CASE REPORT

Neonatal giant exomphalos: A staged approach by external silo and DuoDERM dressing reductions followed by delayed primary repair

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ABSTRACT

We present a 2,440 g male neonate born by caesarian section at 38 weeks of gestational age. Baby had been diagnosed with giant exomphalos during prenatal scans. Due to the giant size of the exomphalos with liver being sac content to more than 50% and severe degree of abdominovisceral disproportion, the decision was made to adopt a staged-treatment approach. We created an external silo supplemented with DuoDERM compression dressings and adjusted it over three weekly sessions. The exomphalos was completely reduced, and the patient underwent delayed primary closure A modified single-layer abdominal wall repair was carried out uneventfully. The post-operative period was uncomplicated and at follow-up 4 years later the patient had no incisional hernia and is thriving well.

Key Words: Abdominal wall defects, Congenital, Delayed primary closure, DuoDERM, Giant exomphalos, Nonsurgical staged closure, Neonatal, Minimal invasive technique

1. INTRODUCTION

There are several types of abdominal wall defects in children, among the most significant of which are: (1) central midline defect with abdominal organs herniated in a sac, i.e., exomphalos; and (2) right paraumbilical defect (rarely, left) without a sac ie gastroschisis.[1–10]Exomphalos is a rare and infrequent congenital anomaly with no standard management strategies. This case concerns a male neonate with giant exomphalos, diagnosed prenatally. We describe procedure and clinical outcome with the aim of demonstrating the ease, safety and efficacy of a staged technique, being sequential gradual reductions using an external silo with DuoDERM dressings for three weeks followed by early neonatal primary fascia closure.

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2. Case report

A prenatally-diagnosed giant exomphalos in a boy of 2,440 g born at 38 weeks of gestation by caesarian-section after an uneventful pregnancy and with normal APGAR scores at birth.

At birth, the baby was active, pink, breathing spontaneously, without any need of supplementary oxygen or ventilatory support and with stable vital signs but exhibiting a giant exomphalos of 15 cm diameter with liver and bowel loops visible within the sac. The rest of the physical examination was normal except for a deformed left ear (see Figure 1A).

The laboratory investigations were all within normal limits. Postnatal ultrasound confirmed giant exomphalos with major content being the liver. No other associated congenital abdominal anomalies were found. Chest radiograph, ECG and echocardiogram were all normal.

Due to the giant size of the exomphalos, liver being > 50%-75% content, the decision was made to create an external silo with DuoDERM compression dressing followed by early neonatal primary fascial closure as a staged approach.

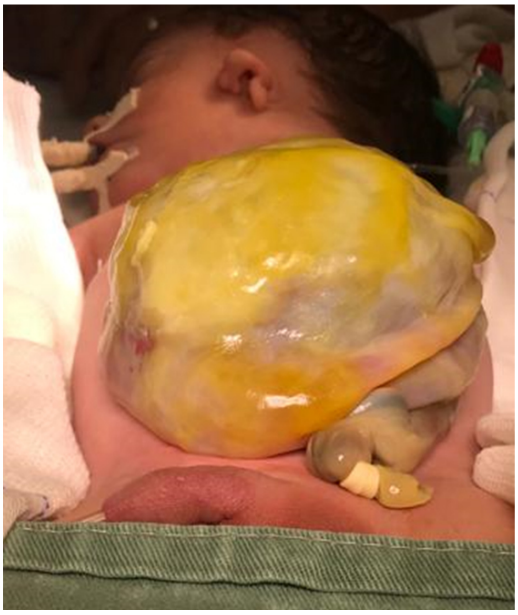
The baby had roomed in with parents to allow bonding, requiring no mechanical ventilation or supplementary oxygen, was on full enteral feeding, and had minimal nonaggressive handling. Vertical silo allowed traction vertically with reduction through gravity, compression of the DuoDERM dressings enhanced surrounding pressure from all sides and the umbilical clamp at the top of the silo contents forced the content to go only one way into the abdominal cavity slowly and steadily. This allowed early and safe reduction at permissible pressures over weeks rather than days. General anesthesia was used to relax the abdominal wall and diaphragmic muscles and to measure intraabdominal pressures and ventilatory settings allowing good spontaneous breathing to minimize the risk of abdominal compartment syndrome. The aim was to assess the ventilatory pressures and stop the reduction at maximum permissible level allowing safe reduction and be evidence based.

At the first stage a vertical external non-surgical silo was applied for the assistance of gravity in gradually reducing the contents was placed at right angles to the abdominal cavity to avoid any secondary injury to the bowel due to kinking at the abdominal wall level (see Figure 1B).

Once the silo was created, a compression DuoDERM dressing was applied to provide lateral compression so that the contents remained in the vertical plane while reducing into the abdominal cavity. The umbilical clamp applied at the top of the contents maintained for a consistent pressure conducive to early reduction (see Figure 2).

For the following 3 weeks, the silo was gradually reduced and DuoDERM dressing changed weekly in the operating room under general anaesthesia under strict aseptic precautions for the reasons mentioned previously. This allowed gradual reductions of the contents over three weeks (see Figure 3).

A Giant exomphalos with large liver and deformed left ear



B First nonsurgical external silo application

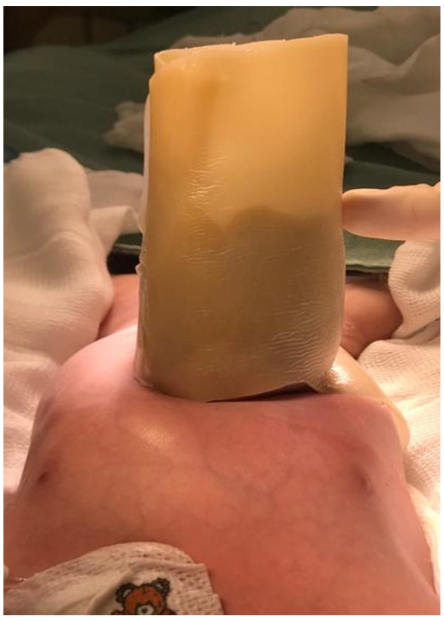


Figure 1. A. Photograph with giant exomphalos and deformed left ear; B. External non-surgical silo application in the operating room.

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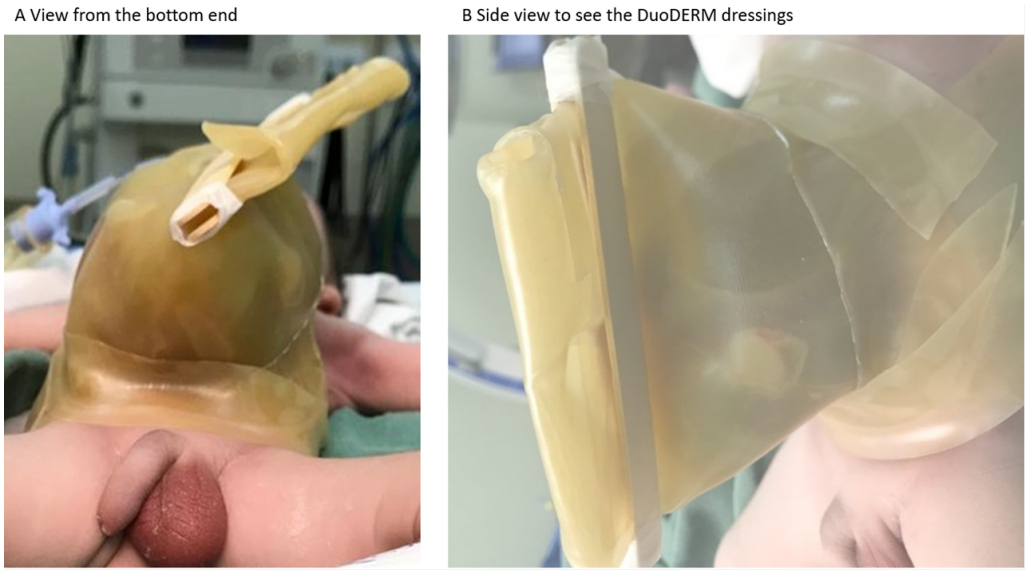

Figure 2. DuoDERM dressing application. A. front; B. side view

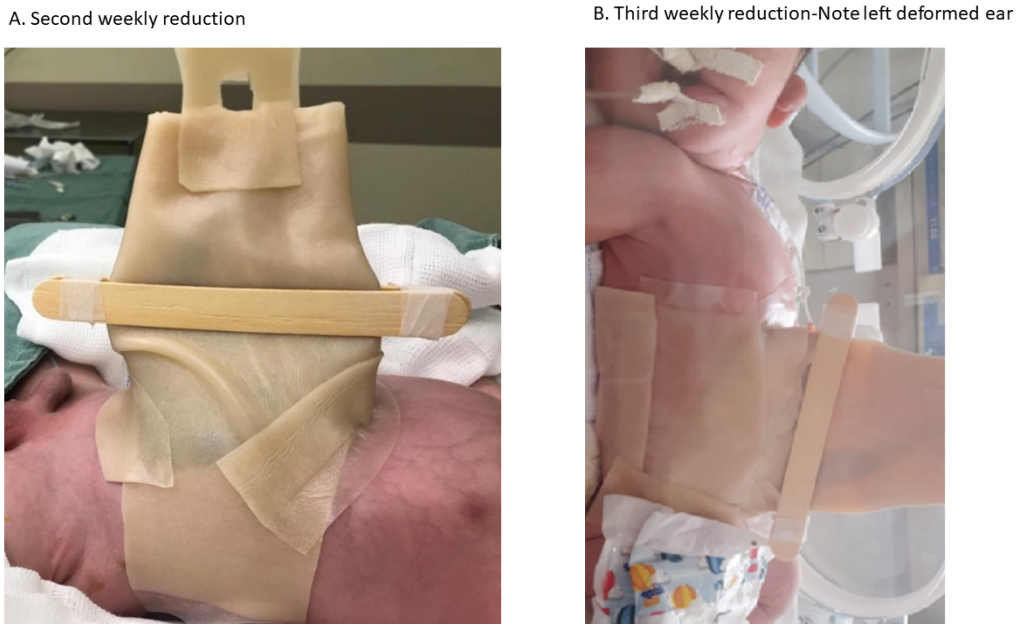

Figure 3. A. Second; B. Third week reductions with DuoDERM dressings application

Three weekly gradually reducing DuoDERM dressings allowed the herniated contents to be fully reduced into the abdominal cavity. The last silo was a flat DuoDERM coverage, as the abdominal cavity has had accommodated all of the abdominal content (see Figure 4A) without any tension in any dimensions (see Figure 4B). The DuoDERM dressing was removed and the abdominal wall being lax and without any tension. The silo and DuoDERM dressing reduction were followed by delayed primary neonatal fascial closure.

The Wharton's jelly acts as a perfect medium for growth of microorganisms especially when the outer layer of the amnion has dried up after losing its placental blood supply. By contrast, the inner layer is the parietal peritoneum which is supplied by blood vessels and is a very strong barrier to infection and rupture. We, therefore, prefer to remove the outer dried up avascular amniotic layer and the Wharton's jelly to avoid inflammation and subsequent infection and to use the parietal peritoneum to close the abdominal wall.

The modified single layer abdominal closure is now well accepted as the best method of closing the abdomen as it not only includes both facial layers of rectus abdominis sheath but also the parietal peritoneum and the linea alba remnants.

This was followed by a separate skin closure using subcuticular sutures to avoid scaring and cosmetically looks good. Minimal undermining to avoid dead space and postoperative collections was used when undermining the skin from the rest of the abdominal wall. Ventilation was only used while attempting optimal reduction, the patient being mainly kept on spontaneous breathing and to assess the ventilatory pressures while attempting more reduction (see Figure 5).

The patient was kept intubated under sedation for 24 hours and prophylactic antibiotics were given for 48 hours postoperatively. The full procedure of external silo application combined with DuoDERM dressings followed by delayed neonatal primary closure was uneventful. The baby was discharged home at 40 days of life. The baby was stable clinically, his upper abdomen was distended but soft and non-tender.

The patient was well and remained asymptomatic at immediate postoperative follow up clinic and at follow up 4 years later. There was no evidence of gastroesophageal reflux disease symptoms, ventral midline, incisional hernias, or the inguinal hernias. The chest radiograph with both domes of the diaphragm were normal (see Figure 6).

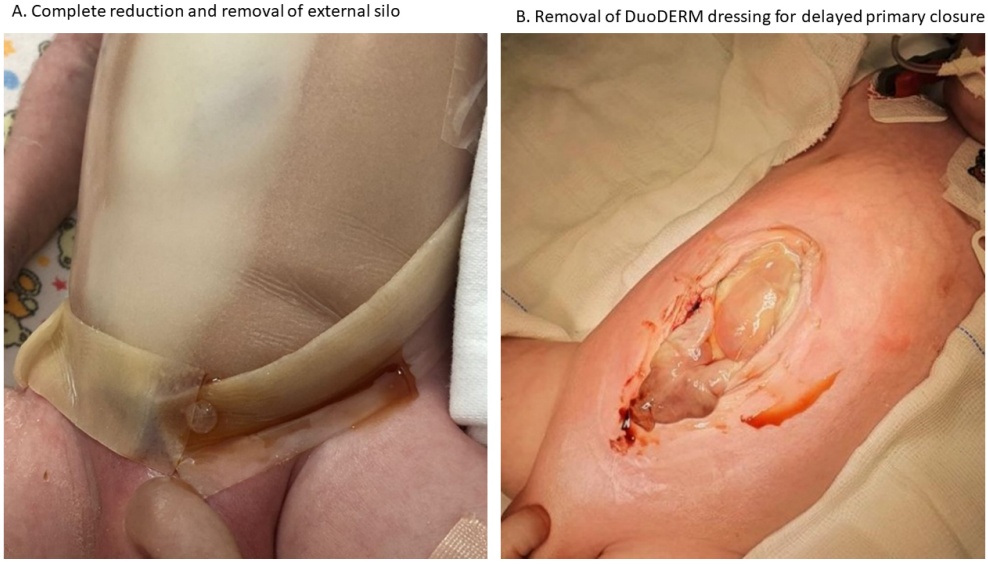

Figure 4. Final appearance after weekly applications. A. Note the complete reduction of contents into abdominal cavity; B. without any tension.

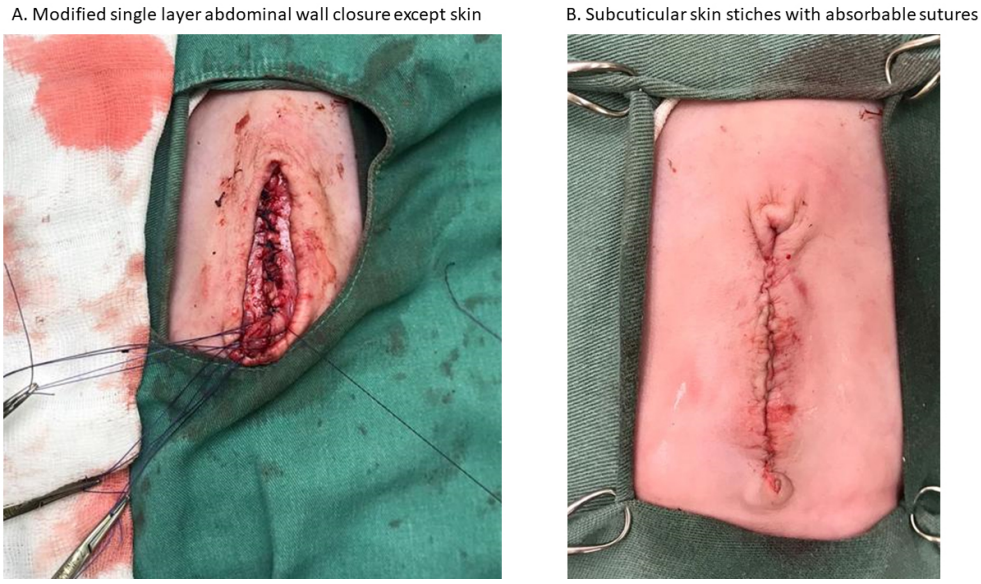

Figure 5. A. Modified single layer abdominal wall closure using interrupted sutures; B. Skin closure using absorbable continuous subcuticular stiches

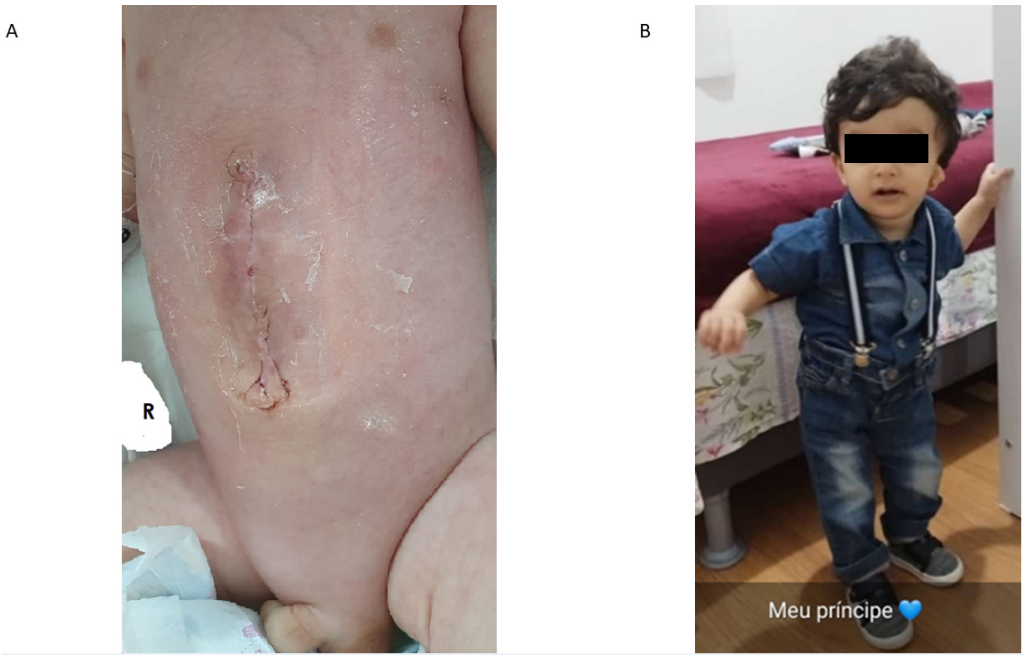

Figure 6. A. postoperative clinic follow up-Note neo umbilicus at the lower end of the scar and well healed midline abdominal scar; B. Follow up at 4 years

3. Discussion

Giant Exomphalos is a complex congenital condition associated with many anomalies and there is no consensus about its accurate diagnosis, classification, treatment protocols and its ideal management. All cases of exomphalos, regardless of size, should be delivered at a specialist tertiary center where expert obstetrics, neonatal, neonatal surgical and NICU care teams are available for the safety of the mother and the baby.

Congenital central midline abdominal wall defects have a spectrum ranging from umbilical cord hernia to giant exomphalos.[8, 11] These congenital anomalies are classified on the basis of percentage of liver as content in the sac, size of the defect, the visceral abdominal disproportion (VAD) and associated malformations. Classification has diagnostic and prognostic significance. Our proposed classification, as shown in Table 1 below, can help in decision-making.

Table 1. Proposed classification of central congenital umbilical anomalies

Anomaly	Liver as sac content	Defect size	VAD	Associated malformations
Hernia of the umbilical cord	None	< 2.5 cm	None	None
Exomphalos minor	None	2.5-5 cm	None	Infrequent, insignificant
Exomphalos intermediate	Edge of the liver, < 25%	5-7.5 cm	Mild	Few, Minor
Exomphalos major	25%-50%	7.5-10 cm	Moderate	Many, Major
Giant exomphalos	> 50%	> 10 cm	Severe	Several, Severe

The main parameter is the percentage of liver as the content of the sac and the size is secondary parameter, abdominal visceral disproportion is the tertiary parameter and associated malformations is the quaternary parameter which may be variable. For a giant exomphalos with > 50%-75% liver as content, minimal acceptable size of the defect is more than 5 cm.

In giant exomphalos, due to the massive herniation of abdominal contents, the abdominal cavity is under-developed. The consequent significant visceral-abdominal disproportion prevents safe and effective primary closure due to the risk of abdominal compartment syndrome and of ventilatory difficulties. Giant exomphalos remains a challenge in neonatal surgery but the advent of prenatal diagnosis now allows for termination of pregnancy in those cases where severe associated anomalies are incompatible with life.[12] The choice and timing of treatment is dependent on the cardiorespiratory reserve of the neonate, number and severity of the associated anomalies, the size of the hernial defect and the severity of viscero-abdominal disproportion.

The established medical treatment of giant exomphalos is an initially conservative approach, being paint-and-wait with later repair of the ventral hernia. This has the limitations of a prolonged period of wound care, infections, rupture of the sac, very difficult repair of the ventral hernia risk of severe adhesions and of bowel or liver injury resulting in hemorrhage.[13]

The second approach is the primary repair at birth. However, factors such as the abdominal-visceral disproportion in newborns, the large diameter of the abdominal wall defect, presence of substantial liver tissue in the sac and of other organ co-existing anomalies make early surgical treatment impractical and requires not only a skilled multidisciplinary approach and but, possibly, greater resources than other options.[14]

The aim of any surgical treatment is to repair the fascial and skin defects while avoiding an intolerable increase of intra-abdominal pressure leading to catastrophic abdominal compartment syndrome. In general, treatment strategies can be classified as the following: primary neonatal repair immediately after birth often inadvisable due to high risk factors;[14] staged-repair with delayed primary closure; delayed repair (paint-and-wait) with secondary closure of abdominal wall hernia.[15]

Recently, a retrospective and multicentric cohort study using hydrocolloid dressings have been reported.[16] However, in this study only 21 female babies were included. Female babies have wider and spacious true pelvis than boys.[17] This fact, together with those placental, maternal and female baby hormones present in the neonatal period which allow muscles to relax, permit the sac-contents to be reduced more easily than in boys. The reductions were done in days as opposed to weeks but, as the growth of the abdominal cavity takes some time, necessitated the use of absorbable and synthetic mesh. Use of such mesh has an associated risk of additional morbidity. Based on our extensive experience and evidence base with gastroschisis challenges, the reductions are easier and faster in gastroschisis due to bowel contents while in giant exomphalos solid liver occupies over 50%-75% as content which takes time to expand and grow abdominal cavity.[1,2,5,7,8,10,18,19]

Despite our advocacy that neither immediate neonatal primary repair in the neonatal period nor paint and wait with later repair should not be adopted, we note that an interesting and novel approach for delayed primary staged closure in the neonatal period using vacuum-assisted reduction and botulinum toxin injections in the course of such treatment has been described, and which might be helpful in selected patients.[20,21]

We believe that if safe reduction of giant exomphalos is to be reliably achieved then closure should not be expected in a day, as in primary closure, or in a few days, as in the female-only cohort study, or as early as is possible in gastroschisis, which is a similar procedure and one perhaps more commonly experienced by paediatric surgeons. The material being reduced being soft and relatively-mobile bowel-and-contents able to expand and fill the cavity rather than, as is the case of giant exomphalos, some 50%-75% of the sac-content being solid liver, which takes time to expand and grow the abdominal cavity.

4. Conclusion

In conclusion, we found it possible in neonates with giant exomphalos to reduce the length of hospital stay through a method which offers faster resolution, namely, a combination of reductions performed at weekly or flexible individualized interval using an external silo with DuoDERM dressings followed by delayed neonatal primary closure once the contents are safely reduced in the abdominal cavity. Our method is, non-invasive, safe, effective and optimizes earliest-possible discharge. We advocate a staged delayed repair as this is easy, very effective, can be repeated in any settings and can be completed in a few weeks without associated mortality or morbidity.

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Authors contributions

All authors contributed to the study, study's conception and design, prepared materials, preparation, collected data collection, and performed analysis were performed by all authors. The first draft of the manuscript was written by RP, and all authors commented on or edited previous versions of the manuscript. All authors read and approved the final manuscript.

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