Seminars in Fetal & Neonatal Medicine 16 (2011) 164–172

Contents lists available at ScienceDirect

Seminars in Fetal & Neonatal Medicine

journal homepage: www.elsevier.com/locate/siny

Neonatal abdominal wall defects

Emily R. Christison-Lagay [a], Cassandra M. Kelleher [b], Jacob C. Langer [a,*]

[a] Division of General and Thoracic Surgery, Hospital for Sick Children, 555 University Ave, Toronto, Ontario M5G 1X8, Canada
[b] Department of Pediatric Surgery, Massachusetts General Hospital for Children, Boston, MA, USA

Keywords:
Fetal diagnosis
Gastroschisis
Genetic counselling
Omphalocele

SUMMARY

Gastroschisis and omphalocele are the two most common congenital abdominal wall defects. Both are frequently detected prenatally due to routine maternal serum screening and fetal ultrasound. Prenatal diagnosis may influence timing, mode and location of delivery. Prognosis for gastroschisis is primarily determined by the degree of bowel injury, whereas prognosis for omphalocele is related to the number and severity of associated anomalies. The surgical management of both conditions consists of closure of the abdominal wall defect, while minimizing the risk of injury to the abdominal viscera either through direct trauma or due to increased intra-abdominal pressure. Options include primary closure or a variety of staged approaches. Long-term outcome is favorable in most cases; however, significant associated anomalies (in the case of omphalocele) or intestinal dysfunction (in the case of gastroschisis) may result in morbidity and mortality.

© 2011 Published by Elsevier Ltd.

1. Introduction

Gastroschisis and omphalocele are the two most common congenital abdominal wall defects (Fig. 1). Described in the literature as early as the first century AD, today these anomalies are frequently detected prenatally due to routine maternal serum screening and fetal ultrasound. Principal differences between gastroschisis and omphalocele are summarized in Table 1. Prognostically, however, the most important distinguishing feature comes not from the defect itself but from the differential rate of associated anomalies; the risk of an associated structural or chromosomal abnormality in an infant with omphalocele exceeds 50%, whereas infants with gastroschisis rarely have associated abnormalities, except for an increased incidence of intestinal atresia. Therefore, the long term outcome for neonates with omphalocele is often determined by its associated anomalies, whereas infants with gastroschisis tend to achieve normal growth and developmental milestones as they progress through childhood.

Gastroschisis occurs in 1 in 4000 live births.[1] The majority of pregnancies complicated by gastroschisis are diagnosed prenatally.[2] An elevated maternal serum α-fetoprotein level may be the earliest indicator of the presence of gastroschisis. Subsequent sonographic visualization of freely floating loops of bowel within the amniotic fluid with an abdominal wall defect to the right of the insertion of the umbilical cord at any point after the normal embryonic return of the intestine to the abdominal cavity at 10 weeks of gestation confirms the diagnosis. Adolescent mothers have an increased incidence of gastroschisis compared to older mothers but recent epidemiological surveillance data have shown a 10–20-fold increase in the overall incidence of gastroschisis in all age groups over the past two decades.[3–6] Preterm delivery is more frequent in infants with gastroschisis, with an incidence of 28% compared with 6% of normal deliveries.[7]

Bowel atresia is the most common associated anomaly in patients with gastroschisis. Recent studies report concomitant atresia and gastroschisis in 6.9–28% of patients.[8,9] Other more unusual associations of gastroschisis include the limb–body wall defect syndrome (amniotic band syndrome). In this rare syndrome, thoracic wall malformations or gastroschisis are found associated with limb abnormalities, meningocele, abnormal genitalia, intestinal atresias, and umbilical cord abnormalities.[10,11]

Omphalocele can also be a cause of elevation of maternal serum α-fetoprotein, though less commonly than gastroschisis. Prenatal diagnosis may be made as early as the first trimester if three-dimensional ultrasonography is available but is more commonly made on routine 18-week two-dimensional ultrasound.[2] The incidence of omphalocele seen on ultrasonography at 14–18 weeks is as high as 1 in 1100, but due to both spontaneous intrauterine fetal death and pregnancy termination, the incidence in live births is ~1 in 4000.[1,12]

A prenatal diagnosis of omphalocele should be followed by a comprehensive fetal ultrasound, including fetal echocardiography

* Corresponding author. Tel.: +1 416 813 7340; fax: +1 416 813 7477.
E-mail address: jacob.langer@sickkids.ca (J.C. Langer).

1744-165X/$ – see front matter © 2011 Published by Elsevier Ltd.
doi:10.1016/j.siny.2011.02.003

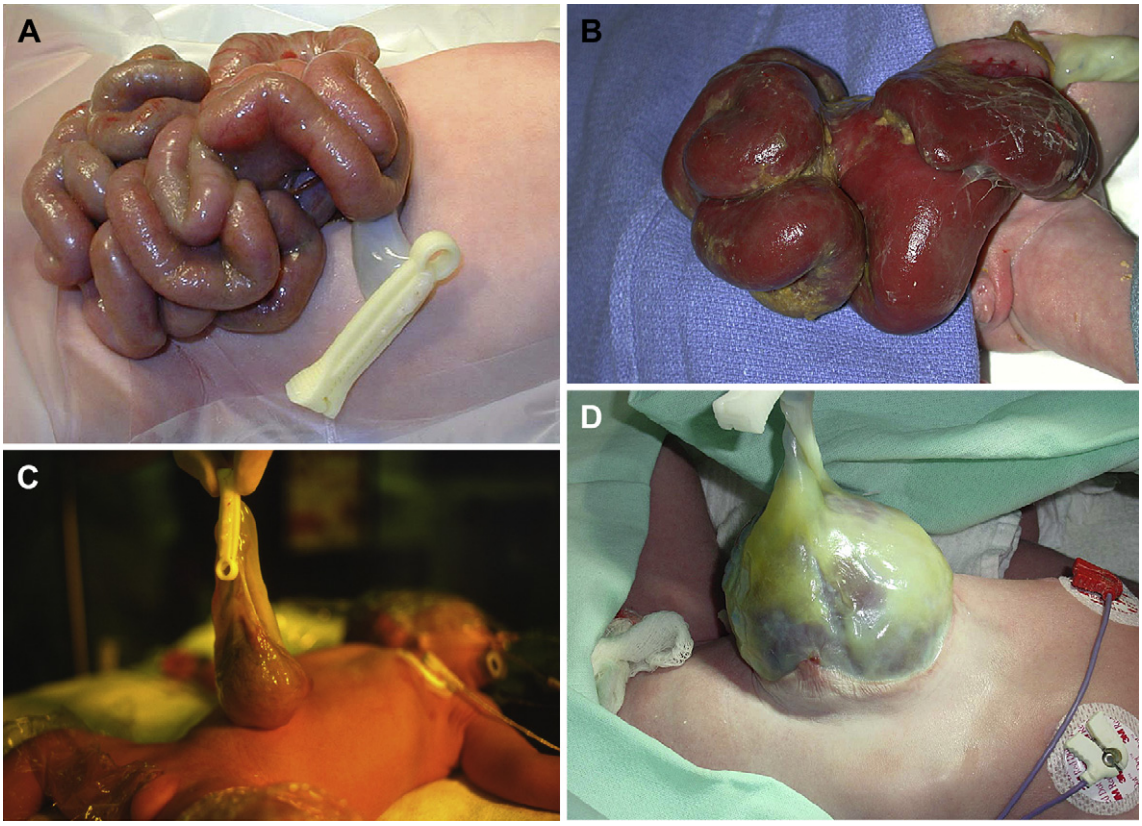

Fig. 1. Typical appearance of gastroschisis and omphalocele. (A) Gastroschisis with relatively normal bowel. (B) Gastroschisis with significantly damaged bowel with evidence of matting, foreshortening, and peel. (C) Small omphalocele. (D) Giant omphalocele with liver out.

as omphalocele is accompanied by an 18–24% incidence of cardiac anomalies.[1] Pulmonary hypoplasia is also commonly associated with giant omphalocele and may result in early respiratory distress requiring intubation and ventilatory support at the time of delivery.[13] Associated syndromes such as cloacal exstrophy, Donnai–Barrow syndrome and pentalogy of Cantrell can also be suggested by fetal ultrasound. Chromosomal abnormalities, most commonly trisomies 13, 18 and 21, occur in up to 49% of fetuses diagnosed with omphalocele.[14] The risk of a chromosomal abnormality appears to be more common in fetuses with a central omphalocele than those with epigastric omphaloceles.[14] Of fetuses with normal karyotypes, nearly 80% have multiple other anomalies.[14] Interestingly, multiple associated anomalies appear to be more common with minor omphalocele ($\leq$4 cm) than giant omphalocele (55% vs 36%).[15] Cardiac defects are the most common anomalies in this group, but such lethal entities as holoprosencephaly and anencephalus, as well as the complete spectrum of VACTERL defects, may be seen.[16]

Table 1
Characteristics of omphalocele and gastroschisis.

	Omphalocele	Gastroschisis
Sac	Present	Absent
Associated anomalies	Common	Uncommon
Location of defect	Umbilicus	Right of umbilicus
Maternal age	Average	Younger
Mode of delivery	Cesarean/vaginal	Vaginal
Surgical management	Not urgent	Urgent
Prognostic factors	Associated anomalies	Condition of bowel

2. Embryology

An omphalocele occurs when the intestines fail to return to the abdominal cavity after normal embryonic herniation into the umbilical cord during weeks 6–10 of development. This is typically attributed to a folding defect in the abdominal wall rather than to the genes involved in gut elongation and rotation.[17,18] Varying amounts of bowel may be contained within the omphalocele sac. Other intra-abdominal viscera including liver, bladder, stomach, ovary, and testis can also be found within the sac. The sac consists of the covering layers of the umbilical cord, which include amnion, Wharton's jelly, and peritoneum. The umbilical cord is attached to the sac itself.

The etiology of gastroschisis is subject to some debate. It is commonly held that the pathogenesis involves an in-utero vascular accident and, along these lines, two theories have been advanced. One theory suggests that involution of the right umbilical vein causes necrosis in the abdominal wall leading to a right-sided defect; a second theory posits that the right omphalomesenteric (vitelline) artery prematurely involutes causing a weakening in the abdominal wall through which the intestinal contents subsequently rupture.[19,20] These theories are supported by the observation that gastroschisis is associated with intestinal atresia, a condition that is also thought to be associated with an ischemic etiology.[21] Additionally, retrospective data have suggested an increased risk of gastroschisis and intestinal atresia with maternal use of vasoconstrictive drugs (ephedrine, pseudoephedrine, or cocaine) as well as with smoking.[22] More recent epidemiological and scientific data suggest that these explanations may be insufficient. Feldkamp et al. note that both umbilical veins degenerate, which does not explain the predominant right-sided occurrence of gastroschisis.[23] Moreover, the body wall derives arterial supply

from a rich arcading network of vessels arising from the dorsal aorta that is neither dependent upon nor intersects with the umbilical or vitelline vessels. A recent large scale epidemiological study measuring the associations between maternal vasoactive exposures, as part of the National Birth Defects Prevention Study, found that vasoactive risk factors play a minor role, if any, in the etiology of gastroschisis in young mothers, but may play a larger role in mothers aged >25 years.[24] Nonvascular explanations for the origin of gastroschisis include failure of incorporation of the vitelline duct into the umbilical cord and abnormal development of the ventral abdominal wall resulting in the failure of midline fusion of the lateral folds.[23,25] In-utero rupture of an omphalocele has also been proposed as a mechanism of gastroschisis formation.[26]

3. Gastroschisis

3.1. Perinatal care

The outcome for infants with gastroschisis is primarily determined by the amount of intestinal damage that occurs during fetal life. The etiology of this injury is likely a combination of exposure to amniotic fluid and constriction of the bowel at the abdominal wall defect and much of the damage seems to occur toward the end of pregnancy.[27,28] Intestinal damage results in impaired motility and mucosal absorptive function which, in turn, lead to prolonged need for total parenteral nutrition and, in some cases, severe irreversible intestinal failure.[29] Prenatal diagnosis provides a potential opportunity to modulate mode, location, and timing of delivery in order to minimize these complications.

The optimal mode of delivery for fetuses with gastroschisis is debated. Proponents of routine cesarean delivery argue that the process of vaginal birth may injure the exposed bowel. However, this philosophy is not supported by published data which have failed to demonstrate a difference in outcomes between cesarean section and vaginal delivery.[30,31] Therefore, the delivery method of a neonate with gastroschisis should be at the discretion of the obstetrician and the mother.

Timing of delivery is also controversial. Some centers advocate early delivery of the fetus with gastroschisis in an attempt to reduce the inflammatory peel on the surface of the bowel. Evidence supports a role of amniotic fluid cytokines and proinflammatory mediators (including interleukin-6 and interleukin-8) in damaging the myenteric nerve plexus and interstitial cells of Cajal in gastroschisis.[30,32–35] Because bowel edema and peel formation increase as pregnancy progresses, early delivery is thought by some to mitigate these effects. However, the literature is mixed in terms of the benefit of preterm delivery. Labor can be successfully induced in a high percentage of cases at 36–37 weeks of gestation in gastroschisis pregnancies, probably because of the inherent tendency toward preterm labor.[8] The argument against early delivery is that low birth weight negatively influences outcome, with neonates weighing <2 kg having increased time to full enteral feeding, ventilated days, and duration of parenteral nutrition compared with those weighing >2 kg.[36] Some authors advocate selective preterm delivery based on the appearance of bowel distention and thickening on prenatal ultrasonography. The presence of dilated fetal bowel has been shown to correlate with poor outcome, including fetal distress and demise in some – but not all – series.[37,38] Many of these data are confounded by non-standardized paradigms for measuring bowel and lack of consensus upon the definition of 'dilated' at any given gestational age.

Most authors advocate delivery at a tertiary perinatal center so as to provide immediate access to neonatal and pediatric surgical expertise. A recent outcomes analysis found that delivery at a perinatal center with immediate access to level 3 neonatal ICU and pediatric surgeon was associated with an overall reduction in risk adjusted morbidity when compared to delivery at a community hospital.[39]

3.2. Neonatal resuscitation and management

Gastroschisis causes significant evaporative water losses from the exposed bowel. Following delivery, appropriate intravenous access should be obtained and fluid resuscitation initiated. Gastric decompression is important to prevent intestinal distension. The herniated bowel should be wrapped in warm saline-soaked gauze, placed in a central position on the abdominal wall with the baby positioned on the right side to prevent kinking of the mesentery, and wrapped with plastic wrap to reduce evaporative losses and temperature instability. A thorough examination of the neonate should be performed to exclude the coexistence of other anomalies with specific attention to the bowel for evidence of intestinal atresia, necrosis or perforation.

3.3. Surgical management

Surgical management of gastroschisis varies from center to center and has evolved over the past several decades, particularly with the introduction of the spring-loaded silo. The primary goal of every surgical repair is to return the viscera to the abdominal cavity while minimizing the risk of damage to the viscera due to direct trauma or increased intra-abdominal pressure. Options include: (i) primary reduction with operative closure of the fascia; (ii) silo placement, serial reductions, and delayed fascial closure; (iii) primary or delayed reduction without fascial closure. In addition, the timing and location of surgical intervention is controversial, ranging from immediate repair in the delivery room, to reduction and closure in the neonatal intensive care unit, to surgical closure in the operating room.[40,41] In all cases, inspection of the bowel for obstructing bands, perforation, or atresia should be undertaken. Bands crossing the bowel loops should be divided before silo placement or primary abdominal closure to avoid subsequent bowel obstruction. Consideration should be given to the early establishment of central venous access, as intestinal hypomotility is invariably present.

3.3.1. Primary closure

Historically, urgent primary closure of gastroschisis was advocated in all cases. This approach was supported by multiple retrospective reviews of primary versus staged closure documenting improved outcomes in those patients undergoing primary closure. Upon closer analysis, however, these results were likely skewed by a significant selection bias because only unstable patients, or those with the greatest intestinal damage or largest defects, were likely to undergo staged repair. Nonetheless, many centers still practice primary closure in neonates considered to possess sufficient intra-abdominal domain to permit full reduction of the herniated viscera. Recent data from the Canadian Pediatric Surgeons Network (CAPSNet) database suggests that infants who are able to undergo immediate primary reduction and closure have a shorter length of parenteral nutrition use and total length of stay when compared with those who require staged reduction and delayed repair.[42]

3.3.2. Staged closure

Originally, staged closure consisted of placing the bowel into a silo constructed of Silastic sheets sewn together and sutured to the abdominal wall. In recent years, the use of a hand-fashioned silo has been eclipsed by the introduction of a prefabricated silo with a circular spring that can be placed into the abdominal defect at the bedside, without the need for sutures or general anesthesia (Fig. 2). In either case, the bowel is reduced once or twice daily into the

abdominal cavity as the silo is shortened by sequential ligation. When the eviscerated contents are entirely reduced, the definitive closure can be performed. This process usually takes between one and 14 days, depending on the condition of the bowel and the infant. The preformed silo can also be used at the bedside to reduce the bowel, with abdominal closure being done immediately without sequential ligation of the sac.

Advocates of delayed closure argue that avoidance of high intra-abdominal pressure reduces the risk of ischemic injury to the viscera and may permit earlier extubation.[43,44] In a prospective study using historical controls, our group demonstrated that the routine use of a spring-loaded silo was associated with a shorter time on the ventilator, lower postoperative airway pressures, shorter hospital stay, lower cost, and lower risk of complications, although other centers have had more variable success.[45–48] The discrepancy of these results in otherwise similarly designed studies suggests that institutional practice protocols, more than the routine use of spring-loaded silo alone, may affect endpoint outcome. In all studies, routine silo management was found to be safe and effective, without an increase in morbidity or mortality over attempted primary closure techniques.

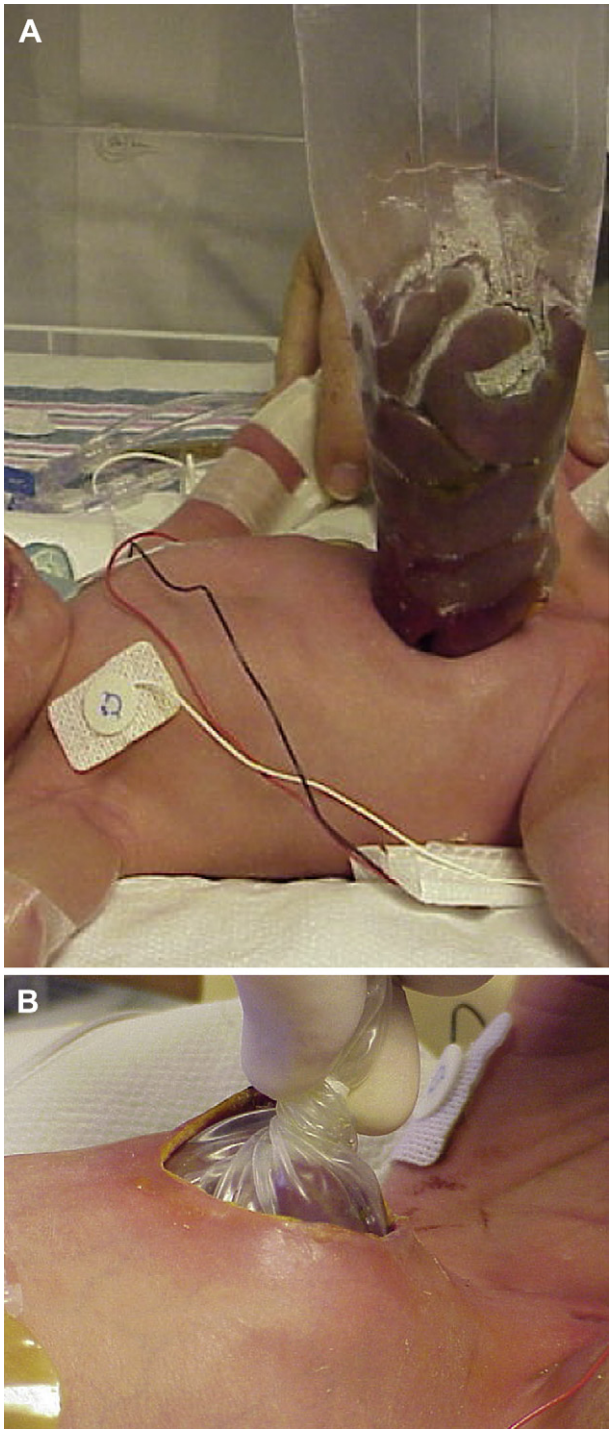

Fig. 2. Preformed silo for the management of gastroschisis. (A) Initial placement of the silo. (B) Complete reduction prior to definitive fascial closure.

3.3.3. Closure techniques

Definitive closure in the operating room consists of raising skin flaps around the fascial defect to facilitate fascial closure followed by subsequent skin closure. Closure of the skin in a linear fashion creates a 'keyhole' appearance with a horizontal scar to the right of the umbilicus. Some surgeons advocate a purse-string suture of the skin around the umbilicus to create a circular scar with improved cosmesis. Recently, the 'plastic closure' method has been introduced in which the umbilical cord, if not too macerated or dry, is tailored to fill the gastroschisis defect and is then covered with an adhesive dressing.[49] This technique allows for a centralized umbilicus without any linear scar component and is best utilized with small defects. If the umbilical cord is not salvageable, the bowel can be directly covered with a non-permeable dressing. This dressing is changed every five to seven days and the wound inspected. Ingrowth of granulation tissue and epithelialization occurs over time. With this technique, an operation can be avoided entirely in many infants. Residual ventral hernia rates are reported to be 60–84%, the majority of which close spontaneously.[50,51]

Multiple methods of closure have been described for children in whom primary fascial closure cannot be achieved. The simplest involves using the umbilicus as an allograft.[52] Prosthetic options include both non-absorbable mesh and biosynthetic absorbable patches such as dura or porcine small intestinal submucosa (Surgisis™, Cook Inc., Bloomington, IN, USA). Another option in selected cases is to reduce the bowel and place a piece of Silastic sheeting under the abdominal wall to prevent evisceration. This technique is useful when the surgeon is concerned about increased intra-abdominal pressure leading to a deterioration in pulmonary function with fascial and skin closure. The Silastic sheet is removed in 4–5 days, and the abdominal wall and skin are closed.

Intra-abdominal pressure, measured either as intravesical or intragastric pressure, can be used to guide the surgeon during reduction. Pressures >20 mmHg are correlated with decreased perfusion to the kidneys and bowel.[53] Similarly, an increase in central venous pressure >4 mmHg has been correlated with the need for silo placement or patch closure during attempted primary repair.[54] Following reduction, careful attention should be paid to physical examination, urine output, and lower limb perfusion with a low threshold to reopen a closed abdomen for signs of abdominal compartment syndrome.

3.3.4. Management of associated intestinal atresia or perforation

Up to 10% of neonates with gastroschisis will have an associated intestinal atresia, most commonly jejunal or ileal. These atresias can be treated at the time of abdominal wall closure with resection and primary anastomosis if bowel inflammation is minimal. If the condition of the bowel makes primary anastomosis inadvisable, the bowel can be reduced with the atresia intact and repair can be undertaken four to six weeks after the initial abdominal wall closure. Stoma creation is another alternative and is particularly helpful in the case of distal atresia.[55] Intestinal atresia should be differentiated from 'vanishing gastroschisis'. This condition is usually associated

with a very small abdominal wall defect that strangulates the extra-abdominal viscera and is characterized by necrosis and disappearance of some or all of the intestine (Fig. 3). Although this happens rarely, it usually results in short bowel syndrome.

Perforation may be managed through many of the same techniques as atresia. If bowel inflammation is minimal, the perforated segment can be resected and continuity established with a primary anastomosis. Other options include ostomy creation and primary fascial closure. If primary closure is impossible, exteriorization of the perforation through a hole in the silo is another option. Once the bowel has been reduced, a formal stoma can be created at the time of abdominal wall closure.

3.3.5. *Postoperative course*

Gastroschisis is associated with abnormal intestinal motility and nutrient absorption, both of which gradually improve over time in most patients. Introduction of enteral feeding is often delayed for weeks while awaiting return of bowel function. During the period of dysmotility, nasogastric decompression and parenteral nutrition is required. Secure central venous access through a percutaneously inserted central catheter (PICC) or tunneled cuffed central line is of utmost importance. Often there is initial feeding intolerance with the need for slow progression of feeds. It is also important to include early oral stimulation, because the sucking–swallowing reflex can be lost while awaiting bowel function and a significant oral aversion may develop.

Gastrointestinal dysmotility is often treated with prokinetic agents in these infants. However, there is little documentation in the literature to support their use. Commonly utilized prokinetics include erythromycin, metoclopramide, domperidone and cisapride. A randomized controlled trial of erythromycin versus placebo showed that enterally administered erythromycin did not improve time to achieve full enteral feedings over placebo.[56] However, a similar randomized trial examining the use of cisapride in postoperative neonates, most of whom had gastroschisis, did show a beneficial effect.[57] Cisapride is available only on a compassionate basis in North America due to its relatively high risk of cardiovascular side effects, and it therefore can only be used in infants with severe dysmotility who have not responded to other agents and who have undergone cardiac evaluation and clearance.

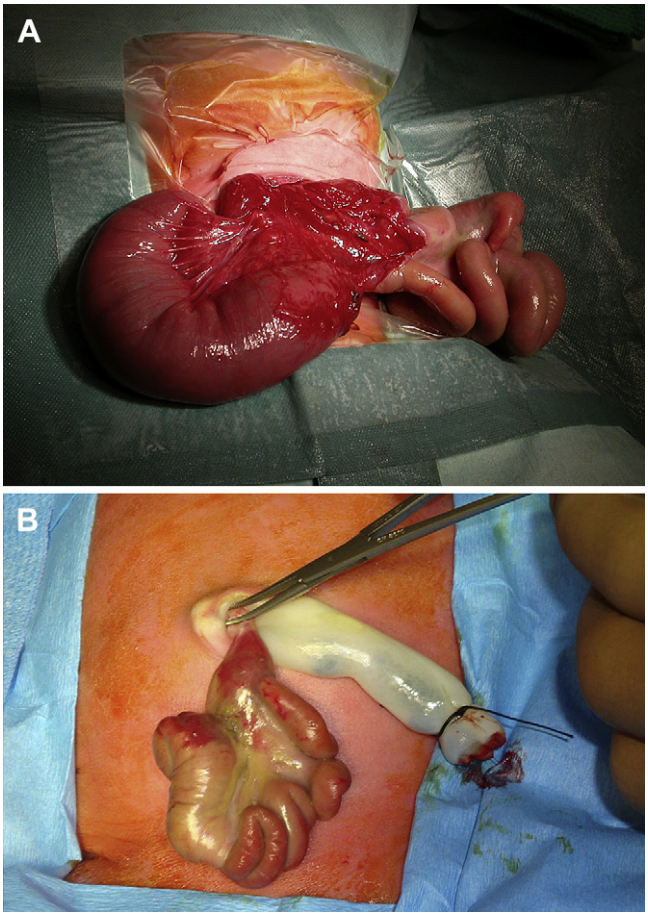

Fig. 3. (A) Gastroschisis with associated intestinal atresia. (B) Gastroschisis with 'vanishing bowel' due to a very small abdominal wall defect, which is usually associated with short bowel syndrome.

Necrotizing enterocolitis has been encountered in full-term infants with gastroschisis in higher than expected frequencies (up to 18.5%).[58] While significant bowel loss from necrotizing enterocolitis can predispose patients to short bowel syndrome and its associated hepatic and septic complications, at least one group found that most gastroschisis patients with necrotizing enterocolitis exhibited a subsequently uncomplicated clinical course.[59]

Attempts to stratify patients with gastroschisis according to risk have found that intestinal damage or complex anomalies (those with atresia, volvulus, necrosis, or perforation) predict a more prolonged hospital course and increased morbidity and mortality.[60,61] Infants with gastroschisis complicated by intestinal atresia, necrotizing enterocolitis, cardiac disease, or pulmonary hypoplasia/bronchopulmonary dysplasia are at a 2–14-fold increased risk of death compared to those with an isolated defect.[62] The ability to risk-stratify gastroschisis patients with respect to increased morbidity and mortality has utility in counseling families, predicting hospital utilization, and identifying a group of patients who would benefit from further strategies to improve outcomes.

4. Omphalocele

4.1. *Perinatal care*

The delivery of patients with omphalocele, like those with gastroschisis, should be dictated by obstetric considerations, because neither vaginal delivery nor cesarean section has been shown to be superior. Nonetheless, most practitioners choose to deliver neonates with large omphaloceles by cesarean section because of the fear of liver injury or sac rupture during vaginal delivery.[16] In addition, most authors advocate delivery at a tertiary perinatal center to allow immediate access to neonatal and pediatric surgical expertise. There is no advantage to preterm delivery for fetuses with omphalocele.

4.2. *Neonatal resuscitation and management*

Initial management in the delivery room for an infant with omphalocele involves careful attention to cardiopulmonary status, since these children may have unsuspected pulmonary hypoplasia that requires immediate intubation and ventilation.[63] A thorough search for associated anomalies should then be undertaken. The high risk of associated cardiac defects mandates a directed cardiac evaluation, including auscultation, four-limb blood pressures, and peripheral pulse examination.[64] Once stabilized, a more detailed evaluation can be provided by echocardiography. Likewise, an abdominal ultrasound should be obtained to evaluate the possibility of associated renal anomalies. Neonatal hypoglycemia should alert the practitioner to the possibility of Beckwith–Wiedemann syndrome.

In preparing infants with omphalocele for transport, risks arising from associated anomalies should be specifically addressed. Adequate intravenous access should be obtained and fluid

resuscitation begun. Infants with omphalocele and an intact sac do not have fluid and temperature losses as significant as those with gastroschisis, but the losses are higher than those with an intact abdominal wall. The omphalocele itself can be dressed with saline-soaked gauze and an impervious dressing to minimize these losses. A nasogastric or orogastric tube should be inserted and placed to suction or gravity drainage. In cases of a ruptured omphalocele, the initial management of the viscera should be the same as described for the infant with gastroschisis.

4.3. Surgical management

4.3.1. Primary closure

Treatment options in infants with omphalocele depend on the size of the defect, gestational age, and the presence of associated anomalies. In infants with small defects, primary closure – consisting of excision or inversion of the sac, with closure of the fascia and skin – may be easily accomplished. This option is predicated on the ability of the patient to withstand an operation if associated cardiac disease is present. Care should be taken when attempting to excise the portion of the sac covering the liver as it is typically densely adherent and excision can tear Glissen's capsule leading to hemorrhage. There is also significant anatomic distortion of the liver in patients with omphalocele and the hepatic veins may be located just under the epithelial/sac interface in the midline, making them susceptible to inadvertent injury. Thus, most pediatric surgeons advocate leaving part of the sac on the liver. Additionally, the inferior portion of the sac covering the bladder can be quite thin, and excision of the sac in this area can lead to inadvertent bladder injury. It is not unusual for an omphalomesenteric duct remnant to be associated with a small omphalocele, and this should be closed during primary repair. As with gastroschisis, intra-abdominal pressure can be measured during reduction in order to avoid abdominal compartment syndrome. In a review of omphalocele treatment at one institution, the authors reported a 12% incidence of complications of increased intra-abdominal pressure after closure, including acute hepatic congestion requiring reoperation, renal failure requiring dialysis and bowel infarction.[65] A high index of suspicion and careful attention to ventilatory pressures, intra-abdominal pressure, urine output, and distal perfusion can minimize these types of complications. Specific to omphalocele, when reducing the contents, attention must be given to the position of the hepatic veins because kinking may result in acute obstruction.

In many cases, the defect is large and the loss of domain in the peritoneal cavity prevents primary closure due to an unacceptable increase in intra-abdominal pressure. Multiple methods have been proposed to achieve primary fascial closure of the abdominal wall in this setting. Flaps that mobilize the muscle, fascia, and skin of the abdominal wall toward the midline and allow midline fascial closure have been used successfully.[66,67] Component separation at the level of the external oblique has also been described.[68] A more recent approach is the use of tissue expanders inside the abdominal cavity to reduce abdominovisceral disproportion.[69] Volume is added to the tissue expander over time until a primary fascial closure can be performed. Some surgeons prefer to place a patch in the abdominal wall and close the skin over the patch. Infection of nonabsorbable patch materials (Marlex, Gortex, Prolene), leading to mesh removal, has prompted investigation into the use of bioabsorbable materials (small intestinal submucosa, dura, or acellular dermis).[70–72]

4.3.2. Staged closure

In 1967, Schuster described the use of a silicone plastic 'silo' to provide staged reduction for children with omphalocele.[73] The sac was excised, and the silo was sewn to the rectus fascia and over the top of the viscera. Some surgeons created a short circumferential skin flap so that the silo was sewn to the fascia only, and some attached the Silastic to the full thickness of the abdominal wall. Some surgeons have recently recommended the use of preformed spring-loaded silos in this setting, but this is usually unsuccessful owing to the relatively large size of the defect that prevents the silo from remaining in place.

Another option for moderate-sized omphaloceles with a relatively thick sac is to use sequential ligation of the sac itself for gradual reduction of the viscera.[74] Serial reductions, similar to that for gastroschisis, are performed on a once- to twice-daily schedule until definitive closure can be obtained. At this time, the infant is returned to the operating theater for definitive closure of the defect. If the fascial edges cannot be approximated at this time, prosthetic closure can be utilized.

'Escharotic therapy', which results in gradual epithelialization of the omphalocele sac, is another form of staged closure that can be used for neonates who cannot tolerate operation due to prematurity, pulmonary hypoplasia, congenital heart disease, or other anomalies (Fig. 4). Historically, mercurochrome was used as both a scarificant and a disinfectant; however, reports of deaths due to mercury poisoning led to abandonment of this treatment option.[75] Povidone-iodine has also been used; systemic absorption of the iodine component during the initial therapy has been associated with transient hypothyroidism. Absorption is negligible after escharification, but infants treated with povidone-iodine as a scarifactant should undergo monitoring of thyroid function.[76] Silver sulfadiazine is the most common topical applicant currently in use. Once initial cicatrization has begun, silver sulfadiazine may be exchanged for an absorbant synthetic fiber such as Aquacel™ (ConvaTec, Québec, Canada) to keep the scarred sac dry while epithelialization gradually occurs. Escharotic therapy usually takes many months for the sac to granulate and epithelialize. Once epithelialization has occurred and the infant is stable enough to undergo anesthesia and surgery, the remaining ventral hernia can be repaired by one of the previously mentioned methods, usually requiring use of prosthetic mesh with skin flap coverage, especially at the upper end of the defect. Tissue expanders have been used at this stage as well as in the neonatal period to create an abdominal cavity big enough to house the viscera.[77]

4.4. Postoperative course

If primary closure has been accomplished, the majority of patients will require mechanical ventilation for a few days postoperatively. During this time, the abdominal wall and bowel wall edema will resolve and the intra-abdominal pressure will decrease. A nasogastric tube should be utilized for gastric decompression. Feeding can begin when the nasogastric output is no longer bilious, the volume is minimal and bowel activity has occurred.

The method of closure (primary, staged with delayed primary closure or prosthetic mesh) has not been shown to affect length of hospital stay. The time to resumption of enteral feeding, however, may be shorter with primary closure although this finding may be biased by omphalocele size and comorbidities.[72]

5. Long-term outcomes

5.1. Gastroschisis

Long-term outcomes for patients born with gastroschisis are generally excellent. Although historically the presence of bowel atresia was felt to be the most important prognostic determinant for a poor outcome, many patients with atresia do very well as long as the bowel is not irreversibly damaged during fetal life.[78] Patients

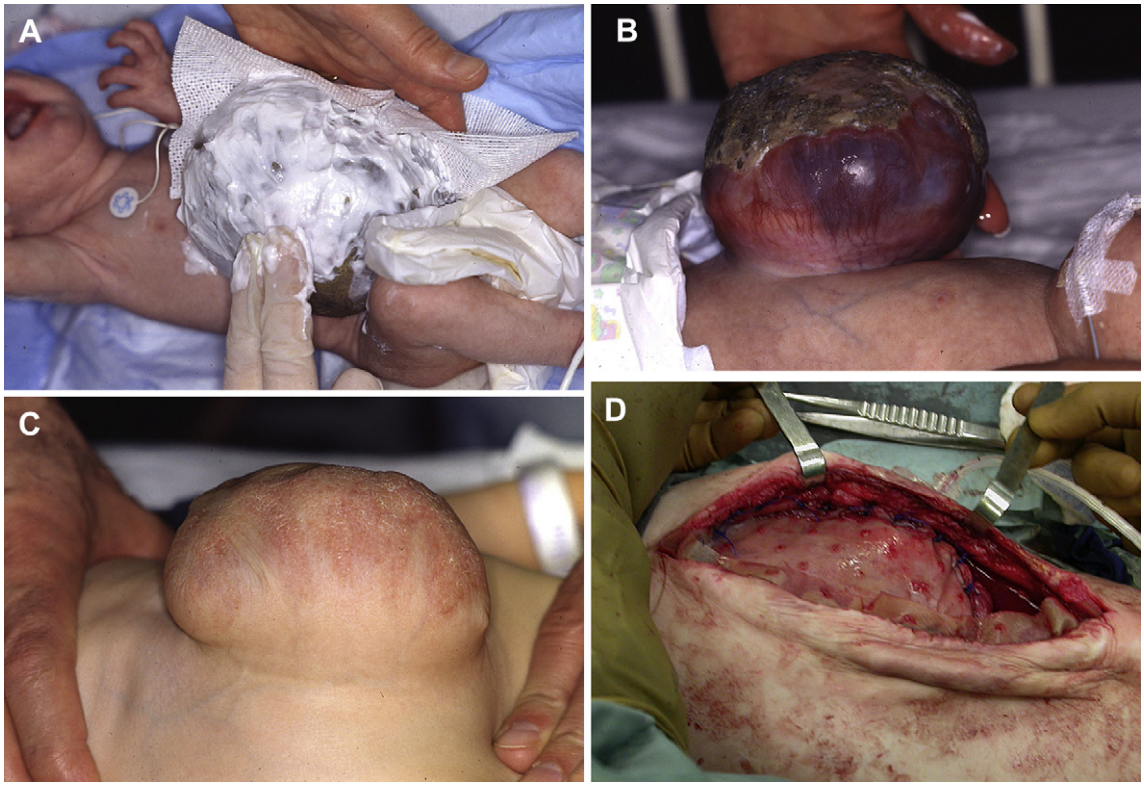

Fig. 4. Escharotic treatment for a giant omphalocele. (A) Initial application of silver sulfadiazine. (B) Mature eschar with partial epithelialization. (C) Complete epithelialization. (D) Ventral hernia repair using porcine intestinal submucosa as a patch in the superior aspect of the defect. This repair should only be done once the child is medically optimized.

born with significant bowel injury are more likely to require prolonged parenteral nutrition with the associated risks of total parenteral nutrition-related cholestatic liver disease and central line-related sepsis. These complications lead to a 20-fold increased risk of death when compared with a patient without associated bowel injury.[50] However, new strategies for the management of parenteral nutrition-associated liver disease and short bowel syndrome (ω-3-rich lipid formulas, serial transverse enteroplasty) may begin to mitigate some of the historical morbidity associated with short bowel.[79]

Cryptorchidism is associated with gastroschisis with an incidence of 15–30%, although it is unclear whether this is secondary to in utero extra-abdominal entrapment of the testis or simply a byproduct of prematurity.[80] Several retrospective analyses have shown that replacement of the herniated testis into the abdominal cavity will result in normal testicular descent into the scrotum in the majority of cases.[81,82] For those in whom the testis does not descend by six months to one year of age, orchidopexy should be performed.

The majority of patients with gastroschisis will achieve normal growth and development after an initial catch-up period in early childhood.[83] If the umbilicus is sacrificed during the repair of the gastroschisis defect, up to 60% of children report psychosocial stress from the lack of an umbilicus.[84] Umbilical reconstruction can be undertaken when the child is healthy, if it is desired by the child or parents.

5.2. Omphalocele

Most infants with a small omphalocele recover well and do not have any long term issues, provided that there are no significant structural or chromosomal abnormalities. This may represent as few as 10% of all omphaloceles diagnosed on prenatal ultrasound.[16] A recent study from The Netherlands found that only 14% of all omphaloceles were isolated.[85] A number of long term medical problems occur in patients with large omphaloceles. These include gastroesophageal reflux, pulmonary insufficiency, recurrent lung infections or asthma, and feeding difficulty with failure to thrive, reported in up to 60% of infants with a giant omphalocele.[86,87] Many of these children may initially require gastrostomy feeding, although some studies report that these difficulties seem to resolve by childhood, with height and weight measurements becoming similar to those of their peer group.[88] The management of severe gastroesophageal reflux can be difficult, especially in the child who is being managed with escharotic therapy in whom a fundoplication may be technically challenging if not impossible. These children may be better served by the use of a naso- or gastro-jejunal tube. Up to a third of patients with omphalocele report intermittent abdominal pain persisting into young adulthood.[15]

The respiratory insufficiency associated with giant omphaloceles may be secondary to abnormal thoracic development with a narrow thorax and small lung area leading to pulmonary hypoplasia. However, a study looking at the long term cardiopulmonary consequences of large abdominal wall defects documented normal lung volumes and oxygen consumption on long-term follow-up, although exercise tolerance was slightly reduced.[89]

The most pervasive concern among patients with omphalocele in long-term follow-up is cosmetic, with nearly one-half of patients expressing dissatisfaction with the lack of an umbilicus and a large abdominal wall scar. Predictably, these complaints were expressed by those with giant omphaloceles at birth to a greater extent than those with minor omphaloceles. Nonetheless, this did not affect the overall quality of life in a recent study.[15] This finding should be borne in mind when counseling parents with isolated omphaloceles – that the long term burden of disease is quite minimal compared to that during early infancy.

Practice points

- Prenatal diagnosis of omphalocele and gastroschisis may influence timing, mode and location of delivery.
- Neonatal resuscitation should focus on temperature stability, fluid resuscitation, placement of a nasogastric tube, and careful physical examination looking for associated anomalies and the condition of the herniated viscera.
- The goal of surgical management is to close the abdominal wall without injury to the viscera either directly or due to increased intra-abdominal pressure. The choice between primary and staged closure should be based on this balance.
- Measurement of intra-abdominal pressure can be helpful in guiding the decision to use primary versus staged closure.
- The main cause of morbidity and mortality in infants with omphalocele is associated anomalies, and the main cause in infants with gastroschisis is the degree of bowel injury.

Research directions

- Prediction of intestinal damage in fetuses with gastroschisis.
- Determination of the role of cesarean section and/or preterm delivery in fetuses with gastroschisis.
- Prediction of pulmonary hypoplasia in fetuses with omphalocele.
- Selection of best closure technique for infants with omphalocele and gastroschisis based on neonatal factors.
- Development of pharmacological therapy for ameliorating hypomotility in children with gastroschisis.

Conflict of interest statement

None declared.

Funding sources

None.

References

1. Baird PA, MacDonald EC. An epidemiologic study of congenital malformations of the anterior abdominal wall in more than half a million consecutive live births. *Am J Hum Genet* 1981;**33**:470–8.
2. Barisic I, Clementi M, Husler M, et al. Evaluation of prenatal ultrasound diagnosis of fetal abdominal wall defects by 19 European registries. *Ultrasound Obstet Gynecol* 2001;**18**:309–16.
3. Rasmussen SA, Frias JL. Non-genetic risk factors for gastroschisis. *Am J Med Genet* 2008;**148C**:199–212.
4. Castilla EE, Mastroiacovo P, Orioli IM. Gastroschisis: international epidemiology and public health perspective. *Am J Med Genet* 2008;**148C**:162–79.
5. Loane M, Dolk H, Bradbury I. Increasing prevalence of gastroschisis in Europe 1980–2002: a phenomenon restricted to younger mothers? *Paediatr Perinat Epidemiol* 2007;**21**:363–9.
6. Alvarez SM, Burd RS. Increasing prevalence of gastroschisis repairs in the United States: 1996–2003. *J Pediatr Surg* 2007;**42**:943–6.
7. Lausman AY, Langer JC, Tai M, et al. Gastroschisis: what is the average gestational age of spontaneous delivery? *J Pediatr Surg* 2007;**42**:1816–21.
8. Kronfli R, Bradnock TJ, Sabharwal A. Intestinal atresia in association with gastroschisis: a 26 year review. *J Pediatr Surg* 2010;**26**:891–4.
9. Arnold MA, Chang DC, Nabaweesi R, et al. Risk stratification of 4344 patients with gastroschisis into simple and complex categories. *J Pediatr Surg* 2007;**42**:1520–5.
10. Russo R, D'Armiento M, Angrisani P, et al. Limb body wall complex: a critical review and a nosological proposal. *Am J Med Genet* 1993;**47**:893–900.
11. Craven CM, Carey JC, Ward K. Umbilical cord agenesis in limb body wall defect. *Am J Med Genet* 1997;**71**:97–105.
12. Blazer S, Zimmer EZ, Gover A, et al. Fetal omphalocele detected early in pregnancy: associated anomalies and outcomes. *Radiology* 2004;**232**:191–5.
13. Kamata SN, Usui N, Sawai T, Nose K, Fukuzawa M. Prenatal detection of pulmonary hypoplasia in giant omphalocele. *Pediatr Surg Int* 2008;**24**:107–11.
14. Brantberg A, Blaas HG, Haugen SE, et al. Characteristics and outcome of 90 cases of fetal omphalocele. *Ultrasound Obstet Gynecol* 2005;**26**:527–37.
15. van Eijck FC, Hoggeveen YL, van Weel C, Rieu PNMA, Wijnen RMH. Minor and giant omphalocele: long-term outcomes and quality of life. *J Pediatr Surg* 2009;**44**:1355–9.
16. Heider AL, Strauss RA, Kuller JA. Omphalocele: clinical outcomes in cases with normal karyotypes. *Am J Obstet Gynecol* 2004;**190**:135–41.
17. Davis NM, Kurpios NA, Sun X, Gros J, Martin JF, Tabin CJ. The chirality of gut rotation derives from left–right asymmetric changes in the architecture of the dorsal mesentery. *Dev Cell* 2008;**15**:134–45.
18. Thieme G. Developmental malformations of the fetal ventral body wall. *Ultrasound Q* 1992;**10**:225–65.
19. deVries PA. The pathogenesis of gastroschisis and omphalocele. *J Pediatr Surg* 1980;**15**:245–51.
20. Hoyme HE, Higginbottom MC, Jones KL. The vascular pathogenesis of gastroschisis: intrauterine interruption of the omphalomesenteric artery. *J Pediatr* 1981;**98**:228–31.
21. Louw JH, Barnard CN. Congenital intestinal atresia; observations on its origin. *Lancet* 1955;**269**:1065–7.
22. Werler MM, Sheehan JE, Mitchell AA. Association of vasoconstrictive exposures with risks of gastroschisis and small intestinal atresia. *Epidemiology* 2003;**14**:349–54.
23. Feldkamp ML, Carey JC, Sadler TW. Development of gastroschisis: review of hypotheses, a novel hypothesis, and implications for research. *Am J Med Genet A* 2007;**143**:639–52.
24. Werler MM, Mitchell AA, Moore CA, Honein MA. Is there epidemiologic evidence to support vascular disruption as a pathogenesis of gastroschisis? *Am J Med Genet* 2009;**149A**:1399–406.
25. Stevenson RE, Rogers RC, Chandler JC, et al. Escape of the yolk sac: a hypothesis to explain the embryogenesis of gastroschisis. *Clin Genet* 2009;**75**:326–33.
26. Shaw A. The myth of gastroschisis. *J Pediatr Surg* 1975;**10**:235–44.
27. Langer JC, Longaker MT, Crombleholme TM, et al. Etiology of intestinal damage in gastroschisis. I: effects of amniotic fluid exposure and bowel constriction in a fetal lamb model. *J Pediatr Surg* 1989;**24**:992–7.
28. Langer JC, Bell JG, Castillo RO, et al. Etiology of intestinal damage in gastroschisis. II. Timing and reversibility of histologic changes, mucosal function, and contractility. *J Pediatr Surg* 1990;**25**:1122–6.
29. Wales PW, Christison-Lagay ER. Short bowel syndrome: epidemiology and etiology. *Semin Pediatr Surg* 2010;**19**:3–9.
30. Salihu HM, Emusu D, Aliyu ZY, et al. Mode of delivery and neonatal survival of infants with isolated gastroschisis. *Obstet Gynecol* 2004;**104**:678–83.
31. Segel SY, Marder SJ, Parry S, et al. Fetal abdominal wall defects and mode of delivery: a systematic review. *Obstet Gynecol* 2001;**98**:867–73.
32. Srinathan SK, Langer JC, Blennerhassett MG, et al. Etiology of intestinal damage in gastroschisis: III. Morphometric analysis of the smooth muscle and submucosa. *J Pediatr Surg* 1995;**30**:379–83.
33. Guibourdenche J, Berrebi D, Vuillard E, et al. Biochemical investigations of bowel inflammation in gastroschisis. *Pediatr Res* 2006;**60**:565–8.
34. Vargun R, Aktug T, Heper A, et al. Effects of intrauterine treatment on interstitial cells of Cajal in gastroschisis. *J Pediatr Surg* 2007;**42**:783–7.
35. Luton D, de Lagausie P, Guibourdenche J, et al. Effect of amnioinfusion on the outcome of prenatally diagnosed gastroschisis. *Fetal Diagn Ther* 1999;**14**:152–5.
36. Charlesworth P, Njere I, Allotey J, et al. Postnatal outcome in gastroschisis: effect of birth weight and gestational age. *J Pediatr Surg* 2007;**42**:815–8.
37. Piper HG, Jaksic T. The impact of prenatal bowel dilation on clinical outcomes in neonates with gastroschisis. *J Pediatr Surg* 2006;**41**:897–900.
38. Langer JC, Khanna J, Caco C, et al. Prenatal diagnosis of gastroschisis: development of objective sonographic criteria for predicting outcome. *Obstet Gynecol* 1993;**81**:53–6.
39. Nasr A, Langer JC. Influence of location of delivery on outcome in neonates with gastroschisis. APSA 42nd Annual Meeting, Palm Desert, CA, USA, 22–25 May 2011 (abstract).
40. Coughlin JP, Drucker DE, Jewell MR, et al. Delivery room repair of gastroschisis. *Surgery* 1993;**114**:822–6.
41. Bianchi A, Dickson AP, Alizai NK. Elective delayed midgut reduction: no anesthesia for gastroschisis: selection and conversion criteria. *J Pediatr Surg* 2002;**37**:1334–6.
42. Skarsgard ED, Claydon J, Bouchard S, et al. Canadian Pediatric Surgical Network: a population-based pediatric surgery network and database for analyzing surgical birth defects. The first 100 cases of gastrochisis. *J Pediatr Surg* 2008;**43**:30–4.
43. Owen A, Marven S, Jackson L, et al. Experience of bedside preformed silo staged reduction and closure for gastroschisis. *J Pediatr Surg* 2006;**41**:1830–5.
44. Allotey J, Davenport M, Njere I, et al. Benefit of preformed silos in the management of gastroschisis. *Pediatr Surg Int* 2007;**23**:1065–9.
45. Minkes RK, Langer JC, Mazziotti MV, et al. Routine insertion of a silastic spring-loaded silo for infants with gastroschisis. *J Pediatr Surg* 2000;**35**:843–6.
46. Pastor A, Phillips J, Fenton S, Meyers R, et al. Routine use of a Silastic spring-loaded silo for infants with gastroschisis: a multicenter randomized controlled trial. *J Pediatr Surg* 2008;**43**:1807–12.

47. Banyard D, Ramones T, Phillips SE, Leys CM, Rauth T, Yang EY. Method to our madness: an 18-yr retrospective analysis on gastroschisis closure. *J Pediatr Surg* 2010;**45**:579–84.
48. Jenson AR, Waldhausen JHT, Kim S. The use of a spring-loaded silo for gastroschisis: impact on practice patterns and outcomes. *Arch Surg* 2009;**144**:1–4.
49. Sandler A, Lawrence J, Meehan J, et al. A "plastic" sutureless abdominal wall closure in gastroschisis. *J Pediatr Surg* 2004;**39**:738–41.
50. Bonnard A, Zamakhshary M, de Silva N, et al. Nonoperative management of gastroschisis: a case-matched study. *Pediatr Surg Int* 2008;**24**:767–71.
51. Riboh J, Abrajano CT, Garber K, et al. Outcomes of sutureless gastroschisis closure. *J Pediatr Surg* 2009;**44**:1947–51.
52. Zivkovic SM. Repair of gastroschisis using umbilical cord as a patch. *J Pediatr Surg* 1991;**26**:1179–80.
53. Lacey SR, Bruce J, Brooks SP, et al. The relative merits of various methods of indirect measurement of intraabdominal pressure as a guide to closure of abdominal wall defects. *J Pediatr Surg* 1987;**22**:1207–11.
54. Yaster M, Scherer TL, Stone MM, et al. Prediction of successful primary closure of congenital abdominal wall defects using intraoperative measurements. *J Pediatr Surg* 1989;**24**:1217–20.
55. Fleet MS, de la Hunt MN. Intestinal atresia with gastroschisis: a selective approach to management. *J Pediatr Surg* 2000;**35**:1323–5.
56. Curry JI, Lander AD, Stringer MD. A multicenter, randomized, double-blind, placebo-controlled trial of the prokinetic agent erythromycin in the postoperative recovery of infants with gastroschisis. *J Pediatr Surg* 2004;**39**:565–9.
57. Lander A, Redkar R, Nicholls G, et al. Cisapride reduces neonatal postoperative ileus: randomised placebo controlled trial. *Arch Dis Child Fetal Neonatal Ed* 1997;**77**:F119–22.
58. Jayanthi S, Seymour P, Puntis JW, et al. Necrotizing enterocolitis after gastroschisis repair: a preventable complication? *J Pediatr Surg* 1998;**33**:705–7.
59. Oldham KT, Coran AG, Drongowski RA, et al. The development of necrotizing enterocolitis following repair of gastroschisis: a surprisingly high incidence. *J Pediatr Surg* 1988;**23**:945–9.
60. Molik KA, Gingalewski CA, West KW, et al. Gastroschisis: a plea for risk categorization. *J Pediatr Surg* 2001;**36**:51–5.
61. Abdullah F, Arnold MA, Nabaweesi R, et al. Gastroschisis in the United States, 1988–2003: analysis and risk categorization of 4344 patients. *J Perinatol* 2007;**27**:50–5.
62. Arnold MA, Chang DC, Nabaweesi R, et al. Development and validation of a risk stratification index to predict death in gastroschisis. *J Pediatr Surg* 2007;**42**:950–5.
63. Vacharajani AJ, Rao R, Keswani S, Mathur AM. Outcomes of exomphalos: an institutional experience. *Pediatr Surg Int* 2009;**25**:139–44.
64. Nasr A, McNamara PJ, Mertens L, et al. Is routine preoperative 2-dimenstional echocardiography necessary for infants with esophageal atresia, omphalocele, or anorectal malformations? *J Pediatr Surg* 2010;**45**:876–9.
65. Dunn JC, Fonkalsrud EW. Improved survival of infants with omphalocele. *Am J Surg* 1997;**173**:284–7.
66. Zama M, Gallo S, Santecchia L, et al. Early reconstruction of the abdominal wall in giant omphalocele. *Br J Plast Surg* 2004;**57**:749–53.
67. Zaccara A, Zama M, Trucchi A, et al. Bipedicled skin flaps for reconstruction of the abdominal wall in newborn omphalocele. *J Pediatr Surg* 2003;**38**:613–5.
68. van Eijck FC, de Blaauw I, Bleichrodt RP, et al. Closure of giant omphaloceles by the abdominal wall component separation technique in infants. *J Pediatr Surg* 2008;**43**:246–50.
69. Tenenbaum MJ, Foglia RP, Becker DB, et al. Treatment of giant omphalocele with intraabdominal tissue expansion. *Plast Reconstr Surg* 2007;**120**:1564–7.
70. Gabriel A, Gollin G. Management of complicated gastroschisis with porcine small intestinal submucosa and negative pressure wound therapy. *J Pediatr Surg* 2006;**41**:1836–40.
71. Rahn S, Bahr M, Schalaman J, Saxena AK. Single center experience in the management of anterior abdominal wall defects. *Hernia* 2008;**12**:345–50.
72. Maksoud-Filho JG, Tannuri U, daSilva MM, Maksoud JG. The outcome of newborns with abdominal wall defects according to the method of abdominal closure: the experience of a single center. *Pediatr Surg Int* 2006;**22**:503–7.
73. Schuster SR. A new method for the staged repair of large omphaloceles. *Surg Gynecol Obstet* 1967;**125**:837–50.
74. Hong AR, Sigalet DL, Guttman FM, et al. Sequential sac ligation for giant omphalocele. *J Pediatr Surg* 1994;**29**:413–5.
75. Yeh TF, Pildes RS, Firor HV. Mercury poisoning from mercurochrome therapy of an infected omphalocele. *Clin Toxicol* 1978;**13**:463–7.
76. Whitehouse JS, Gourlay DM, Masonbrink AR, et al. Conservative management of giant omphalocele with topical povidone-iodine and its effect on thyroid function. *J Pediatr Surg* 2010;**45**:1192–7.
77. Foglia R, Kane A, Becker D, et al. Management of giant omphalocele with rapid creation of abdominal domain. *J Pediatr Surg* 2006;**41**:704–9.
78. Cusick E, Spicer RD, Beck JM. Small-bowel continuity: a crucial factor in determining survival in gastroschisis. *Pediatr Surg Int* 1997;**12**:34–7.
79. Le HD, Fallon EM, deMeijer VE, et al. Innovative parenteral and enteral nutrition for intestinal failure. *Semin Pediatr Surg* 2010;**19**:27–34.
80. Berger AP, Hager J. Management of neonates with large abdominal wall defects and undescended testis. *Urology* 2006;**68**:175–8.
81. Lawson A, de la Hunt MN. Gastroschisis and undescended testis. *J Pediatr Surg* 2001;**36**:366–7.
82. Chowdhary SK, Lander AD, Buick RG, et al. The primary management of testicular maldescent in gastroschisis. *Pediatr Surg Int* 2001;**17**:359–60.
83. Henrich K, Huemmer HP, Reingruber B, et al. Gastroschisis and omphalocele: treatments and long-term outcomes. *Pediatr Surg Int* 2008;**24**:167–73.
84. Davies BW, Stringer MD. The survivors of gastroschisis. *Arch Dis Child* 1997;**77**:158–60.
85. Cohen-Overbeek TE, Tong WH, Hatzmann TR, et al. Omphalocele: comparison of outcome following prenatal or postnatal diagnosis. *Ultrasound Obstet Gynecol* 2010;**36**:687–92.
86. Biard JM, Wilson RD, Johnson MP, et al. Prenatally diagnosed giant omphaloceles: short- and long-term outcomes. *Prenat Diagn* 2004;**24**:434–9.
87. Koivusalo A, Rintala R, Lindahl H. Gastroesophageal reflux in children with a congenital abdominal wall defect. *J Pediatr Surg* 1999;**34**:1127–9.
88. Berseth CL, Malachowski N, Cohn RB, et al. Longitudinal growth and late morbidity of survivors of gastroschisis and omphalocele. *J Pediatr Gastroenterol Nutr* 1982;**1**:375–9.
89. Zaccara A, Iacobelli BD, Calzolari A, et al. Cardiopulmonary performances in young children and adolescents born with large abdominal wall defects. *J Pediatr Surg* 2003;**38**:478–81.