

THE MANAGEMENT OF OMPHALOCELE IN MOSUL

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ABSTRACT

Background: Omphalocele is an anterior abdominal wall defect at the base of the umbilical cord, with herniation of the abdominal contents. The herniated organs are covered by the parietal peritoneum. After 10 weeks of gestation the amnion and Wharton jelly also cover the herniated mass. This defect needed to be closed either by a primary closure or staged closure. **Objectives:** To evaluate the prenatal diagnosis, post delivery management including surgical correction and prognosis in Mosul. **Patients and methods:** A prospective study of twenty patients with Omphalocele was conducted in pediatric surgery center at Al-Khansaa Teaching Hospital in Mosul from January 2005 to January 2008. The medical records of the 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 outcome. 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 was given to all patients as 1mg/kg in single dose. All patients had preoperative treatment with nasogastric tube for decompression of the GIT, kept in the incubator, I.V fluid given, I.V antibiotic and oxygen as needed. **Results:** Twenty full term patients, ten male and ten female. Diagnosis of all of patients was done clinically by inspection, while five of the patients were diagnosed prenatally by ultrasound examination (25.0%). All patients had normal birth weight. Only three of our patients had C\S delivery (15.0%) while the rest were product of normal vaginal delivery. Only one patient had rupture of the sac at presentation (5.0%). A fifteen patients treated successfully by primary closure (83.4%), three patients treated by staged closure (16.6%). two patients died preoperatively (10.0%) secondary to sepsis because of low birth weight and associated anomalies, one patient died postoperatively (5.0%) 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 90.0% of the patients.

KEYWORDS: Management, Omphalocele, Primary closure, Staged closure.

INTRODUCTION

Omphalocele is an anterior abdominal wall defect at the base of the umbilical cord, with herniation of the abdominal contents. The herniated organs are covered by the parietal peritoneum. After 10 weeks of gestation the amnion and Wharton jelly also cover the herniated mass.^[1]

Omphalocele are associated with other anomalies in more than 70% of the cases. Most associated anomalies are chromosomal. The anomaly is detected during routine ultrasonographic examination of the fetus or during an investigation of an increased alpha-fetoprotein

(AFP) level.^[2] Anomalies of intestinal fixation accompany abdominal wall defects, and midgut volvulus is a possible complication. Atypical appendicitis may also occur, if the abnormally located appendix was not previously removed. In addition, these children may have symptoms of gastroesophageal reflux, Hirschsprung disease, or both.^[3]

Abdominal wall defects result from failure of the mesoderm to replace the body stalk, which persists in a region usually occupied by somatopleure.^[4]

In babies with Omphalocele, failure of central fusion at the umbilical ring due to defective mesodermal growth

causes incomplete closure of the abdominal wall and persistent herniation of the midgut. The abdominal viscera are contained in a translucent sac, which is composed of amnion, Wharton jelly, and peritoneum. The umbilical vessels radiate onto the wall of the sac. In 50.0% of cases, the liver, spleen, and ovaries or testes accompany the extruded midgut.^[1]

Factors associated with high-risk pregnancies, such as maternal illness and infection, drug use, smoking, and genetic abnormalities, can be associated with birth of babies with Omphalocele and gastroschisis. These factors contribute to placental insufficiency and birth of premature or small-for-gestational-age (SGA) babies, in whom gastroschisis and Omphalocele are most common. Folic acid deficiency, hypoxia, and salicylates have caused laboratory rats to develop abdominal wall defects, but the clinical significance of these experiments is conjectural.^[5]

Omphalocele can often be detected on fetal ultrasound in the second and third trimesters of pregnancy. A fetal echocardiogram (ultrasound of the heart) may also be done to check for heart abnormalities before the baby is born.^[6]

The anomaly is usually detected during routine ultrasonographic surveillance, during an investigation of a disparity of uterine size with the dates or other obstetric indications, or during an evaluation of an increased maternal serum AFP level. Omphaloceles and gastroschisis are 2 ventral wall defects that are detected by means of AFP measurement. Acetylcholinesterase levels may also be increased.^[1] Ultrasonography is an inexpensive, safe, noninvasive real-time technique that is widely available. It remains the imaging modality of choice for the prenatal assessment of the fetus. In experienced hands, ultrasonography is highly accurate in the diagnosis of most complications associated with pregnancy. It is also used as a guide to intervention in pregnancy.^[6]

At present, the use of MRI in pregnancy is limited, as more experience is gained, fetal MRI may play a greater role in anomaly analysis. Prenatal MRI is at times a useful adjunct to ultrasonography and may enhance fetal anatomic evaluation when complex anomalies are suspected. MRI facilitates perinatal management and parental counseling.^[7]

Although ultrasonography is an accurate and sensitive means for detecting fetal anomalies, it still has limitations, and its dependence on operator skill is a major disadvantage.^[6]

Besides the limited availability of MRI, its safety issues have not been completely resolved. Prudence currently dictates that MRI be used in the first trimester only if a clear medical indication is present and only when it offers a definite advantage over ultrasonography.^[1]

An Omphalocele is diagnosed when a fetal anterior midline abdominal mass is demonstrated. The mass consists of abdominal contents that have herniated through a midline central defect at the base of umbilical cord insertion. The mean size of the defect is 2.5-5 cm. The mass has a smooth surface and contains abdominal viscera, usually the liver and including the bowel and stomach. The covering of the mass, which comprises the peritoneum and amnion, may rarely rupture. The membrane is not always visible. Wharton jelly may be detectable as a hypoechoic lining between the layers of the covering of the membrane.^[8]

The umbilical cord attaches to the apex of the herniated mass, where the umbilical vein can be seen within the mass. The cord may be widened where it joins the fetal skin. Fetal ascitis is common and seen within the herniated sac. Polyhydramnios, and occasionally oligohydramnios, may be present. Other major anomalies may be apparent in as many as 70.0% of cases.^[1]

A number of sonographic features differentiate an Omphalocele from a physiologic midgut herniation. A midgut herniation seldom exceeds 7 mm in diameter, whereas Omphalocele are much larger. Midgut herniation is invariably smaller in diameter compared with the diameter of the abdomen, whereas the diameter of an Omphalocele can be larger than that of the abdomen. The size of the Omphalocele is best measured by using the ratio of the transverse diameter of the Omphalocele to the transverse diameter of the abdomen. A midgut herniation seldom persists after 12 weeks of gestation or in a fetus with a crown-length measurement of more than 44 mm.^[1]

When the ratio of the diameter of the Omphalocele to the transverse diameter of the abdomen is less than 60.0%, the lesion usually contains bowel and not liver. The relative size of the Omphalocele may decrease during pregnancy.^[1,2]

Ultrasonography is a sensitive technique, but it remains operator dependent. Its great advantage is that it can be quickly and frequently repeated as required. A definitive diagnosis of Omphalocele is possible only beyond 12 weeks' gestation when confusion with physiologic midgut herniation no longer exists.^[1]

Fetuses with an isolated Omphalocele should be delivered in a tertiary referral center. The available data do not support a policy of cesarean delivery of babies with abdominal wall defects.^[9] With a large Omphalocele, dystocia may occur and result in injury to the baby's liver; hence, cesarean delivery may be indicated.^[10] Omphalocele diagnosis was clear after delivery by inspection; the investigations were needed only to exclude other anomalies.

The Initial management and resuscitation was included that the infant should be born in (or rapidly transferred

to) a facility with neonatal intensive care and pediatric surgical capabilities. If the Omphalocele are ruptured, the baby is at risk for severe dehydration and hypothermia.^[11] The initial evaluation should include physical examination, chest radiograph, echo cardiogram and orogastric tube is placed for gastric decompression.^[12] Antibiotic are administered, the omphalocele sac is wrapped in a protection dressing and take care to prevent mechanical trauma operative repair may be under taken as soon as the infant condition is stabilized.^[11]

An intact sac will protect the viscera so that the operation is probably not an emergency, but closure or coverage should be performed within 24 hrs, as the sac can crack, separate or tear. From a cardiac view point, perhaps the safest time to operate is immediately after birth while the compliance of the right ventricle has not yet decreased and a left to right shunt across a septal defect or a patent ductus arteriosus was less likely to exist. The sooner the operation, the less chance for bowel distention or edema to develop, and the more likely that a primary repair will be possible.^[13] Ladd's procedure is not performed to manage the intestinal malrotation, the extent and nature of the operation will produce enough adhesion to prevent volvulus.^[14] General endotracheal anesthesia with complete muscle paralysis is recommended for all infants with Omphalocele.^[15] The small to moderate size Omphalocele may contain a small portion of the liver has the umbilical cord insertion to the top of the sac, the sac is sharply removed at the skin fascia edge with careful identification and ligation of the umbilical vessels (umbilical vein above, two umbilical arteries below with urecus between them, all should be ligated), if the liver is firmly attached to the sac and separated can cause bleeding which is difficult to control, then part of the sac can be leaved and introduce to abdominal cavity. The abdominal cavity can be enlarged by manual stretching .The skin is carefully undermined separating it from the deep fascia layers. The fascia is closed with running or interrupted absorbable sutures (poly glactin or poly dioxanone) and the umbilicus is reconstructed. The large Omphalocele was frequently containing most of the liver, are usually not fully reducible at the first operation and staged repair is necessary, after undermining the skin, the skin is closed over the abdominal viscera, producing a ventral hernia that can be repaired six to twelve month later.^[15]

An alternative approach utilize prosthetic closure, Dacron – reinforced polymeric silicone silastic sheets (silo) 0.007 inches thick was used to create a pouch to hold the viscera, with 3-0 mono filament permanent suture in a continuous fashion, one sheet to each lateral edge of the fascia sew and the silastic site secured in contact with the viscera.

Continuing the same suture, the free edges are joint superiorly and inferiorly to form a sac to be used in squeezing the viscera into the abdominal cavity. It may

be helpful to leave long sutures at the apex of prosthetic sac so that it may be suspended from the top of infant warmer to prevent the pouch and its contents falling to one side. Once the sac has been reduced to the level of the fascia (usually one week) a second procedure is under taken to close the fascia.^[14]

Test closure by 3 mattress sutures placed to approximate the fascia with monitoring the transcutaneous oxygen saturation and the end tidal CO₂ to determine if facial closure will be to constrictive; if the patient status is satisfactory with less than 30 mmHg peak respiratory pressure 20 min after the fascia closed; the remainder of the fascia is approximated.^[14] If the defect is difficult to treat by these methods or patient have life threatening cardiac anomalies the defect can be managed by promote epithelization of the sac with secondary closure of the ventral hernia at later date, topical agent have been used are 0.25% merbromin (mrcurochrome), silver nitrate and silver sulfadiazine. The agent is applied to the intact sac once or twice daily and covered with an elastic dressing to apply mild compression of the sac contents; this technique may require many weeks to achieve skin coverage of the defect.^[16]

Early post surgical complications were respiratory distress, wound infection, Post surgical ileus, renal vein thrombosis, dehydration, NEC, lower limb swelling and sepsis. The intermediate post surgical complications were variable; inguinal hernia, gastro esophageal reflux, ventral hernia, small intestinal obstruction, malrotation with obstruction, NEC, and malabsorption. The late complications included growth retardation, low IQ, atypical appendicitis, and intestinal obstruction.

Aim of the study

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

PATIENTS AND METHODS

A Prospective study was conducted from January 2005 to January 2008. Twenty patients with omphalocele were admitted to the pediatric surgery center in Al-khansaa Teaching Hospital in Mosul. Patients were managed by different pediatric surgeons in the center. 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, echo-cardiography and ultra sound of abdomen done for them.

RESULTS

Twenty patients (10 males and 10 females) were involved in the present study with male to female ratio was 1:1. All of patients were full term. Concerning the type of delivery; 17 babies delivered by NVD and 3 by C/S for causes not related to Omphalocele. Nine of the

patients were first born baby. Birth weight was variable; one patient was <2 Kg, 16 patients were 2-3 Kg, and only 3 patients were >3 Kg.

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) |
|-----|--------|-------------|----------------------|------------------|----------------|--------------------|
| 1 | Female | 1 | 36 wk | NVD | 1.75 | 7 |
| 2 | Female | 6 | 36 wk | NVD | 2 | 5 |
| 3 | Male | 1 | 36 wk | C/S | 2.25 | 6 |
| 4 | Female | 5 | 36 wk | C/S | 2.5 | 5 |
| 5 | Female | 8 | 36 wk | NVD | 2.5 | 10 |
| 6 | Male | 1 | 36 wk | NVD | 2.5 | 4 |
| 7 | Female | 6 | 36 wk | NVD | 2.5 | 5 |
| 8 | Male | 4 | 38 wk | NVD | 3 | 3 |
| 9 | Male | 1 | 37 wk | NVD | 3 | 3 |
| 10 | Female | 1 | 37 wk | NVD | 3 | 6 |
| 11 | Female | 1 | 38 wk | NVD | 3 | 5 |
| 12 | Male | 5 | 38 wk | NVD | 3 | 4 |
| 13 | male | 1 | 37 wk | NVD | 3 | 6 |
| 14 | Female | 1 | 36 wk | NVD | 3 | 5 |
| 15 | Female | 3 | 38 wk | NVD | 3 | 3 |
| 16 | Male | 3 | 38 wk | NVD | 3 | 3 |
| 17 | Male | 4 | 38 wk | C/S | 3 | 4 |
| 18 | Male | 3 | 37 wk | NVD | 3.5 | 4 |
| 19 | Female | 2 | 38 wk | NVD | 3.5 | 3 |
| 20 | Male | 1 | 38 wk | NVD | 3.75 | 4 |

Size of the defect was showed in table (2). It elicited that were finding that the size of the defect less than five cm (Omphalocele minor) was found in 10 patients while 5-10cm (Omphalocele major) was found in another 10 patients.

Table (2): Size of the defect.

| Size of the defect | No. of patient | % |
|--------------------|----------------|----|
| <5 cm (minor) | 10 | 50 |
| 5-10 cm (major) | 10 | 50 |

Associated anomalies among the study sample was showed in table (3) and demonstrated that 7 patients had associated anomalies in the form of cleft lip and palate, bladder extrophy, omphalomesentric duct anomalies and one with cardiac problem.

Table (4): The mean of maternal age, gestational age, and birth weight.

| Mean maternal age (years) | Mean gestational age (wk) | mean birth wt. (kg) |
|---------------------------|---------------------------|---------------------|
| 25 years | 37 wk | 2.8 kg |

Treatment techniques was illustrated in figure (1) and showed that 15 patients were treated by primary closure, three patients treated by staged closure; one 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 age three months. One patient was treated by hanging of the defect

Table (3): Associated anomalies.

| Type of anomalies | No. of patient | % |
|----------------------------|----------------|----|
| Cleft lip and palate | 2 | 10 |
| Bladder extrophy | 2 | 10 |
| Omphalomesentric anomalies | 2 | 10 |
| Cardiac anomalies | 1 | 5 |
| total | 7 | 35 |

The mean maternal age among the study sample was demonstrated in table (4) and revealed that the mean maternal age was (25.0±) years and the mean gestational age was (37.0±) weeks. The mean birth weight was (2.8±) Kg.

to the ceiling of incubator and gradual ligation of sac because the size of the defect was large and difficult to close, after 10 days the surgical closure done. One patient was 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.

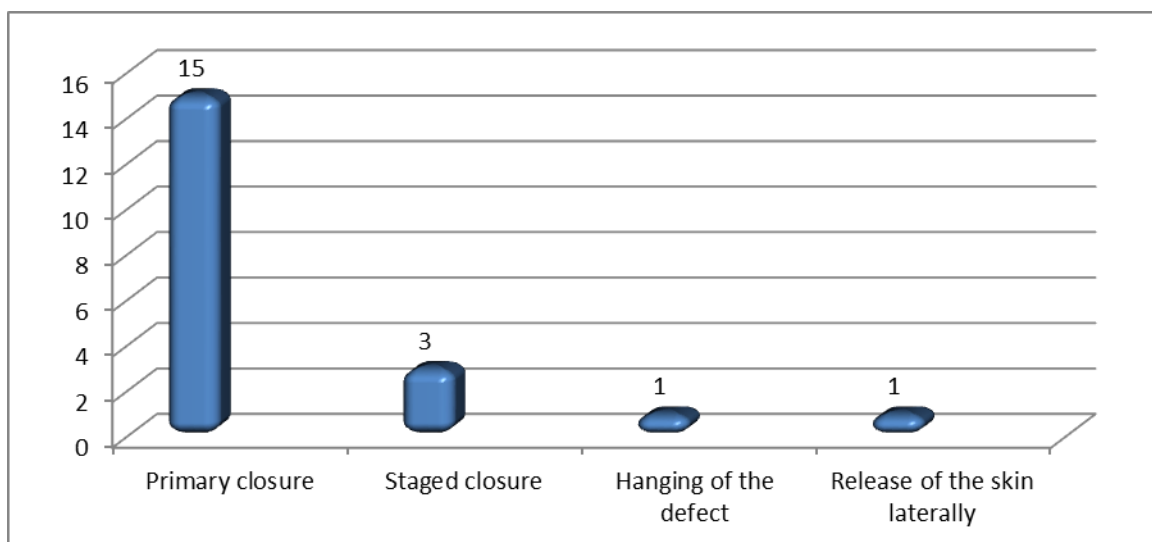


Figure (1): Treatment techniques.

Types of closure was showed in table (5) and demonstrated that the primary closure was done in 83.4% of the cases while the staged closure was done in 16.6%.

Table (5): Types of closure.

| Type of closure | No. of cases | % |
|-----------------|--------------|------|
| Primary closure | 15 | 83.4 |
| Staged closure | 3 | 16.6 |

Necessity of ventilator was showed in table (6) which revealed that most patients have smooth post operative period without complications. Two patients need mechanical ventilator because of an increase of intra abdominal pressure, one of them remain three days and

other for five days on ventilator and both of them have been weaned successfully from ventilator.

Table (6): Necessity of ventilator.

| No. of patients treated | No. of patients need ventilator | % |
|-------------------------|---------------------------------|-------|
| 18 | 2 | 11.11 |

Prognosis and complications were showed in table (7) and it elicited that only one patient among the study sample developed malabsorption one month after operation although he had no interference with the bowel during operation. Other patients develop post operative neonatal sepsis and died.

Table (7): Prognosis and complications.

| No. of patients treated | Type of complication | No. of patients | % |
|-------------------------|----------------------|-----------------|------|
| 18 | Malabsorbtion | 1 | 5.55 |
| 18 | sepsis | 1 | 5.55 |

Table (8) demonstrated the mortality among the study sample and showed that 3 patients were died. Two patients die before operation, one of them was low birth weight and large defect, the other had cleft lip and palate with low birth weight and both develop neonatal sepsis which is the cause of death. One patient died three days after operation due to large size of the defect and associated bladder extrophy with vesicointestinal fissure.

Table (8): Mortality.

| No. of patients | No. of deaths | Mortality |
|-----------------|---------------|-----------|
| 20 | 3 | 15% |

DISCUSSION

Between January 2005 to January 2008; 20 patients presented to Al-Khansaa center. In spite of the limited experience in prenatal diagnoses and the bad situation of this country in the availability of the specific radiological

and laboratory equipment required in the diagnoses of this condition, the current study showed that 25.0% of the conditions diagnosed prenatally and the sensitivity for prenatal diagnoses range from 25.0% to 100.0%.^[17] hence the current study at the lower sensitivity limit, this may be due to some of the sampled patients did not do U/S evaluation regularly because of low social class, low income or may be due to little experience of ultrasound operators with this anomaly; and if 3 dimensions U/S become available in this locality the picture may change.

As the prenatal diagnoses are of low sensitivity; only 3 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, no uterine dystocia or prolonged labor as well as no any rupture of Omphalocele sac were recorded during

delivery (only one patient had rupture of the sac); so there were 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.^[17]

The male to female ratio is 1:1 as shown in the present study and many other studies has the same ratio too. As in a scientific paper of Nawas in which, the ratio was 1:1.^[1]

Regarding birth order, in the current study (45.0%) 9 babies are the first born in the family, although the incidence of the first baby affected by Omphalocele is 27.0% in other studies.^[17]

One of important means of good antenatal care is the birth weight, and found that the main birth weight is 2.8 kg which is the same of analyses published by Tati and Joki published in *Med pregl.* 2006; 59 of 31 patients from 1999-2003, which reflect good antenatal care, the same thing regarding the gestational age.^[18]

Regarding the presence of part of the liver in the sac, in the present study the liver included in the sac in ten patients (50.0%), which is similar to the percentage found in many studies, as 45% stated in pediatric surgery textbook.^[18]

Regarding the size of the defect, in general it ranges from (2-10cm).^[16] in the present study it also range from 3-10 cm which is comparable to others.

Regarding rupture of the sac, only one patient in the current study has rupture of the sac make the percentage 5.0%, which is comparable to 4.0% reported in other studies.^[13]

In the present study associated anomalies seen in 7 of the sampled patients, make the percentage to be 35%, in other studies it range from 40.0-45.0%.^[17] This difference are accepted especially large number of the patients have small to moderate size Omphalocele and unavailability of the technique to diagnose chromosomal anomalies.

The decision to treat patients by primary closure or staged closure depend on the size of the defect and the presence of major associated anomalies which is the same policy which is stated in Ashcraft and Rob and Smith text books.^[14,15]

Fifteen (83.4%) of the sampled patients are treated by primary closure of the defect, which is high percentage and that due to the majority of the patients are small to moderate size Omphalocele which make them prone to primary closure by one stage operation.

A 90.0% of the sampled patients had smooth post operative period, which different from that stated in Ashcraft textbook of pediatric surgery; may be due to 50.0% of our patients have small to moderate size defect and 83.0% of all patients closed by primary closure and none of the patients need prosthetic mesh for closure of the defect; which could increase the morbidity as infection, sepsis and separation of sutures of prosthetic mesh.^[14]

Only one patient, 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 34.0% 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.^[14] The mortality of Omphalocele patient was 52.0% in a study done by Tati and Joki over 5 years.^[18]

Mortality rate in the present study is 15.0% (10.0% pre operatively and 5.0% post operatively), this low percentage because as we said none of our patients treated by prosthetic silo and few of present sampled patients had major associated anomalies, one patient (5.0%) had cardiac anomalies, and two patients (10.0%) had bladder extrophy and that is the cause of this low percentage.

CONCLUSION

Sex ratio was equal. Most 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. Part of the liver found in the sac in 50% of patients. Type of closure of defect depends on the size of the defect and to the associated anomalies. High mortality rate most commonly seen in those have associated anomalies, low birth weight and large size defect. In the present study, 10% need ventilator post operatively.

Recommendations

From the results of present study, the following recommendations were highlighted:

- 1- Prenatal ultrasound is important for all pregnant mothers especially in a family with history of Omphalocele.
- 2- Early resuscitation and rapid transfer to the pediatric surgery centers.
- 3- Associated anomalies should be excluded before operation.
- 4- Early primary closure carries a better prognosis and should be done as early as possible.

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