J Ped Surg Case Reports 3 (2015) 230–233

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Successful three stage repair of a large congenital abdominal region defect

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ARTICLE INFO

Article history:
Received 18 February 2015
Received in revised form
25 March 2015
Accepted 28 March 2015

Key words:
Gastroschisis
Large abdominal wall defect
Omphalocele
A-cell®
Silver-impregnated hydrofiber
Gore-Tex® soft tissue patch

ABSTRACT

We present two infants born with large, right upper quadrant defects which cannot be categorized as either a gastroschisis or omphalocele. We successfully managed one infant with a three stage repair using polytetrafluoroethylene (PTFE) patch, porcine urinary bladder matrix (UBM) and delayed surgical closure. The second infant passed away due to parental consent care withdrawal.

The Centers for Disease Control (CDC) categorizes congenital abdominal wall defects as either gastroschisis or omphaloceles [1]. Classically, an omphalocele is an abdominal wall defect, centered on the umbilicus that has an amniotic sac covering the extra-abdominal organs. An omphalocele may be either small or giant (>8 cm in any dimension with liver present in the sac) and is usually associated with other anomalies involving the musculo-skeletal, cardiac, digestive, genital, CNS, pulmonary and/or reticulo-endothelial systems [2]. In contrast, gastroschisis is a defect of the abdominal wall, usually to the right of the umbilicus that results in evisceration of intestines, but not the liver, without an amniotic sac covering. Our two infants presented with a defect characterized by absence of the right upper quadrant (RUQ) abdominal wall, no amniotic sac covering, and evisceration of all abdominal contents.

Given the defect's characteristics, we cannot classify this condition as either a gastroschisis or omphalocele. Therefore, we coined the term megaschesis, and present the successful management of this condition.

1. Case #1

A 2.7 kg, 35 week-gestation female was born with absence of the RUQ abdominal wall. The defect extended from the right costal margin superiorly, the right anterior axillary line laterally, the umbilicus inferiorly, and the midline medially. All abdominal content including stomach, liver, large and small intestine were extra-abdominal with no amniotic covering. The liver was rotated 90° anteriorly to its normal axis and there were no other anomalies. On day of life (DOL) #0, the child was intubated, placed on prophylactic antibiotics, and transferred to Driscoll Children's Hospital (DCH).

At operation, adhesions between the liver and right coastal margin were taken down with cautery. The liver was 3–4 times larger than normal. Preformed, abdominal silos could not be used due to the large size of the defect. Two sheets of PTFE (PTFE, Gore-Tex® soft tissue patch, W. L. Gore & Associates Inc., Flagstaff, AZ) were sewn to the fascia on either size of the defect as described previously [3]. The two PTFE sheets were trimmed, sewn together and suspended over the baby with mild tension. The PTFE silo was wrapped in betadine moistened Kerlix and wrapped in cellophane. From DOL #2 – 12, the bowel was gradually reduced until it was level with the abdominal wall (Fig. 1A).

On DOL#13, the patient was brought to the operating room for stage 2 of the repair. The PTFE was removed revealing an

Abbreviations: DCH, Driscoll Children's Hospital; DOL, day of life; RUQ, right upper quadrant; POD, post-operative day; PTFE, polytetrafluoroethylene patch; UBM, porcine urinary bladder matrix; CDC, Center for Disease Control.

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http://dx.doi.org/10.1016/j.epsc.2015.03.016

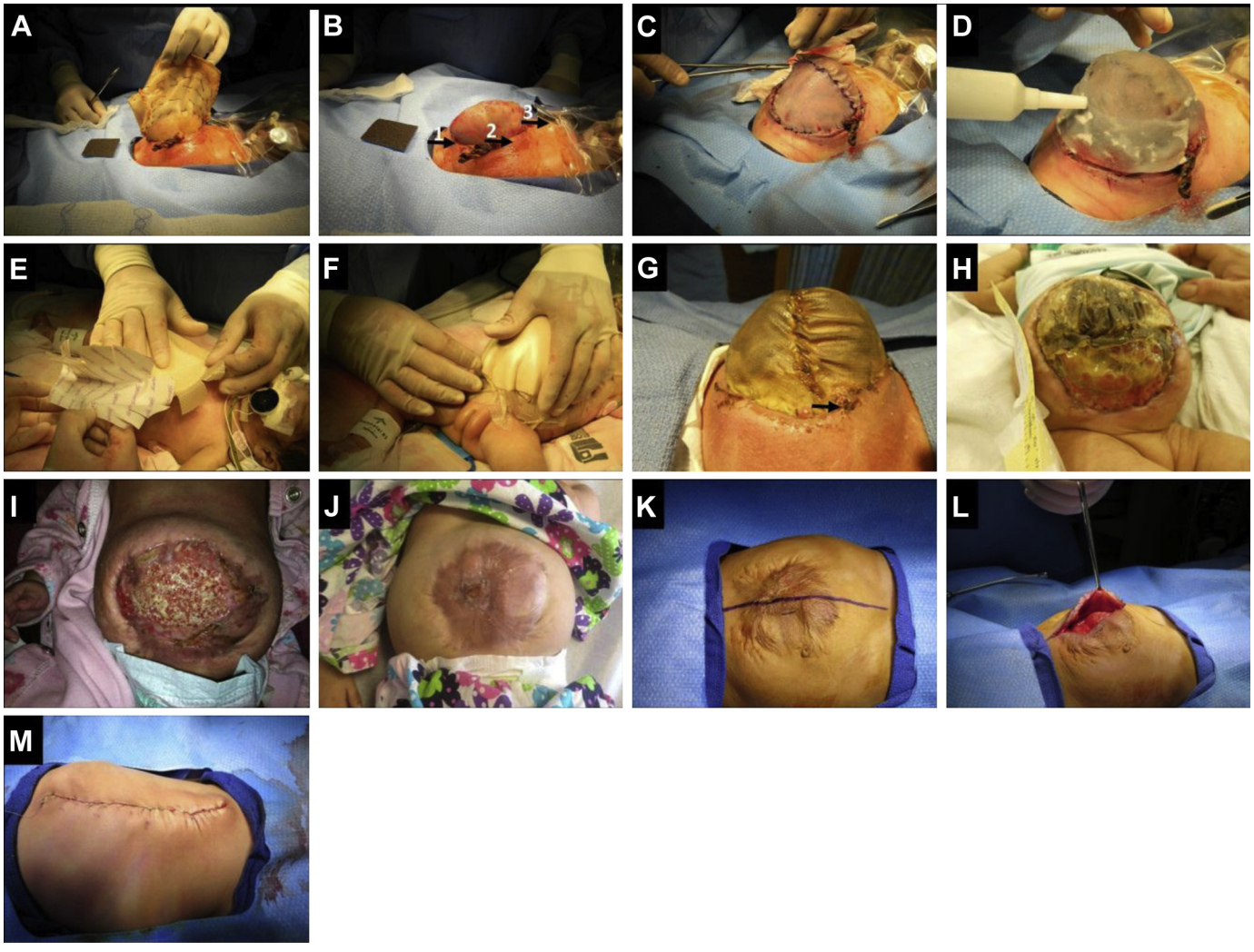

Fig. 1. The management of the infant with a large abdominal wall defect. (A) On DOL#13, the staples across the PTFE silo sac for gradual reduction of the abdominal content are visible. (B) The PTFE silo was removed to reveal the abdominal defect. The 1) Top of bladder, 2) Diaphragm and 3) Nipple line is indicated by arrows. (C) The A-cell is sutured circumferentially around the giant omphalocele. (D) Aquacel Ag was applied to the A-cell to promote epithelization. (E) & (F) Soft silicone foam secondary dressing is applied to the omphalocele. (G) On DOL#30, the granulation is noted at the base of the A-cell membrane. (H) On DOL#73, further sloughing and granulation of the A-cell matrix is noted. (I) On DOL#145, extensive epithelization of the defect is noted. (J) On DOL#243, new fibrotic scar tissue formation noted over the defect. (K) At 10 weeks, 27 days, the infant is brought to the operating room for final surgical closure by first marking the midline on the abdomen. (L) The fibrotic scar is opened followed by eventual (M) surgical closure of the fascial ends.

8 cm diameter defect (Fig. 1B) and the small bowel was irrigated externally with saline (Fig. 1C). The skin and subcutaneous tissue were mobilized circumferentially from the fascia for 2.5 cm. A 6-layer multi-laminate UBM (UBM, MatriStem® Surgical Matrix PSMX, ACell Inc., Columbia, MD) graft was sutured to the fascia with long-term absorbable suture (PDS) to serve as a biological scaffold and facilitate body wall repair (Fig. 1D). Medical grade honey and UBM particles (MatriStem MicroMatrix®, ACell Inc., Columbia, MD) were placed on the wound edges. On DOL#15, she underwent the first dressing change with silver-impregnated hydrofiber [4] (Fig. 1E and F). On DOL#31, the patient was extubated and on DOL#126 discharged with home health assisted silver-impregnated hydrofiber dressing changes [4]. Complete epithelization occurred at 10 months and 2 days of age (Fig. 1G–J).

Final fascia closure was done in the OR at 10 months and 27 days of life. The abdominal wall was marked and the fibrotic scar opened in the midline using sharp and electrocautery dissection. The fibrotic scar was separated circumferentially from the fascia (Fig. 1K). The fascial edges were cleared for approximately 3–4 cm (Fig. 1L). The fascia was re-approximated with multiple, interrupted, non-absorbable sutures. Abdominoplasty and umbilicoplasty were performed and the excess skin sent for pathology [5]. The skin and subcutaneous tissue were closed in layers with absorbable sutures (Fig. 1M). The patient tolerated the procedure well and was discharged without complications on post-operative day (POD) 4.

Pathology revealed benign epidermal, adnexal structures on one edge of the specimen. The specimen was composed of dense fibrous connective tissue, mild chronic inflammation and classified as an omphalocele sac.

At 15 months of age, she is well, without abdominal hernias and gastrointestinal complications.

2. Case #2

A 1.7 kg, 37-week gestation male infant was born with absence of RUQ abdominal wall and all abdominal organs extra-abdominally (Fig. 2). Additional co-morbidities include pulmonary hypoplasia