

Case Reports

Staged closure of a giant omphalocele with amnion preservation, modified technique

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ABSTRACT

يشكل إغلاق الفتق السري الولادي تحدياً وفي الغالب لا يُمارس حفظ السلي عند إغلاقه، نصف في هذا التقرير حالتين لفتق سري ضخمة عولجت بطريقة إغلاق على مراحل ونوضح جدوى هذه التقنية وسلامتها وتعددية استعمال السلي للتكيف مع استراتيجيات التندب حال عدم حصول الإغلاق.

Closure of a giant omphalocele can be challenging. Preservation of the amnion in staged closure is not commonly practiced. Here, we describe 2 cases of giant omphalocele treated with a modified amnion preservation, staged closure technique. This paper demonstrates the feasibility and safety of this technique, and the versatility of amnion to adapt to an escharization strategy if closure is not achievable.

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Giant omphalocele (GO) continues to be a management challenge for pediatric surgeons. These large defects, which typically contain solid viscera, are frequently associated with significant viscerobdominal disproportion and chest wall restriction causing pulmonary hypoplasia. When primary fascial closure is not possible, the options include amnion escharization enabling gradual epithelialization and eventual ventral hernia repair, skin flap closure with the method originally proposed by Gross, or staged

reduction using a variety of prosthetic materials, with or without amnion preservation, leading to delayed closure in the neonatal period.^{1,2} We report our experience with 2 cases of GO treated with a modification of the described amnion preservation techniques.

Case Report. Patient 1. A 2.8 kg male infant with an antenatally diagnosed GO was born at 37 weeks gestation by cesarean section. Prenatal echocardiogram and amniocentesis were normal. Findings at birth included an intact amnion covering an upper midline fascial defect measuring 9 cm, which contained liver, spleen, and small and large intestine. Primary closure was not feasible, and so a staged approach using an extra-amniotic silo was used. The infant was taken to the OR and anesthetized. The amnion and adjacent skin were prepped, and a circumferential skin incision was made a few millimeters on the skin side of the amnio-cutaneous junction. Skin flaps were raised over a distance of 2 cm to expose the underlying fascia, and a mesh reinforced Silastic™ (Dow Corning, Midland, MI) sheet was sewn carefully to the exposed fascia, directly below its junction with the amnion.

Postoperatively, the infant was maintained on mechanical ventilation, and gradual visceral reduction was achieved by sequential suturing of the top of the silo aided by the transverse application of a large bowel clamp. After 6 days, the viscera had been completely reduced, and the infant returned to the OR for silo removal, amniotic excision, and a primary fascial closure with bladder pressure monitoring. After 7 additional days of mechanical ventilation, the infant was extubated, but required nasal cannula oxygen for ongoing pulmonary hypoplasia. Enteral feedings were commenced and the infant was discharged on nasogastric feeds and supplemental oxygen at 6 weeks old.

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Patient 2. A 3.2 kg male infant was born by vaginal delivery without induction at 41 weeks gestation. He was transferred to the neonatal intensive care unit in stable condition. The echocardiogram showed a small ventricular septal defect with patent ductus arteriosus. Chromosomal analysis was normal. Examination of the abdomen revealed a large omphalocele 10 cm in diameter with an intact sac containing liver and bowel (Figure 1). Due to the size of the abdominal wall defect, we elected to attempt staged closure of the omphalocele with preservation of the amnion.

A technique similar to the first case was used. The infant remained intubated and a gradual reduction of the abdominal contents commenced. Unfortunately, on the 5th Postoperative day the patient began to demonstrate increased oxygen requirements, and Echocardiography revealed results revealed severe pulmonary hypertension. No further reduction was performed (Figure 2A & 2B). Two days later the silo began to separate from the fascia exposing the intact amnion. At this time, it became evident that achieving primary closure would not be possible due to the infant's respiratory status. Due to this, the silo was removed and an escherization strategy using silver sulfasalazine was adopted with positive results (Figure 3). However, despite maximum ventilatory support and pulmonary vasodilatory therapy, he died at age 10 weeks from severe pulmonary hypertension.

Discussion. Viscero-abdominal disproportion usually precludes primary closure in cases of GO. Gradual reduction of abdominal viscera over several days allows safe stretching of the abdominal compartment and avoids acute kinking of the hepatic veins and



Figure 1 - Picture of day one of life showing the giant omphalocele, containing liver and bowel.

inferior vena cava. Many techniques of gradual visceral reduction and closure have been described in the literature. Schuster's original description involved the circumferential suturing of mesh directly to the rectus sheath, which was exposed by separating the skin from the amnion. To complete the reduction the sac was reduced by repeated external compression.³ Yokomori et al⁴ and De Lorimier et al⁵ modified Schuster's technique by suturing mesh directly to the skin, facilitating amnion inversion, and eventual delayed primary fascial closure. Both authors noted the risk of mesh detachment due to sutures tearing, if the silo was left too long or if excessive tension was applied. Pacilli et al⁶ reported 12 GO patients treated with staged closure using Schuster's technique modified by the use of a



Figure 2 - Intraoperative pictures showing A) the circumferential exposure of the abdominal wall fascia: the amnion wall fascia the amnion is retracted medially and the skin is dissected from abdominal wall and B) the one sheet of the silo suture to the fascia with 2 rows of interrupted suture to distribute the tension and minimize ischemia.

prolene™ (Ethicon, Somerville, NJ, USA) mesh, fixed with 2 rows of sutures to the exposed rectus sheath. Finally, modified techniques of upwards traction on the amniotic sac maintained by suspension⁷ or sequential sac ligation⁸ have been used to slowly reduce the omphalocele with amnion preservation.

The amnion serves as biological barrier that protects the viscera from mechanical irritation during reduction, as well as temperature and insensible fluid losses. Its preservation also offers protection from microbiological contamination as well as subsequent peritonitis and sepsis. Furthermore, the inversion of the amnion during the process of staged reduction may create additional space in the abdominal cavity. By removing the amnion at the final closure, the space in the abdominal cavity will be larger, which may reduce the risk of postoperative abdominal compartment syndrome.⁹ Apart from this, should the staged reduction not succeed, the amnion can be “painted” after removing the external silo, thereby allowing eschar formation and epithelialization, as occurred with our second case. On the other hand, removing the amnion early may expedite the diagnosis of gastrointestinal anomalies, such as malrotation, and permit consideration of the option of earlier treatment. Earlier amnion removal may also facilitate

the development of adhesions, and while this may increase the risk of future adhesive bowel obstruction, it may also offer the theoretical benefit of reducing the risk of midgut volvulus post GO closure, in patients predisposed by the presence of malrotation. Pacilli and colleagues reported one case of midgut volvulus among their 12-patient series following amnion inversion and, on this basis, advocate elective laparoscopic Ladd’s procedure in survivors shown to be malrotated.⁶

In conclusion, This paper reaffirms the value of amnion preservation, and offers a slight modification from other described techniques in where and how to suture the silo. We observed that the potential benefits of amnion preservation far outweigh the risks, and advocate for its routine use in cases of GO. Attention should be given to expeditious visceral reduction, as long as the infant can tolerate it physiologically. In the event that reduction cannot be tolerated, or if the silo detaches from the fascia, amnion preservation allows the flexibility of switching to an escharization approach.

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Figure 3 - This Picture was taken 4 weeks after removing the Silo with daily application of sulfadiazine. It demonstrates the intact dry amnion with healing skin edges all around.