Approach to the Acute Management of Simple Gastroschisis Clinical Pathway
Johns Hopkins All Children’s Hospital

Approach to the Acute Management of Simple Gastroschisis
Clinical Pathway

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Rationale
1- To provide a standardized clinical approach to the evaluation / management of gastroschisis early in life. The approach to the long-term complications and their management is beyond the scope of this clinical pathway and will be addressed separately.

2- To standardize the choice of and duration of antibiotics coverage, for prophylaxis and treatment purposes in varying clinical presentations.

Background / Published Data and Levels of Evidence

Gastroschisis is a structural defect of the abdominal wall that involves paraumbilical herniation of abdominal organs to the right of a normally inserted umbilicus. It is usually an isolated finding with no more than 10% of cases reported as being associated with either major unrelated defects or recognizable syndromes (1).

Even though it is usually a sporadic defect in an otherwise normal infant, there are reports of affected siblings, and a population-based case–control study from California noted that 6/127 (4.7%) infants with gastroschisis have at least one affected relative (2).

The incidence of gastroschisis is estimated at around 2 to 5 per 10,000 live births.

Its etiology is unknown and thought to be resulting from a vascular compromise of multifactorial etiology involving genes and environmental factors (3). Five prevailing hypotheses have been proposed: (a) Failure of mesoderm to form in the body wall; (b) Rupture of the amnion around the umbilical ring with subsequent herniation of bowel; (c) Abnormal involution of the right umbilical vein leading to weakening of the body wall and gut herniation; (d) Disruption of the right vitelline (yolk sac) artery with subsequent body wall damage and gut herniation; (e) Abnormal folding of the body wall resulting in a ventral body wall defect through which the gut herniates (4).
Prenatal intervention attempts at amnio-exchange to improve the postnatal outcome have not been successful (5).
Available data does not provide evidence to support neither elective delivery prior to 37 weeks gestational age (GA) nor cesarean delivery (6,7).

Prenatal risk prediction
A number of studies have evaluated prenatal risk factors in an attempt to provide prognostication during counselling. A 2015 meta-analysis found that infants with the prenatal ultrasound findings of fetal bowel dilatation (FBD), gastric dilatation and polyhydramnios are at higher risk to develop postnatal complications (9). The absence of FBD is thought to exclude intestinal atresia (10). However, the predictive ability of prenatal clinical findings, in isolation or combined, remains poor.

Postnatal risk stratification
Gastroschisis postnatal outcome varies depending on a multitude of factors such as the size of the defect, constriction of the bowel loops at the level of the defect causing bowel ischemia, the integrity of the intestines, presence of bowel atresia, extent of bowel wall inflammation and severity of the associated dysmotility. Risk stratification has been routinely used to predict outcome.

The most commonly used stratification divides patients into simple (SG) and complex gastroschisis (CG) groups, with the CG defined by the presence of bowel atresia, stenosis, perforation, ischemia or a combination. Numerous studies have shown that infants with CG have an increased morbidity and mortality rate and a more complicated post-operative course. They require a longer duration of mechanical ventilation, have an extended period of adynamic ileus, and a longer time delay before tolerating full enteral feedings (FEF). Their length of stay (LOS) is accordingly longer (11,12).
Those findings were replicated in a population-based cohort study of infants with gastroschisis born in the United Kingdom and Ireland from October 2006 to March 2008 with a significant longer time to achieve FEF (median difference 21 days, 95% confidence interval 9 to 39 days), longer duration of parenteral nutrition (PN) (median difference 25 days, 9 to 46 days) and a longer LOS (median difference 57 days, 29 to 95 days). Infants with CG were also more likely to develop intestinal failure (81% vs 41%; relative risk 1.96, 1.56 to 2.46) and liver disease associated with intestinal failure (23% vs 4%; RR 5.13, 2.15 to 12.3), and were more likely to require unplanned reoperation (42% vs 10%; RR 4.39, 2.50 to 7.70). (14)
Another tool for risk categorization involves the use of the validated gastroschisis prognostic score (GPS) that is based on bowel appearance after birth. It assesses key attributes of bowel injury, specifically intestinal matting, atresia, perforation, and necrosis. A composite score of 2 or more demonstrated significantly worse outcomes compared with scores of 0 or 1 with more prolonged LOS, more days to first enteral feed, and more days on PN. A score of 4 or more demonstrated a significantly worse survival outcome (Fig. 1) (15). Of note is that only intestinal necrosis independently predicted mortality by regression analysis.

<table>
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<tr>
<td>Necrosis</td>
<td>Absent (0)</td>
<td>Present (4)</td>
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**Surgical management**

Initial management is surgical and involves either a primary closure or bedside silo staged closure based on the defect size, bowel appearance, concern for development of bowel ischemia that may result from “too tight” a closure, time/day and patient’s stability (16). Excluding premature infants and those with atresia / stenosis, no differences were seen in outcomes between the 2 approaches in terms of LOS, time to FEF, or ventilator days. (17)

**Risk for infection**

**Short-term risk and reported incidence of early onset infection:**

There has been little research evaluating the need for antibiotic treatment in the immediate post-natal period and little data available regarding the typical incidence of infection in the first week of life. The infection risk is thought to be minimal but because of the exposure of abdominal contents and absence of a protective skin barrier, these infants are often placed on empiric prophylactic antibiotics of ampicillin and gentamicin given their adequate coverage of skin flora.

A 2020 retrospective cohort studies of infants with abdominal wall defects that included 64 patients with gastroschisis reported that all infants were empirically started on antibiotics at birth and none had early-onset or pre-closure infection. (18)

Another 2022 retrospective cohort analysis in the Children’s Hospital Neonatal Database showed that 93.9% of gastroschisis patients received empiric antibiotics after delivery, with median 7 days duration (IQR 3, 9) and only 0.64% of patients had early positive blood culture (19).
To date, there are no reliable inflammatory biomarkers for early-onset infection in infants with gastroschisis. Such biomarkers are invariably influenced by the associated pathologies or interventions such as surgical closure, bowel ischemia, exposure to amniotic fluid, presence of surgical silo and/or manual manipulation. While several biomarkers are available for research use, only C-reactive protein (CRP) has been clinically evaluated and found to be a poor predictor of infection in these patients. Ram et al. found that serum CRP levels were invariably elevated in infants managed with a preformed staged silo closure in the absence of infection, with a median CRP of 8 mg/L at birth (range 6 to 55) and rising to 42 mg/L (range 35 to 68) at the time of closure (20). Another study found similar findings of elevated early CRP levels with no relationship with rates of sepsis, death or adverse outcome (PN days, time to FEF or LOS) (21).

Other studies have evaluated the usefulness of complete blood counts and immature to total neutrophil ratio (I:T) and found that infants with gastroschisis have a higher I:T ratio (> 0.2) as compared to controls (43% vs. 12%, p < 0.001) and that the left shift had no correlation with episodes of culture positive early onset sepsis, thus making the IT ratio not a reliable marker of infection (22).

While initiation of empiric antibiotics at birth is common practice, there remains variation among centers as to antimicrobial coverage during silo reduction with some centers providing continued antibiotics coverage until closure of the defect, while others discontinue the antimicrobial coverage after 48 hours despite the continued presence of a silo (16,23).

There are no studies reported that looked explicitly at antibiotics coverage during silo reduction. A 2019 systemic review and meta-analysis comparing the outcome between primary closure in the operating room (suture closure) versus bedside silo reduction (plastic closure) reported a a significantly lower incidence of wound infection in plastic closure as compared to suture closure (17.6% (6/34) versus 37.2% (35/94) with OR 0.24, 95% CI 0.09–0.69, p = 0.008, I² = 0%) (24). Those results were compatible with previous systemic reviews and RCT that compared the types of closure (25-26).

**Long-term risk:**

Infectious complications remain an important consideration in the long-term management of gastroschisis. They most commonly include abdominal wound infection, late onset sepsis and catheter-related infections (CRI). Analysis of a Canadian, prospective, disease-specific database found that the wound infection (WI) rate was greater in the delayed closure (> 24 hours) group (21.2 vs. 8.2%, p = 0.0006) and that the GPS predicted the subsequent development of an infectious complication (WI + CRI, p = 0.04) (27).
Timing of initiation of enteral feeds

A national database study cohort of 570 cases (16% with "high risk" bowel injury) found that the best outcomes occurred in patients in whom feeds were started 7 days post-abdominal wall closure. Multivariate analyses revealed that PN duration, LOS, and infectious complications were independently predicted by days to FEF (28).

A 2017 systematic review aimed to compare early (≤7 days from birth) versus delayed (>7 days) commencement of enteral feeds (CEF). It found no published randomized controlled trials (RCTs). Meta-regression results from the available 42 observational studies (4,835 infants) indicated that each day delay in CEF was associated with a delay of an additional 1.4 days (95% confidence interval [CI]: 0.95, 1.85) to FEF, 2.05 days (95% CI: 1.50, 2.59) to the duration of PN, and 1.91 days (95% CI: 1.37, 2.45) to the LOS. Sensitivity analysis after excluding studies that provided information exclusively on complex gastroschisis continued to show beneficial effects of early CEF (29).

A 2021 systematic review and meta-analysis of studies published from January 2000 to April 2019 found that the use of standardized feeding protocols resulted in fewer days to first enteral feeding by 3.19 days (95% CI: -4.73, -1.66, p < 0.0001), less complication rates, reduced mortality and better compliance to care. However, the duration of PN and time to full enteral feeding were not significantly affected (30).

Ongoing dilemmas and value of adherence to a consistent approach

An increasing number of institutions have enacted clinical practice guidelines to the approach of gastroschisis as it is a resource-intensive birth defect with wide variability in institutional practice patterns and without consensus regarding optimal surgical and medical management encompassing the optimal surgical repair method, ventilation and paralysis strategies, pain management, antibiotics coverage, central line duration and feeding regimens (31).

A report from the University of California Fetal Consortium (UCfC) sought to examine differences in multi-institutional practices as they relate to outcome. They found that despite wide institutional variation in use of silo, duration of silo, intubation/paralysis strategies, and medication use, there were no institutional differences in the primary outcome of LOS after adjusting for confounding neonatal characteristics. Silo placement was associated with increased ventilator days but there was no effect on age of reaching FF or overall LOS. Additionally, among those infants treated with a silo for <5 days, the amount of time spent in a silo did not correlate with poor outcomes other than increased ventilator days. On the other hand, use of silo for ≥ 5 days was a significant predictor of multiple poor outcomes including prolonged LOS, prolonged ventilation, increased time to FEF, and cholestasis. This is thought to be secondary to those patients having a greater degree of viscero-abdominal disproportion or more severe forms of gastroschisis (16).
Adherence to a clinical pathway for simple gastroschisis across different facilities was shown to be feasible and led to reduction in exposure to mechanical ventilation (median mechanical ventilator days 2 versus 5; \( p < 0.01 \)) and antibiotics days (5.5 versus 9; \( p < 0.01 \)), as well as earlier days to initiation of feeds (12 versus 15; \( p < 0.01 \)). The pathway promoted avoidance of routine intubation and paralysis during silo placement, expeditious abdominal wall closure, discontinuation of antibiotics/narcotics within 48 h of closure, and early initiation/advancement of feeds. However, no differences were observed in LOS (28 versus 29 days; \( p = 0.70 \)) (32)

Clinical Management of simple gastroschisis
This clinical pathway applies to approach of simple gastroschisis. Management of complex gastroschisis as defined on page 4 of Postnatal risk stratification is outside the scope of this document. Its wide heterogenicity makes codifying its approach into one clinical pathway very difficult.

Delivery room management:
- Long umbilical cord (10 cm) requested
- Wear sterile gloves and use a sterile flannel to receive the infant.
- Position patient on back and utilize moistened bowel bag to place infant from the nipple line down into the bag. Do NOT use saline soaked gauze. Utilize sterile gloves when manipulating bowel
- Position patient and bowel on right side to prevent vascular compromise
- Insert gastric tube for decompression
- Ensure adequate thermoregulation - risk of hypothermia related to surface area of exposed bowel
- Establish safe airway and suction as needed - risk of bile aspiration. Routine intubation not indicated
- Regularly assess bowel for color, perfusion or kinks

NICU management:
Pre-Operative NICU care:
- Inform surgical team
- Place oro- or naso-gastric tube on low intermittent suction for gastric decompression
- Establish peripheral intravenous access as soon as possible (preferably in the upper extremities and avoid umbilical lines)
- Maintain fluid and electrolyte balance and initiate clear fluids of D10W @ 100ml/kg/day
• Obtain CBC, blood culture and type and screen
• Initiate ampicillin and gentamicin (Antibiotic therapy to be administered within 60 minutes of silo placement).
• Encourage mother to pump and provide expressed breast milk for storage at the milk depot

Silo placement and reduction at bedside:
• Antibiotic therapy must be administered within 60 minutes of silo placement).
• Administer analgesia prior to reduction if requested by the surgical team
• Routine intubation and mechanical ventilation not indicated
• After silo is placed, wrap bottom of silo with Kerlix gauze to absorb fluid losses
• Secure silo to overhead warmer with umbilical tape to keep silo contents completely perpendicular to infant abdomen.
• Use minimal tension in securement.
• Keep warmer bed in flat position and position infant supine
• Regularly assess peripheral and bowel perfusion post silo placement
• Silo reduction by surgeon at bedside
• Daily and PRN Silo dressing changes by the surgical team (or nursing staff) utilizing Bacitracin and Kerlix gauze.

Surgical closure:
• Monitor work of breathing and assess need to escalate respiratory support given the potential for increased intra-abdominal pressure
• Tylenol PR or IV every 6 hours for 48 hours, then prn.
• Limit the use of opioids to 48 hours post-op as to not further impair gut motility. (Refer to NICU guidelines for post-operative pain management).
• Ensure adequate urine output and give normal saline boluses (15 – 20 ml/kg) as needed to ensure urine output > 2 ml/kg/hr
• Monitor abdominal wall for changes such as distension, erythema or ecchymosis
• Discontinue antibiotics 48 h after abdominal closure. Antibiotics should be continued only for sepsis or positive cultures
• After discussion with the surgical team, place Replogle to gravity when output is <20 mL/kg/day and nonbilious
• After discussion with the surgical team, initiate enteral feeds at least 24 hours after Repogle has been placed to gravity, at 20 ml/kg/day divided every 3 hours and advance per NICU feeding guidelines
• Consent mother for donor milk if she is unable to pump or cannot provide enough EMM
• Consider ID consult for complex gastroschisis or infectious complications as they are outside the scope of these guidelines

Overview of antimicrobial coverage:

• Initiate ampicillin and gentamicin at birth after obtaining blood cultures
• For primary surgical closure: Discontinue ampicillin and gentamycin 48 hours after abdominal wall closure, unless positive blood cultures or clinical instability. Do not rely on CBC or CRP values.
• For silo placement and reduction: Discontinue ampicillin and gentamicin 48 hours after birth if negative blood cultures and initiate cefazolin for wound prophylaxis. Discontinue cefazolin 48 hours after abdominal wall closure.
• Consider ID consult for complex gastroschisis or infectious complications as they are outside the scope of these guidelines

Summary
Gastroschisis is a complex defect with varying clinical course and complications. Adhering to a clinical pathway in its approach is key.
Approach to the Acute Management of simple Gastrochisis Algorithm

Initial Management
- Replogle to LUS
- IV access/feeds
- Blood culture
- Start ampicillin/gentamicin
- Surgery evaluation

Definitive to primary closure?

Yes

Primary Surgical Closure

Post-operative management:
- Respiratory support as needed
- Appropriate fluid resuscitation
- Replogle LUS
- Pain control with Tylenol and morphine per CIC

No

Site Grommet
- Use per pain management plan
- Discontinue ampicillin and gentamicin after 48 hours
- If negative blood culture and initiate oral feeds for wound prophylaxis

Reductions

Delayed Surgical Closure

Discontinue antibiotics unless patient is unstable, or for culture-positive sepsis
Discontinue narcotics

Repllogle output <20mL/kg/day & non-bilious

48 hours post-op

Discontinue Replogle
Initiate feeds at 50mL/kg/day
Glossary
CEF – commencement of enteral feeds
CG – Complex gastroschisis
CRI – catheter related infection
CRP – C reactive protein
FBD – Fetal bowel dilatation
FEF – Full enteral feeds
FF – Full Feeds
GA - gestational age
GPS - Gastroschisis prognostic score
IT – Immature to total neutrophil ratio
LOS – Length of stay
PN – parenteral nutrition
SG – Simple Gastroschisis
WI – wound infection

References


Outcome Measures

1. Compliance with antibiotics recommendations
Clinical Pathway Team

Approach to the Acute Management of Simple Gastroschisis

Clinical Pathway

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Disclaimer

Clinical Pathways are intended to assist physicians, physician assistants, nurse practitioners and other health care providers in clinical decision-making by describing a range of generally acceptable approaches for the diagnosis, management, or prevention of specific diseases or conditions. The ultimate judgment regarding care of a particular patient must be made by the physician in light of the individual circumstances presented by the patient.

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