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Abdominal Wall Defect : A 12 - Year Experience

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Abstract **During the period 1986-1997, 457 infants with abdominal wall defects were treated at the Queen Sirikit National Institute of Child Health. Their clinical data were reviewed. Omphalocele was diagnosed in 115 patients and gastroschisis in 342 neonates. Gastroschisis occurred in thrice as often as omphalocele and was increasing in frequency in recent years. The 71 percent incidence of prematurity in association with gastroschisis was significantly greater than the 51 percent incidence in omphalocele. The greatest hazard in gastroschisis was from small defects because of pressure necrosis and bowel perforation whereas in omphalocele, the greatest hazard was associated with large defects and severe anomalies. In omphalocele, non-intestinal viscus protruding through large defects was limited to the liver. In gastrochisis, the stomach, urinary bladder, uterus and adnexa eviscerated out of the large defects. Omphalocele was frequently associated with specific upper and lower midline anomalies, Beckwith - Wiedemann and trisomy syndromes (26.1%). These syndromes were not encountered in association with gastroschisis. Intracardiac malformations were more frequent in omphalocele (31.3%) than in gastroschisis (2.3%), and they were more complex and serious. Intestinal atresias were recorded in 15 neonates with gastroschisis but was not found in omphalocele. In contrast, 15 cases of imperforate anus occurred in omphalocele while they were not present in gastroschisis. Fourteen of the 15 patients were related to bladder or cloacal exstrophy and one case had imperforate anus only. Omphalocele and gastroschisis also differed with respect to gross appearances, clinical features and response to surgical therapy.**

Omphalocele and gastroschisis are the two most common forms of abdominal wall defect that require surgery during the neonatal period. Although both are congenital defects of the anterior abdominal wall, there are significant differences between the two conditions. Because of the rarity of the report of experience in Thailand, our most recent experience at the Queen Sirikit National Institute of Child Health were reviewed and reported herein.

Materials and Methods

Medical records of the 457 patients with omphalocele and gastroschisis who were admitted to the Department of Surgery at the Queen Sirikit National Institute of Child Health from 1986-1997 were reviewed. Information regarding modes of delivery, hospitals where the babies were born, the patients' condition at arrival, sex, birth weight, sizes of the

defects, eviscerated viscera and associated anomalies were collected. Additionally, types of management, outcomes of the treatment and complications were studied.

Results

Perinatal data

Of the 457 neonates with abdominal wall defects, 115 were omphalocele and 342 were gastroschisis. Among those with omphalocele, 45 were male, 66 were female and 4 had ambiguous genitalia. One hundred and sixty one cases of gastroschisis were male and 181 were female. The male to female ratio was 1:1.2 in both omphalocele and gastroschisis. Twenty three of the 115 neonates with omphalocele and 58 of the 342 babies with gastroschisis were born at Rajavithi Hospital while the remainders were born at other hospitals. During the 1986-1997 period, the total number of neonates born at Rajavithi Hospital was 218,816. The incidence of omphalocele and gastroschisis at Rajavithi Hospital were about 1:9,500 and 1:3,800 live births respectively. Number of the patients in each year was shown in Figure 1.

The average birth weight of infants with omphalocele was 2,680 gm and 58 of them (51%) were prematurity. Infants with gastroschisis had the average birth weight 2,230 gm and 243 of them (71%) had the birth weight less than 2500 gm. The defects of abdominal wall in omphalocele ranged from 2 to 13 cm, and the defects in gastroschisis was from 2 to 4 cm in diameter. Other perinatal data of the two groups of infants were summarized in Table 1.

Associated anomalies

Seventy two patients with omphalocele (62.6%) and 62 patients with gastroschisis (18%) were noted to have other associated congenital abnormalities. Major associated anomalies in omphalocele included cardiovascular malformations (31.3%), bladder or cloacal exstrophy (12.2%) and chromosomal abnormalities (8.7%). Pentalogy of Cantrell and Beckwith - Wiedemann (EMG) syndrome were also found in 3 cases each. Seventy five neonates with omphalocele (65.2%) had diameter of the defects between 2 - 5 cm and 40 of these 75 patients (40%) had serious associated anomalies. Forty cases of omphalocele (34.8%) had a defect larger than 5 cm in diameter and 36 of them (90%)

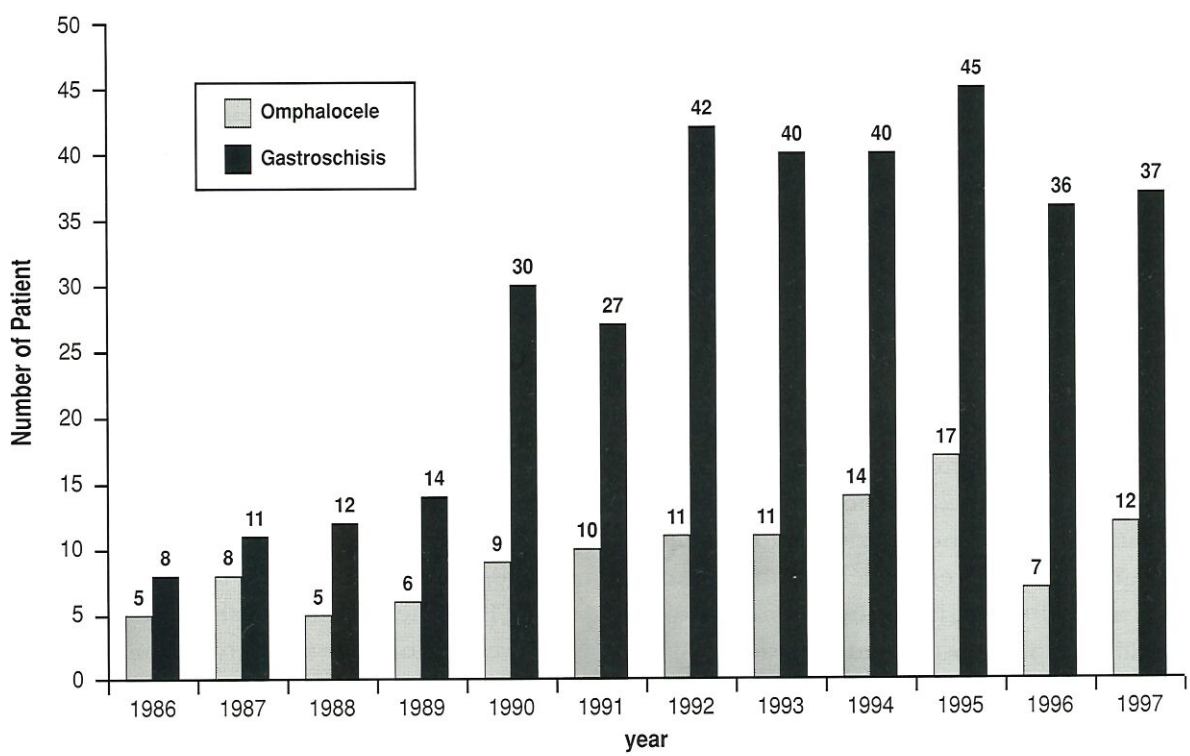

Fig. 1 Number of the patients admitted in each year during 1986 - 1987.

Table 1 Perinatal data from 457 patients with omphalocele and gastroschisis

Perinatal data	Omphalocele	Gastroschisis
Number of patients	115	342
Incidence (at Rajavithi Hospital only)	1:9,500	1:3,800
Male : female ratio	1:1.2	1:1.2
Average birth weight (gm)	2,680	2,230
< 2,500 gm.	58 (51%)	243 (71%)
≥ 2,500 gm	57 (49 %)	90 (29 %)
Vaginal delivery : Cesarean section	79:21	78:22
Location of the defects [related to umbilicus]	central	right
Size of defects (cm)	2-13	2-4
2-5 cm.	75 (65.2%)	342 (100%)
> 5 cm	40 (34.8 %)	-
Sac coverage	present	absent
Non-intestinal viscera herniation	liver	stomach, urinary bladder, uterus and adnexa
Intestinal characteristic	normal	matted and shortened

had serious congenital anomalies. Forty seven of the 58 low birth weight neonates with omphalocele (81%) had severe anomalies while only 29 of the 56 normal birth weight patients (51.9%) did so. The omphalocele patients who had a large defect and low birth weight were associated with severe life threatening anomalies more frequently than the patients with a small defect and normal birth weight ($p<.05$). In gastroschisis, gastrointestinal anomalies (13.5%) were the major problem. Forty four of the 243 patients with low birth weight (18.1%) had serious associated anomalies while 25 of the 99 patients with normal birth weight (15.2%) did so. There was no statistical difference between these two groups ($p>.05$). The details of associated anomalies were presented in Table 2.

Modes of treatment and outcome

Of the 115 patients with omphalocele, 11 neonates (9.6%) who had serious associated anomalies of poor prognosis were treated non-operatively by topical application of 2% mercurochrome or providine - iodine solution and nine of them (81.8%) died. Two neonates survived with complete wound healing. Seventy cases, who had diameter of the defects between 2-6 cm, underwent single - staged or primary repair of the abdominal wall defects and 17 of them (14.3%) died. Thirty four babies, who had the defects between 4-12 cm, underwent two-staged closure or silo reduction technique and 17 of them (50%) died. A silo or artificial sac used in this study was described by Havanonda.[1,2] It is made of stockinette and lined both inside and outside with Steri-Drape® (Figures 2,3). Forty three of the 115 patients with omphalocele died. The overall mortality rate of omphalocele was 37.4%. Twenty six of the 58 neonates with birth weight less than 2,500 gm (44.8%) died while 17 of the 57 neonates with birth weight over 2,500 gm (29.8%) died. Seventy five neonates had the defects of 5 cm or smaller and 19 of them (25.3%) died. Forty patients had diameter of the defects larger than 5 cm and 24 of them (60%) died. The patients with low birth weight and large defects had higher mortality rate than the patients with normal birth weight and smaller defects ($p<.05$).

Of the 342 patients with gastroschisis, one baby died before a definitive treatment could be given. Eighty one cases underwent primary fascial closure and 21 of them (25.9%) died. Two hundred and sixty patients were treated by two-staged closure and 62 of them (23.8%) died. The overall mortality rate of gastroschisis was 24.5 per cent (Table 3). Two hundred and forty three neonates had birth weight less than 2500 gm and 63 of them died. The mortality rate of the low birth weight patients was 25.9 per cent. Ninety nine neonates had normal birth weight and 21 of them died. The mortality rate of the normal birth weight patients was 21.2 per cent. There was no difference in mortality rate between premature and term neonates with gastroschisis ($p> .05$).

Table 2 Associated anomalies in omphalocele and gastroschisis.

Anomalies	Omphalocele (N=115)	Gastroschisis (N=342)
Cardiovascular	36 (31.3%)	8 (2.3 %)
VSD	5	1
ASD	6	1
TOF	6	-
PS	2	-
PDA	10	6
Miscelleneous	9	-
Diaphragmatic hernia	4 (3.5%)	-
Cleft lip and cleft palate	7 (6.1%)	-
Gastrointestinal	42 (36.5%)	46 (13.5 %)
Esophageal atresia	1	-
Persistent omphalomesenteric duct	8	7
Pyloric stenosis	1	-
Meckel's diverticulum	1	3
Intestinal atresia	-	12
Malrotation	7	15
Duplication	2	1
Imperforate anus	15	-
Gastroesophageal reflux	7	8
Genitourinary	21 (18.3%)	15 (4.4%)
Horse-shoe kidney	2	-
Hydronephrosis	1	-
Urachus	1	-
Undescended testis	3	15
Bladder or cloacal exstrophy	14	-
Neurological	7 (6.1%)	3 (0.9%)
Microcephaly	3	1
Meningomyelocele	3	-
Hydrocephalus	1	2
Limb	4 (3.5%)	4 (1.2%)
Pentalogy of Cantrell	3 (2.6%)	-
Beckwith - Wiedemann syndrome	3 (2.6 %)	-
Chromosomal	10 (8.7%)	2 (0.6 %)
Trisomy 13	3	-
Trisomy 18	4	-
Trisomy 21	3	2

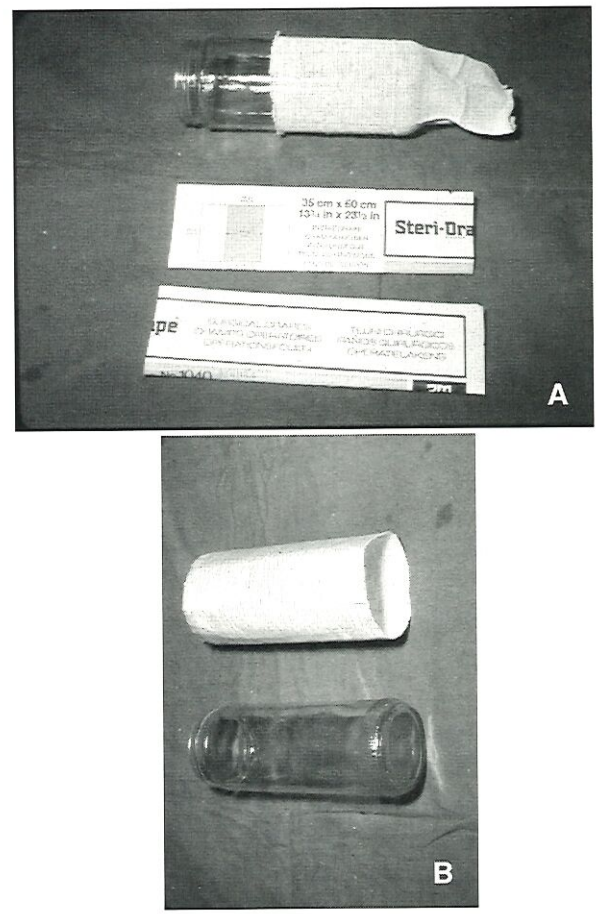

Fig. 2 A: Materials (stockinette and Steri - Drape) for making of an artificial sac.
B: A silo pouch after sealing of both sides with Steri - Drape.®

Common causes of death in omphalocele were severe associated anomalies, sepsis, pneumonia and heart failure. In gastroschisis, common causes of death included sepsis, pneumonia , necrotizing enterocolitis (NEC) and intestinal necrosis (Table 4). Ten patients developed bowel necrosis, 4 associated with small defects and pressure necrosis, 3 due to malrotation and postoperative volvulus and 3 associated with NEC. Prolonged paralytic ileus was noted in gastroschisis more frequently than in omphalocele. Almost all of the patients of both omphalocele and gastroschisis had parenteral nutritional support starting within the second postoperative day until they could well tolerate oral feeding.

Discussion

The etiology of omphalocele is generally thought to involve the failure of the abdominal viscera to return to the abdominal cavity at the end of the 10th week after gestation[3,4]. The etiology of gastroschisis is postulated to be due to a localized teratogenic interference with the differentiation of somatopleuric mesenchyme with consequence reabsorption of the overlying

Table 3 Results of the treatment

Procedures	Omphalocele			Gastroschisis*		
	Patients	Deaths	Mortality (%)	Patients	Deaths	Mortality (%)
Non-operative	11	9	81.8	-	-	-
Operative						
Primary closure	70	17	24.3	81	21	25.9
Staged operation	34	17	50	260	62	23.8
Total	**115**	**43**	**37.4**	**341**	**83**	**24.3**

**One case died before a definitive treatment could be given.*

Table 4 Complications in omphalocele and gastroschisis

Complications	Oomphalocele		Gastroschisis	
	No	Per-cent	No	Per-cent
Sepsis	24	20.9	86	25
Congestive heart failure	23	20	8	2.3
Pneumonia	20	17.4	61	17.8
Respiratory distress syndrome	5	4.3	4	1.2
Hyperbilirubinemia	15	13.0	34	9.9
Prolonged paralytic ileus	8	7.0	52	15.2
Necrotizing enterocolitis	2	1.7	11	3.2
Bowel necrosis	-	-	10	2.9
Enterocutaneous fistula	-	-	9	2.6
Postoperative hepatic hemorrhage	3	2.6	1	0.3
Artificial sac separation	4	3.4	-	-
Wound infection	16	13.9	31	9.1
Wound dehiscence	1	0.8	6	1.7

ectoblastic layer and failure of the muscular migration from the dorsal myotomes completely to invade the splanchnopleure of the embryogenic abdominal wall.[3,4] de Vries[5] suggested that the defects in gastroschisis was the result of infarction due to disturbances in the circulation to the somatopleure at its junction with the body stalk during involution of the right umbilical vein. The gross appearances of the two diseases are obviously different. In omphalocele, the defects involve the central portion of umbilicus and they are covered by a two-layer gelatinous sac from ammion and peritoneum (Figure 3). Almost all cases of gastroschisis, the defects are located to the right side of umbilicus without membranous sac coverage and they are separated from the umbilicus by a small skin bridge (Figure 4). According to the pathogenesis and the clinical findings, omphalocele was classified into cephalic (epigastric), central and caudal (hypogastric) types[3] while gastroschisis was classified into antenatal and perinatal types.[6]

Several authors reported that gastroschisis occurred twice as often as omphalocele did.[7-11] Our data from this study (342 cases of gastroschisis versus 115 cases of omphalocele) revealed that gastroschisis was approximately thrice of omphalocele and it appeared to be increasing in frequency in recent years despite a falling birth rate (Figure 1). The similarities between the two diseases in our patients were the occurrence of sex ratio. Female was slightly predominant to male (1.2:1) in both omphalocele and gastroschisis and this ratio was in contrast to that in the other reports.[12-16] Some series showed that vaginal delivery and cesarean

Fig. 3 Omphalocele with membranous sac and umbilical cord on top.

Fig. 4 Gastroschisis with eviscerated bowel and intact umbilicus.

section had no difference in morbidity and mortality of neonates with abdominal wall defects.[17-19] Cesarean section should be performed when they have proper indications.

The occurrence of prematurity was found in about 30-50 per cent of omphalocele in most reports.[7,9,10,12-14] Prematurity was a frequent finding in cases of gastroschisis, approximately 60-70 per cent, and its incidence was cited in virtually all reports related to gastroschisis.[13,15,16] The greater incidence of prematurity in association with gastroschisis in this study was statistically sigmificant ($p<.05$). In omphalocele, birth weight was considered by many authors to be an important prognostic factor.[17,20-23] Our data from this study showed that omphalocele with low-birth weight frequently had severe associated malformations. In contrast, patients born with gastroschisis seemed to fare as well whether they had low-birth weight or normal birth weight because these patients had few life threatening associated anomalies. It appeared that the important prognostic factor was not the birth weight itself but it was based on the associated malformations.

Table 5 Specific syndromes in omphalocele

Syndromes	
A. Upper midline (Pentalogy of Cantrell)	1. epigastric omphalocele 2. anterior diaphragmatic hernia 3. sternal cleft 4. ectopia cordis 5. intracardiac anomalies
B. Beckwith - Wiedemann	1. omphalocele 2. macroglossia 3. gigantism 4. pancreatic islet cell hyperplasia 5. hemihypertrophy
C. Low midline	1. bladder or cloacal exstrophy 2. imperforate anus 3. colonic atresia 4. vesicointestinal fissure 5. sacral vertebral anomalies 6. meningomyelocele
D. Trisomy	1. trisomy 13 2. trisomy 18 3. trisomy 21

The defects of abdominal wall in omphalocele were larger than in gastroschisis. Small defects in omphalocele were quite benign in comparison to gastroschisis because they could be easily treated by primary closure and rarely had associated malformations. In contrast, the small defects in gastroschisis were associated with a high incidence of pressure necrosis and perforation of eviscerated intestines.[7,24,25] We found this complication in 4 cases from this study. Large defects in omphalocele frequently had associated severe malformations which caused a high mortality rate. The contents in omphalocele with large defects were limited to intestines and liver whereas stomach, urinary bladder, uterus and adnexa eviscerated through the large defects of gastroschisis. Because of the absence of sac coverage over the defects, gastroschisis tended to have heat, fluid and electrolyte losses and greater risk of bacterial contamination on the exposed bowels as compare to omphalocele.[6,9] The eviscerated bowels in omphalocele were normal in appearance because of having sac coverage but the frequently eviscerated bowel in gastroschisis became thickened, shortened and formation of matted loops in varying degree. Spontaneous reversal of the bowel shortening wonderfully occurred after the return of the intestines into the peritoneal cavity for a short period of time.

Omphalocele and gastroschisis sharply differed in the incidence of associated malformations. Grosfeld et al[26] summarized severe anomalies in omphalocele including upper and lower midline syndromes, Beckwith-Wiedeman syndrome and chromosomal abnormality (Table 5). Cardiac defects were more complicated and serious in cases of omphalocele than in cases of gastroschisis. Tetralogy of Fallot, ventricular and atrial septal defects were common in our patients. This was also true in other reports.[9,13,17,26] Intestinal atresia and malrotation were the serious anomalies in gastroschisis. We found that 3 of the 15 cases with malrotation developed volvulus and bowel gangrene after placement of the artificial sac. This serious complication increased the mortality of the patients with gastroschisis.

Omphalocele could be treated conservatively by topical painting with mercurochrome or providine iodine solution. This technique was used in the cases with severe associated anomalies with supposedly poor prognosis. On the other hand, application of these escharotic agents could not be used in gastroschisis because of absence of sac coverage. As was stated by many authors,[12-15,22] primary closure should be done when possible. From this study, primary fascial closure was done in 61 per cent of omphalocele and 24 per cent of gastroschisis. These figures suggested that all cases of omphalocele with small defects could be closed primarily but it could not be done in gastroschisis with the same size of defects because of visceroabdominal disproportion. These resulted in the higher percentage of using two-staged operative technique in gastroschisis than in omphalocele. The results of primary closure in omphalocele and gastroschisis from this study were not significantly different ($p>.05$) but the results of staged operation in gastroschisis was better than in omphalocele ($p<.05$).

The overall mortality rate of gastroschisis was lower than that of omphalocele. The high incidence of severe malformations, particularly cardiac anomalies and other syndromes, remained the main mortality factors for infants born with omphalocele. Fewer serious anomalies may contribute to lower mortality in patients with gastroschisis. Intestinal necrosis, sepsis and prolonged paralytic ileus appeared to be the major hazard in infants with gastroschisis. Many factors can improve the outcome of the surgical management of gastroschisis. These included early protection against heat and fluid losses, protection against bacterial contamination and the rapid transport of these neonates to pediatric surgical center. Prolonged hypofunction of the intestine could be managed with delay in the initiation of oral feeding and administration of parenteral nutritional support.

Summary

This report was a retrospective study of 457 neonates born with abdominal wall defects. One hundred and fifteen had omphalocele and 342 had gastroschisis. The clinical data were analyzed to assess the differences between the two diseases. The obvious differences were the incidence, prematurity rate, gross pathology of the defects and eviscerated viscera. Severe associated cardiac anomalies, chromosomal and syndromic abnormalities occurred in omphalocele more frequently than in gastroschisis. These explained why the result of treatment in gastroschisis was better than that of omphalocele. Poorer prognosis for the

patients with omphalocele was related to the higher incidence of associated structural and chromosomal anomalies. In infants with gastroschisis, outcome was primarily determined by the condition of the exteriorized bowel.

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