Immediate Transumbilical Repair and Umbilical Plasty for Omphalocele Minor

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Abstract

Background/Purpose: Omphalocele is a congenital midline abdominal wall defect that results in herniation of intraabdominal contents. Omphalocles are classified into 'minor' if the diameter of the umbilical defect is less than 5cm or 'major' if more. The objective of this study was to present our experience in treatment of omphalocele minor and to review the literature.

Material and Methods: Over an 8 years period, 38 patients, 20 males and 18 females, with omphalocele minor were treated at pediatric surgery unit, Assiut children university hospital. 26 patients presented the same day of birth and 12 cases presented the following days. Diagnosis was done on clinical basis, antenatal diagnosis proved only in 13. The sac was intact in 33 cases and ruptured sac in 5 cases, one of them had a faecal discharge through a patent vitellointestinal duct. All patients except 4 were passing meconium preoperatively. All patients were prepared for emergency surgery the day of presentation.

Results: 38 patients were operated upon. 3 patients had associated anomalies, one patient had polydactyl, and distal penile hypospadius, one patient had patent vitellointestinal duct and one patient had ileal atresia type 2. There was no associated anomalies that could contraindicate surgery or anesthesia. The mean operative time was 65 minutes. Most patients passed stool and started oral feeding within 24 hours and discharged within 24-48 hours. Minor complications have occurred in 3 patients.

Conclusion:
• Prenatal diagnosis of omphalocele is recommended to detect associated foetal anomalies for and to prepare for post natal management.
• Babies with omphalocles must be examined thoroughly for other congenital anomalies.
• Careful inspection of the base of umbilical cord should be done prior to clamping of the cord because of the possible association of exomphalos minor and a patent vitellointestinal duct.
• Immediate repair of omphalocele minor is the recommended treatment, especially for patients who have irreducible contents, ruptured sac and associated other intestinal anomalies.
• A purse string repair and umbilical plasty is recommended for treatment of omphalocele minor.
• Omphalocele minor has a good prognosis even in delayed presented cases or associated anomalies.

Key Words: Omphalocele — Exomphalos — Umbilical plasty.

Introduction

OMPHALOCELE is a congenital midline abdominal wall defect that results in herniation of intraabdominal contents. The abdominal visceria are surrounded by the Wharton jelly, peritoneum and amnion and contained in a translucent sac. The sac protrudes in the mid-line, through the umbilicus, with a general incidence of 1 per 5,000 live births 111.

Omphalocles are classified into 'minor' if the diameter of the umbilical defect is less than 5cm or 'major' if more. The definitive goal of surgical intervention is to provide complete fascial and skin closure without causing excessive intraabdominal pressure or abdominal wall tension [2]. Operative treatment of omphalocele major is usually challenging for pediatric surgeons, which is reflected by the broad range of approaches described in the literature 131.

'Minor' omphalocles usually contain a loop of the midgut, the peritoneal cavity is adequate and there are no other major associated congenital anomalies and the defects can be closed primarily [2]. This concept forms the basis for our presentation and discussion.

Patients and Methods

Over an 8 years period, 38 patients with omphalocele minor were treated at pediatric surgery unit, Assiut children university hospital. There was 20 males and 18 females.
26 patients presented the same day of birth 17 of them referred from our gynaecological hospital and 9 cases referred from other centers.

12 cases presented the following days and all of them were referred from other centers or come primally to our unit without referral and all of them had irreducible contents.

Table (1): Age of presentation.

<table>
<thead>
<tr>
<th>Day of presentation</th>
<th>Number of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>1st day</td>
<td>26</td>
</tr>
<tr>
<td>2nd day</td>
<td>4</td>
</tr>
<tr>
<td>3rd day</td>
<td>3</td>
</tr>
<tr>
<td>4th to 10th</td>
<td>5</td>
</tr>
<tr>
<td>Total</td>
<td>38</td>
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</table>

Diagnosis was done on clinical basis and through examination revealed intact sac in 33 cases and ruptured sac in 5 cases, one of them had a faecal discharge through a patent vitellointestinal duct. All patients except 4 were passed meconium preoperatively. Antenatal diagnosis Proved only in 13 cases.

All patients were subjected to complete physical examination and routine abdominal sonography to exclude other anomalies

All patients were prepared for emergency surgery the day of presentation after complete laboratory investigations and physical fitness. Prophylactic antibiotics was given as a routine.

Surgical technique:

- Early cases:

The sac is opened on one side, usually the right, midway between the cord and the skin after reduction of the contents if they are reducible or after being sure that the contents are not adherent to the sac at this site if the hernia is irreducible. The contents are examined meticulously for other anomalies. Healthy intestine reduced back into the abdomen. Adherent intestine to the sac is dissected carefully by sharp and blunt dissection on the expense of the sac to preserve blood supply of the intestine. After complete dissection, the contents examined for other pathology. Healthy viscera reduced back into the abdomen. The index or little finger is passed circumferentially around the inner edge of the defect to be sure that the contents reduced completely without adhesions to the abdominal wall. The edges of the sac are excised circumferentially, the umbilical vessels and urachus are dissected and ligated. The edges of the defect are freed circumferentially from the skin for a few millimeters.

The umbilical defect repaired (fascial repair) using 3/0 vicryl as a purse string suture repair.

The umbilicus repaired by an intradermal 5/0 vicryl purse-string suture that is incorporated into the middle of the fascial closure.

The patients were kept on IV fluids until passing stool usually within 24 hours, systemic antibiotic for 5 days and discharged from the hospital 24-48 hours posoperatively.

- Late cases:

The contents were irreducible and the sac adherent to the contents and can’t be pinched. The sac cleaned with saline and local antiseptic. With great care, the sac is gently opened near its dome to avoid injury of the contents. The conained viscera dissected from the wall of the sac and the operation completed as previously described.

Results

38 patients were operated upon. 20 males and 18 females. 3 patients had associated anomalies, one patient had polydactyl, and distal penile hypospadius, one patient had patent vitellointestinal duct and one patient had ileal atresia type 2. There was no associated anomalies that could contraindicate surgery or anaesthesia. The diameter of the defect was variable. The operation was completed successfully in all patients. In 15 patients with a defect more than 3cm, the contents were not adherent to the sac and reduced to the abdomen including 4 cases with ruptured sac. In the other 23 patients with a diameter of the defect less than 3cm, there were adhesions between the sac and the contents which were dissected carefully and reduced to the abdomen. Of those 23 patients one patient had Meckel's diverticulum with a wide base, it was reduced back to the abdomen. One patients had ileal atresia type two. Resection and end to end anastomosis was performed. One patient with ruptured sac had patent vitellointestinal duct, it was mobilized through the umbilicus and treated by wedge resection.

There was no intraoperative complications except a mesenteric tear in the patient who presented on the 10th day due to dense adhesions and friable tissues due to infection and it was repaired. In one patient the caecum was bulky with the defect less than 2cm so reduction was difficult. The defect was enlarged upwards for a few millimeters.
The mean operative time was 65 minutes. Most patients passed stool and started oral feeding within 24 hours and discharged within 24-48 hours. Patients with resection anastomosis and patent vitellointestinal duct were kept on IV fluids for 3 days and discharged after 1 week.

The 1st wound dressing was after 3 days and repeated every 3 days and was left exposed after 10 days. Nice wound healing occurred in 35 patients. Minor complications have occurred in 3 patients from those presented after the 1st day included one patient with superficial wound infection with minimal purulent discharge which responded well to conservative treatment. One patient had a small abscess at the upper edge of the umbilicus, after its drainage a sinus developed. The wound explored and one stitch was extracted and healing occurred within a few days.

One patient had ischaemic edges of the umbilicus, it was trimmed away and healed spontaneously.

### Table (2): Size of the defect.

<table>
<thead>
<tr>
<th>Size of the defect</th>
<th>Number of cases</th>
</tr>
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<tbody>
<tr>
<td>2-3 cm</td>
<td>25 cases</td>
</tr>
<tr>
<td>&lt;3 cm</td>
<td>13 cases</td>
</tr>
</tbody>
</table>

### Table (3): Associated anomalies.

<table>
<thead>
<tr>
<th>Associated anomaly</th>
<th>Number of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypospadius</td>
<td>One case</td>
</tr>
<tr>
<td>Polydactyl</td>
<td>One case</td>
</tr>
<tr>
<td>Ileal atresia</td>
<td>One case</td>
</tr>
<tr>
<td>Vitellointestinal duct anomalies</td>
<td>Two cases</td>
</tr>
</tbody>
</table>
Fig. (2-C): Adhesions between contents and the sac.

Fig. (2-D): Dissection of adhesions.

Fig. (3): Adhesions between terminal ileum and ascending colon, acute ileocolic angle with partial colonic obstruction.

Fig. (4): Purse string repair.

Fig. (5-A): Umbilical plasty, postoperative.

Fig. (5-B): Umbilical plasty, postoperative.
Fig. (5-C): Umbilical plasty, 5 days postoperative.

Fig. (6): Omphalocele minor, 2 days.

Fig. (7-A): Omphalocele minor after 10 days with intestinal obstruction.

Fig. (7-B): Omphalocele minor after 10 days with dense adhesions within the sac.

Fig. (7-C): Omphalocele after 10 days, operative mesenteric tear.

Fig. (8-A): Ruptured omphalocele with eviscerated ileum and colon.
Fig. (8-B): Ruptured omphalocele, ileal atresia.

Fig. (9): Omphalocele minor containing patent vitello-intestinal duct.

Fig. (8-C): Ruptured omphalocele, polydactyl and hypospadius.

Discussion

Exomphalos (from the Greek ex = out; omphalos = umbilicus) refers to protrusion into the umbilicus. Omphalocele (from the Greek omphalos, kele = hernia, tumour), protrusion of small intestine and other viscera, pushing the umbilical cord forward and distending its base into a cystic mass containing the viscera [5].

Exomphalos is a result of failure of formation and closing of the anterior abdominal wall resulting in various degrees of patency of the umbilical ring and could therefore be associated with other forms of impaired organ formation, which will determine the general prognosis [6].

Prenatal diagnosis of gastroschisis or omphalocele is recommended for complex examination and consultation at specialised centres so as to carry out accurate detection and appropriate classification of associated foetal anomalies for guiding the course of pregnancy and to prepare for postnatal management. Attempts to repair the defects intrauterine have not been successful [7,5].

Only 13 cases of our series presented by proved prenatal diagnosis. This could be explained by lack of regular follow-up during pregnancy or sonography done by unexperienced person. In spite of the obvious anomaly, there was delay in presentation in 12 cases (as shown by age at presentation) because many of these children were not diagnosed prenatally, delivered at peripheral health facilities or at home in remote areas. This may increase the morbidity and increases the liability of the sac to rupture.

Incidence of major congenital anomalies varies from 35% to 81% in exomphalos. It is unclear whether these malformations are more common with exomphalos major. Chromosomal anomalies and syndromes occur more commonly in exomphalos minor. This predisposition may help in counseling parents, planning investigations, and organization of multidisciplinary management strategy [8].

Exomphalos minor and major seem to have a predilection for associated anomalies of specific organ systems [8].

Omphalocele minor may be associated with vitellointestinal duct anomalies which are not usually seen in omphalocele major. The association of omphalocele with patent omphalomesenteric duct (POMD) with or without ileal prolapse is a rare anomaly. Only 20 cases have been reported in the literature. Ratan et al., (2007) reported one case of omphalomesentric duct cyst as a content of omphalocele [12,13].

We reported 2 cases with vitellointestinal duct anomalies, one with a wide base Meckel's diverticulum reduced back to the abdomen and the other with patent vitellointestinal duct treated by wedge resection.

Primary closure of the defect is possible in almost all cases of minor omphalocele, but it may need to be delayed after 6 or 7 days in minor omphalocele with an infected sac and oedematous abdominal wall [8]. Yilmaz 2012 advised bedside repair of omphalocele, but this method is suitable only for reducible cases [14].

In early presented cases, the sac is clear, glistening and translucent. The contained viscera are obviously seen through its wall. The sac can be incised safely without visceral injury and adhesions between the sac and contents are dissected easier with minimal bleeding. With time the sac becomes opaque, thick and the viscera could not be seen. Later on it becomes infected. Approaching the contents through incision of sac and dissection of adhesions are more difficult with possible visceral or mesenteric injury and blood loss. Inspite of the preference of many investigators to incise the sac at its cutaneous junction, direct incision of the sac is more safe especially in irreducible cases as the contents are usually adherent at the neck of the sac.

In the published literature, the association of malrotation with exomphalos major has been described, albeit rarely [15,16] but its association with E. Minor is as yet unknown.

In spite of Sinha report of 31% association of malrotation with exomphalos minor which supports the need to actively exclude this abnormality either operatively or radiologically [17], we did not report any cases with postoperative symptoms suggesting malrotation inspite of the fact that caecum is present in the sac in most of our cases and the ascending colon in some cases with mobile caecum and ascending colon. This could be explained by the possibility of absent duodenal obstruction by the Ladd's band.

The umbilicus is aesthetically important, and its absence can be distressing to patients. The appearance of the umbilicus should be acceptable to the patient and family One of the goals of all umbilical surgical procedures is to maintain or restore as normal an appearance as possible [18]. A normal umbilicus is characterized by a depression in which may be found the mamelon (a central eminence that contains the remnants of the solid portion of the umbilical cord) and the cicatrix (dense scar where the intraembryonic and extraembryonic coelom were in continuity). The cushion is the slightly raised margin that surrounds the umbilical depression. Cullen described more than 60 "normal" configurations of the umbilicus [19].

The umbilicus may be retained or reconstructed during the repair of abdominal wall abnormalities. The structures of the umbilical cord may be incorporated into a reconstruction of the umbilicus [20,21].

We repair the defect by a purse string suture to approximates the natural umbilical ligaments which supports the umbilicus. The umbilical plasty is done by a purse string subcuticuar suture aids to invert the umbilical skin inwards resembeling to a great extent the normal umbilicus and supported by the umbilical repair.

After reviewing the literature and according to our results, immediate postnatal management of omphalocele at birth found to be the best management of omphalocele minor for the following reasons:

1- The sac is usually thin and liable to rupture.
2- Most cases especially those with smaller defects, the contents are usually irreducible, adherent with the sac and containing oedematous bulky caecum which is liable to injury in late cases.
3- The operation is easier even with adhesions with low morbidity. Most of the patients’ parents have been satisfied with the results of umbilical repair and plasty.
4- Associated anomalies as atresia, patent vitellointestinal duct and associated obstructed contents are dealt with earlier with better outcome.
5- Usually there are no associated anomalies that could contraindicate surgery or anaesthesia.

Conclusion:
- Prenatal diagnosis of omphalocele is recommend- ed to detect associated foetal anomalies for and to prepare for post natal management.
• Babies with omphalocles must be examined thoroughly for other congenital anomalies
• A careful inspection of the base of umbilical cord should be done prior to clamping of the cord because of the possible association of exomphalos minor and a patent vitellointestinal duct.
• Immediate repair of omphalocele minor is the recommended treatment, especially for patients who have irreducible contents, ruptured sac and associated other intestinal anomalies.
• In babies with omphalocele with a defect 3cm or less, the contents are usually adherent to the sac and should be operated upon immediately.
• A purse string repair and umbilical plasty is recommended for treatment of omphalocele minor. Omphalocele minor has a good prognosis even in delayed presented cases or associated anomalies.

References