A Systematic Review of Intra-abdominal Tissue Expansion for the Treatment of Exomphalos Major and a Case Report Describing a Refinement of the Technique

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Abstract: The management of complex exomphalos major is difficult, and traditional techniques fail to address the visceroabdominal disproportion in the most severe cases. Intra-abdominal tissue expansion is a novel technique and has been used in a small number of patients to safely increase the intra-abdominal volume and allow the reduction of viscera and subsequent closure of the abdominal domain. We review 7 published reports of this technique and add a case report describing our refinement of the technique. We propose that the use of multiple expanders placed in the intra-abdominal preperitoneal space, when expanded slowly, can allow safe reduction of viscera and immediate direct closure of the musculofascial layer of the abdomen.

Key Words: exomphalos major, giant exomphalos, omphalocele, tissue expander

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An exomphalos (also known as omphalocele) is a midline congenital defect of the abdominal wall, abdominal contents lying within an extra-abdominal peritoneal sac. Its prevalence is estimated at 1 in 3000 births.[1,2] Exomphalos major (or "giant exomphalos") is defined as the abdominal defect measuring greater than 5 cm, containing liver.[1,3] The discrepancy between the volume of the extra-abdominal sac and the underdeveloped peritoneal cavity ("visceroabdominal disproportion") creates a significant surgical challenge.

As testament to the surgical difficulty, various methods have been employed to correct exomphalos major in the neonatal period, ranging from silos (a silastic pouch placed over the contents, which is gradually closed), to topical agents, skin grafts/flaps/substitutes, mesh, and more recently external skin closure systems with or without topical negative pressure.[1] In some instances, initial nonoperative management with delayed repair during childhood is preferred, especially if the majority of the liver lies outside the abdomen, which may cause both impairment of venous return through the inferior vena cava and reduction in ventilatory capacity if the liver is forcibly reduced.[4–6]

A novel surgical approach involves the use of tissue expanders within the abdomen to expand the available volume for reduction of the viscera,[7–11] similar to the slow expansion of a pregnant uterus. A child at the Royal Children's Hospital in Melbourne recently underwent this procedure. Herein follows a systematic review of this reconstructive approach and a report of the modified technique applied in our case. We aim to summarize the techniques used and highlight refinements we found useful to help us achieve total musculofascial closure at removal of the tissue expanders.

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CASE REPORT

At 4 years of age, a male child was referred to the Plastic Surgery Department for assistance with the management of his giant exomphalos. This was initially noted on antenatal imaging, and at birth, the sac contained the majority of his abdominal organs with substantial reduction of his abdominal volume. In the neonatal period, he was managed with silver-based dressings until full epithelialization of the peritoneal sac occurred and subsequently required frequent admissions to manage respiratory infections, gastroesophageal reflux disease, and feeding issues. He was also noted to have asymptomatic unilateral renal tract dilatation. After initial assessment by the Plastic Surgery Department, his definitive management was deferred until he was 6 years of age because of the complexity of the procedure and a need for him to develop the maturity required to tolerate the lengthy expansion process. The exomphalos sac measured 14.8 × 12.7 × 8.7 cm, and the defect in his abdominal wall was 7.4 × 6.4 cm on his most recent computed tomography scan. The contents of the sac included the majority of the liver, the subhepatic inferior vena cava, the spleen, gall bladder, pancreas, and loops of the small and large bowel (see sagital view, Fig. 3).

The size of the sac, and the resultant visceroabdominal disproportion, precluded the use of more traditional surgical approaches, such as staged silo closure.[1,12] The volume of the sac was estimated to be approximately 920 mL. Because of the large volume contained in the exomphalos and the child's lack of abdominal domain, intra-abdominal tissue expansion was considered the preferred technique (Fig. 1).

Four tissue expanders (PMT Corporation, Chanhassen, Minn) were implanted. Three were intra-abdominal in a preperitoneal position. All of these were multistage expanders, which allow greater volume for a similar base size. One was placed in the midline (400-mL capacity), suprapubically, via a midline incision, and one each in the flanks (280-mL capacity), via a pararectal incision, with splitting of the external oblique, internal oblique, then transversalis muscles to maintain the muscular nerve supply. A fourth expander was placed subcutaneously in the epigastric region (200 mL), in case further skin was needed for final abdominal closure. Ports were tunneled to remote, subcutaneous positions (Figs. 2, 3). He was discharged on day 5 postoperatively, mobilizing freely, and without complication.

Tissue expansion began 2 weeks postoperatively and continued weekly for the next 6 months. The patient tolerated only 2 or 3 of the expanders to be injected at each visit, resulting in a total expansion time of 8 months. There were no complications during the expansion phase. With planned overexpansion of the 3 intra-abdominal expanders by 50%, the final combined volume of the intra-abdominal expanders was 1410 mL. Only 58 mL was injected in the subcutaneous epigastric expander (Fig. 4).

At the definitive procedure, a midline approach was used, and the sac skin was dissected from the sac contents. The liver was carefully dissected with identification and protection of the inferior vena cava,

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FIGURE 1. Preoperative and final postoperative photographs.

which was at the superior aspect of the exomphalos. The bowel was mobilized, as was the spleen. Once the sac contents were freed, the intra-abdominal expanders were removed via an intra-abdominal approach, and redundant capsules excised. The sac contents were then easily reduced into the abdomen. The abdomen was closed in layers with tension-free approximation of the linea alba with interrupted sutures (Fig. 5), and onlay Biodesign mesh patch (Cook Medical, Bloomington, Ind) as reinforcement superiorly. The excess skin was excised and then closed directly. A closed suction drain was placed in the prefascial plane, and this was removed on day 4 postoperatively. The operation and immediate recovery were uneventful; in particular, he suffered no pressure-related respiratory or intra-abdominal complications. He was discharged on day 6 postoperatively, and at that time point, he was mobilizing freely returning to school at 2½ weeks postoperatively (Fig. 1).

He re-presented within this period with a mild radiologically confirmed communicating hydrocele, which was managed conservatively.

SYSTEMATIC REVIEW

Methods

A PubMed search was conducted using the following title queries: "tissue expander exomphalos" and "tissue expander omphalocele." This search yielded 15 articles. Bibliographies of captured articles were examined to select articles not identified in PubMed, resulting in 2 further articles. One author selected articles from the abstracts and obtained full articles for the review process. Articles selected were in English and were original articles describing 1 or more cases of exomphalos treated with intra-abdominal tissue expanders. Our patient's parents provided written informed consent to be included in this study. A common data set was extracted from the articles, focusing on patient demographics, clinical features of the exomphalos,

FIGURE 2. Surface markings indicating placement of tissue expanders at the conclusion of the first-stage procedure.

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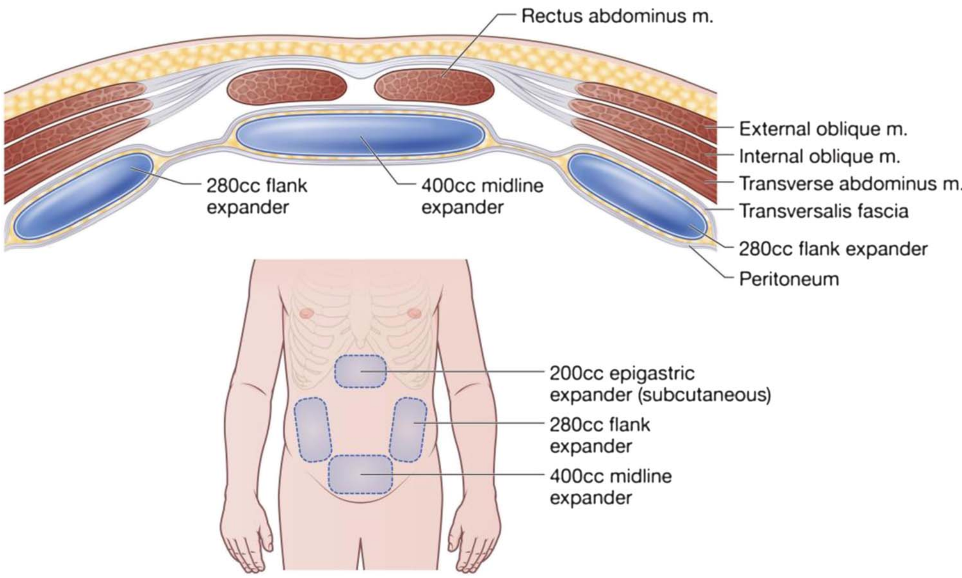

FIGURE 3. Schematic representation of the locations and planes of the expanders.

and finally surgical strategy and outcomes. No funding was involved in this investigation.

RESULTS

A total of 6 articles met the criteria (Fig. 6). These articles were either case reports or small retrospective case series. Two of these articles, published in different journals by separate authors, describe the same 2 patients.[10,11] One published report of 5 cases was not included as the details provided were insufficient for thorough review, and the tissue expansion was only a small part of the surgical process, which predominantly involved an external abdominal muscle traction ("camel litter") apparatus.[13] Another case was excluded because the patient was lost to follow- up after insertion of the tissue expander. Upon representation, they had a deflated expander secondary to detached port; thus, the intra-abdominal expander was not integral to their eventual meshplasty reconstruction.[14] The case described above was included in the systematic review. Table 1 summarizes the cases with respect to demographics, technique used, and outcomes.

Demographics

There were 8 cases in which intra-abdominal tissue expanders have been used in the treatment of exomphalos major. Three patients were younger than 1 month at their first-stage surgery, and the remaining 5 patients were 8 months, 20 months, 4 years, 6 years, and 7 years old. These patients had either tried and failed more conventional treatments (ie, silo) or had been treated nonoperatively.

Surgical Technique and Outcomes

All previously published cases have similarities with respect to stages of repair, placement of the expander, and timing between stages. They have all had a single tissue expander placed within the peritoneal cavity, generally in the lower abdomen or pelvis with or without additional subcutaneous expanders. This has been achieved via minilaparotomy or

FIGURE 4. Coronal and sagittal computed tomography showing the size of the filled intra-abdominal and subcutaneous (epigastric) tissue expanders.

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FIGURE 5. After removal of the tissue expanders at the second-stage procedure, the musculofascial layers are easily opposed without tension or bridging mesh.

laparoscopic surgery. The expansion in the majority of cases took place over a period of 3 to 4 weeks; only Adetayo et al[7] differed in that they describe a 5-month period of filling. Tenenbaum et al[11] and Foglia et al[10] describe using computerized tomographic volumetric scanning as a guide to ensure the expanders volume exceeded that of the sac by 20%. The total expander volume in the previously reported cases was between 180 and 1000 mL. Our case was unique, in that 3 separate preperitoneal expanders were placed, and expansion was over an 8-month period, to a total volume of 1410 mL.

In all previously published cases, either 3 surgical stages have been required to achieve direct closure of the abdominal fascia, or a hernia or alloplastic mesh bridging has been accepted as the final result after 2 stages. The first stage involves placement of the expander or expanders; the second stage, partial or complete reduction of the extra-abdominal viscera but without abdominal fascial direct closure; and the third stage, if performed, reduction of the residual hernia and direct closure of the fascia. The currently described case was the only one in which the abdominal musculofascial layer was closed in 2 stages.

The published reports typically describe a period of mechanical ventilation, either after the placement and on-table fill of the expander, or in the immediate postreduction period. There are no documented cases of major complications secondary to the tissue expanders. One of the patients detailed by Martin et al[8] remained ventilator-dependent 10 months following the surgery, but this was related to their concomitant severe pulmonary disease of prematurity (bronchopulmonary dysplasia and pulmonary hypertension requiring a tracheostomy).

DISCUSSION

Exomphalos major presents a challenging combination of underdeveloped abdominal domain, deficient abdominal wall components, and sparse tissue available for recruitment. Primary closure is seldom feasible because it causes an acute rise in intra-abdominal pressure, as well as obstruction of venous drainage if the extraperitoneal sac involves the vena cava. Furthermore, these patients often have a small thoracic cavity and a resultant pulmonary hypoplasia[9] that is sensitive to increased intrathoracic pressure.

Tissue expanders present a reconstructive tool that are more commonly used to increase the length of tissue, but in this situation, they are used to create extra volume within the abdomen. Traditional subcutaneous placement does not address the abdominal musculofascial wall deficiency and so cannot contribute to successful closure of the abdominal wall.

There have been 6 reported cases of intra-abdominal tissue expansion for the management of exomphalos in the literature to date.[7–11] In the previously reported cases, the operative sequence has involved 3 stages to achieve abdominal fascial direct closure, or the residual fascial defect patched with alloplastic material and accepted as the final result. This is the first case described in which complete closure has been achieved in 2 operative stages. We attribute this to the number, size, and location of the expanders and the gradual inflation of the expanders over a long period.

The suprapubic area is the most effective location for expansion and the most important expander to place correctly. It is the one that mimics an expanding uterus in pregnancy. We supplemented this with 2 flank expanders placed to create extra abdominal width that these children are lacking. All 3 expanders were placed extraperitoneally. The fourth, subcutaneous expander, was placed in case extra skin was required, and in retrospect was of little benefit. We feel the preperitoneal placement of the expanders affords less danger to the intra-abdominal organs and is a very easy plane to develop surgically without causing denervation of the abdominal musculature. Using bellowed expanders (PMT Corporation) and over-expanding them by up to 50% allow greater expansion for the same base size. These are important requirements because of the lack of intra-abdominal size to accommodate large expanders without folding. The total volume of the expanders is the most important factor in planning these procedures and needs to be greater than the volume of the exomphalos to allow tension-free direct closure of the abdominal musculofascial layer on removal of the expanders.

Gradual expansion of the expander is the other important factor both in increasing the volume of expansion that can be achieved and in allowing the lungs to gradually adapt to the change in intra-abdominal volume. The time between tissue expander insertion and removal in previous cases has generally been less than a month,[8–11] but in our case, it was 8 months. The process of tissue expansion involves an immediate stretch ("mechanical creep") followed by actual production of extra tissue components ("biological creep"). Biological creep relies on a longer period of tissue expansion, and new collagen, for example, may not be seen in histological capsule specimens until the seventh week of expansion.[15] While there is no exact time point at which "permanent expansion" takes over from "temporary stretch," it would seem sensible to allow more time for expansion if one endeavored to create more abdominal space without abdominal organ compromise and to maximally avoid immediate recoil of tissue.

CONCLUSION

The surgical dilemma of exomphalos major management relates primarily to visceroabdominal disproportion. Increasing the volume of the abdominal domain is the ideal solution. Our refinements of intra-abdominal expansion focus on the use of multiple expanders in the preperitoneal plane, expanded over a prolonged period of months.

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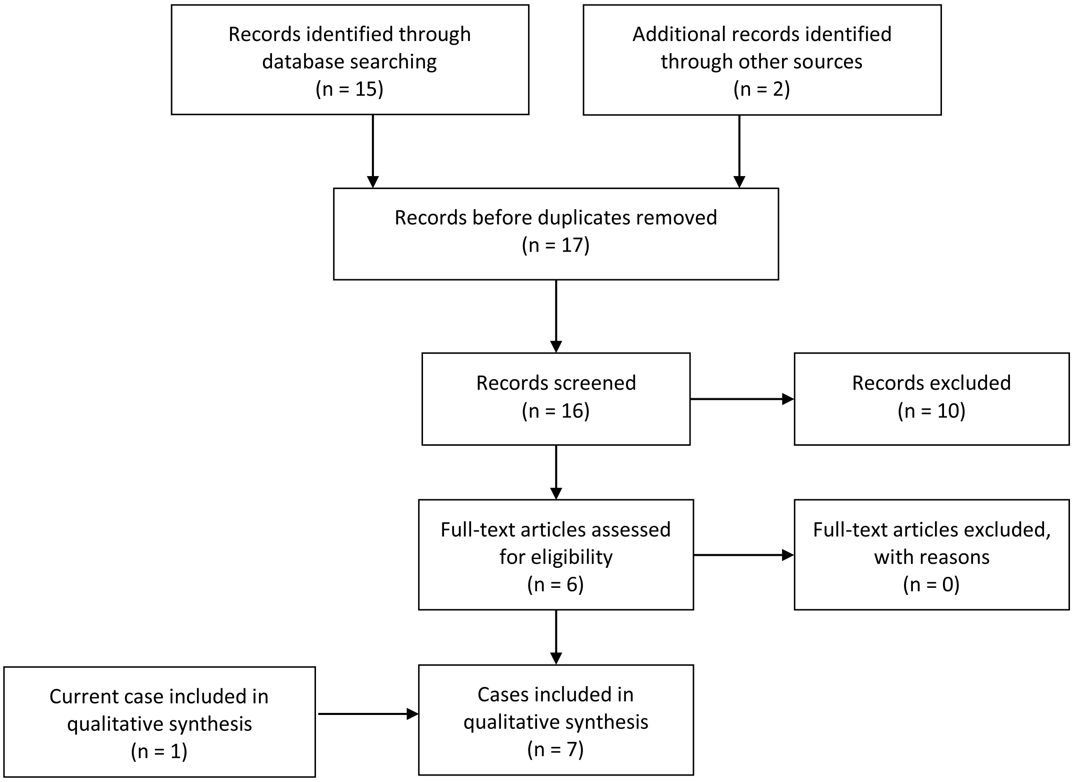

FIGURE 6. Citation attrition diagram for review of the literature.

TABLE 1. Summary of All Cases Where Intra-abdominal Tissue Expansion Has Been Used for the Treatment of Exomphalos Major

Case	Author	Age at 1st Stage	Placement of T/ex	T/ex #	Insert ➔ Remove	Volume, mL	Fascia Closed	Stages	Other
1	Bax et al,[9] 1993	3 wk	Intraperitoneal Pelvic	1	19 d	250	Unclear	2	
2	Foglia et al,[10] 2006 Tenenbaum et al,[11] 2007	Newborn	Intraperitoneal Pelvic	1	20 d	300	Yes	3	
3		4 y	Intraperitoneal Pelvic	1	1 mo	900	No 2 × 7-cm Alloderm (LifeCell Corporation, Branchburg, NJ)	2	
4	Martin et al,[8] 2009	8 mo	Intraperitoneal Pelvic	1	1 mo	1000	No Alloderm patch	2	Still on ventilator dependent 11 mo postoperatively, awaiting ventral hernia closure
5		2 wk	Intraperitoneal Pelvic	1	22 d	180	Yes 2nd stage 2.5 × 2.5 cm	3	
6	Adetayo et al,[7] 2012	20 mo	Intraperitoneal Pelvic	1	5 mo	1000	Yes	3	
7	Authors' case, 2020	6 y	Extraperitoneal	3	8 mo	1410	Yes Onlay Biodesign mesh patch (for reinforcement)	2	Minor hydrocele managed conservatively Return to school 18 d postoperatively

T/ex, tissue expander.

This approach allowed direct closure of the abdomen on removal of the expanders and a successful recovery without complication.

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