschisis may need long term PN. Initial central access should be obtained with percutaneous long lines. Broviac lines should reserved for babies where alternative central access has not been successful or later on in their course. The decision for Broviac line should be taken jointly by neonatal and surgical consultants.

• Enteral feeds -. Gastric dysmotility is very common in cases of gastrochisis which results in intolerance of enteral feeds in early days and thus delaying establishment of enteral feeds. Ensure that parents are made well aware of this and that mothers are encouraged to express breastmilk during this time. Do not use Erythromycin Domperidone or other prokinetics. Oral erythromycin in a prokinetic dose has not been shown to confer any advantage in time to full feeds (Curry et al, 2004) and there have been no studies on the effect of Domperidone on the time to full enteral feeds. Domperidone use should be discouraged in light of a recent MHRA alert (MHRA, 2014).  
• The decision on when to start feeds is made jointly with the surgeons. It takes into account the appearance of the gut prior to surgery, the condition of the gut at the time of surgery, whether the infant is passing stool and the amount of nasogastric aspirate. Parenteral nutrition should be started without delay.There is some evidence that starting enteral feeds early may be beneficial, even if minimal volumes (Walter-Nicolet et al, 2009). Delay in introducing feeds after 10 days appears to be associated with increased rates of sepsis (Singh et al, 2003). Feeds (preferably breastmilk) should be increased using the schedules in the surgical feeding guideline as a basis D10.

• Feed type – Mothers should be actively encouraged to express breast milk as there is a large body of evidence showing that all infants fed exclusively on their own mothers breast milk have less feed intolerance with earlier establishment of full enteral feeds, less time on parenteral nutrition (PN) and less sepsis (Singh et al, 2003), and reduction in necrotising enterocolitis (NEC) (Jayanthi et al, 1998). Enteral feeding with breast milk is known to reduce the stay and time to discharge(Gulack BC et al, 2016). Please refer to ‘Feeding Neonates with Surgical Problems-Guideline D10 for full guidance about feeding.

• In some cases after consultation with surgeons, Glycerin suppository tips may be used to promote bowel action. However this may be avoided if the baby is less than 7 days old or where there is a suspicion of NEC.

• Location of care – This should be on the Neonatal Unit for preterm infants and in the initial post-operative phase for all patients. However, when stable no ongoing respiratory care is needed and further surgical intervention seems unlikely, these babies may need to be transferred to paediatric surgical ward.. This is carefully considered after discussion with the operating surgeons especially in cases where there is significant delay in increasing and establishing enteral feeds., It is
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developmentally more appropriate and convenient for parents too and more applicable to babies with a corrected gestational age of 39 weeks and are at least stayed on NICU beyond 2 weeks to continue their care on the paediatric surgical ward. (See Appendix 1: Care Pathway for Infants with Gastrochisis)

3.6.5 Complicated gastrochisis: Some babies with gastrochisis may have a prolonged stay requiring combined surgical, gastroenterology and neonatology input. These babies can usually be identified early on in their course. Issues related to delayed feeding and prolonged PN including need for a Broviac line, sepsis and liver impairment need to be anticipated and appropriately managed. The families of these babies need clear communication and support.

4. Prognosis and follow up

The majority of babies with gastrochisis should have no significant long-term problems. The mortality of infants with gastrochisis is 5% with a further 5% developing short gut syndrome (Lakhoo, 2009). However, there remains a risk of death, longer duration of PARENTERAL nutrition with its problems, increased incidence of sepsis, NEC and short gut syndrome are in complex Gastrochisis i.e. the cases associated with atresia, necrosis or perforation (Bergholz R et al, 2014).

Other gastrointestinal problems include gastro-oesophageal reflux, malabsorption and faltering growth, obesity, abdominal pain and constipation (Harris EL et al, 2014).

Once discharged there is a lifetime risk of needing further surgery in 10% of these babies. Mainly due to intestinal obstruction secondary to adhesions (Lakhoo, 2009).

Due to the above risks, there should be close surgical follow up must be arranged with the operating surgeon in surgical clinic after 4-6 weeks.

5. Related guidelines

Neonatal Guideline: A5 Resuscitation at Birth A5
Neonatal Guideline: G9 Pain assessment on the neonatal unit
Neonatal Guideline: D10 Feeding Neonates with Surgical Problems

Trent Perinatal Network Guideline: Trent Perinatal Network Referral for Surgical Assessment

6. Audit Points

Time to first surgical intervention
Initiation of enteral feeds and duration of PN
Use of expressed breast milk
Appropriate transfer to paediatric surgical ward if delayed time to full enteral feeds


The purpose of this document is to describe the care pathway for infants with gastroschisis to define when and how their transfer to a paediatric surgical ward should occur to improve the care we offer these infants. This is not intended to be a guideline describing all aspects of their care.

**Stages of Care of Gastrochisis Infants**
The care of infants with gastrochisis can be divided into several stages:

**Stage 1**
The initial period of care for these infants involves peri-operative intensive care often requiring ventilation, fluid / inotropic support, careful fluid balance and intensive monitoring for a period of time (usually 1-5 days). Secure central venous access in the form of a percutaneous long line is usually obtained during this time and PN commenced.

This period of an infant’s care must occur on NICU. The mother is often still an obstetric inpatient this type of intensive monitoring and care requires NICU. Sometimes infants will have a 2 stage repair for their gastrochisis and will then need to return to theatre and will then need perioperative intensive care on another occasion (usually after about 1 week).

**Stage 2**
Following this period infants then enter a stage of waiting for the bowel to start working. NG aspirates are often high as there is an ileus and this will usually require replacement of these losses with IV fluids and careful fluid balance.

**Stage 3**
When the NG aspirates decrease and the bowel is starting to work NG feeding is carefully commenced and feeds are gradually increased over a period of several days / weeks until full enteral feeding is attained. PN is continued until this time.

During stages 2 and 3 depending on the timescale infants could be considered for transfer to the paediatric surgical ward (see below)

**Stage 4**
Establishing sucking feeds and preparing for discharge home

By stage 4 for most infants, if still expected to stay inpatient, will be cared for on the paediatric surgical ward.
Care Pathway for Infants with Gastrochisis

Date of Admission: ……. /……. /…….     Gestational age at birth: ……… Weeks ……… Days

Transfer Criteria

<table>
<thead>
<tr>
<th>Stage</th>
<th>Date</th>
<th>Name / Signature</th>
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<tbody>
<tr>
<td>Care Pathway Booklet given</td>
<td></td>
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<tr>
<td>D35 contacted – staff name</td>
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<tr>
<td>Date (s) of surgery</td>
<td></td>
<td>Primary / Secondary Repair</td>
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<tr>
<td>Parents informed of pathway</td>
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<td></td>
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<tr>
<td>Contact D35 staff by day 5</td>
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<thead>
<tr>
<th>Stage</th>
<th>Date</th>
<th>Name / Signature</th>
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<tbody>
<tr>
<td>At least 39 weeks corrected age 2 weeks old</td>
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<tr>
<td>Over 2kg</td>
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<td>Has had a trial of feeds beyond trophic</td>
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<tr>
<td>Longline / Broviac line in situ for PN</td>
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<tr>
<td>Nursed in a cot</td>
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<tr>
<td>No ongoing cardiorespiratory concerns, desaturations, bradycardias or apnoea’s for &gt; 24 hours.</td>
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<tr>
<td>Mother discharged from obstetric care</td>
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<tr>
<td>Neonatal Service Consultant agree for transfer</td>
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</tbody>
</table>

Planned date for transfer                                 Date: Signed:  
Date transferred                                           Date: Signed: