Management of Gastroschisis

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Abstract

Gastroschisis (GS) is one of the congenital abdominal wall defects, in which the bowel has prolapsed without a covering through a defect adjacent to (and nearly always to the right of) an otherwise normal umbilicus. Proper management of such cases gives them the opportunity to survive and thrive. In this chapter, simplified flowcharts for the initial management of GS, surgical intra-operative decisions and post-operative active follow-up of such cases will be presented and discussed. The first flowchart will discuss how to deal with a GS case from birth till the operative theatre, while the second flowchart will take the lead to guide the surgeon with the available surgical options and how to choose the suitable one for the case. Finally, the post-operative active follow-up fluid management and possible complications are discussed.

Keywords: gastroschisis, AWDs, fluid management, LMIC, complications

1. Introduction

Gastroschisis (GS) or more aptly “laparoschisis” is a congenital abdominal wall defect (AWD) leading to herniation of the gut more commonly to the right of the umbilical cord (Figure 1). It differs from other AWDs in causality, risk factors, and associated anomalies [1–3].

GS incidence is increasing worldwide [4, 5] and is estimated around 1 in 2200 live births [6, 7]. Antenatal scans detect most cases [8], survival in developed countries is excellent [7] and apart from some gastrointestinal dysfunction, long-term problems are rare [9].

This chapter is dedicated to discuss in simplified flowchart-form the initial, operative and post-operative management of GS with emphasis on low-resource settings. In addition it aims to outline salient topics such as fluid management and complications.

2. Etiology and embryology

Though unexplained, a young maternal age and low socioeconomic status are the commonest risk factors for GS [10, 11]. Smoking, drugs, environmental toxins and poor nutrition have also been implicated [12]. A genetic link in the form of homozygous gene polymorphisms has been reported [13] and is substantiated by an increased prevalence among familial cases of birth defects and twins [14].

The embryological origin of GS is still a matter of conjecture. Several theories have been put forward attempting to expound the abdominal wall defect: failed body-wall folding [15]; a vascular insult to the omphalo-mesenteric artery [16] or to the right umbilical vein [17]; a localized disruption of the amniotic membrane [18]
or teratogen-induced mesenchymal failure [19]. None of the theories are fully satisfactory [20]. The right-sided occurrence of the defect has been linked to the position of the yolk sac [15, 21] without clear reasoning as to why. Left-sided defects have also been described [22].

3. Antenatal diagnosis

In high-income countries (HICs) routine antenatal scans may detect more than 97% of cases [23]. A diagnosis can be made as early as 10 weeks of gestation [24] and aids counseling, transfer and delivery [25, 26]. Ultrasound will typically pick up herniated bowel not covered by amnion, to the right of the umbilical cord (Figure 2). In contrast, an exomphalos will be covered by a membrane, lies in the
midline and may involve solid organ prolapse. Ultrasound is instrumental in picking up closing GS which is defined as a worsening ratio of intra vs. extra peritoneal bowel dilatation [27]. Further aids to diagnosis of GS are high levels of maternal serum alpha-fetoprotein (MSAFP) [28], intrauterine growth retardation with or without oligo-/an-hydramnios [7, 29, 30]. As GS is usually an isolated anomaly with very few risks to the mother or child, termination of pregnancy is not habitually offered [2, 31, 32].

4. Timing and mode of delivery

A spontaneous onset of labor will typically occur around 36 weeks gestation and the route of delivery is dependent on obstetric indications [33, 34]. There is a lack of high-level evidence to support early induction of labor in uncomplicated GS cases [35, 36] and a similar lack of evidence to support cesarean section [37]. Early (emergency) delivery is beneficial in closing GS [26].

5. GS in low to middle income countries (LMICs)

LMICs have an overall high mortality rate in neonates with correctable congenital anomalies [38, 39] and suffer from a lack of medical facilities and personnel [40]. Non-governmental and governmental organizations have been criticized for not doing enough [41, 42] though new partnerships are attempting to redress this [43, 44].

Mortality from GS in low-to-middle-income countries (LMICs) can reach up to 80–100% [45–48] which is in sharp contrast to the <10% in HICs [49]. Sepsis is a major culprit in most cases of neonatal mortality in LMICs [48]. The Gastroschisis International (GiT) network has suggested that poor resuscitation combined with sepsis and abdominal compartment syndrome is directly linked to the poor outcome [50].

Antenatal care may not be well developed [51] or mothers may engage poorly with it [52] which risks births in areas far from the reach of the pediatric surgeon. A delay in transfer of the neonate with GS remains a main concern [47, 53] however a recent study from South Africa has suggested that resuscitation at the initial point of care and throughout transfer may be the key to improving the end result [51].

6. Initial management (pre-operative management)

A GS infant is ideally delivered at or near a facility with pediatric surgical support [25]. Conversely, outborn cases have been shown to have worse outcomes such as longer days on parenteral nutrition and longer duration to achieving full feeds [54].

The accepted approach to managing GS is to cover the gut with a sterile bag (Figure 3), nasogastric decompression and fluid resuscitation. Hypothermia is a major risk due to the exposed gut and significant fluid losses [55]. Premature babies are particularly prone to hypothermia because of their high ratio of skin surface to weight and a lower amount of subcutaneous and brown fat. They may also have respiratory issues which impact on their oxygen consumption and heat production [55].

The authors follow the protocol outlined in Figure 4. At the outset doctors and nurses are reminded that the triad of hypovolemia, hypothermia and sepsis are the major threats to this neonate and that resuscitation is directed to mitigating their
Figure 3.
Cling film covering the bowel in gastroschisis.

Figure 4.
Initial management of gastroschisis. *Kinking can be avoided by laying the child on their side or by propping up the bowel with gauze rolls while the child is supine. **ABC of basic resuscitation. Do not forget blood sugar. CBC, complete blood count; UEs, urea and electrolytes; LFTs, liver function tests.
effects. Almost simultaneously, certainly not sequentially, the baby is positioned lengthwise on a resuscitaire or warmer to facilitate access. Any wires, leads or lines are shifted away from the baby and the bowel. Kinking of the bowel is avoided by laying the child on their side or by propping up the bowel with gauze rolls in the supine position. Probes for temperature and oxygen saturation are connected. ECG leads are placed and connected to a monitor. A urinary catheter is placed with an aseptic technique. Resuscitation follows APLS guidelines of airway, breathing, circulation, rapid initial examination while the bowel is covered with cling film. An appropriately-sized nasogastric tube is placed on free drainage supplemented by 2-hourly active aspiration. Peripheral vascular access is secured and bloods are taken for blood sugar (if not done earlier), a complete blood picture, kidney and liver functions, clotting and cross-match. A fluid bolus is then administered followed by maintenance according to body weight. Broad-spectrum antibiotics are given according to the hospital protocol.

7. Intra-operative decision making

The aspired aim is to achieve full reduction of the bowel with muscle and skin closure of the abdominal wall, as cosmetically as possible. Safety of the child and the gut are paramount therefore if a complete primary closure is not possible staged reduction should be considered.

All manipulations should be done in a sterile environment. The authors routinely take all cases to theatre, however bedside procedures are also possible. Central vascular access is secured and a urinary catheter would have been placed during initial resuscitation in all cases.

The authors follow the guideline outlined in Figure 5: cases of simple GS with no obvious visceroperitoneal disproportion (VPD) will undergo primary closure. If very straightforward, sutureless closure with steri-strip dressings is done. On occasion some cases will require division of bands or strands of omentum adherent to the defect and they go on to have formal sutured closure of the defect. Primary (sutured) closure has excellent cosmetic results (Figures 6 and 7). Sutureless closure is associated with a higher incidence of umbilical hernia [56, 57] Guided by ventilation pressures, cases with moderate VPD will undergo a skin closure with the size of the defect determining if the umbilicus, the skin or a prosthetic patch is needed. Marked VPD and high ventilation pressures call for staged silo closure. The authors fashion surgical silos from sterile intravenous fluid bags (Figure 8a–c). Surgical silos can be made from a variety of materials which are summarized in Box 1. Spring-loaded (pre-formed) silos are ready-made and obviate the need for suturing to the abdominal wall [20, 55]. They come in various sizes to allow for the variability in the GS defect (Figure 9). One may rely on gravity alone, active tucking or a combination of both to reduce the contents into the abdominal cavity. There is weak evidence in favor of the routine use of pre-formed silos instead of primary closure [20, 55, 58].

Complex GS is defined as any case with associated bowel atresia, stenosis, perforation or volvulus. In the presence of atresia, the authors’ preference is to plan a delayed repair but a primary resection and anastomosis at the time of abdominal closure is also acceptable if the bowel is healthy and not too edematous. Stoma formation is fraught with high-output stoma complications such as failure to thrive and peri-stoma skin breakdown—therefore is not the surgery of choice in low-resource settings. Closing GS represents a spectrum of disease where the defect has started to narrow down around the prolapsed gut. At its simplest form it can lead to intestinal stenosis but may progress to atresia, gut ischemia up to complete
Figure 5. Options for closure of gastroschisis. *Can be done cotside. **Silo material, see Box 2. ***Delayed repair is around 6 weeks post-operatively. ****Avoid stoma in a low-resource setting. GS, gastroschisis; VPD, viscero-peritoneal disproportion.
disappearance of the prolapsed bowel if the defect closes completely, aka closed GS or “vanishing gut syndrome” [61]. Closing GS is challenging even in HICs and is associated with worse outcomes compared to simple GS. Narrowing or atresia may
lend themselves to resection and primary anastomosis (either at the time of reduction, or delayed). Necrotic gut will require resection (Figures 10 and 11) and vanished gut will indicate an ultra-short intra-abdominal segment. These cases will require either primary or delayed bowel lengthening procedures [62].
8. Post-operative care

The staged reduction process should take between 1 day and 2 weeks and is dependent on the degree of VPD. Enteral feeds are started once the gastro-intestinal system shows signs of resumed function: decreased nasogastric aspirates < 20 ml/kg and bowel motions. Ideally expressed maternal breast milk is used [63–65], but formula feeds are acceptable. Elemental feeds may help protect against necrotizing enterocolitis. GS infants fed at around 7 days post closure seem to have the best outcome [66]. If the bowels do not open within 10–14 days a water-soluble contrast
enema should be done to rule out a bowel atresia. An atresia detected at the time of initial closure or by subsequent imaging may be safely repaired after 3–6 weeks. Albeit uncommon in GS, cases with any associated malformations will require further investigations and management according to the findings.

9. Fluid management in GS

Publications frequently refer to a “consensus” among doctors on the optimal fluids required for GS. However, there is sparse evidence-based literature to guide the perioperative fluid management [65, 66]. Reports of fluid administration have varied from twice to three-times the normal maintenance volumes for neonates [67, 68] and were based on original research by Phillippart et al. in 1972 [69].

Fluid overload in the absence of hypovolemia has been proven to be deleterious in neonates [70]. It affects a patent ductus arteriosus, may cause intracranial hemorrhage, bronchopulmonary dysplasia or may even be fatal [71, 72]. Therefore the mere assumption that GS cases need vigorous volume expansion may be harmful. Preterm neonates may also benefit from fluid restriction according to a recent Cochrane review [73]. It has been suggested that the fluid overload will contribute to intestinal edema leading to a longer hospital stay and longer duration of parenteral nutrition through the increase of total body water and salt [65]. It may also play a part in development of NEC [74].

This practice of over transfusion is routinely carried out postoperatively as well [75]. While it may be of value in cases with a silo where there are ongoing losses of fluid from the base, it has no real justification in cases which undergo primary closure.

Few published sources will give an outright volume to go by. They will always be ranges and the clinician must be guided by continuous assessment of the child. Postnatal diuresis can complicate fluid-balance calculations but a useful milestone to assess the cardiovascular status is after administering 40 ml/kg of fluid. Albumin has been advocated as a volume expander in hypovolemic GS cases. It is not particularly useful in hypoalbuminemia associated with sepsis [76, 77]. An additional tool to help restore insensible water loss incurred through breathing is the humidification of incubator air.

10. Complications

10.1 Abdominal compartment syndrome (ACS)

A large degree of visceroperitoneal disproportion and over-zealous reduction runs the risk of increased intra-abdominal pressure. The latter will result in restricting diaphragmatic movement and compression of the inferior vena cava, which will in turn result in respiratory distress, renal, liver and bowel ischemia, respectively. They manifest as metabolic acidosis, oliguria, renal and liver dysfunction [55].

Frequent monitoring of oxygen saturation/ventilation setting, serial blood gases, urine output, serial abdominal examinations, lower limb perfusion are important in early detection of ACS. Oliguria alone is not a sensitive indicator of ACS as it may be due to hypovolemia. Pressure measurements can be taken using sophisticated transducers used with anesthesia machines or by simply connecting the tubing to a CVP water manometer. Reference values are quoted in Box 2 [78–81].
10.2 Sepsis

Sepsis is a common complication in LMICs. Most common sources are intra-abdominal, silo/wound infections, indwelling central lines or urinary catheters [50, 51].

As always, prevention is better than cure. Meticulous antisepsis protocols and timely use of antibiotics are important first tools. Early suspicion of central line-associated blood stream infection (CLABSI) or urinary tract infection (UTI) should prompt urgent cultures to be sent to the lab. The authors remove the urinary catheter once there is a stable urine output and no further risk of abdominal compartment syndrome.

10.3 Silo complications

Both pre-formed and surgical silos are prone to dislodgement and may cause bowel kinking, ischemia and perforation [58]. It is prudent to keep the silo and its contents visible at all times to allow early detection of any of these complications.

10.4 Pneumothorax

Iatrogenic pneumothorax secondary to barotrauma is an unfortunate complication in ventilated neonates and occurs in around 8.7% of the cases [82]. In GS this may be due to high intra-abdominal pressure after bowel reduction. This is best anticipated in theatre and if pressures exceed 24 cm H₂O, a staged reduction should

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**Box 2. Reference values for safe abdominal closure [78–81].**

- Gastric/urinary bladder pressure < 20 cm H₂O or < 15–20 mmHg
- End-tidal CO₂ < 50 mmHg
- CVP < 4 mmHg or < 5.4 cm H₂O
- Ventilation <24 cm H₂O

**Figure 12.**

Large abdominal wall defect with granulation in long-standing silo.
be the surgery of choice. If post-operative ventilation is unavoidable then positive end-expiratory pressure (PEEP) or high-frequency oscillation ventilation (HFOV) is used. Neuromuscular paralysis may also help reduce ventilation pressures but is not always available in low-resource settings.

A pneumothorax is suspected when oxygen saturation drops and ventilation pressures rise sharply with absent ipsilateral air entry. An urgent plain chest x-ray will confirm this and should be followed by immediate needle decompression then a formal chest drain with an underwater seal. Bilateral asynchronous pneumothoraces are not uncommon [82].

Figure 13.
Entero-cutaneous fistula.
10.5 Others

- **NEC**: It follows the same patterns and risk factors as with non-GS infants. Prematurity, formula feeds, rapid increase in feed volume—have all been implicated. Treatment is standard: nasogastric tube decompression, gut rest and antibiotics will often suffice [83, 84].

- **Large abdominal defect**: The GS defect is seldom large to start with and is occasionally enlarged by the surgeon to facilitate bowel reduction. Hence a large defect is a rare complication which may occur in long-standing cases of staged-reduction (Figure 12). Standard closure techniques include the use of a prosthetic material or plastic surgery techniques such as abdominal wall rotational flaps with or without lateral release incisions [85].

- **Enterocutaneous fistula**: (Figure 13) rare complication which may occur secondary to wound infection, NEC, or a combination of both. Vacuum dressings have been of value in treating such a complication [55, 86]. It is the authors’ experience that vacuum dressings may paradoxically cause an enterocutaneous fistula if incorrectly placed or if the suction is too vigorous. Surgical closure when the infant is in a positive nitrogen balance is beneficial.

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