Outcome differences between simple and complex gastroschisis

Ann. Ital. Chir., 2016 87: 525-530
pii: S0003469X16025926
free reading: www.annitalchir.com

Vesna Milojković Marinović*, Maija Lukač**, Željko Miković***, Blagoje Grujić*, Maja Miličković°, Gordana Samardžija°°, Aleksandra Stojanović*, Dalibor Sabbagh*

** Department of Neonatal Surgery, Institute for mother and child healthcare, Belgrade, Serbia*
*** Department of Neonatal Surgery, University children's hospital, Belgrade, Serbia*
****Obstetric/Gynecology Clinic Narodni Front, University of Belgrade, Belgrade, Serbia*
° Department of Abdominal Surgery, Institute for mother and child healthcare, Belgrade, Serbia
°° Department of Pathology, Institute for mother and child healthcare, Belgrade, Serbia

Outcome differences between simple and complex gastroschisis

BACKGROUND: *We reviewed differences of the outcome of newborn with simple and complex gastroschisis treated at our institution over the past fifteen years.*
METHODS: *A retrospective cohort study was performed on all infants with gastroschisis treated at the Institute for mother and child health care, Belgrade, between 2001 and 2015 (n=70). Premature infants (<34 weeks of gestation) and babies with birth weight less than 1500 g were excluded (n=5). We compared outcomes in infants with simple gastroschisis and those with complex gastroschisis. An outcome analysis was performed for birth weight, gestational age, gender, mode of defect closure, presence of intestinal necrosis or perforation, reoperation, duration of mechanical ventilation and total parenteral nutrition, presence of bowel pseudoopstraction, sepsis, total duration of hospital stay, mortality rates.*
RESULTS: *Of 65 patients, 15 (23,07%) had complex gastroschisis, including atresia 5 patients (33,3%), ischemic complication 9 patients (60%) and one patient with closing gastroschisis (6,66%). Sixty eight percent underwent primary closure.There was difference between the simple and the complex gastroschisis in duration of mechanical ventilation (P= ,000003), total parenteral nutrition (P= ,000019), bowel pseudoobstruction (P= ,00067), reoperation (P= ,00122), sepsis (P= ,0043), hospital stay (P= ,000198). In the simple gastroschisis group 92% of patients survived to discharge. In the complex gastroschisis group 3 patients died in hospital (P= ,338).*
CONCLUSIONS: *More research should be focused to predict complex gastroschisis and to improve prenatal diagnosis and postnatal management, without a significant increase in morbidity and mortality.*

KEY WORDS: Complex, Gastroschisis, Outcomes, Simple, Vanishing

Introduction

Gastroschisis is a congenital abdominal wall defect with protrusion of abdominal organs into the amniotic cavity. The defect is typically located to the right of the umbilicus. Reported mortality is less than 10% in most published series [1-4]. Molik et al. [2] proposed categorizing infants born with gastroschisis into simple and complex cases. Complex gastroschisis was defined as gastroschisis associated with at least one of the following intestinal pathologies: intestinal atresia, perforation, necrotic segments or volvulus. Simple gastroschisis was defined as gastroschisis without any of these additional pathologies. With the development of modern neonatal intensive care and pediatric surgery, the complex gastroschisis are associated with extensive intestinal loss, short bowel syndrome, total parenteral nutrition, liver failure, sepsis and early baby death. The optimal management of this abdominal wall defect remains controversial. Primary clo-

Pervenuto in Redazione Giugno 2016. Accettato per la pubblicazione Settembre 2016.
Correspondence to: Vesna Milojković Marinović Dimitrija Tucović 30, Belgrade, Serbia (e-mail: vesnamilojkovic@yahoo.com)

sure or staged closure using customized silo has been the gold standard for operative approaches [5,6]. At our institution the prefered method of gastroschisis treatment is primary surgical repair, when it is feasible. Last ten years we have moved toward staged reduction of the herniated intestines into the abdominal cavity using a silo in the absence of intestinal anomalies and with complete closure on an elective basis. We reviewed differences of the outcome of newborn with simple and complex gastroschisis treated at our institution over the past fifteen years and tried to identify factors associated with mortality.

Material and methods

A retrospective cohort study was performed on all infants with gastroschisis treated at the Institute for mother and child health care, Belgrade between 2001 and 2015 (n=70). Premature infants (<34 weeks of gestation) and babies with birth weight less of 1500g were excluded (n=5). Infant birth history, demographic and clinical information were obtained from patient records as well as from the neonatal intensive care unit database. Perinatal data included birth age, gestational age, sex, birth weight. Surgical records included presence of intestinal atresia, necrosis, perforation, strictures and method of closure. Outcome data included duration of mechanical ventilation, duration of total parenteral nutrition, pseudoobstruction, sepsis (central line infection), reoperations, length of hospital stay, mortality rates. Cases of complex gastroschisis were defined as infants with gastroschisis and one or more of following anomalies: intestinal atresia, intestinal perforation, ischemic bowel, strictures. Total length of hospital stay was defined as the period of time from the date of first admission to the date of first discharge or transfer to another hospital. Surgical closure method were categorized into 2 distinct eras: pre spring loaded silo (pre SLS, 2001 to 2006) patients undergo primary closure and require only a single operation and spring loaded silo (SLS, 2007 to present) patients undergo staged repair with spring loaded silo and are mandated to have a minimum of two operations.

Data analyses

We compared outcomes in infants with simple gastroschisis (defined as intact continuous bowel that is not compromised at delivery or presentation) and those with complex gastroschisis (defined as the presens of 1 or

Fig. 1: Closing gastroschisis with jejunal and colonic atresia and complete midgut necrosis; abdominal wall defect located to the left side of the umbilicus.

Fig. 2: Closing gastroschisis with proximal and distal atresias at the entry and exit points of the extraabdominal bowel mass, resulting in midgut loss with jejunal and colonic remnants. Abdominal wall defect located to the left side of the umbilicus.

more of the following criteria: intestinal atresia, perforation or intestinal necrosis, strictures at delivery or presentation) using nonparametric methods. An outcome analysis was performed for birth weight, gestational age, sex, mode of the closure of the defect, presence of intestinal necrosis or perforation, pseudoobstruction, reoperation, duration of mechanical ventilation and total parenteral nutrition. Outcome data included presence of sepsis, total length of hospital stay, mortality rates. Data were analyzed using χ^2 test and Mann Whitney U test. P values < .05 were considered significant. All analyses were carried out using SPSS version 12.

Results

There were 70 patients identified during the study period, but only 65 patients were analyzed after excluding the premature infants (<34 weeks of gestation) and babies with birth weight less of 1500 g (n=5). Fifteen patients (23.07%) met the definition of complex gastroschisis. Patient characteristics are summarized in Table I.

Of the 65 patients, 39 were male and 26 were female. There was a preponderance of males in both groups, 54% in simple gastroschisis and 80% in complex gastroschisis group. Primary closure defined as primary fascial repair was performed in 44 (67.69%) patients. Delayed fascial closure using silastic silo was used in 21 (32.30%) patients. We indentified 15 cases of complicated gastroschisis and all of these children were closed with primary fascial repair. Type of complication in complex gastroschisis group are summarized in Table II.

The overall incidence of intestinal atresia was 7,69% (n=5) in our patient population. In 5 patients (33,3%) of the complex gastroschisis group atresia was represented. Ischemic complications such as stenosis, strictures, necrosis and perforation were the main complication in 9 cases (60%). Closing gastroschisis was presented in one case (6,66%) with circumferential closure of the ring around the protruding bowel associated with midgut necrosis.

There was no difference between the simple and the complex gastroschisis groups in gestational age (36.1 ± 1.4 vs 36.16 ± 1.6; P= 0.173) and birth weights (2248.4 ± 507.6 vs 2351.33 ± 633.8; P= 0.319).

These reports demonstrated significant differences in method of closure. However, the cases were quite heterogeneous. In complex gastroschisis group the finding always dictated the method of closure, and all of this patients (n=15) were closed primarily. In the simple gastroschisis groupe primary fascial closure was performed in 29 patients (58%).

Routine silastic bag closure was performed in 21 patients (42%) with simple gastroschisis, and 19 patients (90,47%) of them were with no complications.

In the simple gastroschisis group 4 patients (8%) died in hospital. Two patients with silastic bag treatment had developed bowel gangrena while in silo and two patients with primary fascial repair had abdominal compartment syndrome. Persistent metabolic acidosis, sepsis, poor perfusion, low urine output and respiratory compromitation required performed repeated laparotomies.

In the complex gastroschisis group 3 patients (20%) died. Closing gastroschisis was present in one patient (6.66%

TABLE I - *Patient characteristics of simple and complex gastroschisis groups*

N Gender	Simple n (%); Mean ± SD (n=50)	Complex n (%); Mean ± SD (n=15)	P
Male Female	27 (54%) 23 (46%)	12 (80%) 3 (20%)	P= .071
Gestational age (wk)	36,1 ± 1,4	36.16 ± 1.6	P= .173
Birth weight (g)	2248,4 ± 507,6	2351.33 ± 633.8	P= .319
Primary closure Performed spring-loaded silo	29 (58%) 21 (42%)	15 (100%) 0	P= .0032
TPN duration (d)	13,64 ± 10,8	53.1 ± 42.6	P= .000019 P<0.001
Ventilator duration (d)	7 ± 6,54	24 ± 14.2	P= .000003 P<0.001
Hospital stay (d)	32 ± 15	91 ± 64	P= .000198 P<0.001
Sepsis (n)	19 (38%)	12 (80%)	P= .0043
Reoparation (n)	10 (20%)	10 (66.7%)	P= .00122
Pseudoobstruction (n)	9 (18%)	12 (80%)	P= .00067 P<0.001
Neonatal death (n)	4 (8%)	3 (20%)	P= .338

TABLE II - *Type of complications in complex gastroschisis group*

Complication	n	% of complex gastroschisis	% of all gastroschisis
Closed gastroschisis with midgut necrosis	1	6,66	1.53
Jejunal atresia	1	6.66	1.53
Ileal atresia	2	13.33	3.07
Colonic atresia	2	13.33	3.07
Gaster perforation with jejunal stenosis	1	6.66	1.53
Ileal ischemia	2	13.33	3.07
Ileal and colonic ischemia	2	13.33	3.07
Jejunal and ileal ischemia	1	6.66	1.53
Jejunal and colonic strictures	1	6.66	1.53
Ileal and colonic strictures	1	6.66	1.53
Jejunal stenosis	1	6.66	1.53

of complex gastroschisis and 1,53% of all gastroschisis) with jejunal atresia and complete midgut necrosis of entire extraabdominal bowel mass. We found proximal and distal atresias at the entry and exit points of the extraabdominal bowel mass, resulting in midgut loss with jejunal and colonic remnants. The patient was a boy born in 34th gestational week with abdominal wall defect located to the left side of the umbilicus. Baby boy birth weight was 1900 g, with proximal jejunal atresia 20 cm distal to the ligament of Treitz, and a midgut volvulus progressing to necrosis and liquefaction during the first day of life (Fig. 1, 2). Midgut resection, jejunocolic anastomosis and abdominal fascial closure were performed. The intervention was successful. He died at 20 days of life due to sepsis and multiorgan failure.

Patients with simple gastroschisis were put on enteral feeding earlier than patients with complex gastroschisis and received less parenteral nutrition: 13.64 ± 10.8 versus 53.1 ± 42.6 days; P= .000019 (P<0.001). Also they received less ventilation support duration: 7± 6.54 versus 24 ± 14.2 days; P= .000003 (P<0.001). Patients with simple gastroschisis had a shorter hospital stay: 32 ± 15 versus 91 ± 64 days; P= .000198 (P<0.001).

A total of 21 (32.3%) of the 65 patients developed additional complications. The most common gastroinestinal complications were feeding problems and bowel pseudoobstruction: 9 (18%) in simple gastroschisis group versus 12 (80%) in complex gastroschisis group (P= .00067). Reoperation was needed in 10 (20%) patients of the simple gastroschisis group and also in 10 (66.7%) patients of the complex gastroschisis group (P= .00122).

The most frequent extraintestinal complication was sepsis in 19 (38%) patients of the simple gastroschisis group and in 12 (80%) patients of the complex gastroschisis group (P= .0043).

In the simple gastroschisis group, 4 (8%) patients died in hospital, resulting in 92% survival to discharge. In the complex gastroschisis group 3 (20%) patients died in hospital resulting in 80% survival to discharge (P= ,338).

Discussion

The incidence of gastroschisis has significantly increased over the past two decades [4,5]. The incidence of gastroschisis is as high as 4.4 per 10000 live births [4]. Reviews in large national databases in Great Britain and the United States have shown complex gastroschisis to represent 11,5% and 10,9% of all cases respectively [7,8]. The prevalence of complex gastroschisis in recent publications has been reported as 11% to 31% [2,4,8,9].

We divided cases into simple and complex gastroschisis according to the presence of additional bowel damage such as atresia, perforation, necrosis, as these factors are known to impact on choice of surgical technique as well as outcomes. In our study 77% of infants had simple gastroschisis.

A number of surgical options exist to repair gastroschisis. The goals of surgical intervention in gastroschisis are to minimize the evaporative and thermal loss [10], reduce the bowel back into the abdominal cavity [11], and repair the abdominal wall defect [12]. Primary closure or staged closure using a silastic bag have been the gold standard operative approaches [5,6]. In our cohort study almost 1 in 4 patients had complex gastroschisis. The presence of simple versus complex gastroschisis is the only factor that has been consistently shown to predict poorer outcomes of this anomaly, including duration of total parenteral nutrition, ventilation duration and hospital stay [13,14].

Neonates with gastroschisis have delayed time for full enteral feeding (FEF), possibly due to bowel exposure to amniotic fluid. Antenatal bowel dilatation (bowel diameter ≥ 18 mm) and in particular intraabdominal bowel dilataion (IABD) is prognostically useful for detection of patients with impending necrosis or atresia and other patients who would have bad outcome [15]. In one study [16] 19% complex patients never had IABD and in the other (17) 75% never had extraabdominal bowel dilatation (EABD). Therefore, the absence of bowel dilatation cannot fully exclude complex patients with gastroschisis [18].

Yang CY et al. investigated IABD alone and found an association with prolonged time to full enteral feeding

(FEF) and length of hospital stay (LHS) [15]. These results are not supported by study data of Helen Carnaghan et al. [16] or a systematic review of isolated gastroschisis which shows that neither IABD nor EABD are associated with increased adverse neonatal outcomes [19].
However, the presence of both IABD/EABD or IABD and collapsed extra-abdominal bowel at <30 weeks of gestation proved to be a more accurate predictor of poor outcome. It may be prudent in the presence of such findings to consider early delivery with the aim to salvage necrotic bowel, although these antenatal findings may indicate that the bowel damage has already occurred. Early delivery is associated with prolonged FEF/LHS, suggesting that elective delivery at < 37 weeks is not beneficial. Combined IABD/EABD or IABD/collapsed extra-abdominal bowel is suggestive of complex gastroschisis [17].
The only factor that has been shown to predict poorer outcomes of gastroschisis is the presence of complex gastroschisis. In some studies intestinal complications also predicted higher mortality, as high as 28% [2,8]. In others, as in our study, mortality did not differ significantly between simple and complex gastroschisis [8,9].
Surgical efforts therefore may be best targeted at optimal management of conditions found in complex gastroschisis including atresia, stenosis, necrosis, perforation, short bowel syndrome. We find primary anastomosis as a safe option. Early restoration of bowel continuity and primary fascial closure are associated with much better outcome. Others have reported the safety of both early and late primary anastomosis for atresia associated with gastroschisis [20].
Closing gastroschisis presents with a variety of possible sequelae. The incidence of closing gastroschisis is 6% in series of Houben at al. [21] A review of the more recent literature indentified small numbers of survivors of closing gastroschisis [22-25]. Persistent or progressive intraabdominal bowel dilatation at fetal ultrasound points to closing abdominal ring complication. If there is suspicion of a closing ring, then early delivery must be urgently considered [24].
By experience, surgeons may identify which patients have fascial defects more amenable to primary closure [26]. Our study also favored primary closure with a significant reduction in total parenteral nutrition, days with ventilation support and length of hospital stay. Patients with complicated gastroschisis have prolonged hospital stays regardless of closure method, as the length of stay in complicated gastroschisis is often based on the patients intestinal pathology.

Conclusion

The only factor that has been shown to predict poorer outcomes of gastroschisis is the presence of complex gastroschisis. Surgical efforts therefore may be best targeted at optimal management of conditions found in complex gastroschisis including atresia, stenosis, necrosis, perforation, short bowel syndrome. We find primary anastomosis as a safe option. Early restoration of bowel continuity and primary fascial closure are associated with much better outcome. More research should be focused to predict complex gastroschisis and to improve prenatal diagnosis and postnatal management.

Riassunto

Abbiamo confrontato le differenze di risultato nei neonati con gastroschisi semplice e complessa trattati presso il nostro istituto nel corso degli ultimi quindici anni. Lo studio è stato effettuato retrospettivamente su tutti i neonati con gastroschisi trattati presso l'Istituto per la cura della Salute della Madre e del Bambino di Belgrado tra il 2001 e il 2015, pari a 70 neonati.
I prematuri, nati con meno 34 settimane di gestazione e i 5 bambini con peso alla nascita inferiore a 1500 g sono stati esclusi. Il confronto è stato fatto tra gli esiti nei neonati con gastroschisi semplice e quelli con gastroschisi complessa. L'analisi dell'esito è stata considerata in rapporto al peso alla nascita, all'età gestazionale, al sesso, alla modalità di chiusura del difetto, alla presenza di necrosi intestinale o di perforazione, al reintervento, alla durata della ventilazione meccanica e la nutrizione parenterale totale, alla presenza di pseudo ostruzione intestinale, alla sepsi, alla durata complessiva della degenza ospedaliera ed ai tassi di mortalità.
Dei 65 pazienti, 15 (23,07%) erano affetti da una gastroschisi complessa, di cui 5 per atresia (33,3%), 9 con complicanze ischemiche (60%) e un paziente con gastroschisi occlusiva (6,66%).
Il 68% è stato trattato con chiusura primaria della gastroschisi. Vi sono state differenze di durata della ventilazione meccanica tra gastroschisi semplice e complessa (P<0,001) e della nutrizione parenterale totale (P<0,001); nell'evenienza di pseudo ostruzione intestinale (P = 0,00067); nei reinterventi (P = 0,00122), nella sepsi (p= 0,0043), nella durata della degenza ospedaliera (P<0,001). Nel gruppo della gastroschisi semplice il 92% dei pazienti è sopravvissuto fino a poter essere dimessi. Nel gruppo delle gastroschisi complesse 3 pazienti sono deceduti in ospedale (p = 0,338).
In conclusione la ricerca dovrebbe essere concentrata nella previsione della gastroschisi complessa, per il miglioramento della diagnosi prenatale e della gestione postnatale, per una riduzione della morbilità e della mortalità.

References

1. Holand AJA, Walker K, Badawi N: *Gastroschisis: An update*. Pediatr Surg Int, 2010; 26: 871-78.

2. Molik KA, Gingalewski CA, West KW, et al: *Gastroschisis: A plea for risk categorization.* J Pediatr Surg, 2001; 36: 51-5.

3. Emil SH, Canvasser N, Chen T, et al: *Contemporary 2-year outcomes of complex gastroschisis.* J Pediatr Surg, 2012; 47:1521-528.

4. Owen A, Marven S, Jonson P, et al.: *Gastroschisis: A national cohort study to describe contemporary surgical strategies and outcomes.* J Pediatr Surg, 2010; 45:1808-16.

5. Mayer T, Black R, Matlak M, et al.: *Gastroschisis and omphalocele. An eight-year review.* Ann Surg, 1980; 192:783.

6. McNamara WF, Hartin CW, Escobar MA, et al.: *Outcome differences between gastroschisis repair methods.* J Surg Research, 2011; 165:19-24.

7. Penman DG, Fisher RN, Noblett HR, Soothill PW: *Increase in incidence of gastroschisis in the south west of England in 1995.* Br J Obstet Gynaecol, 1998; 105:328-31.

8. Arnold MA, Chang DC, Nabaweesi R, et al.: *Risk stratification of 4344 patients with gastroschisis into simple and complex categories.* J Pediatr Surg, 2007; 42:1520-525.

9. Eggink BH, Richardson CJ, Malloy MH et al.: *Outcome of gastroschisis: A 20-year case review of infants with gastroschisis born in Galveston, Texas.* J Pediatr Surg, 2006; 41:1103-108.

10. Sandler A, Lawrence J, Meehan J et al.: *A plastic sutureless abdominal wall closure in gastroschisis.* J Pediatr Surg, 2004; 39: 738-41.

11. Riboh J, Abrajano C, Garber K, et al.: *Outcomes of suturekess gastroschisis closure.* J Pediatr Surg, 2009; 44: 1947-51.

12. Weinsheimer R, Yanchar N, Bouchard S, et al.: *Gastroschisis closure-does method really matter?* J Pediatr Surg, 2008; 43:874-78.

13. Payne NR, Pfleghaar K, Assel B, et al.: *Predicting the outcome of newborns with gastroschisis.* J Pediatr Surg, 2009; 44:918-23.

14. Davis RP, Treadwell MC, Drongowski RA, et al.: *Risk stratification in gastroschisis: can prenatal evaluation or early postnatal factors predict outcome?* Pediatr Surg Int, 2009; 25: 319-25.

15. Yang CY, Lien R, Fu RH, et al.: *Prognostic factors and concomitant anomalies in neonatal gastric perforation.* J Pediatr Surg, 2015; 50(8):1278-82.

16. Carnaghan H, Pereira S, James PC, et al.: *Is early delivery beneficial in gastroschisis?* J Peditar Surg, 2014; 49:928-33.

17. Grundi H, Anderson RL, Filly RA, et al.: *Gastroschisis: Prenatal diagnosis and management.* Fetal Ther, 1987; 2:144-47.

18. Haberman S, Burgess T, Klass L, et al.: *Acute bowel perforation in a fetus with gastroschisis.* Ultrasound Obstet Gynecol, 2000; 15:542-44.

19. Tower C, Ong SSC, Ewer AK, et al.: *Prognosis in isolated gastroschisis with bowel dilatation: A systematic review.* Arch Dis Child Fetal Neonatal Ed, 2009; 94:268-74.

20. Kronfli R, Bradnock TJ, Sabharwal A: *Intestinal atresia in association with gastroschisis: A 26-year review.* Pediatr Surg Int, 2010; 26: 891-94.

21. Houben C, Davenport M, Ajayi NA, et al.: *Closing gastroschisis: diagnosis, management, and outcomes.* J Pediatr Surg, 2009; 44: 343-47.

22. Barsoom MJ, Prabulos AM, Rodis JF, et al.: *Vanishing gastroschisis and short bowel syndrome.* Obstet Gynecol, 2000; 96: 818-19.

23. Ashburn DA, Pranikoff T, Turner CS: *Unusual presentations of gastroschisis.* Am Surg, 2002; 68: 724-27.

24. Moir CR, Ramsey PS, Ogburn Pl, et al.: *A prospective trial of elective preterm delivery for fetal gastroschisis.* Am J Perinatol, 2004; 21: 289-94.

25. Kimble RM, Blakclock R, Cass D: *Vanisching gut in infant with gastroschisis.* Pediatr Surg Int, 1999; 15: 483-85.

26. Marinović VM, Lukač M, Miković Ž, et al.: *Gastroschisis with gastric perforation and jejunal stenosis. A rare association of anomalies.* Ann Ital Chir, 2016; 87.