Congenital anterior abdominal wall defects

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Abstract

Anterior abdominal wall defects are a common cause of morbidity and even mortality. These include gastroschisis, usually an isolated defect, and exomphalos, a more fundamental failure of abdominal wall formation often seen in association with chromosomal and other systemic anomalies. The worldwide incidence of gastroschisis has risen inexorably over the past 30 years while the incidence of exomphalos, at least at the time of birth, has been relatively static. This article provides an overview of the key aspects of antenatal, perinatal and postnatal management, highlighting areas of controversy where further study is required.

Keywords Exomphalos; gastroschisis; omphalocele; preformed silo

Introduction

Congenital anterior abdominal wall defects fall typically into two broad groups which are distinct embryologically and aetiologically, and present a different set of problems to the clinician. These are gastroschisis, a full-thickness abdominal wall defect almost invariably to the right of the midline, and exomphalos (also known as omphalocele) where there is an amnion-covered sac enclosing a range of viscera from a few loops of midgut to the entire midgut, liver and spleen. Cloacal exstrophy and bladder exstrophy fall within the clinical spectrum of the latter but will not be considered further.

Such defects were initially described in the first century AD by the Roman physician Aulus Cornelius Celsus, but until the middle part of the 20th century, no real distinction was made between the two. It was held that gastroschisis was purely an exomphalos with a torn or missing sac. Clinical differentiation between these entities only beginning in the 1950s and 1960s.[1]

Gastroschisis

The term, *gastroschisis*, was first used by the Italian pathologist Cesare Taruffi in 1894 and translates from Greek as '*belly cleft*'.

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We now divide the condition into two groups based largely upon appearance and complications.[2]

- **simple gastroschisis (sGS)** (80%) – no evidence of complication
- **complex gastroschisis (cGS)** (20%) – presence of intestinal atresia, necrosis or perforation.

While advances in surgical and neonatal care have achieved a contemporary survival rate in excess of 95% in most societies, it remains a major cause of mortality in sub-Saharan Africa. Indeed there is still significant debate and controversy regarding key aspects of management including timing of delivery, method for visceral reduction and technique of abdominal wall closure.

Epidemiology

Gastroschisis (Figure 1) has a rising incidence which has been noted in all regions where accurate demographic data exist. The reasons are essentially unknown. The current incidence in the UK varies from 1 in 3000 to 1 in 8000 live births, with the higher values seen in urban industrial areas. Figure 2 illustrates the rising incidence over time and geographical variation (Figure 3) for the English county of Kent. There is no specific gender predominance.

There are well-known associations with young maternal age and, at least in the UK, coming from a white, Caucasian background. There are less well-defined associations with maternal drug use (illegal [e.g. cocaine, heroin] and legal [e.g. aspirin]) and occupations involving cooking.[3] There may also be some association with environmental toxins such as the agricultural herbicide Atrazine™, though its use in the European Union was banned in 2002 and this still didn't seem to stop the rising incidence there.[3]

Pathogenesis

The actual mechanism of pathology is not known. Failure of migration and fusion of ventral body folds (particularly from the right) has been suggested together with disruption of the vitelline (omphalomesenteric) artery resulting in infarction at the base of the umbilicus.[4]

Antenatal features

Fetal ultrasound can detect gastroschisis from about 14 weeks' gestation but more typically at 20 weeks' gestation at the time of the universal fetal anomaly scan (at least in the UK). The typical findings are loops of extra-abdominal bowel floating free within the amniotic fluid, herniating through an abdominal wall defect to the right of a normally inserted umbilical cord.

Intra-abdominal bowel dilatation (IABD) is a key sonographic feature as this may reflect varying degrees of abdominal ring closure around the herniating bowel known as *closed or closing gastroschisis*[5,6] (Figure 4). At the most severe end of this spectrum there is complete ring closure and midgut infarction with subsequent intestinal resorption producing a 'vanishing midgut' and an apparently normal appearance of the abdominal wall.[5] Intestinal atresia (jejunal and colonic) occurs at the exit and entry points of the bowel with respect to the abdominal wall defect.

A recent meta-analysis including antenatal data from 2023 fetuses with gastroschisis revealed a strong positive correlation between both IABD and polyhydramnios with bowel atresia.[7] However, bowel atresia has also been documented to develop without abnormal antenatal ultrasound features, suggesting

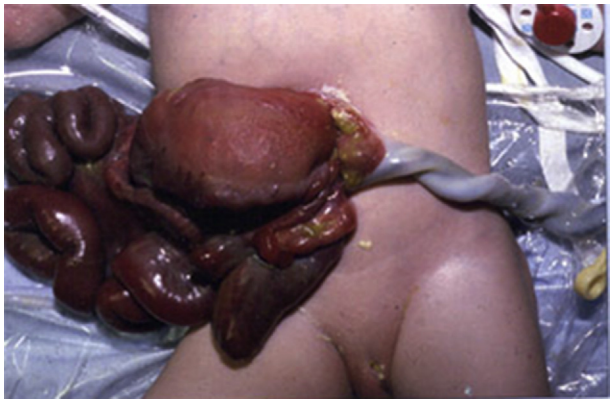

Figure 1 Gastroschisis.

caution in the interpretation of 'normal' scans and potentially the need for more regular imaging. Combined intra- and extra-abdominal bowel dilatation detected prior to 30 weeks has an improved positive predictive value for closing/closed gastroschisis compared with IABD alone.[7,8] IABD was not, however, associated with intrauterine or neonatal death, unlike the presence of gastric dilatation which was significantly associated with neonatal death in a recent meta-analysis published in 2015.[7]

There is a high incidence of intrauterine growth retardation (IUGR), developing from the second trimester and resulting in neonates that are small for gestational age (SGA). Elevated maternal serum and amniotic fluid alpha-fetoprotein (AFP) levels are commonly associated with abdominal wall defects, but second-trimester serum AFP levels with gastroschisis are markedly elevated even when compared with exomphalos pregnancies.

The optimal timing and mode of delivery has been the subject of much debate. Preterm delivery is advocated by those who suggest that prolonged exposure of the extruded bowel to constituents in amniotic fluid results in inflammation and gut dysfunction. Older studies had suggested that there is a reduced time to full enteral feeds and shorter length of stay for infants induced and delivered at $\leq$37 weeks compared to more expectant policy. However, these findings may have been confounded by the administration of antenatal steroids to this group, which may temper bowel inflammation. A recent large retrospective study from two London centres of 246 infants showed a more intuitively correct observation that there was a more prolonged time to full enteral feeds and length of stay in those delivered before 37 weeks' gestation.[8] Finally, third trimester intrauterine death (IUD) is sometimes observed and is probably caused by volvulus.

Vaginal delivery has become the standard method of delivery for infants with gastroschisis after a brief flirtation with elective caesarean section; the latter should be reserved for maternal and obstetric rather than fetal indications.

Postnatal course

Initial management includes maintenance of normothermia, bowel decompression and assessment of the herniated viscera. Secure intravenous access is gained and fluid resuscitation initiated. The herniated bowel is wrapped in 'cling film' to minimize evaporative fluid and heat loss. A nasogastric tube is inserted to achieve decompression, and can be supplemented by evacuation of meconium with rectal washouts.

The bowel is assessed for signs of ischaemia, necrosis, perforation and/or atresia. Often adjacent loops are 'matted' together and there may be an adherent inflammatory 'peel'. It is vital to check that there is no twist of the bowel inducing vascular compromise, and to divide any fibrous bands which may cause obstruction. It is also important to assess at the cot-side whether the abdominal wall defect is too tight and requires immediate division and enlargement under some local anaesthetic.

Risk stratification

The Gastroschisis Prognostic Score (GPS) was developed by the Canadian Pediatric Surgery Network (CAPSnet) as a bedside bowel injury scoring tool to stratify patients into those at either low or high risk of morbidity. This assessment can then guide parental counselling and helps predict resource utilization.[9]

The visual assessment of the degree of matting, presence or suspicion of atresia and identification of necrosis and/or perforation shortly after birth are combined to construct the GPS. Low-

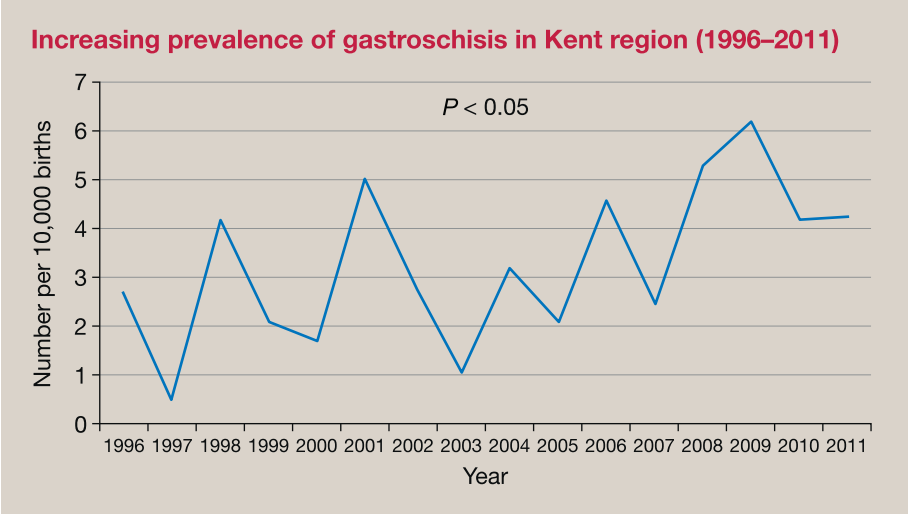

Figure 2

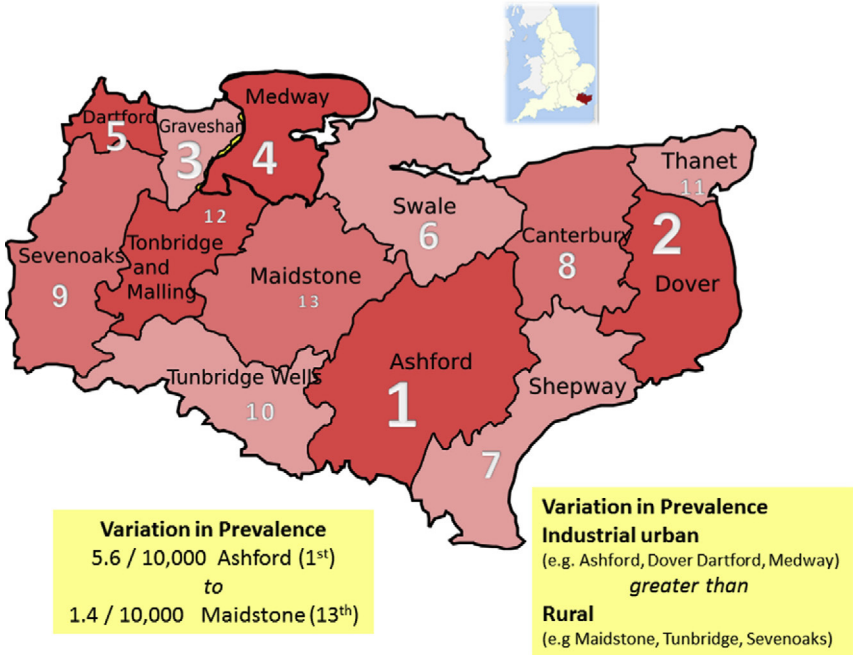

Figure 3 Kent region: variation in prevalence according to district

risk infants have a score ≤1, whereas high risk is indicated by a score of ≥2. This was originally proposed and validated in 2012, and then revalidated in 2017 using a cohort of 343 infants from the 2011–2015 CAPSnet database.

Surgical strategy

The aim is to achieve reduction of the herniated viscera and closure of the abdominal wall, avoiding the development of a compartment syndrome. With time the approach has broadened from emergent operative reduction under general anaesthesia to include several other strategies, notably cot-side application of preformed silos (PFS) without anaesthesia to facilitate a staged reduction.

Primary closure: traditionally primary closure (PC) is performed as soon as practical after birth, with reduction of the herniated viscera in the operating theatre and sutured closure of fascia and skin. If PC is not possible due to viscero-abdominal disproportion, a surgical silo is fashioned from material such as silicone sheeting or Gore-Tex™, sutured to the fascia. Staged reduction is then performed over a period of days back on the neonatal unit. When the intestines are fully within the abdominal cavity then fascial and skin closure can be performed as a separate operation.

Cot-side primary closure without general anaesthesia or sedation has also been described but is only really suitable for completely normal looking bowel without evidence of complication or inflammation.[10] Once the bowel is completely within the abdominal cavity the umbilicus can be taped over the defect avoiding the need for actual suturing.

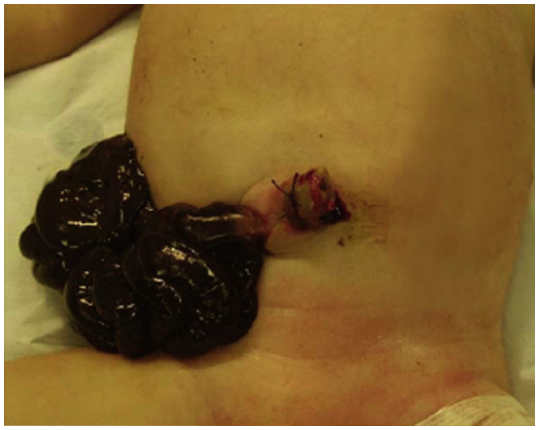

Figure 4 Closed gastroschisis in infant with midgut infarcation (reproduced with permission from Houben et al. [6])

Staged reduction: pre-formed silos are siliconized bags with a collapsible ring at the base. These can be inserted early without anaesthesia under the fascial ring at cot-side. The condition of the herniated viscera can be continuously monitored through the transparent material. Reduction is performed 1–2 times per day, typically being complete in under 7 days.

Sutureless closure: upon complete reduction of the viscera there are two main options for closure – the traditional sutured fascial repair and the sutureless method.[11] This does not require general anaesthesia and uses the umbilical cord as a biologic dressing that is fashioned to fit the individual defect and secured in place with Tegaderm™ (3M Healthcare, Minnesota) dressings. Thus, the umbilicus is centrally located, and the defect closes through secondary intention. It is of course essential to keep the cord long and moist during the staged reduction phase in order to preserve it for closure. This method has been successfully applied both primarily following postnatal reduction of the herniated viscera as well as after staged reduction using PFS. There is a significantly higher incidence of

umbilical hernias following sutureless closure as compared with standard sutured fascial closure and many will still need an operation.

Primary closure versus preformed silo - controversy: use of PFS can obviate the need for anaesthesia in the management of infants with gastroschisis when combined with sutureless closure. Gradual reduction would be anticipated to minimize the risk of intra-abdominal hypertension, subsequent respiratory compromise and/or abdominal compartment syndrome. In line with this theory, a retrospective cohort study highlighted a significantly higher urine output on day 1 of life and reduced requirement for inotropic support in neonates managed with PFS rather than PC.[12] The PC group also required higher mean airway pressures and inspired oxygen concentrations.

The only prospective randomized controlled trial (albeit underpowered) of PFS versus PC showed a trend towards fewer days of mechanical ventilation which can be used as a surrogate marker of raised intra-abdominal pressure, although this did not reach statistical significance.[13] A recent systematic review and meta-analysis incorporating the aforementioned trial and eligible retrospective cohort studies found on subgroup analysis that routine use of PFS was associated with a reduced period of ventilation.[14] Duration of parenteral nutrition (PN) was significantly greater in those managed with PFS; however, there was no difference in time to full enteral feeds or length of stay. Unplanned operations for necrotizing enterocolitis, bowel stricture, obstruction or perforation were performed significantly more often in neonates who had undergone PC.

Management of complex gastroschisis

About 10% of infants have cGS. A 15-year national review showed that of 474 neonates with cGS (11% of series overall), small and large bowel atresia/stenosis were present in 46% and 17%, respectively.[2] Intestinal atresia can be treated simultaneously with abdominal wall closure if the intestinal condition allows. Alternatively, the bowel can be reduced and the defect closed with delayed resection and primary anastomosis performed after a four to six week interval. Some centres form a stoma to permit enteral feeding in the interim.

Postoperative course

Progression to full enteral feeds is typically delayed within this population and a very prolonged need for PN has been termed gastroschisis-related intestinal dysmotility (GRID), although whether it is due to a diminution in the numbers of interstitial cells of Cajal remains speculative. Features include marked small bowel dilatation and extremely delayed intestinal transit times. Infants with complex GS are disproportionately over-represented within this group and due to the prolonged requirement for PN it is one of the most common causes of intestinal failure associated liver disease (IFALD).

Failure of conservative management with prokinetic agents (e.g. erythromycin and where available cisapride), lubricant suppositories and bowel washouts should prompt contrast gastrointestinal studies to exclude a missed atresia or stenosis. Surgical intervention may be required and includes defunctioning ileostomy, and perhaps resection of abnormally dilated small bowel and tapering enteroplasty.

Long-term outcomes

As stated previously, advances in neonatal and surgical care in the developed world have achieved neonatal survival rates for both simple and complex gastroschisis of more than 95%, although there is still significant morbidity related to failure to establish enteral feeds and the short gut syndrome. It is therefore imperative that we understand the long-term sequelae that may develop in this population.

Neurodevelopmental outcomes at one to two years are within the normal range for infants with gastroschisis. Any abnormalities appear to be correlated with being small-for-gestational age rather than the actual abdominal wall defect. A more recent long-term study of gastroschisis survivors aged 5–17 years old showed overall intellectual abilities again within the normal range.[15]

A single-centre study from Denmark evaluated the long-term outcomes of a cohort of 40 children born between 1997 and 2003[16] at a median age of 10.3 years. Of these, 11 children had had at least one episode of adhesional intestinal obstruction at up to 15 years of age, with 7 developing after the first year. Gastrointestinal symptoms prompting hospital admission at any point after neonatal discharge had manifested in 16 (40%) children. Recurrent abdominal pain was also present in 9 (22%) compared to an estimated 12% of normal Danish school children. Vitamin B12 deficiency was identified in one child who had previously undergone a terminal ileal resection.

Overall, four of seven deaths in the cohort occurred after the neonatal period. Two children with complex gastroschisis succumbed to IFALD at 1 and 3 years of age, while two other children with simple gastroschisis had died secondary to small bowel obstruction at 52 days and 6 years of age, respectively.

Exomphalos

Exomphalos, also known as omphalocele, quite literally means swelling (*kele*) of the umbilicus (*omphalos*). It is a congenital anterior abdominal wall defect at the umbilical ring covered by a membranous sac. Unlike gastroschisis, the defect arises directly from the umbilical ring rather than rather than lateral to the umbilical cord.

It is possible to determine two main types of exomphalos based on size of fascial defect and contents:

- **exomphalos minor:** fascial defect <5 cm with contents limited to the intestine
- **exomphalos major** (Figure 5): fascial defect ≥5 cm potentially containing liver, midgut, gonads and spleen. The term '*giant exomphalos*' has been used for an extreme version of this type but is not well-defined.

The membranous sac consists of three layers: an outermost layer of amnion and an innermost layer of peritoneum sandwiching Wharton's jelly. The sac itself protects the intestine which is usually morphologically and functionally normal and can support immediate enteral feeds.

Incidence

The incidence of exomphalos has plateaued since the 1970s, and is now less common than gastroschisis at around 1 to 2.5 per 10,000 live births.[17] However, its incidence when identified antenatally is much higher, 3.8/10,000 in one large 7-year UK

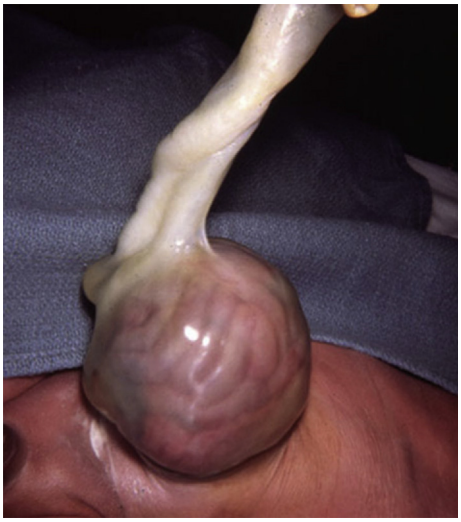

Figure 5 Major exomphalos in a term infant.

study.[18] This is because of the association with chromosomal anomalies and other potentially severe anomalies potentially leading to termination – >85% of those with a chromosomal anomaly were terminated in this latter study. In our own review from the Harris Birthright Fetal Medicine Centre at King's[19] of 445 diagnoses made during the fetal period, 56% had a defined chromosomal anomaly, 30% had a normal karyotype and the rest were not karyotyped due to parental request. Only 18% were actually live-born ($n = 55$).

Embryology

Physiological herniation of the midgut intestinal loop occurs into the base of the umbilical stalk at around 6 weeks to accommodate the increasing size of the liver. On return to the abdominal cavity at 10–11 weeks' gestation, the umbilical ring closes by fusion of the lateral, cephalic and caudal embryological folds. Exomphalos is essentially a failure of the midline fusion of these abdominal wall folds to varying degrees. As the bowel does not reduce, the rotation process is only partially, if at all, complete and as such there is usually an element of non/malrotation in these patients.

Classically, exomphalos is due to a failure of fusion of the *lateral* abdominal folds resulting in a central defect at the umbilical ring. If this defect occurs supra-umbilically (cranial fold), it can be associated with a sternal cleft, anterior diaphragmatic and pericardial defects and cardiac anomalies. This constellation is known as the *pentalogy of Cantrell* (Figure 6). A similar failure of in-folding and fusion of the caudal fold below the umbilical ring is termed *lower midline syndrome* or *cloacal exstrophy*. In these infants, infraumbilical exomphalos is associated with anorectal malformations, bladder exstrophy, epispadias and occasionally sacral defects and diastasis of the pubic rami.

A *hernia of the umbilical cord* is used to describe a much milder variant where a single loop of intestine herniates through a larger than normal umbilical stump. This is due to the failure of the physiologically herniated bowel to return to the abdominal cavity despite complete fusion of all the embryonic folds and as such is a much smaller defect which only ever contains bowel.

All of these lesions are completely distinct from the common *umbilical hernia* which is an entirely post-natal phenomenon related to failure of the cicatrization process following thrombosis of the umbilical vein and arteries.

Aetiology

It is unclear why the failure of fusion of embryonic folds occurs but is likely to be due to disruption of the delicate balance of cellular apoptosis and proliferation during this early stage of development, although the specific insult is as yet unknown. Exomphalos is associated with many other anomalies and syndromes, listed in Table 1.

Depending on the national or social attitude towards termination, most infants with exomphalos will have an associated a structural anomaly. In many this will be a recognized chromosomal abnormality, syndrome or malformation sequence; the most common of these are trisomy 18 and 13 and the Beckwith-Wiedemann syndrome.[19,20] Other associated syndromes are displayed in Table 1 for completeness. Isolated structural malformations are also commonly associated, of which a spectrum of cardiac lesions including coarctation of the aorta to septal defects have the highest incidence of up to 40%.

Management

Antenatal ultrasound as for gastroschisis, and at least in the UK is able to diagnose the majority of cases with elevated levels of AFP being used in conjunction if doubt exists. Differentiation between exomphalos and gastroschisis is by detection of a membranous sac and liver protrusion outside of the abdominal cavity. Studies have attempted to quantify whether antenatal exomphalos-abdominal circumference is predictive of primary closure with some evidence to suggest this may be of use, although in our practice this is not widely used and is perhaps of more academic value.

One of the key features during the antenatal period is to identify other anomalies or chromosomal defects. Chorionic-villous sampling (11–13 weeks) or amniocentesis (>15 weeks) should be offered to assess the karyotype and detailed ultrasound scanning should identify the presence of other system anomalies. Fetal echocardiogram is particularly useful due to the association

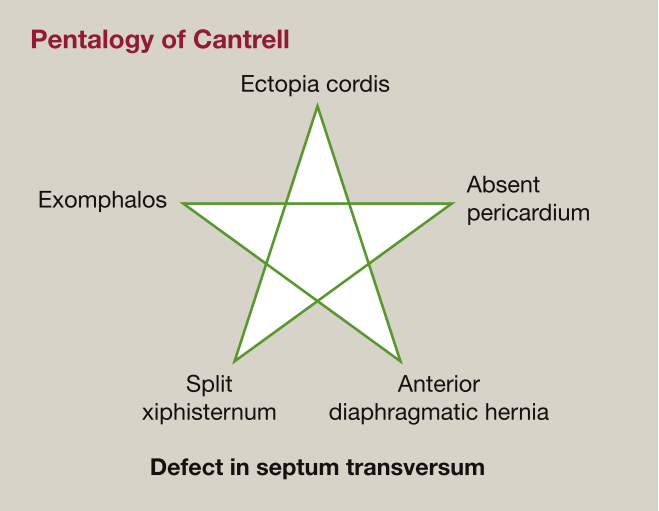

Figure 6

Syndromic associations with exomphalos

Syndrome	Features
Beckwith-Wiedemann syndrome	Macroglossia, gigantism and exomphalos. Increased risk tumour of development.
OEIS complex	**O**mphalocele, exstrophy, imperforate anus and spinal defects
Shprintzen omphalocele syndrome	Craniosynostosis, scoliosis, learning disabilities
Gershoni-Barusch syndrome	Congenital diaphragmatic hernia, radial ray defects and giant exomphalos.
Thoracoabdominal syndrome	Diaphragmatic and ventral hernias, hypoplastic lung, cardiac anomalies, cleft palate

Table 1

with cardiac defects. At this stage there is the potential for termination in cases of severe anomaly. In our practice, as stated before, <10% reached operative repair due to the high termination and fetal death rate.[19] If the choice is made to continue the pregnancy, then prenatal counselling by a neonatologist and paediatric surgeon may be offered to describe the likely postnatal treatment scenarios.

Timing and mode of delivery are determined by ongoing scans assessing growth. Most studies show no specific advantage of caesarean section over normal delivery; therefore the mode of delivery is usually based on obstetric factors. In most centres, however, the antenatal presence of a large exomphalos containing liver would almost certainly be an indication to deliver via caesarean section to reduce the risk of shoulder dystocia and liver damage during vaginal delivery.

Initial management

Prevention of sepsis and hypovolaemia are key considerations but usually not quite as urgent as for gastroschisis. However, sac rupture transforms a possible expedited elective repair into a surgical emergency and can affect outcome. Beckwith-Wiedemann syndrome can result in profound, early hypoglycaemia and as such a blood glucose should be performed as soon as possible post-delivery. Karyotype and array CGH (comparative genomic hybridization) should be sent in all cases if not performed prenatally.

Surgical strategy

There are two broad strategies to achieve surgical closure: primary repair and delayed repair.[20] The indication for either largely depends on the size of defect and degree of viscero-abdominal disproportion. Virtually all cases of exomphalos minor should be treated by primary surgical repair. In exomphalos major, a judgement must be made as to whether one can realistically close the defect early with or without placement of a custom-made silo with staged reduction over a 1–2 week period. The alternative, for those with an intact sac is to sit and wait it out for sac epithelialization to occur. Thereafter one will still have to surgically close the fascial defect, perhaps after a year or so, but at least the operation and situation will be safer.

Primary and silo fascial closure: it can be surgically challenging to close a major exomphalos and certain key points need to be kept in mind.

The liver may be completely outside the abdominal cavity and consequently course of the inferior vena cava (IVC) can be almost like a hump-back bridge. As such, the confluence of the hepatic veins will also be much more anterior and next to the xiphisternum. To separate the sac from the liver capsule one needs to mobilize this vein confluence to allow it, and the IVC, to drop back into the abdomen. The midgut will inevitably be in a state of non-rotation and kinking the porto-mesenteric vascular pedicle is always possible. Great care should be taken having returned this to the abdomen to ensure that the midgut is still well perfused with no venous compromise.

The fascial defect is always wide. Therefore, even if there is full accommodation of the viscera in the abdominal cavity, it may not be possible to close and use of a prosthetic patch use will be likely needed. Absence of skin coverage in these cases is problematic with a real possibility of ischaemic necrosis due to unsupported, thin, undermined skin.

The main and immediate problem following too tight an abdominal wall closure is compartment syndrome leading to renal impairment, acute renal failure, IVC compression, lower limb oedema and intestinal necrosis. This can be life threatening and if suspected the abdomen needs to be opened and a silo fitted. The value of actual measurement of intra-abdominal pressure using intragastric or intravesical pressure monitoring devices is arguable but they are in use in some centres. For infants where full closure cannot be performed, a silo must be sutured to the abdominal wall and the infant returned to the NICU.

Surgical closure will impair ventilation. These infants *always* have a degree of lung hypoplasia and pulmonary hypertension. As such, splinting of the diaphragm and elevation of the ribs further impairs respiratory performance post closure. Acceptance of the need for often a prolonged period of ventilation is part of the surgical strategy.

Conservative strategy

Epithelialization of the sac takes time and this allows any issues with other anomalies to be corrected or if uncorrectable, accommodated. Whether it can actually be speeded up is a moot point but a number of compounds and solutions have been used over the years. These have included solutions with mainly antibacterial properties containing povidone-iodine (Betadine™), silver salts (silver sulfadiazine – Flamazine™) and even mercury (merbromin – Mercurochrome™), although varying levels of toxicity have been reported with all three. More recent compounds such as Manuka honey may also be used. Other solutions, typically alcohol-based, are designed more to dessicate the sac. The whole process may take months to complete and usually this implies a prolonged hospital stay. Establishment of feeds is not usually a problem.

After discharge home, the covered sac needs an abdominal prosthetic device to provide support and sac security which perhaps may also encourage slow visceral return. Definitive surgical fascial closure still needs to be performed perhaps between 1 and 2 years. A small proportion have ongoing major problems with disproportion and there may be a role for

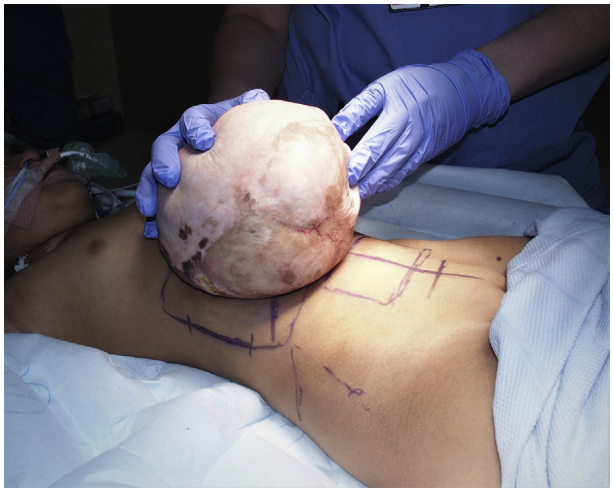

Figure 7 Major exomphalos treated conservatively in 5 year old child. Severe viscero-abdominal dysproportion still evident. [Courtesy of Mr Paul Charlesworth].

subcutaneous tissue expanders as a preliminary device to ensure provision of top-quality skin in the definitive operation (Figure 7).

Prognosis

The main prognostic factor is the presence and severity of associated anomalies, although size of defect, prematurity and sac rupture also have a role to play. In the absence of structural or chromosomal abnormalities, the majority of infants with exomphalos minor have no long-term issues. In a large multicentre series of over 2000 cases over a 10-year period in the USA, the total infant mortality rate was 28.7%, with three-quarters of those occurring in the first 28 days.[17] A hazard ratio of 7.7 was found with chromosomal anomalies, with a similar HR of 7.5 found in low-birth-weight neonates.[17] Long-term morbidity includes gastroesophageal reflux, pulmonary insufficiency and feeding difficulty. Cosmesis can be a concern, more so in those with giant exomphalos, although a recent study from the Netherlands showed that despite this, quality of life was considered good to very good in the majority of patients and comparable with controls.[21]

Outcomes

Single-centre experience can vary depending on factors such as co-location of a fetal medicine centre or paediatric cardiac centre. Our own experience reflects the former with a reported survival rate of >95% in a series of 35 cases all surgically managed with a single death in a preterm infant.[20] The latter report from Great Ormond Street of outcome of 22 infants with exomphalos major.[22] Two deaths occurred, both having undergone conservative management with sac epithelialization in light of significant comorbidities – one with a left congenital diaphragmatic hernia, the other pulmonary hypoplasia, pulmonary hypertension, ventricular and atrial septal defects. The presence of major cardiac anomalies was interestingly not associated with an increase in mortality.

Later midgut volvulus may be a complication of exomphalos being seen in 5 (4.4%) of an international cohort of 115 infants.[23] This occurred at a median age of 250 days (range 21–2109 days) necessitating bowel resection in two. Whether a pre-emptive Ladd procedure is able to reduce this incidence is not actually clear. in a large multistate American cohort of 1069 infants with exomphalos,[24] the Ladd procedure was significantly associated with a higher rate of re-admission due to volvulus than those who had not undergone a Ladd procedure.

Summary

Outcome in both gastroschisis and exomphalos is usually very good with >90% survival in both. The complications are different in both conditions and short bowel syndrome is the major problem in the former while pulmonary hypoplasia, failure to close the abdominal wall and associated conditions (e.g. cardiac and chromosomal anomalies) are the main causes of morbidity in the latter. ◆

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