Omphalocele and Gastroschisis

G. HOSSEIN MAHOUR, M.D.,* JORDAN J. WEITZMAN, M.D.,**
JENS G. ROSENKRANTZ, M.D.***

From the Department of Surgery, Childrens Hospital of Los Angeles and the University of Southern California, School of Medicine, Los Angeles, California 90054

Omphalocele has been defined as an herniation of viscera into the base of umbilical cord with a covering membranous sac of peritoneum-amnion.[1,16,18] In this anomaly, the umbilical cord is inserted into the sac, but the sac may be ruptured pre- or postnatally. When the defect is large, an omphalocele may contain liver or spleen in addition to a large portion of the intestinal tract.

Gastroschisis is a full thickness cleft of the abdominal wall next to the insertion of the cord through which the intestines are eviscerated.[17,18,21] The characteristics which differentiate gastroschisis from omphalocele are the extra-umbilical location of the abdominal wall defect, the normal insertion of the umbilical cord and the absence of a covering sac or its ruptured remnants.[21,22]

Mortality rates for each condition have been high;[1,13,24,26,27] and this has been ascribed to sepsis, starvation during prolonged intestinal ileus, respiratory insufficiency from increased abdominal pressure postoperatively, and to other associated anomalies. With the recent use of staged repair with Silon prosthesis[2,14,25] and total parenteral alimentation for the infant who cannot feed, the outlook for these babies seems to have improved.

The purpose of this paper is to review the experience with these conditions during 10 years at the Childrens Hospital of Los Angeles and to examine in detail the factors affecting mortality.

Submitted for publication March 6, 1972.

* Request reprints: G. Hossein Mahour, M.D., M.Sc. (Surgery), Assistant Professor of Surgery, University of Southern California School of Medicine, Childrens Hospital of Los Angeles, 4650 Sunset Boulevard, Los Angeles, California 90054.

** Assistant Clinical Professor of Surgery, University of Southern California School of Medicine, Los Angeles, California.

*** Professor of Surgery, University of Southern California School of Medicine, Surgeon-in-Chief, Childrens Hospital of Los Angeles, Los Angeles, California.

Material

During the years July, 1960 through June, 1970, 59 patients with omphalocele and gastroschisis have been seen and surgically treated at the Childrens Hospital of Los Angeles (Table 1). In 57 patients, the initial surgical procedure was performed here and in two the initial procedure had been performed elsewhere and the patients were referred here for further treatment. Patients with abdominal wall defect associated with ectopia cordis or exstrophy of the bladder with or without vesicointestinal fistula[6,8,12,23] are not included in this series.

In 22 cases, there was an intact omphalocele. In five additional cases, remnants of the ruptured sac were present; these were classified as ruptured omphalocele. Twenty-three patients had normal insertion of the umbilical cord next to the abdominal wall defect and had no evidence of sac or sac remnant and were diagnosed as gastroschisis. In nine additional instances, the operative description was not adequate to classify the case; these were called Indeterminate (Table 1).

In estimation of mortality rates, death is considered as such when it occurred during the same hospital admission.

Results

Mortality. Of the 59 patients, 30 are alive and 29 are dead—a mortality rate of 49% (Table 1). Mortality rate for 22 patients with intact omphalocele was 23%; whereas, in 37 patients with intestinal evisceration (ruptured omphalocele, gastroschisis plus the indeterminate group) the overall mortality rate was 65% (Chi squared

= 8.19, $p < 0.01$). In the latter combined group, the mortality rate was highest in the patients with ruptured omphalocele: 100% vs. 61% for gastroschisis and 56% for the indeterminate group. There were no significant differences in mortality rate between the gastroschisis and indeterminate groups.

The high mortality in patients with intestinal evisceration has been reduced in recent years. From 1960 through 1965, only four of 22 patients with ruptured sac or no sac survived, a mortality rate of 82%; from 1966 to 1970, 15 such patients were admitted with nine survivors, a mortality rate of 40%; (Chi squared = 5.13, $0.01 < p < 0.05$).

Birth Weight. In 18 patients with intact omphalocele in whom the birth weight was recorded, one weighed less than 2,500 Gm. at birth (this child died). Seventeen patients weighed 2,500 Gm. or more at birth and two of these died.

In 36 patients with ruptured sac or no sac in whom the birth weight was recorded, 24 weighed less than 2,500 Gm. at birth; 15 of these died. Twelve patients weighed 2,500 Gm. or more at birth and 8 of these died.

Size of Defect. Of the patients with intact omphalocele, 11 had defects 4 cm. or less in diameter, none of them with the liver presenting in the defect; two of these died. Eleven had defects more than 4 cm. in diameter; three of these died (Table 3). In the majority of the patients of the latter group, the sac contained liver in addition to the intestines.

Associated Anomalies. Fourteen patients with omphalocele, three with gastroschisis and two patients of the indeterminate group had anomalies in addition to the ventral abdominal defect and coexisting incomplete rotation of the intestine (Table 2).

Four patients with intact omphalocele had congenital heart disease. All of these patients underwent primary closure of the abdominal wall defect. Two of these patients, one with tetralogy of Fallot and the other with ventricular septal defect, are living. The third patient with agenesis of the left atrium, totally anomalous pulmonary venous return, ventricular septal defect, coarctation of the aorta and anorectal agenesis, died 1 day after operation. The fourth patient had transposition of the great vessels, ventricular septal defect and ileal atresia. She died of Pseudomonas sepsis 2 months after operation.

A premature baby with gastroschisis had associated ventricular septal defect and arthrogryposis. This patient died of subarachnoid hemorrhage and sclerema 4 days after skin flap coverage of the eviscerated intestines. Another patient from the indeterminate group had ventricular septal defect; this child is one of the survivors.

Two patients with intact omphalocele had diaphragmatic hernia. Diagnosis of this associated anomaly was made preoperatively in one patient. She died 1 day after repair of the left diaphragmatic hernia, partial hepatectomy, and skin flap coverage of the omphalocele. She also had associated hydronephrosis. The second patient underwent primary closure of the abdominal wall defect. This patient died during early postoperative course and postmortem examination revealed a large right diaphragmatic hernia, which was not recognized at the operation, with herniation of portion of the liver into the right thoracic cavity.

TABLE 1. *Omphalocele and Gastroschisis in 59 Patients—1960–1970*

Diagnosis	No. of Patients M.	F.	Alive	Dead
Omphalocele	15	12	17	10
Sac Intact	12	10	17	5
Sac Ruptured	3	2	0	5
Gastroschisis	15	8	9	14
Indeterminate*	5	4	4	5
Total	35	24	30	29

* Detail of the abdominal wall defect is not recorded.

At operation and after removing the intact sac, diagnosis of small bowel atresia was made in three patients with omphalocele. Three additional patients, two with gastroschisis and one with ruptured omphalocele, had small bowel atresia. In these patients, the presence of this anomaly was recognized between the rubbery, thickened, and edematous mass of bowel at the time of abdominal wall closure.

The chromosome analysis confirmed diagnosis of Down's syndrome in one patient and 18-trisomy syndrome in another. Both patients had repair of the omphalocele with intact sac and both are dead, at ages 2 and ½ years and 1 year respectively, due to unrelated causes.

Treatment

The surgical treatment is outlined in Table 3.

In 16 patients with ruptured sac or no sac, the postoperative course was complicated with sepsis. In nine patients, sepsis was due to *Pseudomonas aeruginosa*, all

TABLE 2. *Additional Significant Anomalies (Incomplete Rotation and fixation of the Intestines Are Not Included).*

Anomaly	Omphalocele (14 patients)	Gastroschisis (3 patients)	Indeterminate (2 patients)
Cardiovascular	4	1	1
Diaphragmatic hernia	2	0	0
Small bowel atresia	4	2	0
Anorectal anomaly	2	0	0
Renal anomaly	3	0	0
Anomalies of extremities	4	1	1
Cleft lip & palate	1	0	0
Down's syndrome	1	0	0
18-trisomy syndrome	1	0	0

TABLE 3. *Surgical Procedures in 59 Patients with Omphalocele and Gastroschisis*

Type	Procedure	No. of cases	Deaths
Intact sac:		22	5
Defect 4 cm. or less		11	2
	One-stage closure	10	1
	Staged procedure*	1	1
Defect over 4 cm.		11	3
	One-stage closure	7	3
	Staged procedure*	4	0
Ruptured sac:		5	5
	One-stage closure	1	1
	Staged procedure*	4	4
Gastroschisis:		23	14
	One-stage closure	5	3
	Staged procedure*	18	11
Indeterminate:		9	5
	One-stage closure	4	2
	Staged procedure*	5	3

* Ventral hernia coverage with mobilized skin (with or without flank relaxing incisions) or prosthetic material as the first stage.

of these patients developed sepsis while on antibiotics such as penicillin, Streptomycin, and Chloromycetin—alone or in combination. Pseudomonas sepsis developed in one additional patient after repair of intact omphalocele and intestinal anastomosis for ileal atresia. This child had transposition of the great vessels and ventricular septal defect.

Three patients with eviscerated bowel developed postoperative fecal fistula after ventral hernia coverage with skin flap.

Omphalocele with Intact Sac. Of the 11 patients with abdominal wall defect 4 cm. or less in diameter, ten had primary closure of the defect (Table 3). The only death in this group occurred in a patient with agenesis of the left atrium, totally anomalous pulmonary venous return, ventricular septal defect and coarctation of the aorta, one day after sigmoid colostomy for anorectal agenesis and repair of the omphalocele. Staged procedure was performed in one patient. This child had hydronephrosis and died 1 day after repair of congenital left diaphragmatic hernia, partial hepatectomy and skin flap coverage of the omphalocele.

Of the 11 patients with abdominal wall defect over 4 cm. in diameter, four patients had staged procedures; all of these patients are among the survivors (Table 3). Primary closure of the abdominal wall was performed in the remainder; three of these patients died. One death occurred during the early postoperative course, and postmortem examination of the patient revealed a large right diaphragmatic hernia, which was not recognized at operation, with some hepatic herniation into the right thoracic cavity. Another patient developed cardiac arrest and pulmonary failure shortly after operation. Her condition improved when tightly closed abdominal fascia was opened, but she died from a second cardiac arrest shortly thereafter. The third patient died with Pseudomonas sepsis 2 months after repair of the omphalocele and intestinal anastomosis for ileal atresia. This child had transposition of the great vessels and ventricular septal defect.

Ruptured Omphalocele. One patient, with this anomaly, had primary closure of the abdominal wall defect; this child died 3 days later with aspiration pneumonitis. Four additional patients had staged procedures; all of these babies were premature, one had associated ileal atresia and all of these patients are dead. Two patients died with Pseudomonas sepsis, one with pulmonary infection, and the last patient died with massive pulmonary hemorrhage.

Gastroschisis. Five patients had one-stage closure of the abdominal wall defect; three of these babies died with Pseudomonas sepsis. One of the survivors was operated upon elsewhere. Two days later, abdominal exploration revealed volvulus due to incomplete rotation and fixation of the intestine.

Eighteen patients had staged procedures; 11 of this group failed to survive. Ten deaths occurred after the first stage of the operation—use of skin flaps or prosthetic material to cover the eviscerated bowel. Sepsis, pulmonary infection and complications of the abdominal crowding were the major contributing causes of deaths. One additional patient died with massive hemoperitoneum and uncontrollable excessive "serosal ooze" shortly after "peeling off" the membrane from the bowel wall and mobilized skin coverage of the eviscerated intestines.

In one of the survivors, the intestinal anastomosis for ileal atresia was left in a "Silon chimney." Forty days later, the Silon sheets were removed and the abdominal wall defect was finally closed. At this operation, the intestines appeared to be normal with no covering membrane and the anastomosis was well healed.

Indeterminate Group. Four patients had one-stage closure of the abdominal wall defect; two of these died. One patient died with aspiration pneumonia and the second patient developed infarction of the intestine 4 days after operation. This last patient died with Pseudomonas sepsis 3 weeks after massive intestinal resection. One of the survivors was operated upon elsewhere. Abdominal exploration on the following day revealed volvulus due to incomplete rotation of the intestines.

Five patients had staged procedures, three of these died. All three patients were premature, two died with sepsis (one Pseudomonas), and the third patient had associated arthrogryposis and died with pulmonary infection and sclerema.

Comments

Omphalocele and gastroschisis are acute emergencies of the neonatal period. As demonstrated in this series,

the high mortality in these patients is due in part to the associated anomalies.

Nonoperative management of large intact omphalocele has been advocated.[10,11,15,28] We have no experience with this type of management, but it appears that surgically correctable intra-abdominal anomalies are frequent enough to justify abdominal exploration in these patients. In this series, three patients with intact omphalocele had small bowel atresia and two other patients had diaphragmatic hernia. Aitken[1] reported a death due to intestinal obstruction from a malrotation which was missed at the original operation. Grob[15] reported 16 patients with intact omphalocele treated with nonoperative management. Two of his patients died from midgut volvulus. Bill[3] reported a patient with midgut volvulus while under expectant treatment. Six patients had to be operated upon after the omphalocele repair so that the malrotation could be corrected.[16]

In patients with gastroschisis, prematurity, incomplete rotation and fixation of an abnormally short midgut is common.[4,19,21,24] In these patients, the eviscerated mass of bowel, similar to that in patients with ruptured omphalocele of long duration, is thickened and edematous. The bowel is relatively rigid, matted together and is covered with a fibrinogelatinous membrane. Peristalsis is usually absent. The spleen and the liver are not protruding through the defect although the stomach, gallbladder, uterus, and adnexa may be seen. Evisceration of the urinary bladder has been reported.[9,21]

We would propose initial operative repair in most cases of omphalocele and gastroschisis and reserve the nonoperative management[10,11,15,28] for the premature infant with large intact omphalocele and multiple associated anomalies, who is a poor risk for operation. Preoperatively, the eviscerated viscera or the intact sac of the omphalocele should be covered with sponges soaked in sterile saline or a plastic bag to avoid heat and electrolyte loss. The exposed viscera should be supported to prevent torsion and kinking.

Considerable judgment must be exercised in selection of the procedure to repair the defect. Tight closure and creation of abdominal crowding should definitely be avoided in all cases. In some patients, manual stretching of the abdominal wall from the inside[17] allows the bowel to reside in the abdominal cavity and the defect be closed without tension. In any patient in whom the primary closure portends abdominal crowding, institution of the staged procedure is vital if high mortality is to be avoided. In some, flank relaxing incisions make the abdominal closure easier and prevent increased intra-abdominal pressure and skin necrosis due to tight skin flaps. We favor utilization of Silon sheets[2,14,25] when staged procedure is undertaken. To prevent the consequences of increased intra-abdominal pressure and skin necrosis produced by tight closure of the flaps, nonoperative management of large intact omphalocele has been advocated with encouraging results.[10,11,15,28] Utilization of the Silon sheets prevents these complications and the other inherent disadvantages of skin covered ventral hernia. Postoperatively, with staged reduction of the content of the Silon pouch and with progressive stretching, the abdominal wall enlarges in a short period of time to accommodate its viscera.

In order to facilitate the abdominal closure, splenectomy and liver resection[5] or intestinal resection[20] should not be done. Removal of the thick fibrinous peel from the surface of the bowel (decortication[21,24]), is difficult, tedious, time consuming and unnecessary. It creates extensive raw surface with its potential immediate and late complications and it may result in perforation. In this series, one patient died due to uncontrollable excessive "serosal ooze" shortly after the operation. In gastroschisis and ruptured omphalocele of long duration, it is extremely important to carry out gentle blunt dissection of the matted-together mass of intestine in search for atresia. Deaths from unrecognized intestinal atresia have been reported.[7] In these patients, the thickened edematous bowel is not capable of peristalsis for many days after operation. We are impressed with distinct advantages of gastrostomy in these cases in helping to control gastric distention, avoidance of ventilatory complications and monitoring the progress of the feedings. Postoperatively, due to resolution of the fibrino-exudate membrane covering of the bowel and exposure of the trapped organisms, early and late sepsis is common. Prolonged administration of antibiotics may be helpful, and the patient should be carefully watched for sepsis. In this series, nine patients developed postoperative Pseudomonas sepsis although they were "covered" with antibiotics.

We attribute our steadily increased survival to frequent employment of the staged procedures, urgent attention to restoring blood volume during and after the operation, prevention of hypothermia and frequent assessment of the electrolytes and correction of acid base imbalance. Careful attention to the nutritional status of the infant with employment of total parenteral alimentation are of considerable importance.

Summary

Fifty-nine cases of omphalocele and gastroschisis are presented—the experience at the Childrens Hospital of Los Angeles during a 10-year period, 1960-1970. On the basis of our experience with these patients, we draw the following conclusions:

1. The high mortality rate in these patients is due in part to the associated anomalies.

2. In patients with omphalocele, surgically correctable

intra-abdominal anomalies are frequent enough to warrant abdominal exploration.

3. Whichever method of treatment is undertaken, creation of abdominal crowding should be avoided in all cases. We favor the ventral hernia coverage with Silon sheets when staged procedure is undertaken.

4. The occurrence of early and late postoperative sepsis, particularly due to *Pseudomonas aeruginosa*, should be anticipated.

References

1. Aitken, J.: Exomphalos, Analysis of a 10-year Series of 32 Cases. Arch. Dis. Child., **38**:126, 1963.
2. Allen, R. G. and Wrenn, E. L. Silon as a Sac in the Treatment of Omphalocele and Gastroschisis. J. Pediatr. Surg., **4**:3, 1969.
3. Bill, A. H., Jr., Omphalocele *in:* Mustard, W. T., Ravitch, M. M., Snyder, W. H., Jr., Welch, K. J. and Benson, C. D.: 2nd Ed., Vol. 1, Year Book Medical Publishers, Inc., pp. 678, 1969.
4. Bill, A. H., Jr.: Gastroschisis in: Mustard, W. T., Ravitch, M. M., Snyder, W. H., Jr., Welch, K. J. and Benson, C. D.: 2nd Ed., Vol. 1, Year Book Medical Publishers, Inc., pp. 685, 1969.
5. Buchanan, R. W. and Cain, W. L.: A Case of a Complete Omphalocele. Ann. Surg., **143**:552, 1956.
6. Cantrell, J. R., Haller, J. A., Jr. and Ravitch, M. M.: A Syndrome of Congenital Defects Involving the Abdominal Wall, Sternum, Diaphragm, Pericardium and Heart. Surg. Gynecol. Obstet., **107**:602, 1958.
7. Cordero, L., Touloukian, R. J. and Pickett, L. K.: Staged Repair of Gastroschisis with Silastic Sheeting. Surgery, **65**:676, 1969.
8. Crittenden, I. H., Adams, F. H. and Mulder, D. G.: A Syndrome Featuring Defects of the Heart, Sternum, Diaphragm and Anterior Abdominal Wall. Circulation, **20**:396, 1959.
9. Diller, W. E. and Travis, B. W.: Eventration of the Abdominal Viscera in a Newborn. Ohio State Med. J., **51**:756, 1955.
10. Dorogi, J.: Improved Conservative Treatment of Exomphalos. Lancet, **2**:888, 1964.
11. Drescher, E.: Observations on the Conservative Treatment of Exomphalos. Arch. Dis. Child., **38**:135, 1963.
12. Duhamel, B.: Embryology of Exomphalos and Allied Malformations. Arch. Dis. Child., **38**:142, 1963.
13. Firor, H. V.: Omphalocele—An Appraisal of Therapeutic Approaches. Surgery, **69**:208, 1971.
14. Gilbert, M. G., Mencia, L. F., Brown, W. T. and Linn, B. S.: Staged Surgical Repair of Large Omphaloceles and Gastroschisis. J. Ped. Surg., **3**:702, 1968.
15. Grob, M.: Conservative Treatment of Exomphalos. Arch. Dis. Child. **38**:148, 1963.
16. Gross, R. E.: The Surgery of Infancy and Childhood. Its Principles and Techniques. Philadelphia, W. B. Saunders Co., 1953.
17. Izant, R. J., Brown, F. and Rothmann, B. F.: Current Embryology and Treatment of Gastroschisis and Omphalocele. Arch. Surg., **93**:49, 1966.
18. Johnson, A. H.: Omphalocele and Related Defects. Am. J. Surg., **114**:279, 1967.
19. Judd, D. R., Wince, L. and Moore, T. C.: Gastroschisis: Report of Two Cases Successfully Treated. Surgery, **58**:1033, 1965.
20. Meltzer, A.: Huge Omphalocele Ruptured *in utero*. J.A.M.A., **160**:656, 1956.
21. Moore, T. C.: Gastroschisis with Antenatal Evisceration of Intestines and Urinary Bladder. Ann. Surg., **158**:263, 1963.
22. Moore, T. C. and Stokes, G. E.: Gastroschisis. Report of Two Cases Treated by a Modification of the Gross Operation for Omphalocele. Surgery, **33**:112, 1953.
23. Mulder, D. G., Crittenden, I. H. and Adams, F. H.: Complete Repair of a Syndrome of Congenital Defects Involving the Abdominal Wall, Sternum, Diaphragm, Pericardium and Heart; Excision of Left Ventricular Diverticulum. Ann. Surg., **151**:113, 1960.
24. Rickham, P. P.: Rupture of Exomphalos and Gastroschisis. Arch. Dis. Child., **38**:138, 1963.
25. Schuster, S. R.: A New Method for the Staged Repair of Large Omphaloceles. Surg. Gynecol. Obstet., **125**:837, 1967.
26. Simpson, T. E. and Lynn, H. B.: Omphalocele: Results of Surgical Treatment. Mayo Clin. Proc., **43**:65, 1968.
27. Smith, W. R. and Leix, F.: Omphalocele. Am. J. Surg., **111**:450, 1966.
28. Soave, F.: Conservative Treatment of Giant Omphalocele. Arch. Dis. Child., **38**:130, 1963.