ORIGINAL REPORT

Staged Repair of Giant Recurrent Omphalocele and Gastroschesis "Camel-Litter Method"-A New Technique

Valiollah Mehrabi[1], Arianeb Mehrabi[1,2], Maliheh Kadivar[1], Mehrdad Soleimani[2], Azadeh Fallahi[1], and Nazila Khalilzadeh[1]

[1] *Department of Pediatric Surgery, Children's Medical Center, Tehran University of Medical Sciences, Tehran, Iran*

[2] *Department of General, Visceral and Transplantation Surgery, University of Heidelberg, Germany*

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Abstract- The aim of this article is to present a new Technique of giant omphaloceles repair in neonatal period and also later in life in patients that the primary repair has been failed. From 1999 to 2006, seven consecutive children (male/female ratio 0.4) with giant omphalocele (n=6) and Gastroschesis (n=2) were underwent this new operation in our center. In this technique, there were two operations. The mean of hospital stay was 38 days (range, 23-42 days), and full enteral feeding was achieved on the 8 to 25 postoperative day (Mean, 14 day). The final closure, in all patients was achieved between the 14 to 32 days after the first operation (Mean, 21 day). Mechanical ventilation was necessary for the mean of 5 days (range, 2-8 days). All patients are alive and have no complication due to the operation (1 month-7 years). Giant omphalocele and Gastroschesis can be safely repaired. The placement of an intraperitoneal tissue expander and traction of abdominal muscles can create the needed space for closure in several weeks in patients with giant omphalocele/ Gastroschesis.

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Introduction

Omphalocele vary in size from very small to "giant". Large or "giant" omphalocele occur in 1 out of every 10,000 live birth. "Giant Omphalocele" and " Gastroschesis" (GO & G) implies an abdominal wall defect that is 6 cm or more in diameter and in most of the cases a part of or even the whole liver is within the defect (hepato-omphalocele). Furthermore, other organs such as spleen and intestines might herniate into the umbilical cord. Consequently, there is a considerable loss of abdominal space that poses a reconstructive challenge for the pediatric surgeon.

Technically GO is often difficult to close due to the abdominal wall defect and a staged repair utilizing prosthetic materials is necessary. The morbidity and mortality rate associated with this entity remain high despite the advances in management techniques. Hospitalization is often prolonged and costly. For the best outcome in managing patients with GO early attention to hypothermia and other metabolic requirements and long-term attention to nutritional needs are important. Techniques of closing the omphalocele should be adapted to the individual characteristics of the defect, but mobilization and stretching of the abdominal muscles should begin in newborn period.

The management of omphalocele has been evolved over the past 4 decades. Early primary fascial closure under tension often leads to the complications related to high intra-abdominal pressure (compartment syndrome), compression of the viscera, and elevation of the diaphragm.

If primary fascial closure cannot be easily achieved, the use of a silo technique with staged closure is standard. However, the management of a GO with most of the liver in an experimental position remains problematic. Many strategies have been used, such as Gross's description of skin-flap closure (1), the use of prosthetic material as a fascial bridge under the skin, sequential sac ligation, techniques to encourage epithelialization of the omphalocele membrane, silo application with gradual reduction of the extraperitoneal viscera (EPV), and sequential clamping without

Corresponding Author: Valiollah Mehrabi
Department of Pediatric Surgery, Children's Medical Center, Tehran University of Medical Sciences, Tehran, Iran
Tel: +98 21 22276198, Fax: +98 21 22912742, E-mail: prof_mehrabi@yahoo.com

prosthetic material (2-5). These techniques all describe the relative advantages of non operative initial management in treating GO (6,7).

Fundamental to the problem with GO is the disproportion between the amount of extraperitoneal viscera and the diminutive size of the peritoneal cavity. All the pediatric surgeons have the opinion that the creation of adequate peritoneal space is the primary aim of the treatment. Our effort was focused on presenting a technique to increase peritoneal cavity to the point that the viscera could be reduced in one operation. Most of the previously described techniques often required multiple-staged operation over a protracted period with the goal of gradual reduction of the extraperitoneal cavity simultaneous to progressive increase of peritoneal cavity.

We want to present a new technique in which we used all modalities including tissue expander and dual PTFE mesh, besides abdominal muscle traction in order to overcome the GO & G.

Materials and Methods

Seven Consecutive children (1999-2006) with giant hepato-omphalocele and Gastroschesis (defect >6 cm) were underwent this new operation technique in our center. There were 5 male and 2 female with the age range between 4 days to 13 years (5 were infants and 2 children). All the patients were checked for the possibility of cardiovascular problems and intestinal atresia and we did not find any other simultaneous anomalies in them. With accompanied anomalies were not referred to our hospital or had died in neonatal period. Four patients were primarily operated in neonatal period with this technique and the other three ones were referred to our center after they had several operations with failure in finding permanent solutions for the repair of abdominal wall defect. Our management's outcomes in these patients have been recorded on the basis of inpatients and outpatients records. The clinical situation of the children and also the process of surgical repair were discussed with their parents and the informed consent was taken.

Surgical procedure

Under the general anesthesia, two orthopedic pins (Figure 1b) were inserted into the rectus muscle, 2-3 cm away from defect and parallel to it, a tissue expander was placed into the abdomen via mini- umbilical. A circumferential elastic dressing was fixed around the defect by glue. Several thick non-absorbable sutures were passed and fixed under the pins and the elastic dressing for traction apparatus. The "Camel Litter" (CL) (Figure 1a), has several holes on top for fixation of the sutures and traction which made after the first and second operation (Figure 1c & d). Before complete recovery from anesthesia, patients were put inside the CL. Tissue expander was inflated to the target volume, over 2 to 3 weeks; traction via sutures and pins were applied during the same period of time and was reinforced as much as possible. Mechanical ventilation was used several days after operation whenever it was needed. At the time of second operation for the closure of the abdominal defect, depending on the age of the patients and the time that the favorite abdominal cavity was provided, the tissue expander was removed allowing the reduction of all viscera into the peritoneal cavity. After identification and separation of skin from the edge of abdominal wall fascia, complete abdominal wall closure with or without Dual (Prolene-PTFE) mesh was achieved. Traction and mechanical ventilation was also used for initial recovery phase after second operation. After healing period, weaning from mechanical ventilation and low residue food tolerance, patients were brought to the operating room again and all traction sutures and also the pins were removed. A Gen or elastic bandage was applied for external support as far as maximum healing been achieved.

Figure 1. Devices used in the new technique. a) Camel Litter, b) The orthopedic pins, c & d) designed "Camel Litter" apparatus: the size of apparatus depends on the age of patients and was estimated before the operation day; the device shown in picture c is for an infant and in picture d is for a child.

Figure 2. a) a Gastroschesis, b) hepato-omphalocele and omphalocele in a newborn after the failure of the traditional technique c) recurrent giant hepato-omphalocele in a 5-year old child d) recurrence of a giant omphalocele in a 13-year old girl

Figure 3. a & b) antro-posterior and lateral views of an omphalocele before extension c & d) repositioning the tissue into the abdominal cavity after 3 weeks of retraction and extension e) 6 months after operation

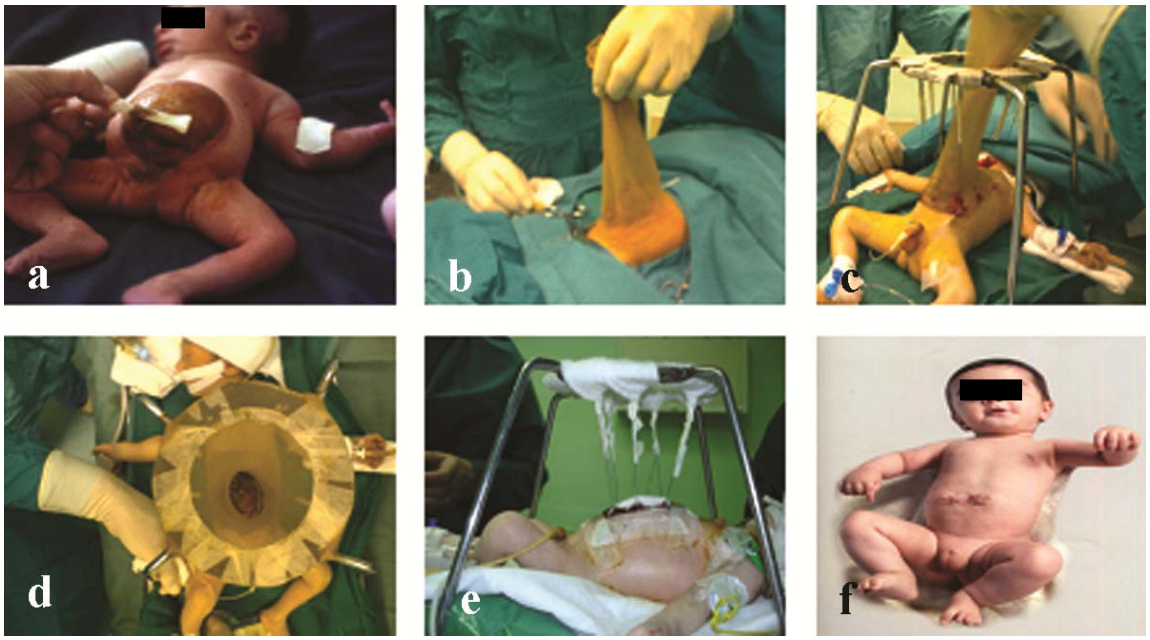

Figure 4-Group A. Technical procedure and the result in newborn; a) hepato-omphalocele before extension b) connection of an elastic dressing around the edge of the omphalocele with glue c & d) extension with Camel-Litter, e) connection of nylon sutures with elastic Gen to the Camel-Litter f) 6 months after the final operation.

Patients were on liquid diet between the two operations for providing maximum room for reduction of viscera. Therefore, the patients could use full enteral feeding several days after the second operation.

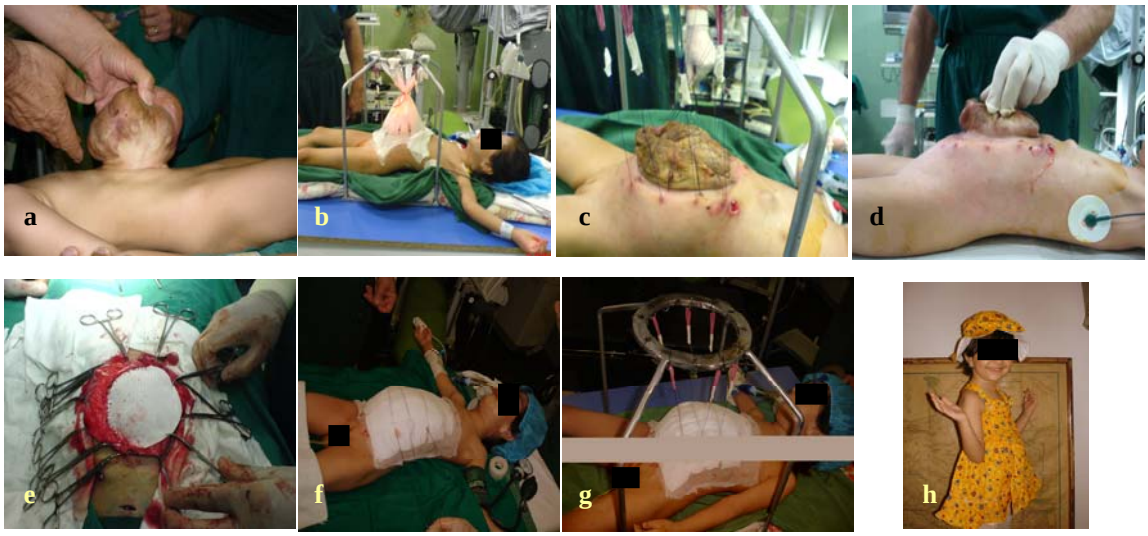

Figure 4-Group B. Technical procedure and results in a 5-year old kid; a) lateral view of omphalocele b) insertion of pins and extension with Camel-Litter c) 3 weeks after extension d) reposition before operation e) insertion of a 6x6 cm mesh f & g) after closure h) one month after the final operation.

Results

The final closure, in all patients was achieved between the 14 to 32 days after the first operation (Mean, 21 day). Mechanical ventilation was necessary in all patients for the mean of 5 days (range, 2-8 days); the mean of hospital stay was 38 days (range, 23-42 days), and full enteral feeding was necessary and achieved after 8 to 25 days (Mean, 14 day). There were no major complications due to surgical procedure and we only confronted with mild erythematic skin and serous discharge from wound that was handled with IV and topical antibiotics prescription. All patients are alive and have no complication due to the operation (1 month-7 years) and three of them are now 4-7 years postoperative with a near normal abdomen.

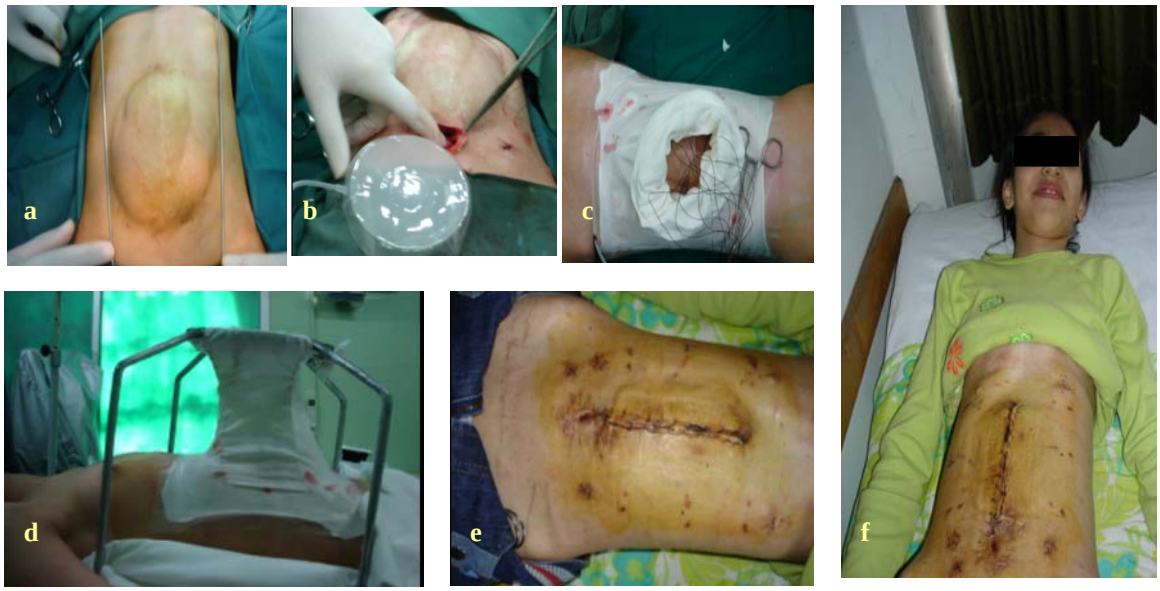

Figure 4-Group C. Technical procedure in a 13-year old girl; a) demonstration of the way of insertion of the pins b) insertion of the tissue expander in the abdominal cavity (the same is necessary in all patients) c) after fixation of the elastic dressing and sutures to the Camel-Litter (above view) d) lateral view of traction with Camel-Litter e) 2 weeks after the operation f) the final result.

Discussion

The outcomes in infants born with omphalocele have improved tremendously in the past 40 years because of medical and surgical advances. The development of neonatal intensive care units, infant ventilator, and hyper alimentation has dramatically improved survival rate. The use of prosthetic material to assist the closure of the abdominal wall was a turning point in the management of these children. Ideally, omphalocele repair would consist of separate primary repairs of both the fascial and skin layers. However, in complicated cases, especially in the management of the child with the GO & G, either neonate or an older child, who has never been repaired or has had a failed repair, any intervention is complex and still remained as a challenging. The fundamental problem with the GO & G is the insufficient peritoneal space in most of the cases.

Generally, two kinds of management can be applied for the treatment of difficult omphaloceles: 1- There are some methods for gradual reduction of extraperitoneal viscera; application of silo with different material and external compression. Silo placement and staged closure is a well-known solution for the mentioned difficult condition. It makes a gradual reduction of the extraperitoneal feasible, by increasing intra-abdominal pressure, causing a slow increase in peritoneal volume. Most of the aforementioned techniques involve a series of operation over several weeks to months to slowly and gradually increase intra-peritoneal space. Often, the patients may not be able to be fed enteraly during this period, needing long-term sedation and artificial ventilation support; also there are potential complications with silo dehiscence, infection, and compromise of blood flow to visceral organs. Furthermore, in patients with GO, there is a potential problem because of angulations or obstruction of hepatic venous and of the retro-hepatic inferior vena cava may result from direct pressure on the hepatic venous outlet after visceral reduction and final abdominal wall closure (20). 2-There are methods to increase peritoneal cavity space without using the viscera as the mechanism to achieve an increase in intra-abdominal pressure. We have chosen to focus on them:

2-a) in some patients with GO, tissue expander had been placed intra-peritoneally (8,14). This might be advantageous in these patients with GO because the peritoneal cavity could be increased without using the viscera as the source of pressure. The use of an intra-peritoneal tissue expander (IPTE) has several advantages: (I) it can precisely control the pressure; (II) it is less invasive than gradually constricting or compressing the silo where the viscera are used to increase intra-abdominal pressure; and (III) with the information from the multi-detector CT scan, one can calculate the extraperitoneal volume and therefore make a reasonable projection of how much volume the tissue expander should be inflated. This technique has worked well both in newborns and also older children. Sander *et al.* reported the usage of elastic bandage and the application of subcutaneous tissue expander to facilitate the primary closure (21). We have used this elastic support for abdominal wall repair in our technique. 2-b) Schuster first described the usage of prosthetic sheet, using artificial material to bridge the abdominal muscle wall defect and then to mobilize skin flaps in order to cover the sheet (2). Different authors reported various techniques of using prosthetic material either to bridge the fascial defect, but not to cover the prosthetic material with skin, or to use the prosthetic material as a silo to allow for coverage of the extraperitoneal volume and subsequent staged closure (22,23). These techniques resulted in favorable outcomes for most of the children. It should be mentioned that, the prosthetic carries an inherent risk of infection. This risk alone may warrant its removal a few years later when primary repair may be accompanied after the child growth.

We described 7 cases of GO & G in whom a new surgical technique was used for closing the abdominal wall. We have got ideas from our colleagues and combined them to find a better solution to resolve the problems in patients that have experiences of several operations that all failed or in cases that was not considered for another operation after the first operation with creation of ventral hernia. The presented technique was traction of the muscular layers of the abdominal wall besides the use of intraperitoneal tissue expander to induce the expansion of abdominal cavity (esp. in lateral dimension), use of prosthetic mesh to overcome abdominal wall defect and continuing abdominal wall support with external traction and elastic dressing to prevent recurrence of hernia and to support the abdominal breathing in kids. The functional and cosmetic results appear superior compared with other suggested treatments used for these abdominal wall defects. A progressive stretching of the abdominal wall and enlargement of the abdominal cavity enabled the defect to be closed 1-3 weeks later. In comparison to the classic method, the presented modification of staged repair of active enlargement of the abdominal cavity makes possible the earlier reduction of the viscera into the abdomen; it does not seem to be associated with a

higher risk of mechanical or infectious complications. In conclusion, GO and Gastroschesis can be safely repaired. However, sophisticated staged operations should be applied with the use of all modalities and facilities. Intraperitoneal tissue expander placement and traction of abdominal muscles can create the needed space over several weeks in GO. Use of synthetic mesh in some cases helps to repair the abdominal wall defect, therefore, eliminate the unwanted and unnecessary rising intraperitoneal pressure and subsequently the chance of repair failure and hernia formation.

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