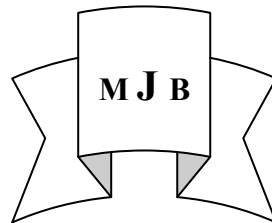


A Study of 40 Cases of Omphalocele

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Abstract

Background: Congenital abdominal wall defects (probably omphalocele) were described as early as the first century. Successful repair of omphalocele was first reported in 1803.[1] Excising sac and covering the viscera with skin flaps was described in 1948[2]. In 1967, the staged repair of omphalocele with artificial material sewn to the facial edges was introduced [3].

Objectives: To evaluate the prenatal diagnosis, post delivery management including surgical correction and prognosis, in Iraq.

Setting: All of our patients were managed in pediatric surgery center at children welfare teaching hospital in Baghdad between may 2004 to may 2007.

Patients and methods : The medical records of our patients were recorded for prenatal diagnosis, birth order, sex, weight, type of delivery, maternal age, gestational age, size of the defect, associated anomalies, treatment and out come.

All patients had been examined to exclude associated congenital anomalies.

All patients investigated for blood group, blood glucose and urea, echo-cardiography and ultra sound of abdomen done for them. Vitamin K were given to all patients as 1mg/kg in single dose.

Results: forty full term patients, twenty male and twenty female. Diagnosis of all of patients were done clinically by inspection, while ten of the patients were diagnosed prenately by ultrasound examination(25%).

six of our patients(15%) had C\S delivery while the rest were product of normal vaginal delivery. Only two patients(5%) had rupture of the sac at presentation. thirty patients(83.4%) treated successfully by primary closure, six patients (16.6%) treated by staged closure. four patients(10%) died preoperatively secondary to sepsis because of low birth weight and associated anomalies, two patients(5%) died postoperatively for the same reasons.

Conclusion: All our patients were full term infants. No difference in male to female ratio. Omphalocele minor not interfere with birth weight. C\S have no additional benefit over normal vaginal delivery. Mortality is high in patients have major associated anomalies or big defect. The treatment is surgical include removal of the sac and primary closure or staged closure. The post operative period was smooth for 90% of the patients.

دراسة مستقبلية لأربعين حالة من قروء السرة الولادي OMPHALOCELE

الخلاصة

الهدف من الدراسة: دراسة تحليلية حول كيفية التشخيص والمعالجة الاولى والجراحية لمرضى قروء السرة وتقييم نتائج العلاج ومقارنة النتائج مع دراسات اخرى.

التصميم: دراسة مستقبلية تتضمن أربعين حالة مصابة بقروء السرة .

موقع الدراسة: جميع المرضى عولجوا في مستشفى حماية الأطفال التعليمي في مدينة الطب للفترة ما بين حزيران ٢٠٠٤ - حزيران ٢٠٠٧

النتائج: كانت نسبة الذكور الى الاناث ١ : ١ ، ٢٥% من المرضى شخضوا قبل الولادة بواسطة فحص السونار ، ١٥% من المرضى كانت ولادتهم قيصرية .

كان وزن معظم المرضى ضمن المعدل الطبيعي ، كان حجم قروء السرة اقل من ٥ سم في ٥٥% من المرضى.

٨٣,٤% من المرضى عولجوا بغلق قروء السرة جراحيا بمرحلة واحدة بينما ١٦,٦% من المرضى احتاجوا لأكثر من مرحلة علاجية .

معدل الوفيات من جراء قروة السرة ومضاعفات التداخل الجراحي هي ١٥% .

الاستنتاجات:

- ١- تبين ان قروة السرة هي حالة نادرة من التشوهات الولادية .
- ٢- لا يوجد فرق في نسبة الذكور الى الاناث .
- ٣- الولادة القيصرية ليست افضل من الولادة الطبيعية لمرضى قروة السرة من ناحية مستقبل الحالة وعلاجها .
- ٤- يمكن تشخيص الحالة قبل الولادة بفحص الامواج فوق الصوتية (السونار) .
- ٥- علاج قروة السرة هو جراحي ويتضمن مرحلة واحدة او اكثر حسب حجم جيب قروة السرة وحسب وجود تشوهات ولادية اخرى .
- ٦- معدل الوفيات يزداد في حالة وجود تشوهات ولادية اخرى مصاحبة لقروة السرة.

Introduction

Omphalocele is an anterior abdominal wall defect at the base of the umbilical cord, with herniation of the abdominal contents. After 10 weeks of gestation the amnion and Wharton jelly also cover the herniated mass [4]. Omphaloceles are associated with other anomalies in more than 70% of the cases. The anomaly is detected during routine ultrasonographic examination of the fetus or during an investigation of an increased alpha-fetoprotein (AFP) level [5]. In addition, these children may have symptoms of gastroesophageal reflux, Hirschsprung disease, or both [6]. Small omphaloceles occur with a rate of 1 case in 5000 live births. Large omphaloceles occur with a rate of 1 case in 10,000 live births [7]. The occurrence of omphalocele in relation to male:female ratio is 1.5:1 [4]. The associated anomalies includes: Chromosomal anomalies (40-60%) [8], Cardiac defects (16-47%), Gastrointestinal anomalies (40%) [9], Genitourinary anomalies (40%) [11], Neural tube and head and neck anomalies (10%), Musculoskeletal anomalies (10-30%) [10], Beckwith-Wiedemann syndrome (5-10%) [11].

Aim of the Study

To evaluate the prenatal diagnosis, post delivery management including surgical correction and prognosis.

Patients and Methods

Prospective study from May 2004 to May 2007. Forty patients with omphalocele were admitted to the pediatric surgery center in children welfare teaching hospital in Baghdad.

Medical records of these patients were recorded for prenatal diagnosis, birth order, sex, weight, maternal age, gestational age, type of delivery, size of the defect, associated anomalies, treatment and outcome.

All patients had been examined to exclude associated congenital anomalies.

All patients investigated for blood group, blood glucose and urea, echocardiography and ultra sound of abdomen done for them.

We have 40 patients in our study, most of them had prenatal U/S, only ten of them had been diagnosed by U/S to have abdominal wall defect, and after delivery they found to had more than 5cm defect (omphalocele major).

All patients had preoperative treatment with nasogastric tube for decompression of the GIT. Kept in the incubator. Intravenous fluid given. Vitamin K given I.M (at 1mg/kg in single dose).

Treatment of omphalocele were done under General anesthesia with endotracheal complete muscle paralysis.

Small to moderate size omphalocele were treated with primary repair, by which we were sharply remove the sac at the skin-fascia edge with careful identification and ligation of the

umbilical vessels (umbilical vein above, two umbilical arteries below with urachus between them, all should be ligated) . The contents of the sac were reduced inside the abdominal cavity. The fascia was closed with running absorbable sutures (poly glactin or poly dioxanone) and the umbilicus was reconstructed. These were done for 30 patients.

large omphalocele staged repair were used for 6 of patients; each patient was treated by different methods either by undermining the skin, the skin was closed over the abdominal viscera, we were produce a ventral hernia, to be corrected at later age. This was done for two of our patients because the defect was large and fascia were difficult to close.

or the defects in other two patients was difficult to treat by this method not due to large size defect, but patient have had life threatening cardiac anomaly, the defect was managed by promote epithelization of the sac to form a ventral hernia; topical agent had been used was silver sulfadiazine. The agent was applied to the intact sac twice daily and covered with an elastic dressing to apply mild compression of the sac contents.

While other two patients with large size defect we were used the intact sac as silo for reduction of the contents of the sac; and after 8 days we took the patient to operating room and close the defect.

Postoperatively naso gastric tube removed 2-3 days postoperatively, start oral feeding after 2-3 days when the bowel sound became positive.

Four patients need mechanical ventilation because of increased intra abdominal pressure, two of them remained for 3days and the other two for 5days and all were weaned from ventilator successfully.

Results

Forty patients (twenty male and twenty female), the male to female ratio is 1:1, all of our patients are full term, thirty four babies delivered by NVD and six by C/S for causes not related to omphalocele, eighteen of our patients are first born baby, two patients less than 2 Kg, thirty two patients 2-3 kg and six patients more than 3Kg (table 1).

We see from table one that increase in the size of defect is associated with decrease in the birth weight.

Ten of our patients (25%) were diagnosed prenatally by U/S.

We found that the size of the defect were less than five cm (omphalocele minor) in 20 patients (50%) and five to ten cm (omphalocele major) in the other 20 patients (50%).

Twenty patients had the liver present as part of the sac contents (50%). Only two patients had rupture of the sac (5%).

Table 1 Relation of the size of the defect with birth weight

No.	sex	Birth order	Gestational age (wk)	Type of delivery	Birth wt. (kg)	Size of defect(cm)
2	Female	1	36 wk	NVD	1.75	7
2	Female	6	36 wk	NVD	2	5
2	Male	1	36 wk	C/S	2.25	6
2	Female	5	36 wk	C/S	2.5	5
2	Female	8	36 wk	NVD	2.5	10
2	Male	1	36 wk	NVD	2.5	4
2	Female	6	36 wk	NVD	2.5	5
2	Male	4	38 wk	NVD	3	3
2	Male	1	37 wk	NVD	3	3
2	Female	1	37 wk	NVD	3	6
2	Female	1	38 wk	NVD	3	5
2	Male	5	38 wk	NVD	3	4
2	male	1	37 wk	NVD	3	6
2	Female	1	36 wk	NVD	3	5
2	Female	3	38 wk	NVD	3	3
2	Male	3	38 wk	NVD	3	3
2	Male	4	38 wk	C/S	3	4
2	Male	3	37 wk	NVD	3.5	4
2	Female	2	38 wk	NVD	3.5	3
2	Male	1	38 wk	NVD	3.75	4

Fourteen patients had associated anomalies in the form of cleft lip and palate, bladder extrophy

,omphalomesentric duct anomalies and two with cardiac problem .

Table 2 Associated anomalies

Type of anomalies	No. of patient	%
Cleft lip and palate	4	10
Bladder extrophy	4	10
Omphalomesentric anomalies	4	10
Cardiac anomalies	2	5
Total	14	35

The mean maternal age 25 years,mean birth weight was 2.8 kg and mean gestational age was 37 weeks.

Treatment:

The treatment of omphalocele is to close the defect by primary surgical closure or staged closure .

Four patients died before operation, Thirty patients(83.4%) treated by

primary closure, six patients (16.6%) treated by staged closure two of them treated by promote epithelization by topical agents because of the risk of operation due to associated anomalies, then the defect closed surgically at the age of three months. Two patients treated by hanging of the defect to the ceiling of incubator and gradual ligation of sac because the size of the defect are large and difficult to close, after 10 days the surgical closure done. Two patients treated by release of the skin laterally and suturing anteriorly and formation of ventral hernia also because the defect size is too large, the ventral hernia of this patient not close yet.

Most patients have smooth post operative period with out complications. Four patients (11.11%) need mechanical ventilator because of an increase of intra abdominal pressure, two of them remain three days and other two patients for five days on ventilator and all of them have been weaned successfully from the ventilator.

Two of our patients (5.55%) develop malabsorption one month after operation although they had no interference with the bowel during operation. Other two patients (5.55%) develop post operative neonatal sepsis and died.

In our study 6 patients (15%) died. Four patients died before operation, two of them are low birth weight and large defect, the other two have cleft lip and palate with low birth weight and all developed neonatal sepsis which was the cause of death.

Two patients died three days after the operation due to large size of the defect and associated bladder extrophy with vesicointestinal fistula

Discussion

Between May 2004 to May 2007; 40 patients presented to our center.

In spite of our limited experience in prenatal diagnoses and the critical situation of our country in the availability of the specific radiological and laboratory equipment required in the diagnoses of this condition, our study showed that 25% of the conditions diagnosed prenatally; and as we know the sensitivity for prenatal diagnoses range from 25% to 100% as in other studies [12,13] make our study at the lower sensitivity limit, this may be due to some of our patients did not do U/S evaluation regularly because of low social class, low income or may be due to little experience of our radiologist.

In our study only six mothers had C/S which also might be due to reasons other than omphalocele. The normal vaginal delivery is a safe mode of delivery to all patients with omphalocele as it never cause any interference with normal delivery, we did not record any uterine dystocia or prolonged labor as well as no any rupture of omphalocele sac during delivery (only two patient had rupture of the sac); so we think that no need to change the mode of delivery when there is omphalocele (except in case of giant omphalocele), which also supported by many European and American studies which show that the mode of delivery is a decision to be made by the obstetrician on the basis of obstetric indication [14,15,16]

The male to female ratio is 1:1 which is similar to a study done by Dr. Ali Nawas.

In our study (45%) 18 babies are the first born in the family, although the incidence of the first baby affected by omphalocele is 27% in other studies [17].

One of important means of good antenatal care is the birth weight, we found the main birth weight is 2.8 kg which is the same of analyses published by Med pregl. [18], which

reflect good antenatal care and the same thing regarding the gestational age[19].

In our study the liver included in the sac in twenty patients (50%), which is similar to the percentage found in many studies, as 45% stated Grant H.[19]

In our study the size of defect range from 3-10 cm which is similar to Arnold G.[7]

Two patients in our study have rupture of the sac make the percentage 5%, which is comparable to 4% reported in other studies[20].

In our study associated anomalies seen in 14 of our patients, make the percentage to be 35%, in other studies it range from 40-45% [11,12] This difference are accepted especially large number of our patients have small to moderate size omphalocele; and we didn't have the technique to diagnose chromosomal anomalies but our result similar to Rob and Smith textbook[21]. 90% of our patients had smooth post operative period, which different from that stated in Ashcraft textbook of pediatric surgery; may be due to 50% of our patients have small to moderate size defect and 83% of all patients closed by primary closure and none of our patients need prosthetic mesh for closure of the defect; which could increase the morbidity as infection, sepsis and separation of sutures of prosthetic mesh[4].

Only two patients, who was treated by primary closure, died 3 days post operatively because of large size of the defect and associated bladder extrophy.

Patients with omphalocele in other studies had 34% mortality; almost exclusively due to associated anomalies; overall increased mortality was noted with the use of prosthetic silos, usually secondary to sepsis as stated in Ashcraft Pediatric surgery textbook [4]. The mortality of

omphalocele patient was 52% in [18] and 34% in Michael D.[4] while the mortality rate in our study is 15% (10% pre operatively and 5% post operatively) and our followup for one year, this low percentage because as we said none of our patients treated by prosthetic silo and few of our patients had major associated anomalies, two patient (5%) had cardiac anomalies, and four patients (10%) had bladder extrophy and that are the causes of this low percentage.

Conclusion and Recommendations

Male to female ratio was equal, All of the babies were full term.

Most of the patients had a good birth weight and it decrease with increase in the size of the defect.

Except giant omphalocele, C/S delivery had no benefit over NVD in the outcome of omphalocele management.

Patients with omphalocele had 35% chance of associated anomalies. The liver was found in the sac in 50% of patients.

High mortality rate most commonly seen in those have associated anomalies, low birth weight and large size defect and 10% need ventilator post operatively.

Prenatal ultrasound is important for all pregnant mothers specially in a family with history of omphalocele.

Early resuscitation and rapid transfer to the pediatric surgery centers.

Associated anomalies should be excluded before operation.

Early primary closure carries a better prognosis and should be done as early as possible.

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