Gastroschisis: putting the bowel back safely

Gastroschisis is an anterior abdominal wall defect which is an increasing problem in the UK. This article outlines the rationale of the most common postnatal surgical management strategies to safely reduce the eviscerated bowel and close the defect.

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Key points

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1. The overall survival of gastroschisis in developed countries is greater than 90%.
2. Preformed silo staged closure makes good physiological and economical sense and has similar outcomes to immediate operative fascial closure.
3. Complex cases (intestinal atresia, perforation, necrosis, stenosis, volvulus) have worse outcomes than simple cases.
4. Further research of both the epidemiology and postnatal managements are required to understand and thus prevent gastroschisis or minimise morbidity.

Gastroschisis is characterised by eviscerated bowel herniated through a congenital abdominal wall defect invariably to the right of the umbilicus, which is distinctly different from exomphalos whereby the bowel is covered by a membrane (**FIGURE 1a and 1b**). Worldwide, gastroschisis has become a growing concern and in the last three decades there has been a steady increase in the prevalence of gastroschisis; with the UK being no exception. Currently, for all maternal ages the prevalence of gastroschisis in the British Isles is 4.0 per 10,000 live births. However more worryingly is the unexplained increase in incidence of gastroschisis in the last 10 years from 8.9 to 24.4 per 10,000 live births to mothers younger than 20 years of age[1].

Normal development of the anterior abdominal wall involves complex cell differentiation, growth and migration during the first trimester of pregnancy, converting the embryo from a flat disc of germinal cells into a cylinder by the fourth week of gestation. By the tenth week the anterior abdominal wall fuses in the midline on return of the bowel to the abdomen following 'physiological herniation of the midgut' into the umbilical cord[2]. Unfortunately, the pathogenesis of gastroschisis remains poorly understood. The most widely accepted theory is the premature regression of the omphalomesenteric artery at an early embryonic stage, causing a full thickness defect approximately 2cm in diameter in the anterior abdominal wall[3].

The postnatal management of gastroschisis has evolved considerably over the last fifty years and overall 90% of neonates are expected to survive, which is most likely attributable to advances in neonatal intensive care and the development of parenteral nutrition. Now, the focus of management is shifting away from reducing mortality, towards reducing morbidity. A vast array of surgical techniques have been described and continue to evolve in an effort to provide the best possible outcome but a number of key principles remain constant:

- Reduce the eviscerated bowel safely
- Close the defect with a cosmetically acceptable outcome
- Support nutrition until full enteral feeds are established
- Identify and treat associated anomalies
- Recognise and treat abdominal, wound and bowel complications.

Pre-operative management

As a result of antenatal screening programmes in the UK over 97% of neonates born with gastroschisis are diagnosed antenatally[4], providing an ideal opportunity for a paediatric surgeon to adequately inform the parents of this rare

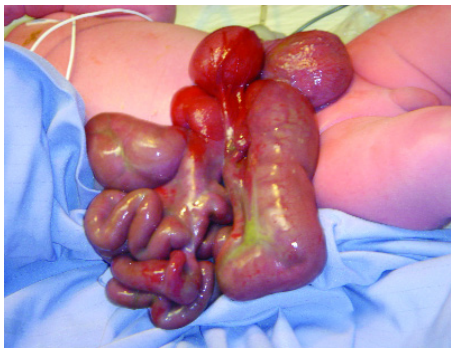

FIGURE 1a Eviscerated bowel in gastroschisis.

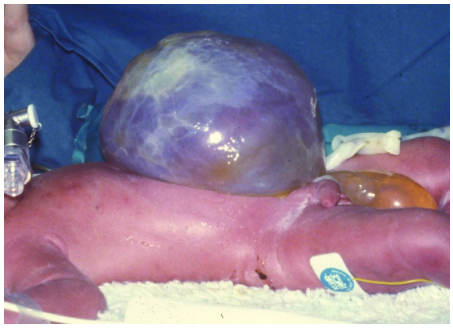

FIGURE 1b Membrane covering bowel in exomphalos.

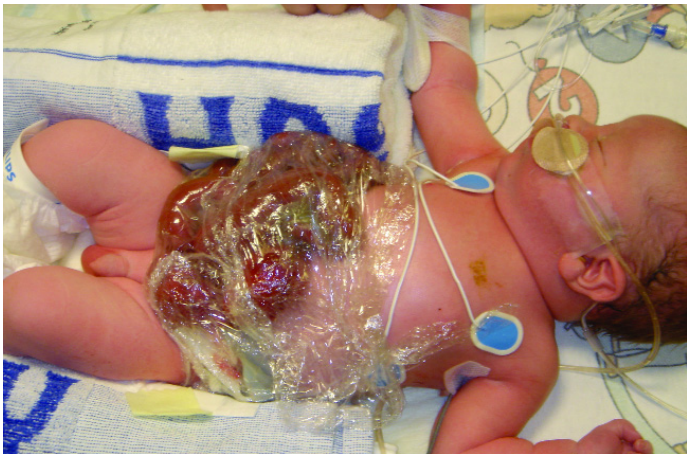

FIGURE 2 Eviscerated bowel wrapped in cling-film supported centrally on the abdomen with the neonate supine.

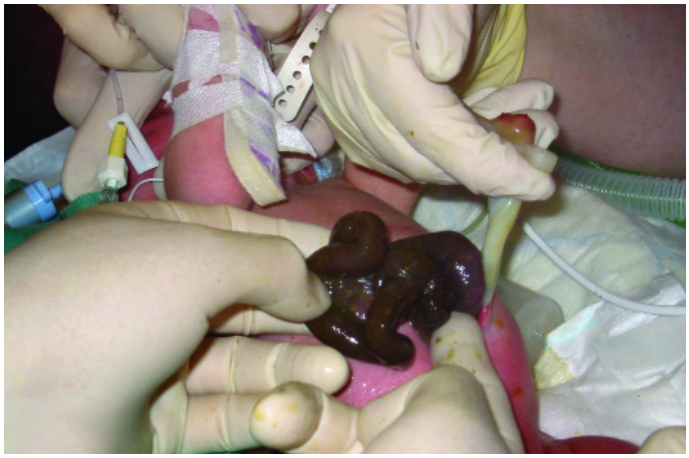

FIGURE 3 "Ward reduction" technique for gastroschisis.

condition, expected treatments, outcomes and complications. Simultaneously, arrangements should be made to ensure delivery in a tertiary centre where unforeseen complications can be dealt with by the relevant expertise and unnecessary transfers can be avoided. It makes good sense to transfer *in utero* from peripheral hospitals if possible. Presently, there is no evidence to suggest any benefit of caesarean section for delivery[5] and in the UK most centres elect for vaginal delivery at 37 weeks' gestation to avoid late fetal loss – caesarean is only indicated for maternal or fetal complications.

Immediately postpartum the child should be stabilised as for any other delivery. In addition, gastric decompression is mandatory and a large bore nasogastric tube should be inserted at an early stage. The eviscerated bowel must be covered and supported by wrapping it in cling-film which acts as a protective barrier and serves to minimise heat and fluid loss (**FIGURE 2**). Whilst applying the cling film it is essential the evsicerated abdominal contents are supported centrally on the abdomen so that neither the bowel nor the mesentery is tensioned or twisted in order to maintain bowel perfusion.

Alternatively, if the infant is haemodynamically stable, a preformed silo can be put in position by a paediatric surgeon in the delivery room, instead of using cling-film. The silo provides superior central stability and protection, as the bowel does not need to be exposed again until the defect is closed. Fluid lost from the bowel is rich in protein and the authors routinely use human albumin solution 4.5% to replace these losses and for resuscitation when required. Antibiotics should be started empirically and continued until the bowel has been reduced and the defect closed.

Reduction of the viscera

At birth the macroscopic appearance of the bowel is highly variable ranging from almost entirely normal to severely inflamed, thickened, shortened bowel that is non-compliant and difficult to handle. Bowel injury may be a consequence of injurious substances within the amniotic fluid in which the eviscerated bowel is bathed, or secondary to bowel hypoperfusion resulting from constriction of the mesentery at the defect, or a combination of both[6,7]. Reduction of the viscera is largely dependent on the degree of viscero-abdominal disproportion that usually worsens with the severity of bowel injury and decreasing abdominal capacity, which is frequently underdeveloped.

In an effort to increase the abdominal domain some authors have described milking the bowel of its contents and performing bowel washouts but these have been shown to be ineffectual[8]. However, as previously mentioned gastric decompression during reduction is mandatory to aspirate both fluid and swallowed air and this does aid bowel reduction.

Significant viscero-abdominal disproportion encountered at the procedure makes abdominal closure following reduction difficult or 'tight'. A sudden rise of intra-abdominal pressure may compromise bowel perfusion and cause cardio-respiratory embarrassment, which if unrecognised quickly progresses to abdominal compartment syndrome and its potentially devastating physiological complications of renal failure, sepsis, bowel ischaemia and wound complications.

Reduction of the eviscerated bowel and defect closure may be achieved primarily or as a staged procedure, either of which can be achieved under general anaesthesia (GA) or at the bedside, but the single most important factor is safety and the avoidance of abdominal compartment syndrome.

Primary reduction of gastroschisis

Until recently it has been widely accepted that emergency surgery and operative fascial closure (OFC) under GA offers the greatest chance of survival with minimal morbidity and only when OFC is unsafe is a staged technique employed[6,7]. Reduction of the eviscerated contents describes the first stage of OFC and is most frequently performed under GA within a few hours of life in the emergency setting.

At this early stage the bowel is usually oedematous and reduction can be difficult due to the large volume of bowel. Frequently the defect requires enlarging to reduce the oedematous bowel and stretching of the abdominal wall has been described to increase the abdominal domain. However, the resulting inflammation and oedema from the incision and stretching most likely negate these efforts and only serve to increase pain. In addition the neonate is often paralysed during OFC allowing the abdomen to accommodate the bowel with greater ease, but this makes assessment of the neonate's behavioral characteristics impossible with reliance predominantly on cardiac and respiratory parameters to assess the intra-abdominal pressure.

This lack of information prompted Bianchi to develop ward reduction (WR) whereby reduction and closure are performed with the neonate conscious (**FIGURE 3**) observing both physiological

and behavioral parameters to prevent over enthusiastic reduction[9]. However, he was not the first to describe this practice; William Fear in 1878 described the first successful management and survival of a child born with gastroschisis. Obviously the procedure was performed without anaesthesia and closure achieved "with a skein of thread"[10]. Despite initial encouraging results with WR others reported unsatisfactory outcomes and this technique is used infrequently[11].

Staged reduction of gastroschisis

Staged techniques temporarily increase the abdominal domain by using a silo, or rarely a patch, allowing the bowel oedema to reduce and the newborn time to gradually acclimatise to the increasing abdominal contents thus avoiding the sudden increase in intra-abdominal pressure observed in OFC. Traditionally, 'custom silos' have been fashioned from a wide variety of materials, most commonly Silastic® or Gortex® sheets, and sutured to the fascial edge, invariably after the defect has been enlarged (**FIGURE 4**). The bowel is gradually reduced within the silo over a number of days and the abdomen closed, frequently resulting in an uneven crimped scar and poor cosmetic outcome. Manufactured or 'preformed silos' (PS) are not a new invention and were first introduced by Shermeta in 1975[14] but have only become popularised within the last decade. PS do not require cutting or suturing to the fascia and have the advantage of being able to be sited in the delivery room or the neonatal unit, obviating the need for a GA. At the bedside the PS can simply be slipped over the bowel and the distal lower ring of the silo gently pushed through the defect, fixed in place with dressings and a ligature secured around the silo above the bowel to provide gentle downward compression. Using both gravity and compression the bowel is sequentially reduced twice a day as tolerated, until complete reduction has been achieved (**FIGURE 5a, b and c**).

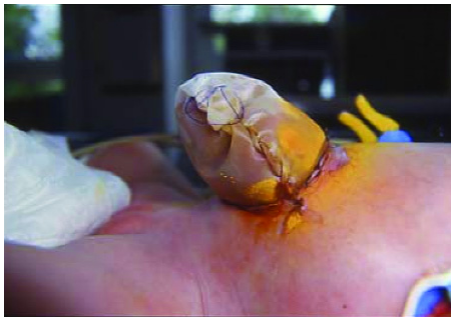

FIGURE 4 'Custom' made silo fashioned from silicone sheets sutured to the fascia.

As previously mentioned staged techniques have traditionally been employed only when primary OFC has been deemed unsafe or inappropriate. Historically, staged closure has been associated with significant increases in time to establish full enteral feeds, length of hospital stay, and complications including higher rates of sepsis and problems with silo detachment[9,10]. However data from a large multi-institutional study by Singh (2003) and more recently a randomised controlled trial by Langer and colleagues (2008) do not support these findings and demonstrated no significant differences between OFC and staged techniques for the aforementioned outcomes[15,16].

Closure of the defect

Closure of the defect following successful return of the eviscerated bowel to the abdomen may be managed using operative or non-operative methods. Whatever the strategy, preservation of the umbilical cord is essential for a satisfactory cosmetic outcome. Following primary or staged reduction of the bowel under GA the defect is usually closed with fascial sutures thus approximating the fascia and the skin above. Suture closure at the bedside without GA is more problematic due to the small volumes of local anaesthetic that can be administered and movement of the newborn during closure.

The defect seen in gastroschisis has a tendency to close naturally in the absence of high intra-abdominal pressures, intervening bowel and sepsis. Sutureless or plastic closure of the defect, using only adhesive dressings, takes advantage of this phenomenon and this technique of closure is frequently used at the bedside without GA. The authors routinely use preformed silos at the bedside without GA and perform a plastic sutureless closure where appropriate. Once the PS has been removed the umbilical cord, which has been preserved, is gently pulled to the contralateral side to appose the skin edges. Adhesive strips, then slit transparent IV dressings secure the skin edges ensuring the defect remains closed and these are left in place for 7-10 days[17]. The umbilical cord is allowed to protrude through the centre of the dressing and dry out which encourages the defect to cicatrise and close the abdomen (**FIGURE 6a and b**).

Fortunately patches are rarely used for the management of gastroschisis as the defect invariably can be closed following silo bowel reduction. Patches are fashioned from a variety of synthetic and biomaterials. Synthetic materials including polypropylene mesh, Gortex® and reinforced silicone, are used as a temporary measure with the aim of removing the patch and closing the defect ideally at the fascial level. However, synthetic materials sutured to the fascia cause a variable degree of inflammation and oedema making closure difficult with a subsequent poor cosmetic appearance. Biomaterials such as porcine-derived materials or acellular human allografts of dura or dermis are used when skin coverage is desirable and

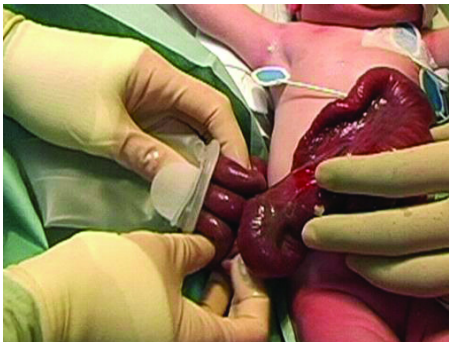

FIGURE 5a Application of preformed silo.

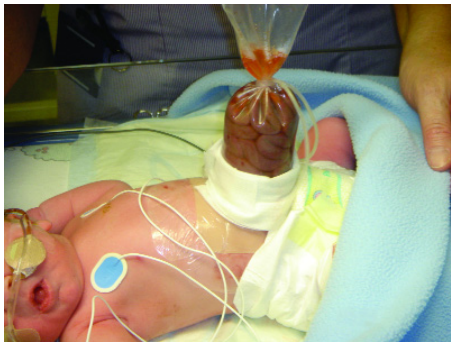

FIGURE 5b Preformed silo secured with ligature tied above bowel to provide downward compression.

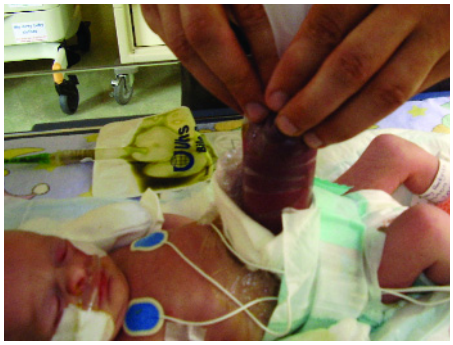

FIGURE 5c Preformed silo sequential bowel reduction.

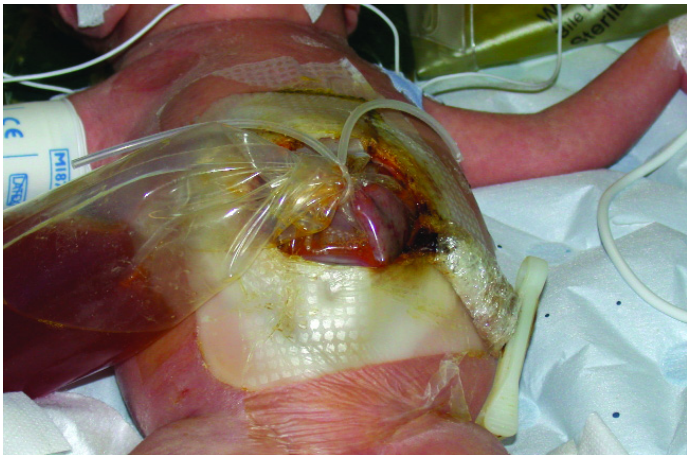

FIGURE 6a Fully reduced bowel following preformed silo sequential reduction.

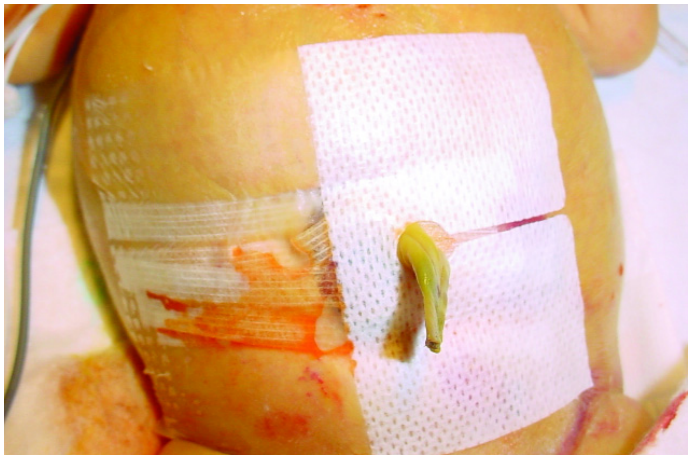

FIGURE 6b Plastic sutureless closure of gastroschisis.

are designed to encourage ingrowth of vascular tissue so they can be left in permanently[18-20].

Simple and complex gastroschisis

Simple gastroschisis describes intact bowel that is not compromised or breached. In contrast complex gastroschisis is defined by the presence of one of five criteria: intestinal atresia, necrosis, perforation, stenosis or volvulus. Fortunately, most reports refer to a proportion of 90% for simple and only 10% for complex cases of gastroschisis. Needless to say, complex cases have a significant effect on outcome with inpatient mortality rates of 2.9% for simple and 8.7% for complex, with a median length of hospital stay of 28 *vs.* 67 days respectively.

Complex gastroschisis tests a surgeon's expertise and ingenuity to the limit and in an effort to optimise outcomes a combination of the aforementioned techniques are utilised and frequently a number of procedures are required. The loss of intestinal length seen mainly in complex cases may result in short bowel syndrome whereby the bowel is unable to absorb the required nutrients to sustain life. Prolonged periods of parenteral nutrition are required whilst the bowel undergoes adaptation and numerous procedures to increase the bowel length have been described. However, despite these measures some cases will require small bowel transplantation if they meet the relevant criteria to undergo this extremely extensive surgery. Unfortunately, the mortality for small bowel transplantation is very high; 50% of those on the waiting list die before transplantation and 50% receiving donor bowel die within five years. The impact of these treatments and poor outcomes on the family unit cannot be underestimated.

Discussion

The epidemiology of gastroschisis most likely will evade understanding and hence prevention for many years to come. Only in 2004 did the Chief Medical Officer issue a report on the growing concern of gastroschisis and recommended that more research be commissioned to establish the causes of gastroschisis and investigate the increasing prevalence[1]. In response and for the first time, a collaboration between the National Perinatal Epidemiology Unit (NPEU) based in Oxford and the British Association of Paediatric Surgeons (BAPS) has enabled capture of pre- and postnatal gastroschisis data throughout the UK[4]. The study is ongoing but will hopefully help unravel the causes of gastroschisis that are most likely multifactorial.

Until then, gastroschisis will continue to test paediatric surgeons worldwide to achieve the highest survival rates and lowest morbidity, using an armoury of techniques for both simple and complex gastroschisis. Due to the ever-expanding array of surgical techniques and materials used to tackle gastroschisis it is difficult to ascertain which method should be used when and for whom. In short, a 'one size fits all' approach does not apply.

Robust evidence in the form of randomised controlled trials is distinctly lacking and often treatment is instigated on surgeon preference and then tailored or modified on an individual basis. Thus, management strategies for gastroschisis within a tertiary centre, let alone nationally, are not standardised making recruitment into protocolised studies very difficult. With this in mind the large observational studies or randomised trials required to identify superior methods may not be forthcoming.

The postnatal surgical management of gastroschisis continually evolves and seems to have gone full circle with staged reduction becoming more popular. Although the evidence is lacking, staged reduction seems to make good physiological sense allowing the bowel oedema and inflammation to reduce prior to definitive abdominal wall closure, thus reducing the risk of abdominal compartment syndrome. Preformed silos can safely be placed at the bedside out of normal working hours without the need for GA, which may also have significant financial implications.

Finally, in today's climate aesthetics are also an important consideration for both patients and parents, but safety and good surgical practice should never be compromised at the expense of cosmesis, as both may be compromised in the long-term.

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