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Original Article

Clinical Outcomes of Omphalocele: An Analysis of 124 Patients

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Abstract

Background: **Omphalocele is defined as an anterior midline defect of the abdominal wall through which various viscerae herniated into an avascular sac. Currently, the mortality rate is not decreasing because of high incidence of severe associated abnormalities.**

Objectives: **The aim of this study was to review long-term outcomes of patients with omphalocele in the recent 10-year period.**

Materials and Methods: **Medical records of patients with omphalocele who were treated at the Queen Sirikit National Institute of Child Health (QSNICH) during 1998-2007 were collected. Patient's data including demographics, modes and results of the treatment were collected and analyzed.**

Results: **One hundred and twenty-four babies were treated for omphalocele at the QSNICH during the study period. An incidence of omphalocele among neonates born at the Rajavithi hospital was about 1:2,800 live births. Sizes of the omphalocele defects ranged from 2 to 13 cm. in diameter (average 4.5 cm.). Musculoskeletal and cardiovascular defects were the most common abnormalities in this study. Eight patients succumbed before the definitive treatment. Twenty-two patients (17%) who had severe associated anomalies and suspected incompatible with life were conservatively treated by topical antiseptic therapy. Only 2 of the 22 neonates (9%) survived. Ninety-four patients were operated on and primary fascial closure was feasible in 61 cases (49%). Forty-four of the 61 patients (72%) survived. The duration from closure to the first oral feeding ranged from 7 to 14 days. The remaining 33 neonates (27%) with omphalocele defect larger than 6 cm. in diameter were managed by staged operations and only 21 patients (63.6%) survived. Of the total 124 patients, 67 cases survived. The overall survival rate was 54%. Fifty-seven patients (41%) died because of severe associated anomalies and related complications including sepsis, pneumonia and congestive heart failure.**

Conclusions: **The important factor which influences the mortality of omphalocele is severe associated anomalies. The survival rate might be improved if these anomalies could be corrected.**

Keywords: abdominal wall defect, congenital anomalies, omphalocele

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Introduction

Omphalocele is defined as an anterior midline defect of the abdominal wall through which various viscera herniated into an avascular hernial sac. Omphalocele is also known as exomphalos[1], with an incidence of 17 per 100,000 or 1:5,880 live births in Western countries[2]. Unlike gastroschisis, the herniated abdominal organs are always covered in a protective membrane (Figure 1). The umbilicus inserts into the sac[3]. There are unknown specific risk factors. In the literature, several theories have been proposed to explain the pathogenesis of omphalocele by many authors[4-8]. According to Gray and Skandalakis[9], omphalocele is the result of developmental arrest at the time of physiologic herniation of the gut into the celomic cavity of the umbilicus. The defect involves the central portion of umbilicus and is covered by a two-layer gelatinous sac. The outer surface is derived from amnion and the inner layer is the derivative of the peritoneum. The content between these two layers is composed of Wharton's jelly. Duhamel[10] classified omphalocele into cephalic (epigastric), central, and caudal (hypogastric) types. The size of the defects varies from small (2-3 cm) to large size (>10 cm). Abdominal viscera within the sac commonly consists of the intestines, stomach, liver, and urinary bladder. The mortality rate of omphalocele is not decreasing because of the high incidence of severe associated abnormalities such as Beckwith-Wiedemann syndrome, cloacal exstrophy, OEIS complex (omphalocele, exstrophy, imperforate anus and spinal anomalies), Cantrell's pentalogy and trisomy syndrome[11,12].

Omphalocele can also be diagnosed prenatally by ultrasonography with a sensitivity of 75-79 %[11]. Prenatal diagnosis is advisable because of the associated syndromes. Cardiac anomalies are described in 13-15%[2,13]. Fetuses with omphalocele appear to have an increased risk not only of congenital heart disease (CHD) but also of perinatal cardiac abnormalities, especially persistent pulmonary hypertension of the neonates (PPHN)[14]. Operative technique for repair is similar to that for gastroschisis. During the subsequent decade, primary repair of the abdominal wall is performed when feasible, but staged repair is utilized when needed to prevent increased abdominal pressure.

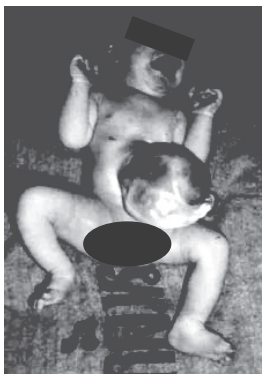

Figure 1

Although there are improvements of antibiotics, mechanical ventilatory support and parenteral nutritional administration in this era, but results of the treatment in omphalocele are not satisfactory because of severe associated anomalies. The purpose of this report was to review the results of management of infants with omphalocele seen in the 10-year period, from January 1998 through December 2007 and to compare the survival rate of the previous report in 1998 at our institution[15].

Materials and Methods

Medical chart records of patients with omphalocele who were admitted at the Queen Sirikit National Institute of Child Health (QSNICH) during 1998-2007 were reviewed. The following data were obtained: birth weight, types of delivery, associated anomalies, modes of treatment, postoperative therapy and clinical outcomes of the patients. The study was performed after permission from the Institutional Review Board of QSNICH (Document No. 51-060).

Results

Incidence and perinatal data

One hundred and twenty-four babies with omphalocele (60 males and 64 females) were available for the study. Male and female ratio was 1:1.07. Of the 124 neonates, 32 had been born at the Rajavithi Hospital and the others had been born at other hospitals. During the study period, 89,070 neonates were delivered at the Rajavithi Hospital. An incidence of omphalocele among neonates born at the Rajavithi Hospital was about 1:2,800 live births.

Birth weights varied from 950 to 4,300 g. (average 2,630 g.). Prematurity was noted in 57 patients (45%). The sizes of the omphalocele defects ranged from 2 to

13 cm in diameter (average 4.5 cm). In the vast majority of cases, eviscerated viscera in the sac included intestines, stomach and liver. In the cases with lower midline defects, exstrophy of bladder or cloaca was common. Seventy-three of the 124 neonates (58%) were born by transvaginal delivery while the remaining 49 babies(39%) were delivered by cesarean section. Two neonates were noted to have rupture of the omphalocele sac before their admissions to QSNICH.

Associated anomalies

Musculoskeletal defects were the most common anomalies in this study (Table 1). These abnormalities included clubfeet, syndactyly, polydactyly and limb dysplasia. Congenital heart diseases (CHD) were the second most common anomalies including ventricular septal defect (VSD), atrial septal defect (ASD), tetralogy of Fallot (TOF), pulmonary artery stenosis (PS) and patent ductus asteriosus (PDA). Chromosomal abnormalities were found in 7 neonates and all of them were proven to have trisomy[13]. Neurological anomalies were noted in 6 patients with microcephaly, myelomeningocele and spinal defect into 2 cases of each anomaly. Beckwith-Wiedemann or EMG syndrome characterized by omphalocele (exomphalos), macroglossia, gigantism and pancreatic islet cell hyperplasia were present in 2 patients. One neonate had the most severe anomaly of pentalogy of Cantrell including epigastric omphalocele, anterior diaphragmatic hernia, sternal cleft, ectopia cordis and intracardiac anomalies (Figure 2).

Table 1 Omphalocele with associated anomalies (n = 124)

Associated anomalies	No. of patients (case)	Percentage (%)
Musculoskeletal	31	25
Cardiac	26	21
Genitourinary	12	9.7
Gastrointestinal	10	8.1
Chromosomal	7	5.6
Neurological	6	4.8
Others	4	3.2

Figure 2

Modes and results of treatment

Eight neonates (6.5 %) succumbed within 3 hours after admission. They were in moribund status during arrival with endotracheal intubation due to congestive heart failure and respiratory failure from CHD and other severe associated anomalies. All of the 8 neonates died before the definitive treatment. Twenty two patients (17.7%) with severe associated anomalies such as CHD, trisomy 13, pentalogy of Cantrell were conservatively treated by topical therapy. The omphalocele sacs were daily painted with povidone iodine solution. Only 2 of the 22 patients (9%) survived (Table 2).

Primary fascial closure of the abdominal wall defects was feasible in 57 patients. The defects varied from 2 to 6 cm. in diameter (average 3.5 cm.) The survival rate of primary fascial closure was 72.1% (44 in 61 patients). The interval from primary closure to the first oral feeding was noted in 7-14 days (average 9 days). The remaining 33 cases with abdominal wall defects over 6 cm. in diameter were managed by staged closure or silo-reduction technique. We used an artificial sac or silo pouch created by Havanonda[16]. This artificial sac was prepared by using Steri-Drape® covering both surfaces of a stockinette silo (Figure 3, 4). The time period from placement of artificial sac to abdominal defect closure was within 7 days (Figure 5). The interval from abdominal closure to the first oral feeding varied from 8 to 20 days (average 14 days). Only 21 of the 33 patients survived. The survival rate of the staged operation was 63.6%.

Of the total 124 patients, 67 cases survived. The overall survival rate of patients with omphalocele in the present study was only 54%. Causes of death were related to severe associated anomalies and severe complications including pentalogy of Cantrell, trisomy[13], CHD, respiratory distress syndrome (RDS), pulmonary hypoplasia and pulmonary hypertension, septicemia, pneumonia and congestive heart failure. Of the 6 cases with exstrophy of cloaca, 2 died before

Table 2 Modes of treatment and results

Procedures	No. of patients (cases)	Survivors (cases)	Survival rate (%)
Non-operative			
Povidine-iodine application	22	2	9
Operative			
Primary closure	61	44	72.1
Staged operation	33	21	63.6
Death before treatment	8	0	0
Total	**124**	**67**	**54**

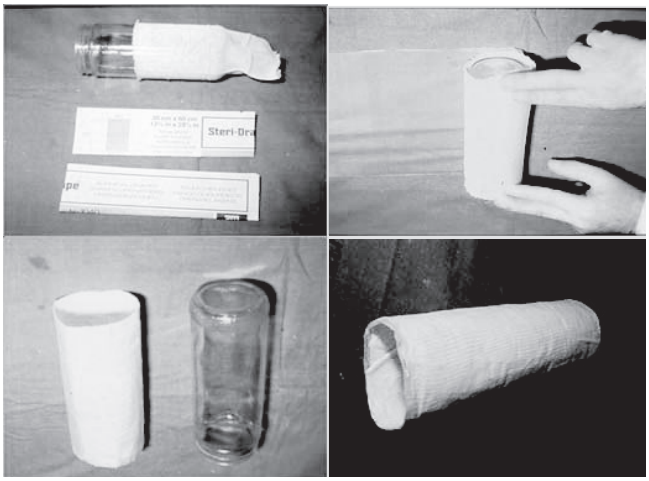

Figure 3

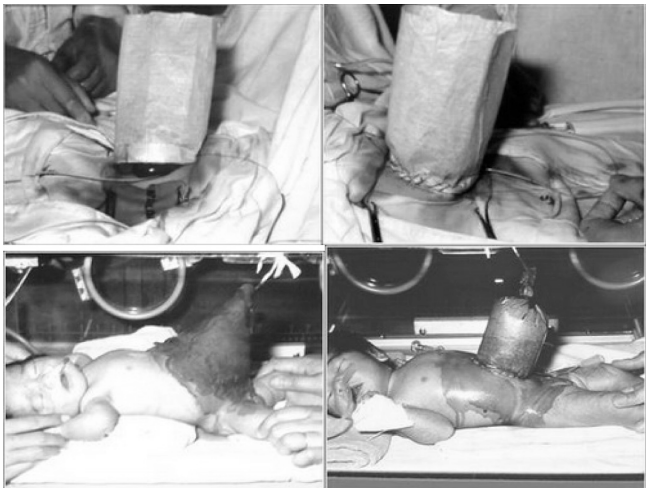

Figure 4

the definitive surgical treatment. The remaining 4 cases underwent surgical correction of cloacal exstrophy and primary fascial closure and only 2 cases survived.

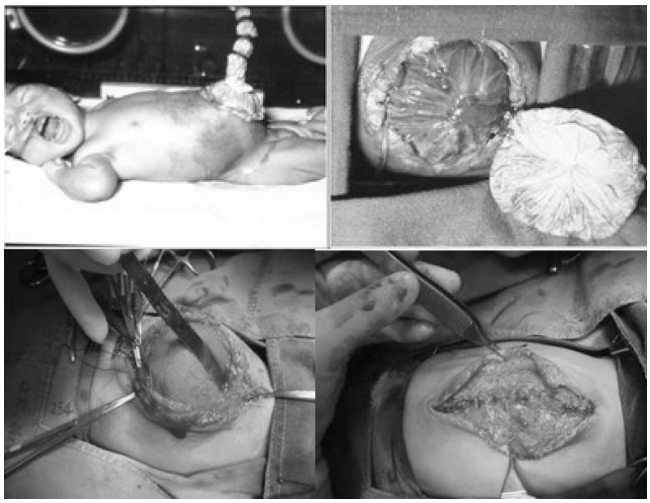

Figure 5

Discussion

The incidence of omphalocele occurred in approximately 1 of 6,000-10,000 live births in Western countries[2,17]. From the present study, the occurrence rate or incidence of omphalocele was about 1:2800 live births comparing with 1: 9,500 liver births in the previous study at our institute[15]. The number of cases of omphalocele in each year has been steady in QSNICH while the incidence from Rajavithi Hospital has been increased each year as a result of modern prenatal sonogram. Mothers with antenatally diagnosed omphalocele fetuses were increasingly referred to have delivery at the tertiary care center as Rajavithi Hospital. Data from the previous study of our institute (1983-1989 and 1986-1997)[16,17] indicated that approximately 10 cases with omphalocele were admitted per year. This incidence has not changed in the recent years.

Knight et al[17] reported that prognostic factors of omphalocele included size of the defect, preoperative

rupture of the sac, delay in treatment, low birth weight and the presence of associated anomalies. Prematurity was considered by Stringel and Filler[18] to be an important prognostic factor because of the presence of severe associated anomalies in the low birth weight neonates. It is indicated that the important prognostic factor of omphalocele is not only the low birth weight but also the severe associated anomalies[19]. The life-threatening abnormalities included cardiovascular, chromosomal and cranionervous system anomalies[18,19]. The syndromes in which an omphalocele is a common component are pentalogy of Cantrell, Beckwith-Wiedemann syndrome, the lower midline and trisomy 13 and 18 syndrome[20,21].

Because of severe anomalies in our patients, we reserved non-operative treatment with escharotics for those whose survival was not expected. Povidone-iodine solution is used instead of 2% mercurochrome in this era in order to prevent the hazard of mercury toxicity. The primary goal of omphalocele therapy is the successful and satisfactory enclosure of the exposed viscera. It is clear that primary abdominal closure whenever possible is the ideal treatment. When this cannot be achieved, the second choice would be the staged-operative procedure by using artificial sac or silo. This technique remains a good procedure for those whose skin closure cannot be achieved primarily. The precaution for operative treatment , either the primary closure or the staged operation, is to leave some parts of the omphalocele sac adherent to the liver capsule in order to avoid postoperative hepatic hemorrhage[22].

Our data from the present study revealed that the patients who were treated by primary fascial closure had the survival rate higher than those treated by staged-operative procedure ($p < 0.05$). This was probably due to the smaller defect and rarer severe associated anomalies in the former group. The size of the defect was an important prognostic factor for the survival whereas preoperative rupture of the omphalocele sac was another high prognostic factor for mortality. The overall survival rate of 54% in this study was significantly lower than that of 62 % in the previous study[15] ($p < 0.005$) because of having more severe associated anomalies than those in the previous study period.

Prenatal diagnosis of omphalocele should not automatically dictate the need for cesarean section. Suita et al[23] demonstrated no effect of the mode of delivery on the prognosis of omphalocele. Elective cesarean section should be indicated after antenatal ultrasound revealed the presence of a giant omphalocele containing part or all of the liver. Prenatal ultrasonography is useful in detection of other malformations so that the overall prognosis can be evaluated earlier. Improvement in the case of neonates born with omphalocele will have to come from better management of the associated anomalies.

Conclusions

Our data from the present study suggested that mortality rate of patients with omphalocele is usually higher in low birth weight infants with severe associated malformations and in those having large defects of abdominal wall. Selection of modes of treatment should be based on assessment of severe associated anomalies and abdominovisceral proportion in each patient. The survival rate of patients with omphalocele might be improved if these anomalies are less severe and feasible for correction.

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