Open Access Original Article

Early Outcome of Patient Born with Gastroschisis and Omphalocele

DOI: https://doi.org/10.32007/jfacmedbagdad.6431901

Salar S. Berdawd* FIBMS
Abdulrahman O. Taha** FIBMS

This work is licensed under a Creative Commons Attribution-Noncommercial 4.0 International License

JFac Med Baghdad
2022; Vol.64, No . 3
Received: March. 2022
Accepted: Sept. 2022
Published: Oct. 2022

Abstract:

Background: Gastroschisis and omphalocele are the most common congenital defects of the anterior abdominal wall which need urgent and prompt intervention post-operative time outcomes of gastroschisis and omphalocele differ between different countries, especially between developed and developing countries.

Objectives: Is to determine the early outcome of patients born with gastroschisis and omomphalocele

Patients and Methods: An analytical study including 30 patients (11 gastroschisis and 19 omphaloceles) were treated in Erbil city during 2015 -2018. Patients with associated bladder and cloacal exstrophies were excluded.

Results: Female to male ratio was 1.3:1, and the mean age at presentation was 11.7 hours. The Mean weight was 2.86 kg. The Majority of patients25 (83.3%) were term. In Nineteen cases (63.3%)were delivered vaginally (9 gastroschisis and 10comphalocele), prenatally diagnosis was done in 9(30%). The Mean size of the defect was 3.7cm, and the content was mixed in 7(23.3%). Primary repair has performed in 18(60%), a silo in 8(26.7%), and 4(13.3%) treated none surgically (all were omphalocele). Operations have been done within 24 hours of arrival in 16(61.5%). Associated anomalies have been found in 11(57.9%) patients with omphaloceles and only 2(18.2%) patients with gastroschisis. The mean period of hospital stays was6 days. Mortality among the gastroschisis patients were 8(72%), but only 4(21.1%) for omphalocele patients.

Conclusion: Gastrochisis and omphalocele constitute a considerable part of neonatal surgical problems and their management is still challenging. Omphalocele cases were more likely to have associated congenital anomalies and gastroschisis, if the associated anomalies were confined to the gastrointestinal tract. There was a high mortality rate, especially in patients with gastroschisis and ruptured omphalocele due to a lack of intensive care units, facilities, and trained personnel to look after such high-risk patients.

Keywords: Post-operative outcome, Gastroschisis, Omphalocele, Abdominal wall defect, Congenital anomaly

Introduction:

Management challenges of infants with gastroschisis and omphalocele begin in the prenatal period and can extend many years into the child's life. Prenatal diagnosis provides the opportunity to influence the outcome through changes in the management of the pregnancy and by education and counseling of the family.1 Gastroschisis, is characterized by an intact umbilical cord and evisceration of the intestine through a defect in the abdominal wall to the right of the umbilical cord, with no membrane covering while omphalocele, is characterized by protrusion of the bowel, liver, and other organs into the umbilical cord and covered by membranes.2 Three major advances led to a marked improvement in survival in babies with gastroschisis: parenteral nutrition, the ability to use a silo when primary closure was not possible, and advances in perinatal care in the intensive care unit (NICU).3

*Corresponding Author: Hawler Medical City salar.sabeh@hmv.edu.kurd
** Raparin Teaching Hospital for Children abd.omer79@gmail.com

The Estimated rate of the incidence of gastroschisis is 1 in 10 000 births and omphalocele is 2.5 in 10 000 in Western countries.4 Although the incidence of omphalocele has remained generally stable over the past 20 years4, reports from Europe, the United States. and Japan suggest that the incidence of gastroschisis has increased as much as 10-fold.5,6 The mortality for patients with omphalocele is increased significantly with chromosomal syndromes or a cardiac defect. Similarly, giant omphaloceles and those associated with pulmonary hypoplasia have a worse outcome.7 The high-risk group of gastroschisis includes infants with intestinal atresia, perforation, or volvulus, whereas the low-risk group had no intestinal anomalies. The high-risk, complex group had increased morbidity and mortality characterized by long periods of mechanical ventilation, long ileus, time to tolerate oral feeding, long stay in the hospital, and an increased complication and mortality rate.3

Patients and methods:

An analytical study includes 30 patients (19 omphaloceles and 11 gastroschises) managed in

Raparin Teaching Hospital for children from January 2015 to December 2018. All cases with associated bladder and cloacal exstrophies had excluded from the study. Most patients had preoperative treatment with a nasogastric tube for gastric decompression kept in the incubator. Intravenous fluid has been given. Vitamin K is administered, and Parenteral antibiotics are given as needed. In cases of gastroschisis and ruptured omphalocele, the eviscerated bowel is kept in a sterile surgical pack soaked with saline, supplementary oxygen is provided according to the need of the patients. Preoperative investigations of abdominal ultrasound, echocardiography, and x-ray were done according to the type of defect and general condition of the patient. No chromosomal study had been performed for any case in this study, and no preoperative ventilation and TPN administration. Postoperative management was different according to the type of the defect and the way of management (primary or staged closure), generally including the followings:

1- Patients were kept on IV fluid, electrolyte supplementation, and parenteral antibiotics.
2- NPO with NG tube until the gastric drainage has reduced and bowel motion started.
3- Urinary catheter for follow-up of urine output.
4- Kept in the incubator with oxygen supplementation.
5- Daily compression of silo and reduction of its contents (in staged reduction).
6- Blood, plasma, and albumin were given when indicated.

No postoperative assisted ventilation was used in this study.

Statistical analysis:

Statistical Package for the Social Sciences (SPSS-version 22) package software program has used for statistical analysis. Descriptive statistics (numbers and percentages) have calculated for all variables, analytical statistics were done to find the relations between variables by using, Chi-square, and fisher exact test. A p-value < 0.05 was considered as significant

Results:

Among 30 patients included 17(56.7%) were females and 13(43.3%) were males.

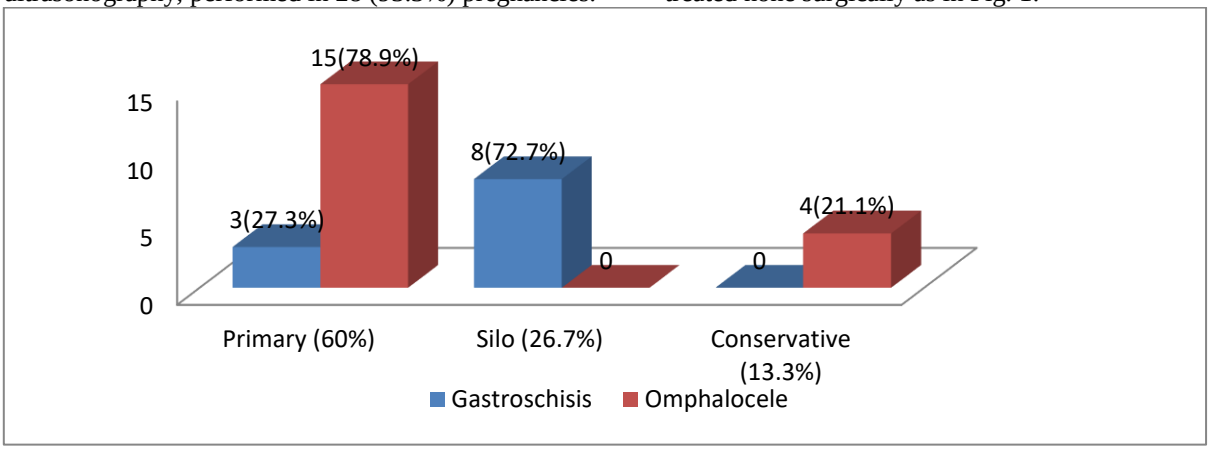
Age at presentation varied between half an hour to 70 hours with a mean age of 11.7 hours and they were classified into two groups, twenty-three (76.7%) of patients entering the first group which covers those presenting immediately within 12 hours of life. As shown in Table 1.

Table 1: age at presentation.

	Immediate (within 12 hours)	Late (after 12 hours)	
Gastroschisis	11 (100%)	0 (0.0%)	
Omphalocele	12 (63.2%)	7 (36.8%)	P =0.025
Total	23 (76.7%)	7 (23.3%)	

The mean weight for the patients was (2.86 kg) ranging from 1.2 – 5.0 kg. Twenty-five (83.3%)patients of them were term babies (≥ 37 weeks of gestation), and 5patients were preterm (16.7%). Out of 30 patients, 19(63.3%) were delivered by vaginal delivery and 11(36.7%) by Caesarian section. Prenatal diagnoses were possible only in 9 (30%)patients using ultrasonography, performed in 28 (93.3%) pregnancies.

The size of the defects ranged from 2 cm to 9 cm with a mean of 3.7 cm. In 27 (90%) cases the size was less than or equal to 5 cm. The content of the defect was loops of the small and large bowel in 23 (76.7%) cases and solid organs (liver) with bowel loops in 7 (23.3%) cases.Primary repair was done in 18 (60%) patients, the silo was performed in 8 (26.7%),and 4 (13.3) have treated none surgically as in Fig. 1:



P= 0.001 **Fig. 1: Distribution of mode of therapy.**

Age at repair ranged from 3.5 hours to 85 hours with a mean of 29 hours and in 16 (53.3%) repair was done within the first 24 hours of life as shown in Table 2 .

Table 2: Age at repair.

	Age at repair		
	Within 24 hr	After 24 hr	
Gastroschisis	11(100%)	0 (00.0%)	P=0.001
Omphalocele	5 (33.3%)	10 (66.7%)	
Total	16 (61.5%)	10 (38.5%)	

Associated anomalies were recorded in 11 (57.9%) omphalocele patients and only in 2(18.2%) patients with gastroschisis (p=0.034). As shown in Table3.

Table3: Incidence of associated anomalies.

Associated anomalies			
Omphalocele 11 (57.9%)		Gastroschisis 2 (18.2%)	
Cardiac	6 (31.5%)	Intestinal atresia	1 (9.1%)
Genitourinary	2 (10.4%)		
Gastrointestinal	2 (10.4%)	Meckel's diverticulum	1 (9.1%)
limb anomalies	2 (10.4%)		

Most of our patients stayed in the hospital for five days or less in 18 (60%) patients with a range between 20 hours to 32 days and a mean period of 6 days.

There were a total of 18 (60%) survival and 12 (40%) deaths, the mortality among gastroschisis patients was 72% but only 21.1% for omphalocele patients (p=0.005). see Table 4:

Table 4: Time and possible causes of death among cases of AAWD.

Type	Cause of death	Age at death
Gastroschisis	Sepsis	13 days
Gastroschisis	Sever prematurity with respiratory insufficiency	21 hours
Gastroschisis	Sepsis	4 days
Gastroschisis	Malnutrition (ileostomy complication)	32 days
Gastroschisis	Intestinal obstruction with sepsis	38 days
Omphalocele	Sepsis with prematurity	6 days
Omphalocele	Associated anomalies	37 days
Omphalocele	Anomalies with sepsis	4 days
Gastroschisis	Sever prematurity with respiratory insufficiency	35 hours
Gastroschisis	Severe respiratory insufficiency	20 hours
Gastroschisis	Severe respiratory insufficiency	45 hours
Omphalocele	Lethal associated anomalies	38 hours

Discussion

The Increasing incidence of abdominal wall defects has been reported to necessitate a review of management approaches to reduce morbidity and mortality.8

The present prospective study is an attempt to study the early outcome of gastroschisis and omphalocele in 30 neonates at Raparin Teaching Hospital for children which is the main pediatric hospital serving Erbil Governorate. The female/male ratio affected by this condition was found to be 1.3:1 by S.Askarpour et al in their study9 and a ratio of 1.1:1 by Abdur-Rahman et al 10 studies which were near to the present study (1.3:1) but Selma Aličelebić et al 11 found more common in males than females with a ratio of 1.5:1. The mean birth weight in the current study was 2860 g (range, 1200-5000 g), the mean birth weight of gastroschisis (2427 g) was lower than that of omphalocele (3115 g), and there is no significant correlation between the type of the defects and, birth weight in our study (p=0.091), Barisic et al12

Study show means the birth weight of 3074 (range, 1250– 4600) g for omphalocele and 2393 (range, 1050–3800) g for gastroschisis with significantly lower birth weight in cases of gastroschisis compared to omphalocele(P < 0.01), they also found the mean gestational age at birth of 38.3 gestational weeks for omphalocele and significantly lower gestational weeks for gastroschisis at 36.3 (P < 0.01). we found gestational age of gastroschisis (mean 37.5) was lower than that of omphalocele (mean 38.4) but without a significant difference between them(p=0.245). Age at presentation differs between gastroschisis and omphalocele in which the former is present earlier than the latter (if not ruptured). Abdur-Rahman et al10 found age at presentation ranging from 2-169 hours (median 23.5 hours). In the present study the age at presentation varied between half an hour to 70 hours (median 3.5 hours) with the majority (76.7%) presenting immediately within a few hours of age. This earlier presentation in the current study returns to the

fact that most of our patients were delivered to the hospital (83.3%) and referred soon to our department.

Abdominal wall defects are diagnosed by prenatal ultrasound done for routine examination or for obstetric indications. Henrich K et al13 studies showed 35/40 children (88%) with gastroschisis and 18/26 children (69%) with omphalocele, had been diagnosed prenatally, Murphy FL et al14 found 28/53 (53%) with gastroschisis and 15/43(34%) with exomphalos diagnosed prenatally which was much higher than our results 2/11 (18.2%) with gastroschisis and 7/19 (36.8%) with omphalocele. Although most of the patients in the present study had prenatal US (93.3%), diagnoses were missed in the majority of them (70%), this calls for a need to upgrade ultrasound scans for prenatal diagnosis of these defects so that in utero transport to and possible intervention at tertiary centers can help in reducing postnatal complications and improve outcome. From those 9 (30%) patients diagnosed prenatally only, 4 (13.3%) of them were delivered by CS and studies failed to find significant correlations between perinatal outcome and mode of delivery. Omphalocele is associated with other anomalies in up to 72% and gastroschisis in 10–15% of cases.15 In this study we found that 11(57.9%) patients with omphalocele had associated anomalies, while it is found in 2(18.2%) patients with gastroschisis. A similar result was recorded by Tatić M et al16 in their study with Coexisting anomalies in 57.1% of omphalocele cases and 16.7% of gastroschisis cases. Henrich K et al study13 shows congenital abnormalities in 28% of gastroschisis cases (limited to the gastrointestinal tract like in the present study) and 81% of omphalocele cases. This difference may be due to the higher number of cases in their study (40 children with gastroschisis and 26 with omphalocele) and better equipments for diagnosing of congenital malformations. The decision to treat patients by primary closure or staged closure depends on the type of the defect, degree of viscero-abdominal disproportion, size of the defect, whether the membrane is intact or ruptured (in case of omphalocele), and the presence of major associated anomalies. Primary closure of the abdominal wall defect was reported by Henrich K et al13 in 31/40 (78%) of the gastroschisis cases and 15/26 (58%) of the omphalocele cases, primary closure was possible in 3/11 (27.3%) of the gastroschisis and 15/19 (78.9%) of the omphalocele cases in the present study. This high percentage of primary closure of omphalocele cases in the current study is because of the majority of our patients (84.2%) were small to moderate size omphalocele (<5cm), which makes them prone to primary closure, Conversely, we report a low percentage of primary closure of gastroschisis cases because this mode of therapy is not standardized in the present center because of the lack of the facilities to measure the IAP and elective ventilation postoperatively. Four cases (21.1%) with omphalocele were treated non-surgically in this study, 3 (15.8%) of them were due to the large size of the defect and in 1(5.2%) case of omphalocele minor, the condition of the patient was not stable to undergo the GA. Abdur-Rahman et al10 in their study treated 19/49 (38.7%) cases of omphalocele with non-operative management (2 cases (4.1%) were minor, 17 cases (34.6%) were major). This high rate of non-operative management in their study was due to the large number of omphalocele major 34/49 (69.4%) cases in the study. The mortality of children with abdominal wall defects is now reduced to less than 10% in developed countries, especially in cases without chromosomal anomalies or major organ malformation.17 The overall mortality rate in the present study was 40% (72% for gastroschisis and 21.1% for omphalocele). There was a significant correlation between the type of anomaly and outcome (p=0.005). Our results are comparable with reports in developing countries as shown in Table.

Table: Mortality rate of AAWD among different studies in developing countries.

	Present study N=30	Abdurahman et al10 N=56	S.Askarpour et al9 N=42	Tatić M et al16 N=13
Gastroschisis	72%	57.1%	80%	66.7%
Omphalocele	21.1%	32.4%	20%	52%

Sepsis was the leading cause of death, especially in cases with eviscerated bowel (gastroschisis and ruptured omphalocele) accompanied by respiratory insufficiency, prematurity, and associated anomalies.

The overall mortality rate is much higher in developing countries than the developed countries. This higher mortality in developing countries is due to a lack of primary care for the neonate and appropriate transport to specialized healthcare centers. Also a lack of neonatal intensive care facilities and personnel's skills in dealing with such high-risk patients.

Conclusion:

Gastrochisis and omphalocele constitute a considerable part of neonatal surgical problems, and their management is still challenging. Omphalocele cases were more likely to have associated congenital anomalies and gastroschisis if the associated anomalies were confined to the gastrointestinal tract. There was a high mortality rate, especially in patients with gastroschisis and ruptured omphalocele due to a lack of intensive care units, facilities, and trained personnel to look after such high-risk patients.

Recommendations:

for better outcomes for patients delivered with gastroschisis and omphalocele, we recommend good prenatal workup to make early suspicion of diagnosis and better postnatal care and prognosis. Trained personnel and facilities in the delivery rooms for adequate dealing with such cases and early referral to a specialized center for definite therapy. Having a neonatal intensive care unit in the pediatric surgical ward provided with ventilator support is a crucial step in the management of neonates with gastroschisis and omphalocele.

Authors' contributions:

The Author conceived of the presented idea, carried out the experiment, performed the computations, wrote the manuscript with support from coauthor, and supervised the findings of this work. The Corresponding Author helped supervise the project, planned and carried out the simulations, contributed to sample preparation, to the interpretation of the results.took the lead in writing the manuscript. Both authors provided critical feedback and helped shape the research, analysis and manuscript, discussed the results and contributed to the final version of the manuscript

References:

1. Corey KM, Hornik CP, Laughon MM, McHutchisonK, Clark RH, Smith PB. Frequency of anomalies and hospital outcomes in infants with gastroschisis and omphalocele. Early Hum Dev. 2014; 90(80):421–4.

2. Nguyen, W., Belani, K. (2018). Omphalocele and Gastroschisis. In: et al. Anesthesiology. Springer, Cham. https://doi.org/10.1007/978-3-319-74766-8_41

3. Christison-Lagay ER, Kelleher CM, Langer JC. Neonatal abdominal wall defects. Semin Fetal Neonatal Med 2011;16:164-72.

4. Kong JK, Yeo KT, Abdel-Latif ME, Bajuk B, Holland AJA, Adams S, et al. Outcomes of infants with abdominal wall defects over 18 years. J Pediatr Surg. 2016; 51(10):1644–9.

5. Rankin J, Dillon E, Wright C. Congenital anterior abdominal wall defects in the north of England, 1986-1996: occurrence and outcome. Prenat Diagn 1999; 19:662-8.

6. Suita S, Okamatsu T, Yamamoto T, Handa N, Nirasawa Y, Watanabe Y, et al. Changing profile of abdominal wall defects in Japan: results of a national survey. J Pediatr Surg 2000; 35:66-71.

7. Corey KM, Hornik CP, Laughon MM, McHutchison K, Clark RH, Smith PB. Frequency of anomalies and hospital outcomes in infants with gastroschisis and omphalocele. Early Hum Dev. 2014; 90(80):421–4.

8. Emily R Christison-Lagay , Cassandra M Kelleher, Jacob C Langer Neonatal abdominal wall defects. Semin Fetal Neonatal Med 2011 Jun;16(3):164-72.

9. Askarpour S, Ostadian N, Javaherizadeh H, Chabi S. Omphalocele, gastroschisis: epidemiology, survival,
10. and mortality in Imam Khomeini Hospital, Ahvaz-Iran, Polski przeglad chirurgiczny 2012;82:82–85.

11. Abdur-Rahman LO, Abdulrasheed NA, Adeniran JO. Challenges and outcomes of management of anterior abdominal wall defects in a Nigerian tertiary hospital , African Journal of Paediatric Surgery, May-August 2011; 8:2.

12. Aličelebić S, Mornjaković Z, Ćosović E, Šahinović M, Kapić D. Frequency of congenital anterior abdominal wall defects in surgically treated infants at Sarajevo region of Bosnia and Herzegovina: A nine-year follow-up, Folia Med. Fac. Med. Univ. Saraeviensis 2014; 49: 99-103.

13. I. Barisic M, Hausler M, Gjergja R, Kern J, Stoll C. Evaluation of prenatal ultrasound diagnosis of fetal abdominal wall defects by 19 European registries, Ultrasound Obstet Gynecol 2001; 18: 309–316.

14. Henrich K, Huemmer HP, Reingruber B, Weber PG. Gastroschisis and omphalocele: treatments and long-term outcomes, Pediatr Surg Int. 2008 Feb; 24:167-73.

15. Murphy FL, Mazlan TA, Tarheen F, Corbally MT, Puri P. Gastroschisis and exomphalos in Ireland 1998-2004. Does antenatal diagnosis impact on outcome? Pediatr Surg Int. 2007 Nov; 23:1059-63.

16. Steven W. Bruch and Jacob C. Langer. Omphalocele and gastroschisis. In: Puri P, editor. Newborn Surgery. 2nd edition. London: Arnold; 2003. P.605-613.

17. Tatić M, Jokić R, Bukarica S, Borisev V. Clinical analysis of congenital abdominal wall defects--omphalocele and gastroschisis, Med Pregl. 2006 Jul-Aug;59:347-55.

18. Venugopal S, Zachary RB, Spitz L. Exomphalos and Gastroschisis: A 10- year review. Br J Surg 1976; 63:523-5.

النتائج المبكرة للمرضى المولودين بانشقاق البطن الخلقي و قروة السروة

د.سالار صباح بيرداود
د.عبدالرحمن عمر طه

الخلاصة:
الخلفية : تختلف نتائج ما بعد جراحة الإنشقاق البطن الخلقي و قروة السروة بين البلدان المختلفة وخاصة بين البلدان المتقدمة والنامية.
الأهداف :الهدف من هذه الدراسة هو تحديد النتيجة المبكرة للمرضى المولودين إنشقاق البطن الخلقي و قروة السروة.
الموضوعات والطرق: شملت هذه الدراسة التحليلية 30 مريضًا (11 مريضًا إنشقاق البطن الخلقي و 19 مريضًا قروة السروة). تم علاج المرضى في مدينة اربيل خلال الفترة 2015-2018. تم استبعاد المرضى الذين يعانون من انقلاب المثانة وانقلاب مجرور.
النتائج: كانت نسبة الاناث الى الذكور 1.3: 1 ومتوسط العمر عند الوصول 11.7 ساعة. كان متوسط الوزن 2.86 كجم. وكانت الغالبية العظمى من المرضى ولدوا في نهاية فترة الحمل الطبيعي (83.3 ٪) وتمت الولادة في 19(63.3 ٪)حالة عن طريق المهبل. وقد تم التشخيص قبل الولادة في 9 (30 ٪)حالات. كان معدل حجم الخلل 3.7 سم و كانت مختلطة المحتوى في 7(23.3 ٪) حالات. تم إجراء الإصلاح الأولي في 18 (60٪) حالة ، والصومعة في 8 (26.7٪) ، و 4 (13.3٪) حالات لم يتم علاجها جراحيًا. تم إجراء العمليات في غضون 24 ساعة من الوصول في 16 حالة (61.5٪). العاهات الولادية المصاحبة وجدت في 11(57.9 ٪) حالة من قروة السروة و حالتان فقط(18.2 ٪) من إنشقاق البطن الخلقي . كان متوسط فترة الإقامة في المستشفى 6 أيام. وكانت نسبة الوفيات بين مرضى المصابين بإنشقاق البطن الخلقي 72 ٪ ولكن 21.1 ٪ فقط للمرضى المصابين بقروة السروة.
الاستنتاج: إنشقاق البطن الخلقي و قروة السروة يشكلان جزءًا كبيرًا من المشاكل الجراحية لحديثي الولادة ، ولا تزال إدارتها صعبة. يعد التشخيص قبل الولادة ضروريًا للحد من مضاعفات ما بعد الولادة من خلال التحضير والمشورة قبل الولادة بشكل أفضل. مرضى قروة السروة لديها نسبة عالية من التشوهات الخلقية المرتبطة بها ومرضى انشقاق معدي التشوهات المرتبطة بها محصورة في الجهاز الهضمي. كان هناك ارتفاع معدل الوفيات بين المرضى الذين يعانون من إنشقاق البطن الخلقي وتمزق قروة السروة.
الكلمات المفتاحية: نتيجة ما بعد الجراحة ، انشقاق المعدة ، القيلة الأمينية ، عيب جدار البطن ، الشذوذ الخلقي.