Chirurgia (2013) 108: 509-515
No. 4, July - August

Considerations on Gastroschisis Repair

S. Ionescu[1], B. Andrei[1], S. Tirlea[1], B.M. Bunea[1], E. Licsandru[1], C. Cirstoveanu[2], M. Bizubac[2], M. Ivanov[1], M. Shelleh[1], A. Gurita[3], R. Tabacaru[3]

[1]Department of Pediatric Surgery, "Marie S. Curie" Emergency Children Hospital, Bucharest, Romania
[2]Newborn Intensive Care Unit, "Marie S. Curie" Emergency Children Hospital, Bucharest, Romania
[3]Anesthesiology Unit, "Marie S. Curie" Emergency Children Hospital, Bucharest, Romania

Rezumat

Considerații asupra tratamentului gastroschizisului

Introducere: Deşi închiderea primară a gastroschizisului este posibilă în multe cazuri, au fost descrise diferite strategii şi materiale pentru a acoperi viscerele abdominale, pentru protecția viscerelor şi reducerea pierderii de căldură şi lichide, în cazul gastroschizisurilor ireductibile primar. Au existat sugestii de acoperire ale defectului cu alografe de piele, dura mater liofilizată, transplant liber de corium şi autografe de piele-mesh (1,2).
Scopul studiului: prezentăm o metodă alternativă de reparare a gastroschizisului, în cazul în care există o disproporție între mărimea viscerelor eviscerate şi cea a cavității abdominale hipoplazice. În cazul închiderii primare a peretelui abdominal, diferența de volum poate determina o creştere importantă a presiunii intraabdominale
Metodă: În unele cazuri, atunci când închiderea primară completă nu este posibilă, am folosit o metodă alternativă de reparare a defectului parietal folosind patch de cordon ombilical.
Rezultate: prin această tehnică se creează o suprafață mezotelială în contact cu intestinele. Urmărirea acestor pacienți la distanță a demonstrat rezultate excelente.
Concluzii: Această tehnică este uşor de aplicat şi foarte utilă în cazul nou-nascuților cu gastroschizis când închiderea primară a defectului nu este posibilă. Utilizarea de material autolog, cum este cordonul ombilical are mai multe avantaje printre care: disponibilitatea, o rată mai scăzută a infecțiilor şi costuri semnificativ reduse.

Cuvinte cheie: Gastroschizis, tratament, cordon ombilical

Abstract

Background: Although primary closure of the gastroschisis is possible in many cases, there have been various strategies published and materials used to cover the eviscerated bowel when the abdominal wall defect cannot be closed in one step, providing bowel protection and reduction of heat and fluid loss. There have been suggestions of coverage materials such as skin graft, lyophilized dura mater graft, free flap corium and meshed skin graft (1,2).
Purpose: We highlight an alternative repair method of gastroschisis in those cases where there is a disproportion between the amount of eviscerated organs and the hypoplastic abdominal cavity. If in this case primary closure of the abdominal wall is chosen, the difference in volume can cause a significant increase in intraabdominal pressure.
Method: In some cases, when complete primary closure was not possible, we used an alternative method to repair the parietal defect using umbilical cord patch.
Results: This technique creates a mesothelial surface in contact with the bowel. Remote tracking of these patients showed excellent results.

Corresponding author: Sebastian Ionescu, MD
Department of Pediatric Surgery
"Marie S. Curie" Emergency Children Hospital
62 Constantin Brancoveanu Blvd., sector 4
Bucharest – 041451, Romania
Tel/fax: 021/4601040
E-mail: nsionescu@yahoo.com

This paper represents an original article, all the authors had the same contribution.

Conclusions: This technique is easy to apply and very useful for infants with gastroschisis especially when primary closure is not possible. The use of autologous material, in this case as the umbilical cord, has several advantages, including wide availability, a lower rate of infection and significantly reduced costs.

Key words:gastroschisis, surgical treatment, umbilical cord patch

Introduction

Laparoschisis was first described, in 1940, by Barnstein, under the name of gastroschisis, the name coming from Greek language and meaning abdominal cleft. The incidence of gastroschisis is assessed at 1/4000-6000 births and has a very severe prognosis (3). (*Fig. 1*)

The abdominal cavity is underdeveloped. The wall defect is always on the right side of the umbilicus, with a diameter of 3-4 cm; the umbilical cord is normally inserted and complete. The small bowel and the colon, which has not undergone the normal rotation, from the duodenum all the way down to the rectum, is exposed and not covered by peritoneal sac. It floats freely in the amniotic fluid which has an irritating action on the intestinal wall, causing an inflammatory reaction (4). The intestinal wall is thickened, swollen, dark red, changes that are secondary to ischemia and to the contact with the amniotic fluid; there are numerous adhesions and the intestine is covered by a pseudo membrane containing collagen (5). This produces shorter intestinal length, which creates significant functional disturbances. The mesentery is short, thickened and there is a risk of strangulation when passing through the parietal defect ring. There is a disorder of the bowel peristalsis and significant malabsorption (6). (*Fig. 2*)

The most widely accepted pathogenic theory is that of intrauterine vascular occlusion of the right umbilical mesenteric artery with infarction, rupture of the umbilical ring and intestinal evisceration. This theory explains the location of the defect on the right, in most cases, and sometimes the association with intestinal atresia (7).

The diagnosis of the parietal defect must be recognized early in the intrauterine life, by performing foetal morphology ultrasound, starting in the second trimester of pregnancy and by an abnormal high level of alpha-fetoprotein in the maternal blood (level much higher than for omphalocele) (8). It is unacceptable to leave the diagnosis of this condition to chance, given the complications, difficulties in treatment and the vital and functional unfavourable prognosis.

The ultrasound parameters to be followed are: abdominal defect size, presence or absence of intestinal peristalsis, the presence of superior mesenteric artery Doppler flow, foetal biometry, amniotic fluid quantity and quality (hydramnios may be a sign of intestinal obstruction). The hypotrophy and foetal distress, the look of the intestinal loops (over 3 mm thick wall, hiperechoic) and the quality of the mesenteric vascularization are important factors that could indicate the necessity of preterm birth induction for improved prognosis. (*Fig. 3*)

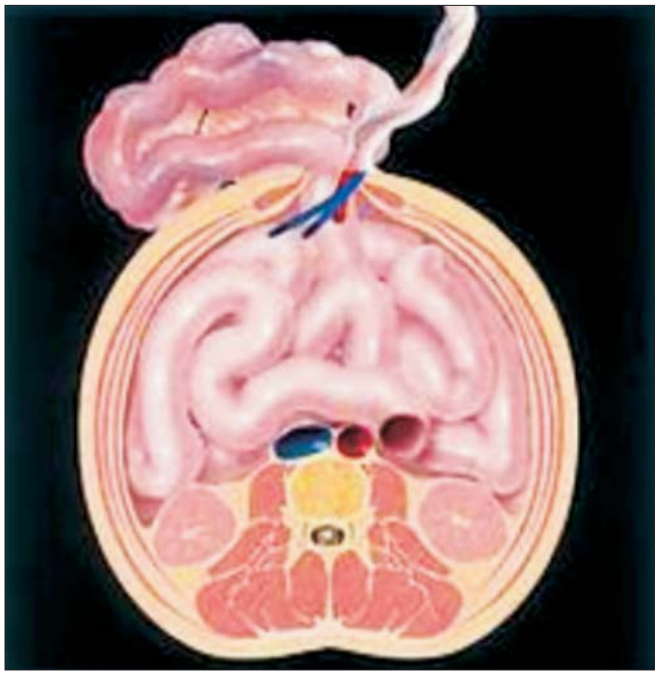


Figure 1. *Cross-section of a foetal abdomen with gastroschisis*

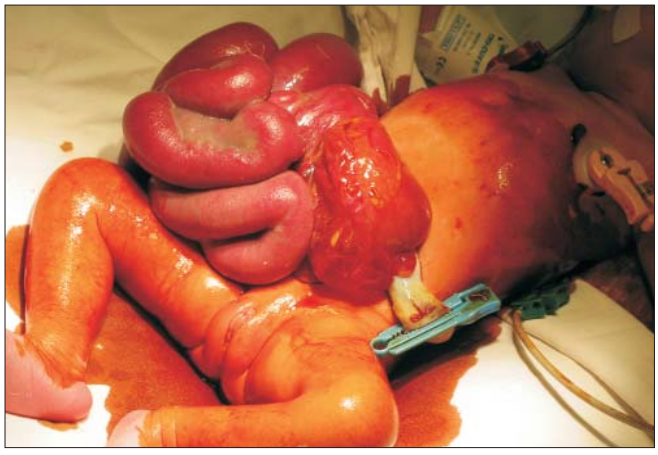


Figure 2. *Gastroschisis*

Sometimes it can be associated with moderate hypotrophy and foetal distress, which can be aggravated by severe oligohydramnios. The appearance of the bowel loops and quality of the mesenteric vascularization are important prognostic factors: the diameter of the herniated bowel loops over 17 mm and thickness of the bowel wall of more than 3 mm, with hyperechoic aspect is a sign of intestinal distress.

The size of the parietal defect rarely exceeds 4-5 cm. Not very often, associated with eviscerated intestines, the hernia pouch can contain the liver (5 cases), gall bladder (3 cases), stomach (5 cases), adnexa in girls (3 cases) or ectopic testis in boys (1 case).

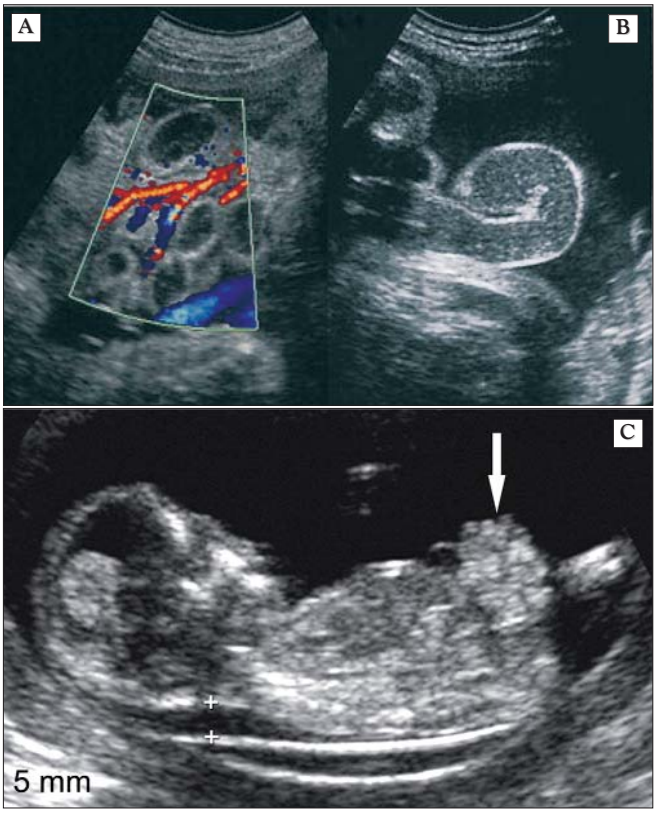


Figure 3. *2D ultrasound in a patient with gastroschisis: Doppler flow is observed in the mesenteric artery (**A**) and the intestinal loops floating in the amniotic fluid (**B**) foetus with gastroschisis - 13 weeks old (**C**)*

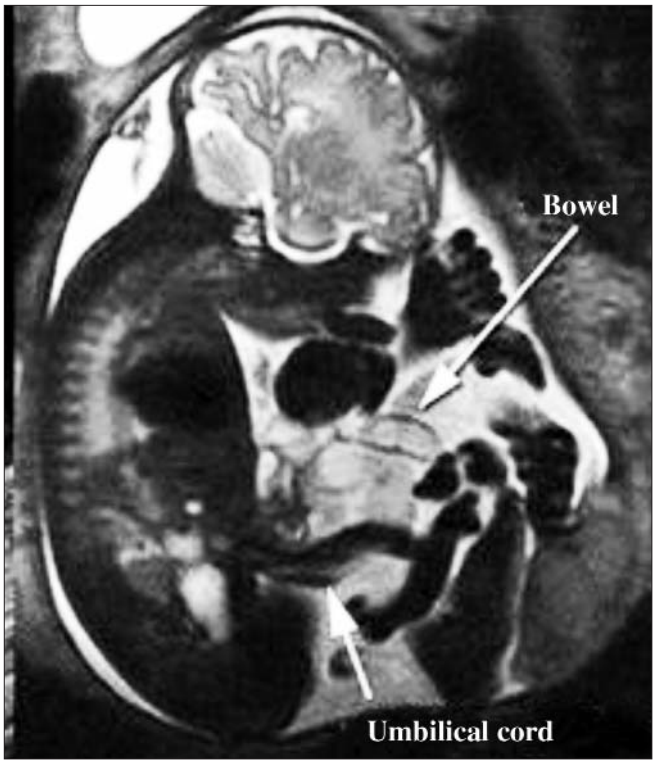


Figure 4. *Foetal MRI – normal insertion of the umbilicus and intestinal loop floating in amniotic fluid*

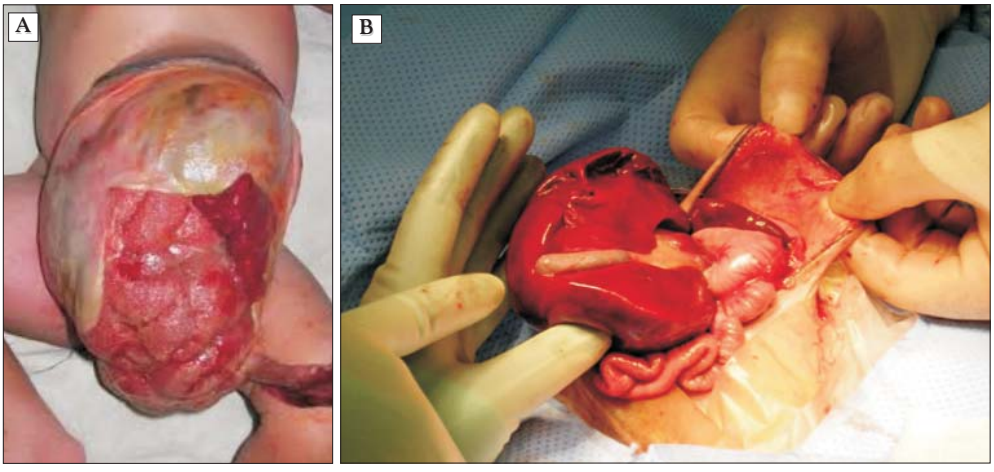
For a more accurate diagnosis a foetal MRI can be done. (*Fig. 4*)

The differential diagnosis must be done with a ruptured omphalocele, but usually there are sufficient elements for the diagnosis of the gastroschisis: examination of the viscera, the paraumbilical defect, the normal aspect of the umbilicus (4). (*Fig. 5*)

Treatment

Probably the most important aspect in the treatment of this condition is the quality of care. Given the possibility of antenatal diagnosis of this condition there have been discussions regarding the preferred method of birth, but there is no proof of positive outcome if cesarean delivery is chosen in order to prevent mechanical injury to the exteriorized intestinal loops compared to natural birth (8). It is very important to assess the possible antenatal complications or

Figure 5. *Ruptured omphalocele*



foetal distress, which would be indications for cesarean section. Preterm birth of these children was proposed in order to limit the bowel lesions through prolonged contact with amniotic fluid, but there have been no proven benefits. It is essential to reduce the time between birth and integration of the viscera into the abdomen.

In gastroschisis, the shock of expulsion is very high; the child is generally low weighted and has important electrolyte loss from the exposed intestinal loops. Sometimes, the small and fibrous parietal wall defect can be a source of strangulation and needs to be enlarged to allow a good blood flow. The intestinal loops can be twisted during labour, and should be uncoiled after birth with as little aggression as possible, not allowing the loss of fluids. Placing the child on the side with the defect seems to improve the vascularization of the bowel, which also needs to be wrapped in sterile gauze soaked with warm physiological saline solution; the baby should be wrapped in a sterile bag up to the chest in order to preserve the humidity and temperature, and after a short preoperative preparation directed to the OR (9).

The therapeutic principles for gastroschisis repair remain the same, visceral reintegration and primary fascial closure of the defect or progressive reduction achieved with a Silastic silo (Silon) (9,10). Using this mesh closure was not a successful method, as previously thought (11). Surgical treatment has evolved greatly from the first therapeutic success of Grob, Gross and Schuster (12). There are still cases when the reduction is considered unsafe, physically impossible due to viscero-abdominal disproportion, or dangerous because of abdominal compartment syndrome. Abdominal compartment syndrome is defined by increased intra-abdominal pressure and its consequences on the cardiovascular system, lungs and kidneys. There are 2 methods for measuring the intra-abdominal pressure: monitoring the intragastric or the intravesical pressure. Both techniques measure the intermittent abdominal pressure indirectly. Reducing of the eviscerated bowel into the abdomen can lead to abdominal compartment syndrome, increased intra-abdominal pressure causing cardio-respiratory compromise/failure with lifting of the diaphragm and abdominal organ hypoperfusion secondary to compression.

Often, if there is no surgical intervention, the condition can be fatal. The pressure in the stomach and bladder with a value of 20 cm H_2O correlates with values of 9 - 9.4 cm H_2O direct intra-abdominal pressure (13).

The use of the umbilical cord as an autologous material in the repair of abdominal wall defects has been known since the early 1970s. Compared with free flaps and synthetic Silo, the human umbilical cord was associated with lower inflammatory reaction, lower incidence of wall dehiscence, faster healing and highest overall survival rate in experimental abdominal wall defects in rats (14,15). The first successful repair in humans was published in 1974, when the umbilical cord was used to cover a 2 cm^2 defect in a patient with gastroschisis. Closure was completed using this material at the top (16). However, the final closure of the primary defect using only the umbilical cord resulted in ventral hernia (17). A Spanish group reported a primary closure technique in two premature newborns with a split umbilical cord and reinforced by polypropylene mesh attached to the fascia and covered with skin (18). Other groups have used a "sutureless" approach after progressive reduction of the eviscerated bowel covering the defect with the umbilical cord and applying a dressing for secondary epithelialization. All of these techniques result in a higher incidence of ventral hernia, but the rate of complications related to intra-abdominal pressure and use of heterogeneous materials is much lower (19,20).

Materials and Method

This study includes 12 infants with gastroschisis. The following parameters were evaluated: preoperative preparation of the newborn, gestational age, birth weight, other associated malformations, the surgical technique used, hemodynamic tolerance after primary closure, parenteral nutrition, pre-and postoperative intraabdominal pressure, mechanical ventilation, postoperative scar healing, length of hospitalization.

We used the umbilical cord patch to close the abdominal wall defect, creating a mesothelial surface in contact with the intestines. This technique is very useful in infants with gastroschisis when primary closure is not possible.

In preparation for this operative technique, obstetricians are usually instructed to keep the cord in its full length. The umbilical cord is cut at its insertion into the placenta and placed in a sterile plastic bag together with the eviscerated bowel. After stabilization of the newborn, it is transferred to the operating room. A nasogastric tube is placed for gastric decompression and an enema with normal saline is ordered preoperatively to facilitate evacuation of the meconium. The abdominal wall and the bowel are prepared with povidone iodine solution. Under general anesthesia the bowel is carefully inspected for intestinal atresia and other possible malformations. Manual milking of the small intestine from proximal to distal direction including the colon is performed in order to decompress the intestine. If the abdominal wall defect is too small for bowel reintegration, one can use cranial or caudal incisions around the defect. This direction of incision extension is important when we use the umbilical cord patch.

Bowel reintegration is performed manually, while monitoring the pressure in the bladder. When the pressure reaches 20 cm water, reintegration must be stopped to prevent abdominal compartment syndrome.

The umbilical cord is cut longitudinally on the right side in all its length. A curved clamp is carefully inserted in the incision and gently separated (*Fig. 1*). The two umbilical arteries and umbilical veins are exposed in full-length; they are ligated with 4-0 silk at the base, and excised. This avascular membrane cord which is being created comes from the left side of the defect. The three sides of the membrane are sutured with 4-0 Vicryl to the edges of the parietal defect, covering it, followed by skin suture and dressing. (*Fig. 6, 7*)

The newborn is transferred to the neonate intensive care unit. Intravesical pressure should be monitored for another day. Newborn extubation is possible when breathing is effective.

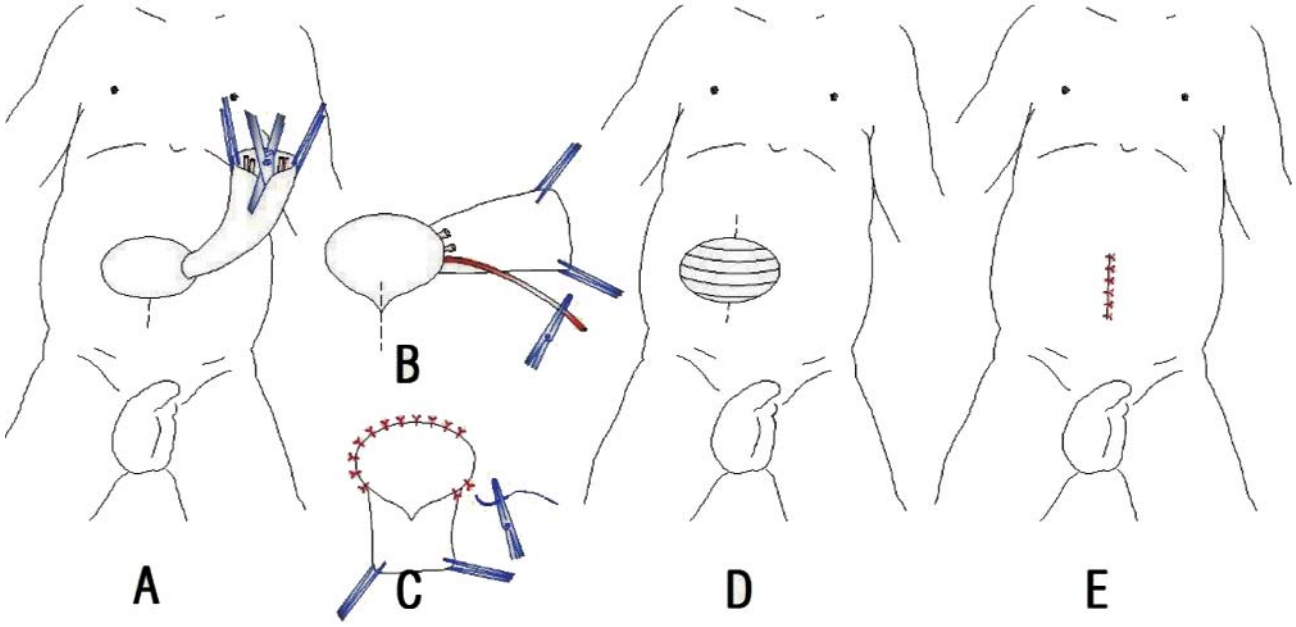


Figure 6. *Surgical technique. After the stretching of the abdominal wall and partial reduction of the viscera in the abdominal cavity (**A**) the umbilical cord is cut on the midline and opened like a book, this way providing a sufficient area to cover the defect. The dotted line on the abdominal wall marks the line for further enlargement of the initial wall defect. (**B**) the umbilical vessels are ligated and excised. (**C**) The free edges of the created membrane are sutured to the wall defect*

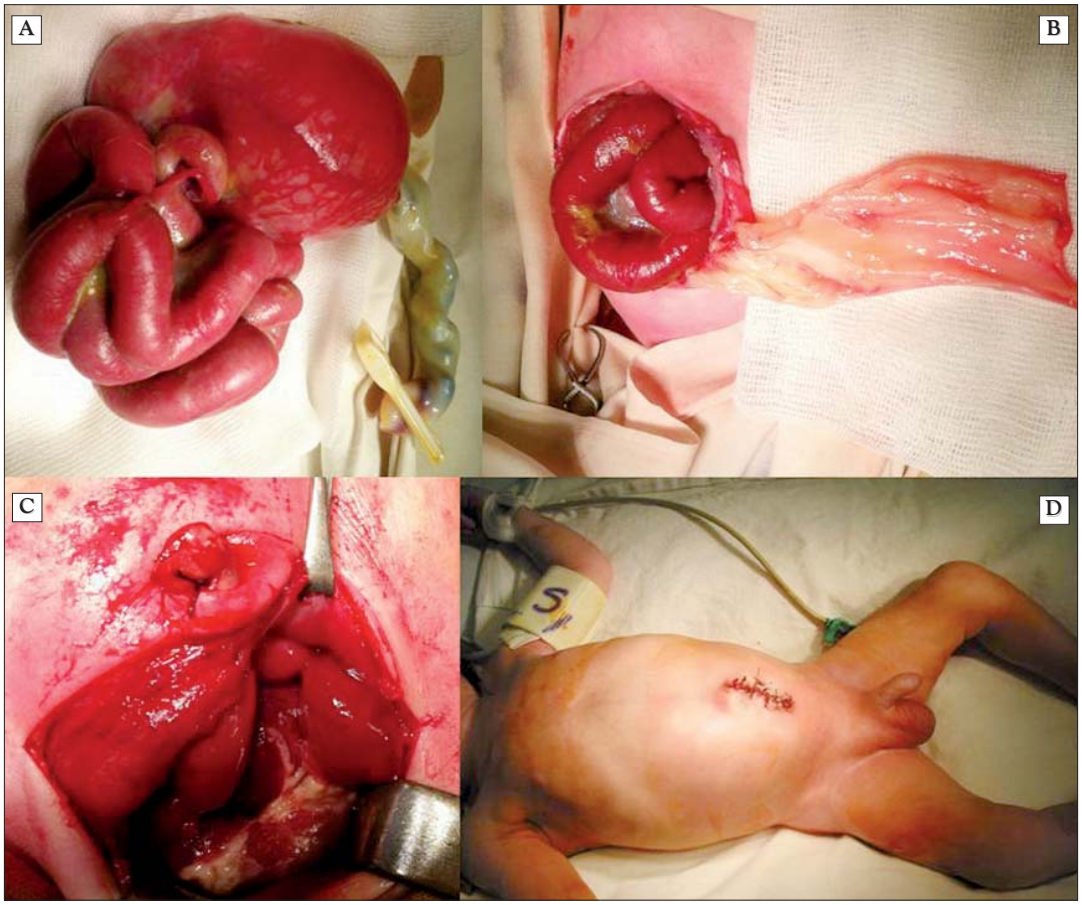


Figure 7. *Intraoperative aspects of the gastroschisis*

After the surgery the nasogastric tube is maintained with total parenteral nutrition, via peripheral or central veins. The initiation of enteral nutrition is encouraged to decrease the gastric reflux and to stimulate the bowel transit. Child's position is irrelevant.

Usually after 6 to 8 weeks, patients were discharged from the hospital with a small ventral hernia.

Results

Between 2008 and 2011 we had 12 cases of gastroschisis (4 girls and 8 boys). The gestational age of these children ranged from 33 to 38 weeks. Birth weight ranged between 1500 g and 3300 g. Only 3 of 12 patients with gastroschisis were diagnosed by antenatal ultrasound, the rest were unscreened pregnancies. There was one case with associated malformations, namely agenesis of the ascending and descending colon.

In 7 cases, we tried to use primary abdominal wall closure when the abdominal cavity volume allowed reintegration of the intestinal loops without increasing the abdominal pressure or ventilatory pressure. The intra-abdominal pressure was monitored intravesically or intragastric not exceeding 20 cm H_2O to prevent abdominal compartment syndrome.

It was necessary to increase the size of the abdominal wall defect for the reintegration of the protruding bowel in 3 cases.

In 5 cases out of 12, primary abdominal closure was not possible (important intestinal edema in 3 cases, small abdominal cavity in 2 cases) so we decided to close the defect using the following methods:

a) umbilical cord patch at the aponeurotic defect in 2 cases;
b) the progressive closure method: over several days, using a Silastic silo bag attached to the margins of the aponeurosis in 3 cases. This "silo" was closed progressively from top to bottom, thus favoring gradual reintegration of the viscera in the abdomen while intestinal edema regressed. Median time to complete closure of the wall was 7 days. The duration of parenteral nutrition ranged from 7 to 39 days, especially in those with low gestational age and low birth weight (1500g - 1700g) and with extensive intestinal edema. Duration of mechanical ventilation ranged from 3 days to 13 days as a result of increased pressure limiting diaphragm excursion during respiration. Average length of stay was 6.6 weeks (range: 23 -135 days).

There were no cases of infection due to umbilical cord patch or the postoperative wound dehiscence. There were 2 patients who died.

The first death was a male newborn with a gestational age of 39-40 weeks and birth weight of 2700 g. The 3D ultrasound performed at 21 weeks of age was apparently normal. The repair was performed using a synthetic mesh. On postoperative day 2, alloplastic prosthetic defect reduction is performed, splitting the defect from the right edge and suturing the aponeurotic edge of the defect and the suture aponevrotic edge to reduce the size of the mesh. After 3 days the defect is finally closed with the suture of the aponeurosis defect by joining the edges and keeping a portion of 0.5 mm width of the mesh. Skin is sutured on the midline. After 8 days we explored the suture and the next day the patient died from cardiac arrest, septic shock with multiple organ failure from a Candida infection, respiratory shock, mechanical ventilation.

The second death: female neonate with a gestational age of 38 weeks and birth weight of 2300 g, natural birth, resuscitated at birth. We performed gastroschisis repair using a synthetic mesh. The evolution of the patient is severe with wound bleeding, abdomen distension, cyanosis of the skin, generalized edema, heavy bleeding from puncture sites, TGP 4839 U / L, ALT 7700 U / L. The patient died due to cardiac arrest, respiratory distress, sepsis, interstitial pneumonia.

Discussion

Primary closure results are much better compared to procedures using Silo bag, free flaps, lyophilized dura mater and other techniques for coverage of the wall defect and intestine (2). Primary fascial closure is possible in 90% of infants and the mortality rate is about 10% (21).

A tight closure during primary fascia repair can increase the intra-abdominal pressure and lift the diaphragm increasing the need for mechanical respiratory support. On the other hand, we must also take into consideration the morbidity associated with mechanical ventilation (22). In some patients, due to the direct pressure on the newly reintegrated bowel and the diminished blood flow from the superior mesenteric artery to the intestinal wall vessels, perforation or diffuse necrosis of the bowel and even death can occur (2,23,24). Another intra-operative problem that can occur with primary closure of the wall defect is inferior vena cava obstruction/compression with severe cyanosis in the legs. In some of these patients early surgical reintervention might be necessary with the use of the Silo technique (12). Morbidity associated with Silo use is caused by sepsis, enteric fistula, prolonged ileus, and the need for more surgeries. The Silo bag can often lead to an infection along the suture line. It cannot create any symphysis between the Silo and patient tissues (23).

Our repair method of the gastroschisis is a combination of procedures used in primary closure and patch technique. Almost the entire exposed bowel is reduced into the abdominal cavity, except for a few loops, which will be covered with an umbilical cord patch. We used this technique only in neonates with gastroschisis in whom the defect could not be closed primarily. By this we tried to avoid compromising the respiratory status, venous return and bowel ischemia (24). To prevent abdominal compartment syndrome, one should monitor the intravesical pressure, during the operation and at least one more day after (24,25,26). The maximum allowed value is 27 cm H_2O.

The umbilical cord patch is an autologous tissue. The umbilical cord patch and the edges of the abdominal wall defect, subcutaneous tissue and the skin heal completely within a few days when the defect was extended laterally. We had no cases of surgical site infection. On the other hand, this natural patch prevented the formation of adhesions between the intestine and the umbilical cord patch. It forms a ventral hernia, which is usually small and easy to fix.

Conclusions

Whenever it is possible to preserve the umbilical cord we recommend this technique because of its availability, easy technique and perfect tolerance of the autologous tissue. Infection rate is low and mechanical complications are uncommon. There is only one disadvantage: the umbilical cord patch should be prepared in the operating room. Primary closure of the umbilical cord patch was applied successfully in all neonates with gastroschisis, the postoperative intravesical pressure not exceeding the mentioned values. We intend to collect more cases to verify the benefits of this procedure and to provide statistical power.

References

1. Rahn S, Bahr M, Schalamon J, Saxena AK. Single-center 10-year experience in the management of anterior abdominal wall defects. Hernia. 2008;12(4):345-50.
2. Henrich K, Huemmer HP, Reingruber B, Weber PG. Gastroschisis and omphalocele: treatments and long-term outcomes. Pediatr Surg Int. 2008;24(2):167-73. Epub 2007 Nov 6.
3. Lewis Spitz, Arnold G. Coran. Operative Pediatric Surgery, 6th edition. Hodder Arnold 2006.
4. Zamfir T, Basca I, Jianu M, Puiu S, Balanescu R, Patrancus T, Ulici A. Chirurgie viscerala, urologie si ortopedie pediatrica. Bucuresti: Ed. Stiintifica; 1996.
5. Nicolae Angelescu, Florian Popa, Sebastian Ionescu. Caiete de tehnici chirurgicale - vol 3. Bucuresti: Ed Medicala; 2011.
6. Radu Balanescu, Sebastian Ionescu. Principii de diagnostic si tratament in chirurgia si ortopedia pediatrica. Bucuresti: Ed. Universitara Carol Davila; 2011.
7. Ashcraft K. W, George W. Holcomb III, J. Patrick Murphy. Pediatric Surgery, 5th Edition. Philadelphia: Saunders-Elsevier; 2009.
8. Arnold G. Coran MD, N. Scott Adzick MD, Thomas M. Krummel MD, Jean-Martin Laberge, Robert Shamberger, Anthony Caldamone MD. Pediatric Surgery - 7th edition. Philadelphia: Mosby Elsevier; 2012.
9. Snyder CW, Biggio JR, Brinson P, Barnes LA, Bartle DT, Georgeson KE, et al. Effects of multidisciplinary prenatal care and delivery mode on gastroschisis outcomes. J Pediatr Surg. 2011;46(1):86-9.
10. Sabetay C, Jona JZ, Maloş A, Stoica A, Zavate A, Ciobanu O, et al. The gastroschisis: clinical and therapeutic considerations. Chirurgia (Bucur). 2006;101(2):147-50. Romanian
11. Lansdale N, Hill R, Gull-Zamir S, Drewett M, Parkinson E, Davenport M, et al. Staged reduction of gastroschisis using preformed silos: practicalities and problems. J Pediatr Surg. 2009;44(11):2126-9.
12. McNamara WF, Hartin CW, Escobar MA, Lee YH. Outcome differences between gastroschisis repair methods. J Surg Res. 2011;165(1):19-24.
13. Engum SA, Kogon B, Jensen E, Isch J, Balanoff C, Grosfeld JL. Gastric tonometry and direct intraabdominal pressure monitoring in abdominal compartment syndrome. J Pediatr Surg. 2002;37(2):214-8.
14. Tănase M, Marin S, Amza S, Toader A. Chirurgia (Bucur). 2012;107(2):195-8. The use of allografts in the surgical treatment of enlarged hiatal hernias. Romanian
15. Heaton FC, Thomas CG Jr, Owen J. The use of umbilical cord for reconstruction of abdominal wall defects. Surg Forum. 1970;21:56-7.
16. Samii AM, Jafroudi Y. Gastroschisis: use of umbilical cord in its repair. J Med Liban. 1974;27(4):473-7.
17. Zivković SM. Repair of gastroschisis using umbilical cord as a patch. J Pediatr Surg. 1991;26(10):1179-80.
18. Hernández Siverio N, M López-Tomassetti Fernández E, Mario Troyano Luque J. Gastroschisis: primary closure using umbilical cord strengthened by a polypropylene mesh. J Perinat Med. 2007;35(3):249-51.
19. Riboh J, Abrajano CT, Garber K, Hartman G, Butler MA, Albanese CT, et al. Outcomes of sutureless gastroschisis closure. J Pediatr Surg. 2009;44(10):1947-51.
20. Werbeck R, Koltai J. Umbilical cord as temporary coverage in gastroschisis. Eur J Pediatr Surg. 2011;21(5):292-5.
21. Wesson DE, Baesl TJ. Repair of gastroschisis with preservation of the umbilicus. J Pediatr Surg. 1986;21(9):764-5.
22. Christison-Lagay ER, Kelleher CM, Langer JC. Neonatal abdominal wall defects. Semin Fetal Neonatal Med. 2011; 16(3):164-72.
23. Henrich K, Huemmer HP, Reingruber B, Weber PG. Gastroschisis and omphalocele: treatments and long-term outcomes. Pediatr Surg Int. 2008;24(2):167-73.
24. Mirza B, Ijaz L, Saleem M, Sheikh A. Postnatal intestinal ischemia in a patient with gastroschisis: a sinister problem of home delivery and improper transportation. J Pediatr Surg. 2010;45(11):2289-90.
25. Santos Schmidt AF, Goncalves A, Bustorff-Silva JM, Oliveira-Filho AG, Miranda ML, Oliveira ER, et al. Monitoring intravesical pressure during gastroschisis closure. Does it help to decide between delayed primary or staged closure? J Matern Fetal Neonatal Med. 2012;25(8):1438-41.
26. Olesevich M, Alexander F, Khan M, Cotman K. Gastroschisis revisited: role of intraoperative measurement of abdominal pressure. J Pediatr Surg. 2005;40(5):789-92.