Operative Techniques

Management of giant omphalocele with a simple and efficient nonsurgical silo

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ABSTRACT

Introduction: Giant omphaloceles can be a challenge for pediatric surgeons and neonatologists worldwide. It is a rare and low-frequency congenital anomaly with no standardized management schemes or treatment protocols. Over the past few decades, we have developed a simple and efficient staged management for giant omphaloceles that allows definitive closure in the neonatal period, the results of which we outline in this report.

Material and methods: With IRB approval, a retrospective and multicentric cohort study was carried out between 1994 and 2019 with patients with giant omphalocele defined as an abdominal wall defect greater than 5 cm in diameter and/or that contains more than 50% of the liver within the sac. We included all patients managed with the nonsurgical silo technique. Data on demographics, gestational age, associated malformations, amniotic fluid reduction and inversion time, anatomic closure, requirement of a mesh, intra- and post-silo complications, mortality and follow-up were collected. The technique consists of the construction of a silo with an adhesive hydrocolloid dressing (Duoderm®) to achieve an omphalocele staged-reduction until complete abdominal reintegration of the liver and bowel preservation of the amniotic sac. This also enables the simulation of abdominal closure before definitive surgical closure, being managed in the neonatal intensive care unit (NICU).

Results: Forty patients, 21 of whom were female, were managed with this technique. The average weight was 2900 gs (890–3900), and the median gestational age was 38 weeks (28–40). In total, 37.5% of cases had an associated comorbidity. The average silo reduction time was 7.3 days (0–35), the average time of amniotic fluid inversion was 5 days (2–9), and the average time to closure was 14.6 days (6–38). Anatomical closure was achieved in 95% of cases. In 4 patients, an absorbable mesh was used to reinforce the anatomical closure, and in 2 patients (5%), a mesh (Dualmesh®) was required to achieve an abdominal closure. There was no mortality associated with this nonsurgical silo technique. The average follow-up time was 60 (6–288) months.

Conclusion: The staged silo management of giant omphalocele in this series is safe and effective and reduces the time to closure and potential morbidity and mortality compared with traditional surgical or medical management.

1. Introduction

Omphalocele is a congenital midline abdominal wall defect at the base of the umbilical cord with a sac containing the herniated contents. Since herniation occurs early in gestation, there is often an underdeveloped abdominal cavity [1].

Its incidence is estimated to be 1:6000–10,000 live births and is probably progressively declining due to termination of pregnancy of patients with complex genetic disorders and malformations [2].

Omphalocele is commonly associated with additional congenital anomalies (30–40%), such as cardiac malformations and chromosomal disorders (Beckwith–Wiedemann syndrome, trisomy 13 and 18) [3].

Early prenatal diagnosis allows further workup to establish prognosis and a delivery plan in a fetal center with pediatric surgeons and neonatal intensive care support available [4]. The main causes of perinatal death are associated malformations and polyhydramnios [1,5].

With umbilical cord hernias and/or omphaloceles with abdominal wall defects smaller than 5 cm without solid viscer, early primary surgical closure is recommended [1]. Omphaloceles are generally considered giant when the defect is larger than 5 cm in di-
Giant omphaloceles always present a challenge. It is a rare and low-frequency congenital anomaly with no standardized management schemes or treatment protocols. Primary closure is often impossible because an early reduction with a reduced abdominal domain would lead to high intra-abdominal pressure, abdominal compartment syndrome and death [1,7]. Gradual reduction before closure is the alternative that reduces the associated morbidity and mortality in the management of these patients. In this study, we describe the results of our management protocol with early reduction and definitive abdominal closure in the neonatal period.

2. Methods

After obtaining institutional approval, we conducted a retrospective review of a 3-center cohort study from 1994 to 2019. All giant omphaloceles in Cmipediatra International since 1994 and in Hospital Exequiel González Cortés and Clínica Las Condes since 2017 were treated with the same standardized management protocol. We included patients with giant omphalocele defined as wider than 5 cm and/or containing >50% of liver who were managed with our nonsurgical silo technique because primary surgical closure was impossible to perform.

Demographic data included gestational age, birth weight, sex, associated malformations, amnios indemnity and the presence of pulmonary hypertension. Treatment variables included silo reduction and amnion inversion time, anatomic closure, use of mesh and the need for additional treatment such as Botox® injection and the component separation technique. Outcomes included pre- and post-silo complications, mortality and follow-up data.

Statistical analysis was performed with STATA-16® using Fisher's test for dichotomous variables and the Mann-Whitney U test for quantitative variables. Significant results were considered with a p value of 0.05 or less, with 95% confidence.

2.1. Technique

The technique used was the construction of a silo with an adhesive hydrocolloid dressing (Duoderm®) to achieve the reduction of the omphalocele in stages until the complete intra-abdominal reintegration of the liver and bowel into the abdominal cavity, preserving the omphalocele sac. After complete silo reduction and prior to abdominal closure, we performed what we called “inversion of the amnion”, which simulates definitive anatomical surgical closure.

All of the patients were managed at the neonatal intensive care unit under muscle relaxation, ventilatory support and intra-abdominal pressure monitoring; immediate postnatal care includes covering the omphalocele with a sterile plastic dressing, naso- or orogastric tube, sedation or muscle relaxation with intubation, and broad-spectrum antibiotics.

We describe 3 phases for this technique.

2.2. Phase 1: Silo reduction

Under relaxation, all patients are subjected to a “visceroabdominal disproportion test” that consists of manual reduction with no amnion disruption to exclude the possibility of closure. We build a silo with 2 plaques of Duoderm® that have already been cut with a “T” shape, and paper backing is removed. The Duoderm® dressing is fixed to the skin lateral to the abdominal defect with a vertical part of the T shape, and the horizontal free edges wrap the omphalocele sac from that side. The procedure is repeated on the other side in the same way, which completes the 360° silo. Duoderm® adheres to the skin but not to the amnion, allowing the amnion to be reduced further. If the amnion sac is ruptured, it is important to suture it before building the Duoderm® silo. Tongue depressors are placed on each side of the top of the silo and fixed to each other without compressing the omphalocele contents and without trapping the umbilical cord (Images 1-7). It is important to build the silo as soon as possible for faster reduction of the omphalocele; it is key that the amnion remains soft and reducible. If we delay the silo construction and the amnion becomes thick, it could become attached to the liver capsule, increasing the risk of bleeding during the reduction.

After 24 or 48 h and depending on the hemodynamic and respiratory stability of the newborn, we initiate the sequential reduction of the silo every 1–2 days. The 2 wooden tongue depressors at the free edge of the silo are used to gently reduce the omphalocele contents with compression toward the abdominal wall defect. The maximum compression allowed is 20 cm of H2O of intra-abdominal pressure (IAP) or manifestations of high IAP such respiratory compromise, decreased urine output, or lower limb re-...
duction of oxygen saturation. The timing of this reduction will be given by how the neonate tolerates the sequence of silo compression. If the neonate develops any of these signs 4 h after silo reduction, the silo can be easily loosened to decrease the IAP.

The physical features of Duoderm® keep the amnion and skin edges moisturized; in most cases, the amnion and edges remain wet even after a few days and lose adherence. We change the adherent dressing if it becomes dislodged or wet. Otherwise, it is routinely changed every 4–5 days to check the amnion status. Phase 1 is complete once the liver and entire bowel are returned to the abdominal cavity (flat silo reduction).

2.3. Phase 2: Amnion inversion

Phase 2 initiates once the reduction of the liver and the bowel is complete into the abdominal cavity (flat silo reduction) (Image 9). If the patient is stable, the surgeon inverts the amnion and approximates the skin edges (Images 10-12), and Duoderm® is applied to maintain the skin edges approximated. This replicates the pressure expected from definitive surgical anatomical closure. This enables the surgeon and neonatologists to evaluate whether the neonate will tolerate a definitive closure.

A few patients tolerate amnion and viscera reduction but not amnion inversion; in these cases, the surgical closure is not attempted due to compartmental syndrome risk, but they start from the flat silo stage and amnion inversion.

2.4. Phase 3: Surgical closure

The third and last phase is definitive surgical closure after proving that the neonate tolerates phase 2. This phase starts with resection of the amnion with caution for adhesions to the liver. Additionally, if needed, exploration of the abdominal contents for other
anomalies is performed. Anatomical closure is performed by suturing 3 layers: the peritoneum, muscle, fascia, and skin.

Additionally, umbilicoplasty in virgin and intact skin has better esthetic and functional results, without the need for secondary-intention wound healing. If anatomical closure cannot be achieved, we use Botox® injections, a component separation technique or a mesh (Dual mesh®) to achieve abdominal closure. For component separation, we use a longitudinal fasciotomy of the external oblique muscle lateral to the linea semilunaris on both sides, followed by a lateral submuscular dissection in the avascular plane between the external and internal oblique muscles. Once the external oblique muscle is released bilaterally, the rectus, the internal oblique and the transverse abdominis muscles are mobilized to the midline [8]. If additional advancement is required for tension-free closure, the rectus muscles are released from the posterior sheath. The advancement flaps are then closed primarily at midline with a mesh when needed [9]. If possible, the umbilical cord is preserved and included in the closure.

This process of sequential reduction and closure is performed under a strict protocol in the NICU, with metabolic and hemodynamic monitoring, endotracheal intubation and muscle relaxation. During the reduction, the intra-abdominal pressure cannot exceed 20 cm of H2O, and cardiorespiratory stability and urinary debit must be maintained.
The success of this technique is important to do the following.

a) Maintain the amnion intact to preserve a nonadherent interphase with the intra-abdominal viscera due to the risk of hemorrhage and hemoperitoneum during the reduction of the silo. It is important to set the silo and start the reduction in the first 24–48 h.

b) Change the adherent dressing when needed (dislodge, wet) or every 4–5 days.

c) When sequential reduction is being made, care must be taken to avoid damaging the hollow viscus between the tongue depressors. The omphalolecele vertex is not always the umbilical cord, and it must be reoriented when the silo is placed.

d) When reducing the liver, the surgeon must be aware of perfusion and blood pressure.

e) Always maintain the intra-abdominal pressure under 20 cmH2O. Measure the IAP with an intravesical or inferior cava vein catheter. If not possible, clinical parameters can be used as an indirect measure of IAP.

f) Permanent sedation or muscle relaxation and intubation is required. As sequential reduction takes place, endotracheal intubation and muscle relaxation will eventually be needed.

g) Parenteral nutritional support

h) Broad spectrum antibiotics are used due to long-term exposure to tracheal intubation, foley catheters and central lines.

3. Results

Forty patients were managed with our technique with a nearly equal sex distribution. The majority were full term, but 37.5% had an associated congenital anomaly (see Table 1).

At the moment of birth, 37 patients (92.7%) had an intact sac, 65% were hospitalized between 15 and 30 days, and more than 65% of newborns had a silo reduction and sac inversion within 5 days, with a final abdominal closure in less than 15 days in 73% of cases (see Table 2).

In 10 cases, a pressure monitor was used through the bladder; the rest were clinically monitored.

Four patients (10%) tolerated a complete manual reduction of the omphalolecele the day of birth at silo instalment after muscle relaxation and intubation. These patients were managed with a flat silo without the need for tongue depressors. After a couple of days, they proceeded directly to phase 2.
Compliations during reduction were observed in 9 patients (22.5%). Three patients who underwent Duoderm® silo after 72 h developed a mild hemoperitoneum (due to the reduction of the silo and bleeding from the liver capsule), with no treatment required; one patient had severe bleeding that required a blood transfusion. Another 3 patients presented with signs of local skin infection related to the silo that required intravenous (IV) antibiotic therapy and a change of the silo. Two patients developed high intra-abdominal pressure with periods of transient oliguria. These patients were managed with diuretics and dopamine, and the silo was mildly loosened. The high pressures occurred during the first 4 h of adjustment of the silo’s tongue depressors.

In one patient, the silo reduction would not progress to phase 3 due to the patient’s malformations and comorbidities. After 3 weeks, he did not tolerate amniocentesis, and closure was allowed to occur by secondary intention, completing a final abdominal closure within one month with a small ventral hernia.

Complications following abdominal wall closure were observed in 4 patients (10%). One patient developed abdominal compartmental syndrome due to progressive ascites. One patient developed a Pseudomonas aeruginosa wound infection. One patient suffered a 2-cm area of abdominal wall necrosis after component separation, and one patient with very low birth weight presented an early contained eventration after final abdominal closure with mesh interposition.

Seventeen patients (42.5%) developed a central line-associated bloodstream infection. Four patients (10%) died; one presented with transposition of the great vessels, two presented with pulmonary hypertension, and one presented with severe sepsis due to central line-associated bloodstream infection. None of these deaths were related to the silo management protocol. Regarding omphalocele characteristics in these patients, all had an intact omphalocele sac, and 50% had a minor technique-related complication.

### 4. Discussion

This series outlines the results of our minimally invasive management protocol for neonates with giant omphalocoeles. Our population is heterogeneous with a wide range of birth weights and gestational ages. We also observed a high incidence of associated anomalies, which may be related to the fact that in our country, therapeutic termination of pregnancy policies effectively does not exist. Despite this, the overall mortality rate was low (10%) compared to other series (13–25%) [10] and not related to the technique.

Many strategies for giant omphalocoele have been described, but no standard of care exists [10]. Giant omphalocoele management can be summarized as surgical and nonsurgical closures. The staged surgical closure techniques include different types of sutured silos, intra-abdominal tissue expanders and closure tech-
niques, such as interposition of synthetic mesh, skin flaps [11], skin grafts and dermal matrix [10]. All of these involve multiple operations under anesthesia. The final goal is closure of the abdominal wall fascia or covering of the liver and bowel with skin.

Nonsurgical closures, such as the one described in 1957 by Grob [12], involve the application of topical chemicals on the omphalocele membrane to produce an eschar, which is followed by granulation tissue and neoeplithelialization. Due to the protracted course of treatment, this has been associated with high infection rates and still leaves the need for surgical repair of the remaining moderate-to-severe ventral hernia [10,12].

The rationale behind our method was based on biomechanical forces. The adherent silo has at least the same effect as the traction from sutured silos. The force is applied over the entire area and not just the line of sutures. Force vectors are vertical during phase 1 and then horizontal in phase 2. These forces are distributed without sutures and without damaging the amnion or skin. Skin has a biomechanical property that allows it to gradually stretch beyond the limits of its extensibility [13,14]. This is due to collagen fibers strengthening in the direction of the stretching force until fibers are parallel and resist further extension. Others have described techniques with amnion sac inversion [5] and progressive external compression of the sac without a sutured silo [15].

The Duoderm® silo is a simple silo solution that is easily built at the bedside with accessible and inexpensive materials. Additionally, because it does not require surgery, it diminishes the morbidity related to anesthesia, the surgical procedure itself and the long exposure of the abdominal cavity. Amnion inversion is a real-time therapeutic test that will provide information about how the neonate will tolerate surgical and definitive closure. This decreases the risk of high intra-abdominal pressure complications after definitive closure. Respiratory, urinary and hemodynamic instability can easily and quickly be managed by loosening the tension of the Duoderm® silo when required during the silo reduction phase at any time.

Even if the amnion sac is ruptured, the Duoderm® silo can be used by previously repairing the omphalocele by simply suturing it. In this series, 3 newborns (7.5%) with a ruptured sac were managed with the Duoderm® silo with acceptable times of silo reduction and amnion inversion. We did not have any ruptured sacs after applying Duoderm®, which we think is due to the gentle of the material.

There were 4 patients who did not require reduction of the silo because even if they had giant omphalocele, the liver and bowel could be reintegrated into the abdominal cavity immediately after birth, and the protocol started from inversion of the sac in phase 2. These patients had giant omphalocele, but they probably had a lower viscero-abdominal disproportion and tolerated the simulated closure better.

The Duoderm® silo is safe to be used as long as needed to manage viscero-abdominal disproportion. No fatalities related to the time of silo reduction and amnion inversion were reported. The fact that 3 of the 4 deceased neonates had longer treatment times is related only to the complexity of their associated malformations and comorbidities and the size of the abdominal wall defect. The time for final abdominal closure ranged from 6 – 35 days, which is far less than the topical approaches that can take months or years before a final repair. There was one patient who underwent 35 days of silo reduction prior to surgical closure. Due to cardiac comorbidity, the silo was slowly reduced every 2–5 days with good results moving forward; it was one of the largest giant omphaloceles treated. We decided not to do “paint and wait” because of the potential complications and because the newborn had a favorable evolution, though slightly slower. We consider that all patients with giant omphalocele should be treated at the beginning with a Duoderm® silo and that their ability to tolerate it can be subsequently determined. We acknowledge there might be some patients with giant omphalocele that will need a “paint and wait” management, but we have not find that patient yet.

As outlined in the results, there were 3 main groups of complications: infection, abdominal hypertension, and intraperitoneal bleeding. The silo infection rate was 7.5%, lower than that reported in other series, which are as high as 20% [10]. This may be because there is no manipulation or exposure of intra-abdominal viscera and because the period until the final abdominal closure is shorter than other techniques that require secondary-intention wound healing. Additionally, the Duoderm® silo is gentle to the amnion and skin tissue, preventing inflammation and scarring. All 3 patients with silo infection weighed less 1500 gr, and they had only local skin infection; we assume that this was due to minor erosion of the Duoderm®.

We did see a high rate of central line-associated bloodstream infections (42.5%), which can be improved with quality improvement initiatives around line care in the NICU.

This study describes 3 neonates with abdominal hypertension, one of them with abdominal compartment syndrome after abdominal closure, which we will discuss further. The 2 newborns who underwent abdominal hypertension during silo reduction were easily and quickly managed by loosening the Duoderm® silo. No fatalities related to this complication occurred.

Four neonates (10%) developed hemoperitoneum, among whom one required a blood transfusion. This was due to detachment of the amniotic sac from the liver's Glisson capsule or due to a falci-form ligament tear because of inadequate umbilical cord traction at the moment of silo reduction. This complication was seen in patients referred from outside centers with delayed management (after 72 h), probably with a thicker amnion that can get rapidly attached to the liver capsule. We suspect that this can be avoided by an immediate silo construction after birth and a careful reduction of the silo.

Complications following final abdominal closure were seen in 4 patients (10%). One patient developed abdominal compartment syndrome that required management with a component separation technique and mesh reinforcement. After that, presented progressive ascites that required paracentesis and abdominal drainage for 72 h, reverting the abdominal compartment syndrome. The patient who presented with Pseudomonas aeruginosa wound infection was managed with IV antibiotics. Another patient presented abdominal wall necrosis (2 cm in width) after a component separation technique that was managed with antibiotics and a second-intention closure with hydrocolloid dressing. A neonate with very low birth weight presented an early contained eversion after final abdominal closure with mesh interposition that was managed with different botulinum toxins at one year of age.

There were no other complications among those described for other techniques, including entero-cutaneous fistula, or complications related to topical medicine (silvadene: seizures, nephrotic syndrome, leukopenia, and elevated liver enzymes; iodine: thyroid dysfunction).

We acknowledge this study has statistical limitations. It will be necessary a larger number of patients to validate this technique. We are already optimistic with the preliminary results with this standardized management protocol.

5. Conclusion

The staged nonsurgical silo management of giant omphalocele in this series is safe, effective, simple, easy, and low cost. It shortens the time to closure and reduces potential morbidity and mortality compared with traditional surgical or medical management.
References


