

Omphalocele and Gastroschisis

Analysis of a 6 year Series of 23 Cases

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ABSTRACT

Twenty-three cases of omphalocele or gastroschisis managed at the Padiatric Surgical Department of Juntendo University Medical School, Tokyo, Japan during the period 1973-78 are analysed. There were 5 cases with hernia into the cord, 13 with omphalocele proper and 6 with gastroschisis. The overall mortality was 26%. Both omphalocele proper and hernia into the cord had high incidence of concomitant malformation. Survival could be closely correlated with severity of associated congenital anomalies. Patient with birth weight lower than 2000gm and/or with defect larger than 8 cm was also at great risk. T.P.N. and assisted respiration evolved the management of omphalocele and gastroschisis. Significant differences in associated anomalies, birth order and birth weight between hernia into the cord and gastroschisis tends to rebut

that both are the results of same congenital accident.

KEY WORD: Omphalocele, Gastroschisis.

T.P.N. = Total Parental Nutrition.

INTRODUCTION

Omphalocele is a hernition of viscera through the open umblical ring into the base of umblical cord with a covering membraneous sac of peritoneum-amnion. The sac is usually intact but may be reaptured at any time prior to, during or after delivery. In these cases, remnants of the sac with the umblical vessels within its walls will be found, thus differentiating the ruptured omphalocele from a gastroschisis. The embryogenesis of omphalocele have been lucidly discussed. In small omphalocele or hernia into the cord, around which the umblical ring is less than 4cm in diameter and a sac that contains only loops of small intestine is usually regarded as resulting simply from failure of complete return of the extracoelemic

midgut to the abdomen with secondary failure of the umbilical ring. On the other hand, the giant omphalocele or omphalocele proper represents a true morphological failure of the process of closure of embryo body by its component folds.

The embryology and the definition of gastroschisis have been varied and confused. In this article, the term is used for designating which has an uncovered anterior abdominal defect with umbilical cord originating from abdominal wall. The effect may be immediately left to the cord or separated from it by a strip of skin. Gastroschisis has been postulated as a developmental failure of one lateral abdominal plate of the somatopleure. However, because no one has described the presence of any muscle mass intervening between the defect and the cord, and the defect is always between the medial borders of the rectus muscles, recent authors consider that gastroschisis is resulted from intrauterine rupture of the amniotic membrane of extraembryonic coelom.

Although there is no unanimity of opinion of the therapeutic mode, the survival rate for infants with omphalocele and gastroschisis has been generally improved. The purpose of this paper is to analyse the factors that affect prognosis when newer methods of patient management are used, and also to determine the clinical difference between omphalocele proper, hernia into the cord and gastroschisis.

FIG. I



Gastroschisis.

FIG. II



Repaired Gastroschisis.

MATERIAL AND METHODS

During the period 1973–78, twenty-three cases of omphalocele or gastroschisis were admitted to the Pediatric Surgical Department of Juntendo University School. Patients with omphalocele associated with ectopia cordis or cloaca extrophy are not included in this series of 17 cases with intact sac; five had defects 4cm or less in diameter, these were classified as Hernia into the cord, twelve had defects greater than 4cm in diameter and were considered as Omphalocele proper. In additional 6 babies, absence of a covering sac, intestines eviscerated through a full-thickness defect in the abdominal wall left to the insertion of the cord, were diagnosed as Gastroschisis. The presence of a narrow skin bridge interposed between the medial margin of the defect and the insertion of the cord was clearly found in only one of them.

From retrospective review of the charts of each case, difference in clinical features between each group were compared. Factors affecting the prognosis were estimated, and result of treatment was discussed, the need for nutritional support by total parenteral alimentation and postoperative respiratory support by assisted respiration were assessed too.

RESULTS

(a) Sex.

There were 13 males and 10 females.

(b) Birth Order.

Birth order showed that 50% of infants with omphalocele proper and 67% of infants with gastroschisis were first born. No infant with hernia into the cord belonged to first born. Difference in birth order between infants with gastroschisis and those with hernia into the cord is significant (Chi squared test: $P < 0.01$).

(c) Birth Weight.

The birth weights of each group and its relation to mortality are listed in Table 1.

TABLE 1

Relation of Birth Weight to Mortality

B.W. gm Type	2000	2000 2500	2500	Total	Mortality Rate
H			5(1)*	5(1)	20%
Om	1(1)	2	9(3)	12(4)	33%
G	1(1)	4	1	6(1)	16%
Total	2(2)	6	15(4)	23(6)	26%
Mortality Rate	100%	0	26%	26%	

(* Number in parenthesis indicates cases of death).

H: Hernia into the cord.

Om: Omphalocele proper.

G: Gastroschisis

All of the 5 cases with hernia into the cord had birth weight more than 2500gm. In contrast, 5 of the six babies with gastroschisis weighed less than 2500gm at birth ($P < 0.01$). Three of these were preterm. Extremely lower birth weight would affect the survival rate. Both of the children whose birth weights below 2000gm died. Only 4 of 15 babies who weighed over 1500gm succumbed. This difference in mortality is highly significant ($P < 0.001$).

(d) Size of defect.

In Table 2, mortalities in infants with different size of defect are compared. Difference in mortality between infants with defect less than 4cm (20%) and those more than 8cm (50%) is not statistically significant ($P < 0.1$) because of the small sample of patients.

(e) Protruded or Eviscerated Organs.

Only loops of intestine were herniated in cases with hernia into the cord.

TABLE II

Relation of Size of Defect to Mortality

Size cm Type	2-4	4-6	6-8	8-10
H	5(1)			
Om		2(1)	8(2)	2(1)
G	5(1)	1		
Total	10(2)	3(1)	8(2)	2(1)
Mortality	20%	33%	25%	50%

In infants with omphalocele proper, the liver with or without intestine was seen through its sac in 9 cases, 4 of these died.

In almost all babies with gastroschisis only part of stomach and intestine eviscerated through the defect, the eviscerated guts were oedematous, thickened and covered by fibrogelatinous material in all cases. One baby in whom liver evisceration was noted survived.

The overall mortality in patients with liver herniation and those without liver herniation were 40% and 13% respectively ($P < 0.1$).

(f) Associated Anomalies

Malrotation of the midgut was noticed in 7 infants with omphalocele proper, in all of the 6 babies with gastroschisis and in no one with hernia into the cord.

Other than malrotation, 3 cases with hernia into the cord (60%) and 4 infants with omphalocele proper (33%) had additional associated anomalies, these are listed in Table 3. No associated anomaly other than malrotation was discovered in gastroschisis patients in this series. This difference in frequency of associated anomalies other than malrotation between infants with gastroschisis and hernia into the cord is significant ($P < 0.025$).

As shown in Table 3, associated anomalies in patients with omphalocele proper or hernia into the cord were severe and complicated. All these 4 deaths were directly contributed to associated anomalies. In cases with intact sac, the 57% mortality in infants with major associated anomalies compared with a mortality of only 9% in infants with minor or no associated anomalies. This difference is statistically significant ($P < 0.005$).

DISCUSSION

It is well-known that omphalocele is frequently associated with other anomalies. In our series, only infants with omphalocele proper but also those with hernia into the cord had a high incidence of concomitant malformation. There was no correlation

TABLE III

Relation of Associated Anomalies to Mortality in Hernia into the Cord and Omphalocele Proper

Number of Associated Anomalies	Outcome
1. Asymmetrical conjoined twin	Survive
2. ASD, VSD and left superior Vene Cava	Survive
3. Trisomeal-D syndrome	Death
4. Multiple cardica anomalies, Horse-shoe kidney, Hydroureter and Cryptorchism	Death
5. Multiple cardiac malformations*	Death
6. T.F. and Microcephalus	Death
7. E.M.G. Syndrome	Survive

between the frequency of associated anomalies and the size of the defect in omphalocele.

Gastroschisis is characterised by rarely concomitant anomalies and prematurity. Gastroschisis also is predominately first born. Our analysis showed that differences in associated anomalies, birth order and birth weight between infants with hernia into the cord and those with gastroschisis were statistically significant. This feature tends to rebut that gastroschisis and hernia into the cord are the result of same congenital accident.

(b) Requirement of T.P.N. and Assistant Respiration

Eight cases with omphalocele proper and 5 cases with gastroschisis received total parenteral nutrition. Peripheral hyperalimentation was employed in 12 cases and only one whose peripheral venous route been infeasible received central hyperalimentation. The median duration of T.P.N. from all cases was 16.5 days. The median weaning period of parenteral nutrition was 8.27 days after starting of gastric feeding.

There were 11 infants, 5 gastroschisis patients and 6 cases of omphalocele proper, whose respiratory functions were not adequate post-operatively, as demonstrated by general conditions as well as the blood pictures, required artificial respiratory therapy in median duration of 4 days. Of these; 6 were treated with prosthetics, 4 were primary layer closure, 1 was managed by skin flap technique. Pneumonia in 2 cases was related to the prolonged use of respirator, 3 deaths occurred in this group. However, no death could be contributed to the use of assisted respiration.

(a) Treatment

The modes of treatment for our cases with omphalocele and gastroschisis are summarized in Table 4.

Of the 5 cases with hernia into the cord: 2 had primary closure of the defect; one of these died 4 months later with Trisomy-D syndrome, 3 were treated successfully by painting the sac with mercurochrome.

Of the 12 patients with omphalocele proper: 2 babies received mercurochrome application; one expired 2 days after admission from multiple cardiac anomalies, the survivor underwent hernioplasty for ventral hernia at age of one and a half year. 3 infants had one-stage closure of the defect; one of these did not survive attributing to associated anomalies. Four cases were treated with the skin-flap closure technique, the resulting large ventral hernia required eventually second staged repair in two and fourth staged repair in another two of this group, all of these are alive. Three infants had staged repair using prosthetics; 2 of these died, one death occurred 3 months later secondary to microcephalus and cardiac malformation, then another case died on month later due to sepsis and pneumonic, the survivor complicated with ventral hernia and inguinal hernia required subsequently correction. All of the three had wound infection and premature separation of prosthetics during the course of treatment.

Of the 6 cases with gastroschisis, primary layered closure was feasible only in one baby. Five infants underwent silon pounce staged repair; completed closure of the defect was achieved at, 5th and 6th day respectively in three cases, the fourth case had wound infection and premature separation of the prosthetics resulted ventral hernia, the only death in this group was 1800gm birth weight baby who developed anuria and died 2 days after operation.

Factors affecting prognosis including low birth weight, associated anomalies, liver herniation and size of the defect have been thoroughly discussed. Among our 6 mortality: 4 were directly related to associated anomalies, 2 were extremely lower birth weights and only one who had defect larger than 10cm died of sepsis. As demonstrated in results, omphalocele infants with associated anomalies had a higher mortality rate than those without associated anomaly. Difference in mortality between patients with birth weight lower than 2000gm and those weighed over 2500gm was significant, patients with

TABLE IV

Mode of Treatment for Omphalocele and Gastroschisis.

Method Type	Primary Closure	Skin Flap	Staged c Prosthetics	Conservative Treatment
H	2(1)			3
Om	3(1)	4	3(2)	2(1)
G	1	5(1)	5(1)	
Total	6(2)	4	8(3)	5(1)

a large defect also usually associated with liver herniation and had a poorer outcome than those with a small defect.

Mortality in infants with severe associated anomalies, with birth weight lower than 2000gm and with defect larger than 8cm were 57%, 100%, and 50% respectively. Despite current therapy, it seems that those infants with a defect larger than 10cm, or with multiple associated anomalies and those with a birth weight less than 2000gm still to be poor risks.

The defect of hernia into the cord could be treated simply either by primary layer closure or by painting with drying agent. The only one death in this group was secondary to associated anomalies. On the other hand, management of the giant omphalocele, especially those with multiple anomalies, has been and still is a challenge to us. There was no mortality in infants treated with gross procedure. But all of those resulted in massive ventral hernia presenting technical problems at the time of secondary repair. Our result using prosthetics in treatment of omphalocele proper was unfavourable, all of the three had premature separation of the prosthetics and all of them had wound infection, primary layer closure seems to be less complicated, but this method if undue compression of viscera to be avoided, only could be utilized for selected cases. No-operative method had been re-advocated by recent authors.

Because of time consuming, we reserved this method only for infants with high surgical risk.

In treatment of gastroschisis, with vigorous muscle stretching maneuver and aided by assisted respiration, the defect could be closed completely either immediately or within a week using the silon pouch stage repair.

Peripheral Hyperalimentation was employed routinely for infants with omphalocele proper and gastroschisis. As stressed by many authors gastroschisis always complicated with prolong ileus that needs T.P.N. to omphalocele proper, the chief objective of T.P.N. is to provide reparatively nutrition support during the digestive dysfunction period as well as to facilitate the process of abdominal wall closure by mean of placing the gastrointestinal tract at rest and decreasing the abdominal distension.

Respiratory failure has been the most common cause of immediately post-operative health in infants with omphalocele and gastroschisis. As managed with assisted respirator, no death was due to respiratory failure in this series. Current respiratory therapy has tremendously benefited those patients with acute respiratory. By the way of increasing

functional residual capacity and decreasing veno-arterial 1 shunt, the hypoxia resulting from acutely increased intra-abdominal pressure can be improved by the application of the positive pressure breathing.

It appears that by carefully using the peripheral hyperalimentation and the mechanical respiration as adjuncts in the management of infants with omphalocele or gastroschisis to be satisfactorily facilitating the stage of the abdominal wall closure by application of tolerable pressure.

The overall mortality was 26%, we attributed improved survival rate to diligent overall care, especially early peripheral hyperalimentation and intensive respiratory therapy post-operatively.

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