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Staged repair of giant exomphalos major using tissue expanders

R.S. Shah, S.V. Parelkar, B.V. Sanghvi, R.K. Gupta, K.P. Mudkhedkar

Department of Pediatric Surgery, Seth G.S. Medical College & K.E.M. Hospital, Parel, Mumbai, India

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ABSTRACT

Giant exomphalos, also called hepato-omphalocele, is a major exodus of abdominal viscera. Due to the large discrepancy between abdominal domain and the volume of extra abdominal organs, these defects present a significant challenge to pediatric surgeons. A 10 month old boy with antenatally diagnosed exomphalos major had a giant exomphalos 15 × 15 × 10 cm in size. Investigations revealed significant visceroabdominal disproportion, in view of which staged repair of the exomphalos was planned. An intraperitoneal silicon tissue expander was inserted for this child in the infra-umbilical abdominal cavity with the flat surface in the recto-vesical pouch through pfannenstiel incision & gradually inflated. Subsequently, subcutaneous expanders were placed in both flanks using minimal access technique to get adequate healthy skin cover prior to final ventral hernia repair. At eight years of age, the patient underwent exploratory laparotomy with ventral hernia repair with meshplasty using dual surface mesh & had an excellent and prompt recovery. There are numerous surgical techniques for giant omphalocele closure, which fall into the categories of staged, and delayed closure. Uniqueness of this case is combined use of both intraperitoneal and subcutaneous tissue expansion with the aid of minimal access technique in placement of subcutaneous expanders. The combined use of both intra-abdominal & subcutaneous expanders has not yet been reported in children.

Giant exomphalos, also called hepato-omphalocele, is a major exodus of abdominal viscera [1]. Large or “giant” omphalocele occurs in 1 out of every 10,000 live births [2]. It is defined as a defect that measures more than 6 cm and a sac that contains most of the abdominal viscera including the liver, resulting in significant loss of abdominal domain, visceroabdominal disproportion and an underdeveloped peritoneal cavity. Due to the large discrepancy between abdominal domain and the volume of extra abdominal organs, these defects present a significant challenge to pediatric surgeons [3].

In this report we present a unique method for gradual correction of giant exomphalos using tissue expanders without causing abdominal compartment syndrome.

1. Case report

A 10 month old boy with antenatally diagnosed exomphalos major presented to us after being managed conservatively with povidone iodine dressings elsewhere. On examination he had a giant exomphalos 15 × 15 × 10 cm in size (Fig. 1). Plain X-ray abdomen revealed presence of large soft tissue shadow outside the abdomen (Fig. 2). Ultrasound showed the entire liver, gall bladder, portal vein, hepatic artery, small & large bowel loops herniating outside the abdominal cavity through an abdominal defect of 6 × 4.5 cm with stretched out mesenteric vessels & ascites thus was aptly a giant exomphalos major as defined earlier. CT scan confirmed the findings and CT Volumetry revealed sac volume of 427 cc & abdominal cavity volume of 80 cc (Fig. 3). Because of significant visceroabdominal disproportion, staged repair of the exomphalos was planned. The child followed up at 3-years age for the surgery.

1.1. STAGE I (3-years)

CT Volumetry was repeated and showed persistent visceroabdominal disproportion with sac volume – 1321 cc & abdominal cavity volume – 619 cc.

As the disproportion was very large, we planned expansion of the intraperitoneal space using an expander. An intraperitoneal silicon tissue expander (volume 850 cc, 11 × 9 cm) was inserted for this child in the infra-umbilical abdominal cavity with the flat surface in the recto-vesical pouch through pfannenstiel incision. The injectable chamber was brought out through a separate

E-mail address: rujutas84@gmail.com (R.S. Shah).

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Fig. 1. Clinical photograph at presentation.

incision lateral to the incision & inflated with 25 cc saline on table (Fig. 4). Followed by 65 ml saline inflation weekly for 11 weeks to reach a final volume of 740 ml. On CT Volumetry after full expansion, Volume of Expander was 732 cc & Volume of Exomphalos sac was 2001 cc (Fig. 5). However, the expander had to be deflated by 150 cc in view of tense abdomen, large infected trophic ulcer & left inguinal hernia. At 6-years, CT volumetry showed exomphalos sac = 1284 cc, abdominal volume without expander = 1210 cc and volume of expander = 602 cc. The child was lost to follow up and only returned at 7-years of age with a deflated intraperitoneal expander due to disconnection of port.

Fig. 2. X-ray abdomen erect.

1.2. STAGE II (7-years)

At this time, subcutaneous expanders were placed in both flanks to get adequate healthy skin cover prior to final ventral hernia repair. Silicon rectangular tissue expanders (550 cc each) measuring 13 x 7 x 7 cm were used. Subcutaneous tunnels of 14.5 × 8.5 cm were created using minimal access surgery with 10 mm central & two 5 mm lateral ports. The expanders were then placed in the subcutaneous spaces with the flat end downwards (Fig. 6).

Raw areas over the exomphalos were dealt with by split thickness skin grafts. After 5 months, subcutaneous expanders were removed in view of excessive thinning of skin over the expanders. The intraperitoneal expander had to be removed 2 months after subcutaneous expander removal in view of purulent discharge from the port site.

To summarize:

Age	Abdomen	Exomphalos sac	Expander
10 months	80	427	–
3-years	619	1321	–
4-years	850	2001	732
6-years	1210	1284	602

1.3. STAGE III (8-years)

At eight years of age, the patient underwent exploratory laparotomy with ventral hernia repair with meshplasty using dual surface mesh (Figs. 7 and 8). The contents of the sac were mainly liver which could be easily reposited into the expanded abdomen. The child had an excellent and prompt recovery (Fig. 9).

2. Discussion

An omphalocele is a congenital abdominal wall defect caused by failure of the cephalic, caudal, and lateral folds to fuse at the

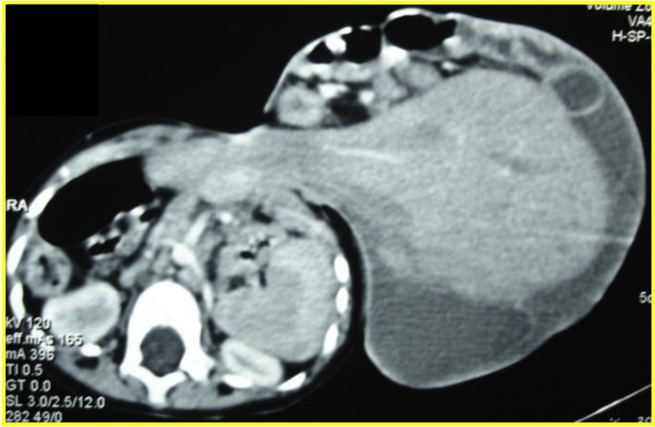

Fig. 3. CT scan abdomen.

umbilical ring during the fourth week of gestation [3] due to failure of the midgut loop to return to the abdominal cavity, after the physiological herniation about the 4th–6th weeks of embryonic life [4]. Due to the anatomical complexity like disproportionate visceroabdominal domains, laterally displaced rectus muscle, abnormally developed liver in exomphalos sac and associated comorbidities, surgical tension-free closure of tissues over the defect remains challenging and problematic [5].

Potential pitfalls encountered when trying to return extra-abdominal organs to the peritoneal domain include wound infections, compression of abdominal viscera or major intraabdominal vessels, and respiratory and cardiovascular compromise because of elevated diaphragms and high intraabdominal pressure [3,6]. G. Belloli et al. reported a case in which external compression of the herniated viscera within the abdomen was carried out with elastic bandaging under careful control of central venous pressure & ventilation followed by delayed primary closure of the defect [7].

There are numerous surgical techniques for giant omphalocele closure, which fall into the categories of staged, and delayed closure. Primary closure can be considered for smaller exomphalos.

A delayed closure involves epithelization of exomphalos sac using silver sulfadiazine cream or other escharotic agents, use of skin flap with delayed ventral hernia repair.

Staged techniques of closure require slow reintroduction of the herniated viscera back into the abdominal cavity using external silo reduction, sequential sac ligation, progressive pneumoperitoneum, vacuum assisted closure systems & use of tissue expanders: subcutaneously, intraabdominally, or intramuscularly [8].

Reports of the use of tissue expanders in the subcutaneous space, intramuscular space, or intraabdominal cavity have illustrated the usefullness of this technique to provide biologic (anatomical) closure of abdominal wall defects [3].

Most traditional methods of treating giant omphaloceles have concentrated on allowing coverage of the defect with a resulting large ventral hernia to be repaired later in life. The basic aim of all the methods described for management of giant omphaloceles emphasizes on decreasing the visceroabdominal disproportion. Because the viscera are fixed, increasing intraperitoneal space becomes the goal and tissue expanders offer the ideal solution to this problem [3]. In addition to visceroabdominal disproportion in patients with giant exomphalos, there is a potential problem of angulation or obstruction of hepatic venous drainage through the suprahepatic inferior vena cava after reposition of the herniated liver [9,10]. Tissue expanders can be used as an additional therapeutic modality in the treatment of these complex abdominal wall defects.

The location for placement of a tissue expander depends on the medical condition and specific reconstructive needs of the patient [1]. INTRAPERITONEAL: Intraperitoneal placement of tissue expanders offers the advantage of creating “tissue flaps” that include all layers of the abdominal wall, including skin, muscle, and peritoneum [3]. The advantages of this technique are that the peritoneal domain can be increased without using the viscera as a source of pressure, one can precisely control the amount of expansion, and it is less invasive than gradually constricting or compressing the silo where the viscera are used to exert increased intraabdominal pressure. Using information from the multidetector CT scan, one can calculate the extra peritoneal volume and therefore make a reasonable projection of how much volume the tissue expander should be inflated [2].

Expansion of the abdominal cavity can cause undesirable effects such as increase in intrabdominal pressure, impairment of respiratory function, and visceral ischemia [11]. Furthermore, the blood supply to the viscera, which is commonly displaced in these patients, is susceptible to kinking and compression [12]. Another concern with placement of a tissue expander within the abdominal cavity is the potential for displacement of viscera into the omphalocele sac [1].

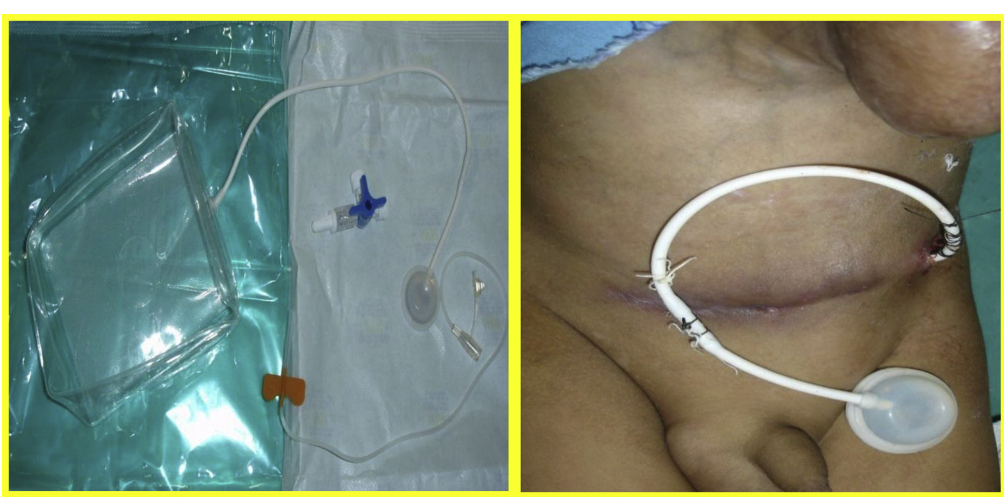

Fig. 4. Intra abdominal tissue expander.

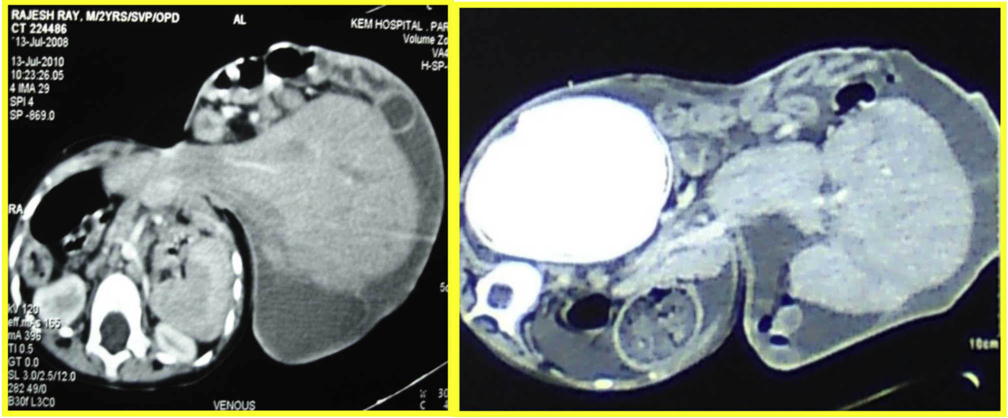

Fig. 5. CT scan following intra abdominal tissue expander inflation.

Fig. 6. Subcutaneous tissue expander placement using minimal access technique.

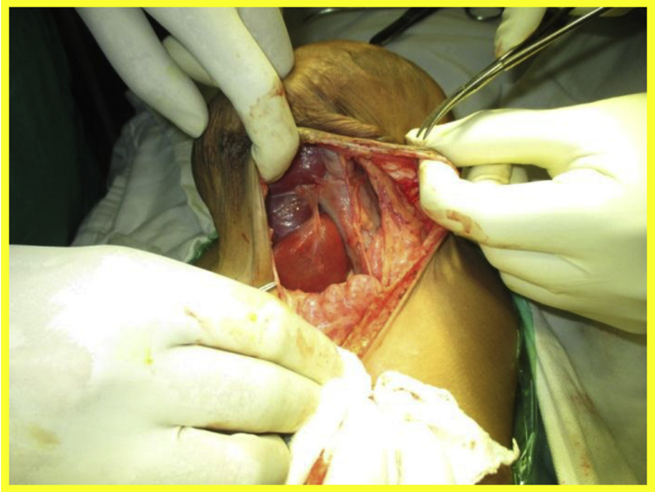

Fig. 7. Intra operative photograph during definitive repair.

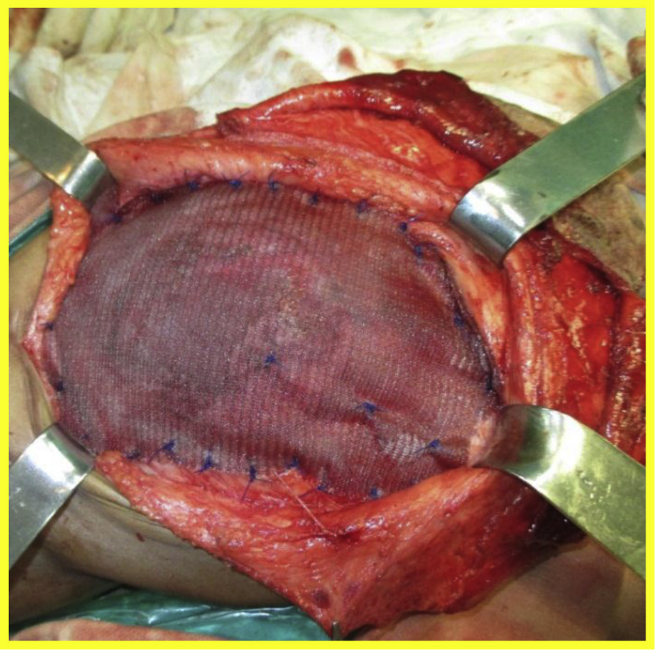

Fig. 8. Meshplasty.

Fig. 9. Post operative at 3 months follow up.

2.1. Subcutaneous/Intramuscular

The placement of crescent-shaped tissue expanders within the abdominal wall in a plane between the internal oblique and transversus abdominis obviates the need for laparotomy and has several practical advantages. An expander with a hard bottom surface is used to help direct pressure on the abdominal wall upward and outward. This helps to stretch the abdominal wall fascia with minimal effect on the intraabdominal pressure. Another benefit of this location is the creation of a 2-layered abdominal wall [13]. In a very small infant, placement of a silo may be still be necessary to allow reduction of the viscera before the skin and muscle flaps could be closed over the defect [3]. In cases of giant omphalocele complicated by respiratory insufficiency, placement of tissue expanders within the abdominal wall helps avoid laparotomy and is a safe and anatomically logical approach for reducing the degree of visceroabdominal disproportion [1].

The relatively superficial location of the expander medially can increase the risk of thinning the skin and result in extrusion. However, this complication can be dealt with safely by early removal and placement of a new expander at a later date. Another potential complication of tissue expander placement within the abdominal wall is disruption of the nerve or blood supply to the abdominal wall. The intercostal nerves travel perpendicular to the incision, and their injury could lead to denervation of the rectus abdominis. No functional abnormalities of the abdominal wall have been reported with this procedure [13].

Progressive pneumoperitoneum technique has also been found to be eminently suitable for setups where ventilatory support, total parenteral nutrition and intensive care are not available. Dr. Gharpure reported 6 patients with this technique in which peritoneal catheter was inserted under general anesthesia & progressive pneumoperitoneum was created and increased using air followed by reduction of contents and closure after adequate expansion [11].

The vacuum assisted closure (VAC) device is another technique which consists of a sponge applied directly to the bowel and liver, covered with impermeable transparent dressing, and attached to a low negative pressure system. It was associated with rapid shrinkage and reduction of the viscera, cleansing of the wound, excellent granulation, maintenance of a sterile environment, and ease of use, with changes possible at the bedside [7].

The main challenges in our case were discrepancy of more than 700 cc between abdominal cavity and exomphalos sac, the entire liver as the main content, and the need to increase the abdominal cavity volume to accommodate structures within the exomphalos sac without causing abdominal compartment syndrome.

Intraperitoneal placement of tissue expander of adequate volume and its gradual expansion increased the volume of abdominal cavity as well as use of subcutaneous expanders helped in obtaining healthy skin cover in our case. Uniqueness of this case is combined use of both intraperitoneal and subcutaneous tissue expansion with the aid of minimal access technique in placement of subcutaneous expanders. The combined use of both intraabdominal & subcutaneous expanders has not yet been reported in children.

3. Conclusion

Giant exomphalos major in children necessitates use of individualized & innovative surgical procedures to deal with the visceroabdominal disproportion in children. We recommend the use of tissue expanders (either alone or in combination depending on amount of visceroabdominal disproportion) as another therapeutic modality in the management of giant exomphalos.

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