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The factors affecting survival in newborns with omphalocele and gastroschisis *

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Summary

A survey of 52 newborns witn abdominal wall defects was carried out to distinguish the factors which possibly increase mortality. There were 36 newborns with omphalocele (21 boys, 15 girls) and 16 newborns with gastrochisis (7 boys, 9 girls). The mortality rates were 50 % in omphalocele group and 56.2 % gastrochisis group. In omphalocele group mortality was higher in patients with associated major congenital anomalies (75 %), Large defect (83.3 %) and patients treated conservatively (83.3 %). In the gastrochisis group delayed referral greatly increased mortality (85.7 %). We conclude that the main factors relative to survival are the size of defect and associated major anomalies for omphalocele, and the improper early management of newborns for gastrochisis.

Key words: *Omphalocele, gastroschisis, mortality*

Introduction

Congenital abdominal wall defects include eviscerated internal organs not covered by skin [14]. There are two distinct types: Omphalocele and gastroschisis. Until late 1960s, many children born with these defects did not survive.

Children with this condition are expected to lead normal lives today, in the absence of associated major congenital malformations or syndromes [7,8,15].

Certain perinatal factors may increase the mortality rate of these patients. The purpose of this study is to determine the factors affecting survival in newborns with omphalocele and gastroschisis.

Materials and Methods

The charts of the newborns with omphalocele and gastroschisis treated at the Department of Pediatric Surgery between February 1987 and February 1995 were reviewed retrospectively. Primary diagnosis, sex, gestational age, maternal age, parity, period between birth and admission to the hospital, associated congenital anomalies, the size of the abdominal defect, the extent of evisceration, management modalities and outcome were assessed.

Treatment was planned regarding the size of defect and/or presence of associated major anomalies in the newborns with omphalocele. A conservative approach was preferred if the diameter of abdominal defect was greater than 7 cm and in the presence of any life-threatening associated anomaly. The primary repair of defect was performed immediately in all other patients with omphalocele and in newborns with gastroschisis.

The significance of the differences between the two groups were assessed by Fischer's Exact test. Differences between two groups were considered significant if $p<0.05$.

Results

There were 36 newborns with omphalocele (group 1) and 16 newborns with gastroschisis (group 2). The results are summarized in the Table I. Gastroschisis was more common in very young or primiparous mothers ($p<0.01$ for both), and the ratio of prematurity was significantly higher in this group ($p<0.0001$). The mortality rates of premature newborns were similar in both groups.

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Tablo I. Factors relative to mortality in newborns with abdominal wall defects

	Omphalocele (group 1)				Gastroschisis (group 2)			
	Patients		Mortality		Patients		Mortality	
	n	%	n	%	n	%	n	%
Total numbers of patients	36	69.2	18	50.0	16	30.8	9	56.2
Sex: boys	21	58.3			7	43.7		
girls	15	41.6			9	56.2		
Maternal age								
20>	33	91.7			9	56.2		
20<	3	08.3			7	43.8		
Parity								
1.	6	16.6			9	56.2		
2>	30	83.3			7	43.8		
Gestation								
Premature	3	08.3	1	33.3	8	50.0	3	37.5
Mature	33	91.7	17	51.5	8	50.0	6	75.0
Place of birth								
in province	23	63.9	13	56.5	10	62.5	5	50.0
out province	13	36.1	5	38.4	6	37.5	4	66.6
Period between birth and admission								
0-6 hrs	16	44.4	11	68.7	9	33.3	3	33.3
6-24 hrs	13	33.3	7	53.8	7	66.6	6	85.7
24 hrs>	7	22.2	-		-			
Assoc. anomalies								
major	8	22.2	6	75.0	2	12.5	-	
minor	1	02.7	-					
chromosomal	3	08.3	-					
none	24	66.6	12	50.0	14	87.5		
Size of defect								
7 cm<	24	66.6	8	33.3				
7 cm>	12	33.3	10	83.3				
Management								
surgical	30	83.3	13	43.3				
conservative	6	16.6	5	83.3				

All of the patients with gastroschisis presented within the first 24 hours after birth, but 7 patients in this group presented later than the first 6 hours. Time period between birth and admission to the hospital; and place of birth had no influence on mortality ($p>0.05$).

Eight patients with omphalocele (22.2 %) had major associated anomalies (4 VSD 2 ASD, one multicystic kidney and one type 3 ileal atresia) and 2 of them (25 %) survived. In addition, one patient had minor associated defect (pes equinovarus) and 3 (8.3 %) had chromosomal anomalies (Down's syndrome) in this group. Two patients had major associated anomalies (type 2 and 3 ileal atresia) in group 2, but no mortality occurred in these patients.

Total mortality rate was 50 % in group 1, and 56.2 % in group 2. In group 1, mortality rate was 33.3 % in patients with small defects (<7 cm), and 83.3 % in patients with defects greater than 7 cm in radius ($p<0.05$). Also in this group, mortality rate was 43 % in surgically treated patients, and 83.3 % in conservatively treated patients ($p<0.05$). The death in surgically treated patients was due to associated anomalies (5 patients) and sepsis (3 patients). In 6 patients with large defects, surgical treatment was performed in group 1, and 5 of them died. The causes of death in these patients were cardiorespiratory embarassment (4 patients) and sepsis (1 patient). In conservatively treated patients with omphalocele, the causes of death were associated anomalies (3 patients) and sepsis (3 patients).

Discussion

The incidence of omphalocele is about 1 in 4000 live births, while this rate is 1 in 10000 for gastroschisis [3,5-9,11-13]. In the past decade, gastroschisis was reported in increasing numbers, but the rates

for omphalocele remained relatively unchanged [12]. Whether this trend is a selection of diagnoses or represents an actual increase in rate of gastroschisis is uncertain [14]. Race, maternal age, parity, and social habits have been suggested to influence the relative incedences of omphalocele and gastroschisis. It is difficult to support the role of these factors on the incidence of abdominal wall defects with current data [14]. The occurence of gastroschisis in very young primiparous mothers begs explanation [7,12]. In our series, 7 patients with gastroschisis (43.8 %) had a mother younger than 20 years of age and 9 (56.2 %) were born in first gestation.

Transportation, if improper, may lead to hypothermia and dehydration and increase the risk of sepsis [9,12]. In our series, mortality of patients received in first 6 hours after birth was significantly lower in group 2. The overall low mortality of gastroschisis babies born in our province underline the significance of these factors. Naturally the risks of contamination and dehydration are much less in omphalocele.

Associated anomalies are known to occur in a greater percentage of newborns with omphalocele (30-66 %) than gastroschisis (5-24 %) [3,6,7,11]. Fetuses with omphalocele have a high mortality due to the frequency of concurrent malformations (50-70 %) and chromosome abnormalities (30-40 %) [3,4,6]. The commonest associations with omphalocele are cardiovascular, anorectal, genitourinary anomalies, musculoskeletal deformities and Beckwith-Wiedeman syndrome [8,11,12]. In this series of patients cardiovascular anomalies in omphalocele had a comparable incidence (75 %) and mortality of these patients were 75 %. The incidence of associated anomalies (12.5 %) for group of gastroschisis patients is in the predicted range [8,9,12].

The contents of the omphalocele may be an indicator of the karyotype: The absence of liver in the omphalocele was strongly associated with an abnormal karyotype [10]. The observations of Benacerraf et al [2] are in compliance with our findings. Three patients with Down's syndrome in our series had small defects and their liver had not herniated.

Treatment of giant omphaloceles is difficult as a result of the disproportion between the volume of eviscerated organs and the relatively volume of the abdominal cavity. This disproportion has a prognostic importance [1,13,14]. The prognosis of newborns with giant omphaloceles is poor. The outcome in these newborns is influenced particularly by the type of malformations frequently found in association. Technical difficulties in management also add to morbidity.

The mortality rate in 12 newborns with giant omphaloceles were 83.3 % in our series. The main factors that determine outcome in gastroschisis are prematurity and intestinal malformations [8,12]. However, half of the patients in our group 2 were premature, but their mortality (37.5 %) were no higher than the cumulative mortality rate of this group (50 %). This may be attributed to the absence of intestinal malformations in these patients.

In summary, we conclude that the main factors affecting survival were the size of defect and life-threatining associated anomalies for omphalocele, and the conditions of initial management and transport for gastroschisis.

References

1. Bax NMA, Zee DCV, Gunne AJP, Rövekamp MH: Treatment of giant omphalocele by enlargement of the abdominal cavity with a tissue expander. J Pediatr Surg 28:1181, 1993
2. Benacerraf BR, Saltzman DH, Estroff JA, Frigoletto FD: Abnormal karyotype of fetuses with omphalocele: Prediction based on omphalocele contents. Obstet Gynecol 75:317, 1990
3. Brown DL, Emerson DS, Shulman LP, Carson SA: Sonographic diagnosis of omphalocele during 10th week of gestation. AJR 153:825, 1989
4. Curtis JA, Watson L: Sonographic diagnosis of omphalocele in the first trimester of fetal gestation. J Ultrasound Med 7:97, 1988
5. Festen C, Severijnen RSVM, Staak FHJM: Gallbladder embedded in an accessory liver lobe in umbilical cord hernia. J Pediatr Surg 23:978, 1988
6. Hughes MD, Nyberg DA, Mack LA, Pretorius DH: Fetal omphalocele: Prenatal US detection of concurrent anomalies and other predictors of outcome. Radiology 173:371, 1989
7. Lafferty PM, Emmerson AJ, Fleming PJ, et al: Anterior abdominal wall defects. Arch Dis Child 64:1029, 1989
8. Loder RT, Guiboux JP: Musculoskeletal involvement in children with gastroschisis and omphalocele. J Pediatr Surg 28:584, 1993

9. Moretti M, Khoury A, Rodriguez J, et al: The effect of mode of delivery on the perinatal outcome in fetuses with abdominal wall defects. Am J Obstet Gynecol 163:833, 1990
10. Nyberg D, Fitzsimmons J, Mack L, et al: Chromosomal abnormalities in fetuses with omphalocele: Significance of omphalocele contents. J Ultrasound Med 8:299, 1989
11. Sugarman ID, Malone PS: Exomphalos major, oesophagel atresia and tracheooesophageal fistula- A rare combination of major malformations. Eur J Pediatr Surg 4:178, 1994
12. Torfs C, Curry C, Roeper P: Gastroschisis. J Pediatr 116:1, 1990
13. Tucci M, Bard H: The associated anomalies that determine prognosis in congenital omphaloceles. Am J Obstet Gynecol 163:1646, 1990
14. Tunnel WP: Omphalocele and gastroschisis in Ashcraft KW, Holder TM (eds): Pediatric Surgery, WB Saunders Co, Philadelphia, Pennsylvania 1993, p.546
15. Yokomori K, Ohkura M, Kitano Y, et al: Advantages and pitfalls of amnion inversion repair for the treatment of large unruptured omphalocele: Results of 22 cases. J Pediatr Surg 27:882, 1992