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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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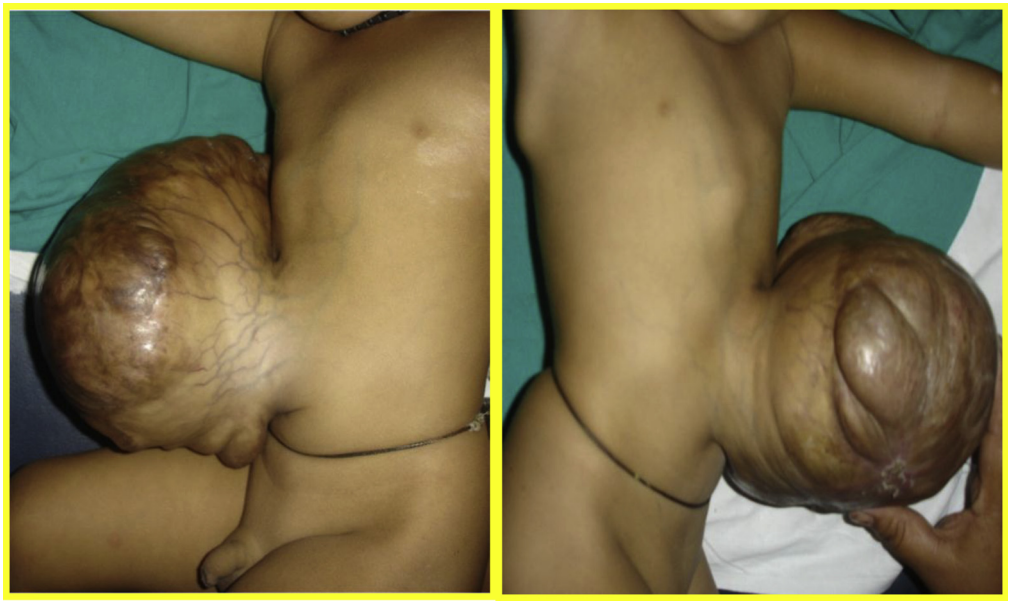

Fig. 1. Clinical photograph at presentation.

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.

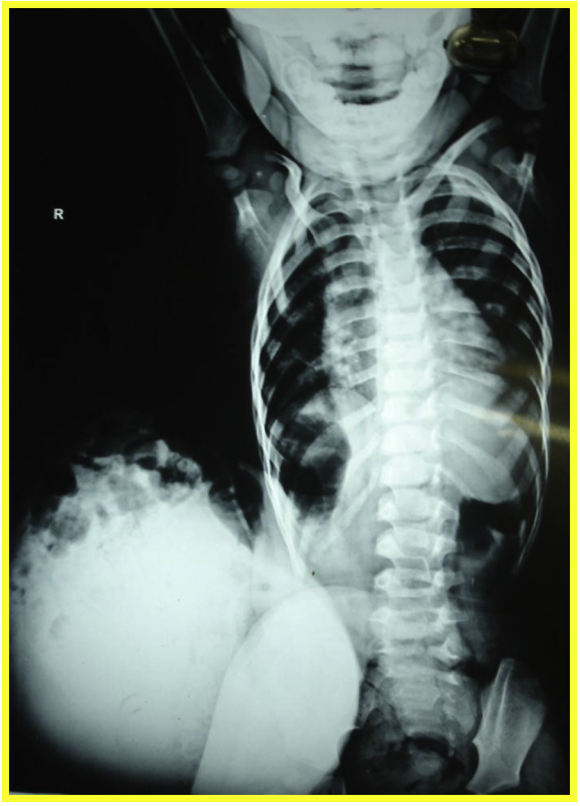

Fig. 2. X-ray abdomen erect.

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