Management of omphalocele using the sac as supportive material

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ABSTRACT

Objective: To document retrospectively the outcome of a newly modified procedure for closure of large omphalocele, where the sac was conserved and used as autogenous supportive material.

Methods: The medical records of 10 patients with omphalocele major admitted to Al-Noor Specialist Hospital in Makkah, Kingdom of Saudi Arabia in the last 15 years, October 1990 to October 2005, were retrospectively reviewed. The defects were considered by the same treating surgeon too big to be closed primarily so the sac was preserved to reduce the contents gradually.

Results: In the first stage, complete reduction was achieved in all the cases followed by second stage, the surgical closure. There was no mortality related to this intervention.

Conclusion: This proposed technique is safe, simple and effective for the treatment of large omphalocele.

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The reported annual incidence of omphalocele in an international collected series is 1 in 3300 to 6600 live births,[1] whereas, a local retrospective study, carried out at the Al-Noor Hospital, Makkah in Saudi Arabia, aiming to document the incidence of congenital malformations requiring emergent surgical intervention, reported the incidence of omphalocele of 1/16000 live births.[2]

Omphalocele was classified by Benson et al[3] in 1945 of 2 types: type one, hernia of the umbilical cord when the umbilical defect is less than 4 cm and type 2 omphalocele (amniocele), when the umbilical defect is greater than 4 cm. A small to medium sized defect can be repaired at one stage. In contrast, larger omphalocele cannot be closed at birth due to considerable discrepancy between the capacity of the abdominal cavity and the volume of the herniated organs.

The surgical management of patients with omphalocele has evolved over the past decades, while Gross[4] in 1948 covered the membrane with mobilized skin, Ravitch and Goni[5] used a progressive pneumoperitoneum technique to enlarge the abdominal space, in another study in 1956 Buchanan and Gain[6] resected the spleen and large portion of the right lobe of the liver to close the skin, while partial hepatectomy was carried out to aid the closure of the abdominal wall in 1968 by Kleinhause et al.[7] All these surgical techniques which interfere with the abdominal organs are to be deplored and are not acceptable at the present time.

There are 2 classical strategies currently stated for treating these giant defects,[8] conservative and operative techniques: both have their own advantages and disadvantages. The operative treatment described by Schuster[9] 1967 consists of 2 stages: First stage is the immediate excision of the sac and the use of Silastic sheet stitched to the abdominal wall with gradual reduction of the herniated organs, followed by second stage of removal of the Silastic sheet and secondary surgical closure of the abdominal defect.The conservative management (initial non-operative) depends on the epithelialization of the membrane followed by repair of the residual ventral hernia. This procedure was first described by Ahlfeld[10] in 1899 using alcohol dressing, then modified by Grob[11] in 1957 who used Mercurochrome 2%. Currently 0.5% Mercurochrome solution in 65% alcohol is recommended making the monitoring of mercury serum level necessary due to the danger of mercury toxicity.[12] Alternative topical agents include, silver sulphadiazine or 0.5% silver nitrate solution, or 70% alcohol.[13]

Few papers were published in the last decade aiming to gradually increase the size of the abdominal cavity to accommodate the herniated organs or gradually reduce the content of the sac followed by surgical closure. Bax et al[14] used tissue expander to enlarge the abdominal cavity in one child with omphalocele major followed by successful closure of the defect. Hong et al[15] described a technique in 3 patients by sequential ligation of the sac for reducing the herniated organs into the abdominal cavity. Delayed primary fascial closure was achieved in the patients.

Brown and Wright[16] reported the treatment of 6 patients with large omphalocele by the application of external bandage compression for several days to reduce the omphalocele keeping the sac intact, followed by surgical closure of the defect.

Therefore, the purpose of this study is to describe a non-invasive modified technique in the management of large omphalocele keeping the sac intact and using it as a supportive material with an interesting variation for the gradual reduction of the herniated organs into the abdominal cavity in the first stage followed by the second stage which is surgical closure of the defect.

Methods. We retrospectively reviewed the medical records of 10 patients with omphalocele treated from October 1990 to October 2005 in Al-Noor Specialist Hospital, Makkah. The average gestational age was 36 weeks. The dimensions of the defects were evaluated by the attending surgeon, the mean length of the defects was 5.1 x 5.9 cm and judged that they could not be closed primarily. Parts of the liver or stomach were situated outside the abdominal cavity in 5 cases. (**Figure 1**)

Technique. The patients were electively ventilated. Eight to 10 separate stitches of monofilement prolene 0 were applied on the cutaneo-fascial junction keeping the threads long. The sac is covered with a sterile gauze and every 2 opposite stitches anchored and tightened together to partially reduce the contents of the sac (**Figure 2**). The stitches were then re-tightened every 2 days, pushing the sac and its contents into the abdominal cavity to obtain gradual reduction smoothly. After every sitting of stitches-tightening the patients were kept under close monitoring of renal, cardiovascular, respiratory and gastrointestinal systems.

Complete reductions were obtained within 5-14 days (average 8 days), at which point the babies were operated under general anesthesia, where the prolene stitches were removed, the sacs were resected, and primary fascial and skin closures were carried out (**Figure 3**).

Postoperatively, patients were kept on ventilator on average of 5 days.

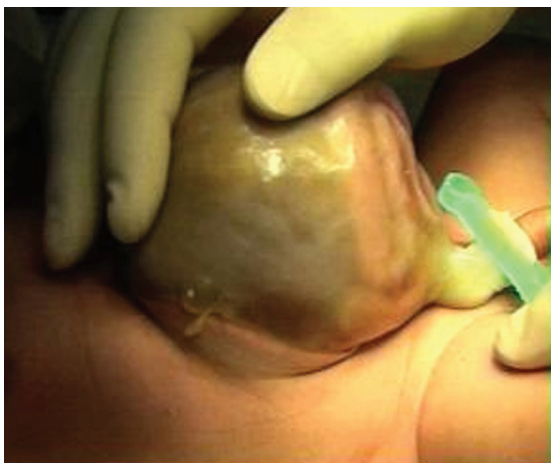

Figure 1 - Omphalocele major (the liver and intestine are herniated in the sac)

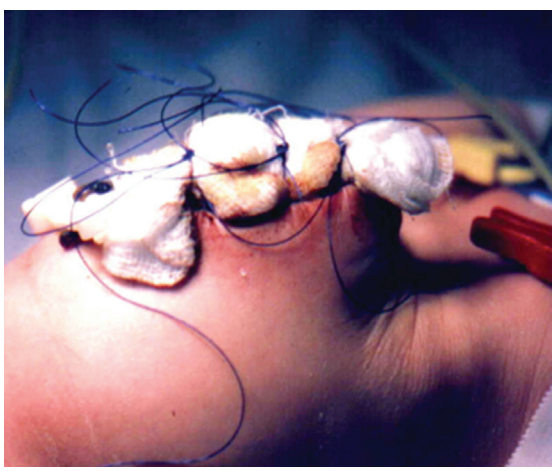

Figure 2 - The herniated organs are reduced by the tightening of the stitches.

Figure 3 - Closure of the skin.

Results. The sac remained intact in all the cases during the initial stage of reduction, no infection or injury to the abdominal contents occurred. While superficial necrosis of the skin occurred in 2 cases, wound infection occurred in another 2 cases, one case had intestinal obstruction for which laparotomy was performed and one case developed small incisional hernia needed later repair. No serious complication was seen related to the procedure.

Associated malformations found in 4 cases. In one case, cystic ileal dilatation, in the second case, patent omphalo-mesenteric duct, while the third case had complex congenital heart disease (atretic mitral valve, hypo-plastic left ventricle, over-riding aorta), the fourth case was associated with Potter' syndrome (hyperbiluribinemia, cleft palate, low–set ears, dysmorphic features, congenital heart disease). And the last case had bilateral undescended testes for which orchidopexy was performed later on.

The mean age of discharge was 25 days. For follow-up, 2 patients were followed up to 2 months, 4 patients were followed up to 6 months, 2 patients were followed up to 2 years. Two patients had associated cardiac anomalies: one patient was transferred to cardiac surgery center for advance cardiac surgery, while the other patient died due to severe cardiac anomaly.

Discussion. Surgical treatment of giant omphalocele is difficult due to discrepancy between the larger volume of the omphalocele and the small abdominal cavity.

The 2 classical strategies: the staged surgical repair and the initially conservative are still adopted. Dunn and Fonkalsrud[17] from Los Angeles, California, used surgical repair by the means of Silo chimney initially in 13 cases with large defects amongst a total of 31 cases. The worst complications of Silastic sheet placement are tension and infection of the fascia with disruption of the suture line. Once infection or premature disruption developed the closure of the defect is very difficult if not impossible. On the other hand, Nagaya et al[8] operated on 17 patients using the silo chimney technique and applied the conservative management with epithelialization in 45 cases, concluded that the conservative treatment was the most reliable therapy for omphalocele.[8] The disadvantages of this management is the long hospital stay (10-19 weeks), the premature separation of the sac and the need for a later repair stage for the giant ventral hernia. Other authors consider conservative treatment of omphalocele is appropriate for very premature infants with acute respiratory distress syndrome or in infants who have other life-threatening conditions.[13]

The target of the new techniques aiming to preserve the sac, is the avoidance of the initial placement of a prosthetic material reducing the risk of infection associated with the foreign body, thus, Bax et al[14] used tissue expander to enlarge the abdominal cavity. However, this technique needs 2 operations, the first is to place the expander in the abdomen and the second operation is to remove the expander and to close the defect of the abdominal wall.

The idea of Hong et al[15] consisting of ligation of the sac sequentially has the advantages of keeping the sac intact in the 3 cases, thus, avoiding the initial placement of a prosthetic material and avoiding first stage operation, but in one case complete reduction could not be achieved after 3 weeks and temporary placement of a silastic sheet was required.

The new concept reported by Brown and Wright[16] using delayed external compression to reduce the omphalocele aims to keep the sac intact reducing the risk of infection associated with the prosthetic material and avoid first stage operation. It proved to be a safe and effective alternative to Silastic silo placement in moderate and large size omphaloceles that cannot be closed primarily.

The technique proposed by us depends on a similar idea, with a different mechanism; the pressure applied by the tightening of the stitches over the sac reduces the herniated contents gradually into the abdominal cavity by a simple manner, at the same time enlarges the abdominal cavity by the reduced viscera, thus, achieving the target of preserving the sac and using it as a support with the advantages of avoiding the use of prosthetic material, and decreasing the number of operations needed from 2 to one .

This technique proved to be simple, safe, effective and inexpensive for gradual reduction of large omphaloceles and may be considered for babies born with this anomaly. However, a controlled study may be needed to provide further evidence on the effectiveness of this treatment.

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