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The tale of two omphaloceles: Same treatment, different outcomes

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Abstract

Omphaloceles are one of the most common abdominal wall defects, and unfortunately multiple long-term medical problems can occur as a result. Specifically, other structural and chromosomal abnormalities may be present in the setting of an omphalocele. Treatment options vary depending on the size of the defect, with a common option being the "paint and wait" technique, in which a topical agent is applied that allows epithelialization over the amnion sac. Here, we discuss two such cases of omphalocele, and although both received the same treatment, very different outcomes occurred.

Introduction

The two most common congenital abdominal wall defects requiring neonatal intensive care are gastroschisis and omphalocele [1]. However, these two entities are vastly different in regard to pathogenesis, with omphalocele often occurring with other anomalies including chromosomal abnormalities [2]. Specifically, omphalocele is associated with Beckwith-Wiedemann syndrome, OEIS complex (Omphalocele, Exstrophy, Imperforate anus, Spinal) and pentalogy of Cantrell [3]. As many as 77% of the patients with omphalocele will have other congenital anomalies [4].

Omphalocele has an estimated incidence of 1 per 3000-4000 births [3] and is a congenital malformation that results from incomplete body wall folding during embryogenesis with incomplete reduction of the physiologic intestinal herniation, leading to an often midline abdominal wall defect with a thin membrane surrounding any protruding organs and tissue [1,3]. The sac covering the defect is a three-layered membrane made of peritoneum, Wharton's jelly, and amnion [1]. The size of the omphalocele and the presence of the membrane allow an omphalocele diagnosis to be stratified into small, giant, or ruptured [5]. Small typically describes the abdominal wall defect of ≤5 cm, with giant describing a defect of at least 5 cm with inclusion of the liver [4]. Interestingly, a retrospective study found that of the 111 patients with omphaloceles, those with a small defect were more likely to also have additional congenital anomalies [4].

Following initial stabilization of a patient with an omphalocele, the treatment goal is reduction of the abdominal contents and closure of the defect. However, depending on the size of the defect and the amount of herniated tissue, closure may need to occur in stages to allow time for compensation due to increasing abdominal pressure. Prior to the use of parenteral

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nutrition, as many as 80% of infants with omphaloceles would die secondary to complications from the prolonged intestinal ileus and starvation [4]. Now, conservative management is possible and has improved the morbidity and mortality within the population. Specifically, a technique called "paint and wait" is often utilized, in which topical agents are applied to the area and covered with a dressing, which allows for epithelialization. This treatment is continued until the membrane is gone and the skin has covered the defect [2,6]. Here we discuss two cases of omphaloceles with similar treatments but different outcomes and the incidence of complications occurring with omphaloceles.

Case Report

Patient Presentation #1

Baby Boy M (BBM) was diagnosed with an omphalocele on ultrasound completed at 15 weeks' gestation; repeat ultrasound at 26 weeks' gestation confirmed the omphalocele, which was considered giant and contained liver, stomach, and spleen. He was born at term (39 weeks' gestation) via a repeat cesarean section with APGAR scores of 5, 9 and 9 at 1, 5 and 10 minutes, respectively. BBM required intubation in the delivery room due to apnea after delivery. An orogastric tube (OG) was placed to decompress the stomach. The infant was transferred to the Newborn Intensive Care Unit (NICU) for further management.

Examination was significant for the presence of an omphalocele in which the liver, stomach, intestines, and inferior portion of the pericardium were included. A small serosal tear was observed in the otherwise intact sac. Pediatric surgery evaluated the patient and opted for the "paint and wait" technique. The defect was covered with Silvadene and covered with Kerlix. Additional anomalies identified after delivery included dextrocardia and undescended testes. Pediatric genetics was consulted due to the various anomalies, after which a microarray was completed with normal results showing no alterations of the loci tested.

BBM continued to have significant respiratory distress and failed several extubation attempts. He was suspected to have a central diaphragmatic hernia; subsequently, an exploratory surgery was completed at 34 days of life in an attempt to confirm the diagnosis as well as repair a hernia, if present. The thoracoscopy was difficult secondary to adhesions between the lung and the pericardium, but the area of the diaphragm visualized appeared to be intact. The patient developed a chylothorax 1 week post-op and required chest tube placement. The infant was transitioned to a fat free formula until resolution of the chylothorax, after which he transitioned back to breast milk. Likely secondary to the pulmonary abnormalities, BBM was not able to extubate. A tracheostomy was placed on day of life 48, which was well tolerated.

Oral feeding was attempted, but feeds were unsuccessful secondary to oral aversion. A gastrostomy tube was discussed; however, the anatomical positioning of the stomach was unclear, and thus the patient was not a good candidate for the procedure. Therefore, BBM was continued on nasogastric feeds, which were continued at discharge.

BBM was discharged home after 118 days in the NICU. He was referred to the Pediatric Ventilator Clinic for home management of respiratory support and to Early Intervention services for continuing home therapy. Multiple subspecialists followed him after discharge, including: pediatric surgery, pediatric cardiology, pediatric ear, nose and throat, and pediatric pulmonology.

He was re-admitted at 3 years of age for an exploratory laparotomy, during which there was lysis of adhesions, repair of the central diaphragmatic hernia with prosthetic patch, an abdominal wall reconstruction with component separation closure with prosthetic patch, and an inversion appendectomy. Approximately 6 months later, he underwent tracheostomy decannulation, which was well tolerated. He has been hospitalized for respiratory infections and pneumonias but remains on room air without supplemental respiratory support required. He also is able to take some oral nutrition, with the gastrostomy tube used for additional nutrition and hydration.

Patient Presentation #2

Baby Girl B (BGB) was diagnosed with an omphalocele at 12 weeks' gestation. The prenatal course was complicated by a dichorionic, diamniotic pregnancy, with BGB being twin "B". Prenatal labs were also significant for group B streptococcus positive, hepatitis C positive, and gonorrhea positive in the first trimester with a negative test of cure. The mother of BGB had a history of poly-substance abuse including methamphetamines, heroin, cocaine, and marijuana. BGB was born at 34 weeks' gestation via an elective cesarean section due to intrauterine growth restriction in the setting of an omphalocele of BGB. The APGAR scores were 4, 7, and 8 at 1, 5, and 10 minutes, respectively, with the infant requiring Positive Pressure Ventilation (PPV) due to poor respiratory effort and bradycardia. An OG was quickly placed to decompress the stomach. She was transported to the NICU on Continuous Positive Airway Pressure (CPAP) due to increased work of breathing with grunting and retractions observed.

Pediatric surgery was called to evaluate the patient, and a "paint and wait" approach was taken. Silvadene was applied to the defect and covered with Kerlix. BGB was started on feeds, and able to tolerate oral feeds once her respiratory distress resolved. However, at three months of age the patient developed a septic ileus. The sepsis evaluation revealed a urinary tract infection with Enterococcus faecalis. During this period, the sac covering the omphalocele ruptured with evisceration of 15 centimeters of small intestine. BGB had surgery completed in which the intestine was returned to the abdominal cavity and the defect was covered with a Vicrylpatch. At four months of age, the patient returned to the OR for excision of an eschar, adhesion lysis, ileal resection, and silo placement. A few days later she required further debridement of the necrotic ileostomy at which time a Vicryl mesh closure of the abdominal wall occurred. A fistula developed with small bowel obstruction, at which time negative-pressure wound therapy was placed followed by split thickness grating from the left thigh. Concern arose for a graft site infection and BGB was treated with antibiotics; she had the ileostomy closed and gastrostomy tube placed at 11 months of age, which was used at time of discharge.

Throughout the hospital course, BGB was intubated for procedures and initially was able to tolerate brief extubation periods. However, with each extubation attempt it became more difficult for the patient to tolerate non-invasive modes of ventilatory support. At four months of age she had significant respiratory distress and failed extubation, with a bronchoscopy showing airway edema leading to placement of a tracheostomy. Additional complications occurred including volume overload requiring intermittent courses of Lasix and colonization of the

tracheostomy with Pseudomonas aeruginosa and Klebsiella pneumoniae.

BGB was discharged home after 529 days in the hospital. She was noted to have hyperopia and was globally developmentally delayed. At the time of discharge, she could sit independently, roll and pull toys and medical devices to her body. At 4.5 years of age she underwent laryngotracheal reconstruction with a right inferior rib cartilage graft and was able to extubate and remains stable in room air with no additional supplemental oxygen support required. She continues to utilize the gastrostomy tube for nutritional support.

Discussion

Although omphaloceles only occur 1 per 3000 – 4000 births [7], the defect is one of the most common abdominal wall defects in infants. The use of topical agents to aide in epithelialization is well established, as described in the care of both cases. The main benefits of delayed closure, due to topical therapy and epithelialization, are the ability to feed the infant more quickly after delivery and the potential for reduced needs of mechanical ventilation from the avoidance of increased abdominal pressure as observed in primary closure [8]. The "paint and wait" technique includes applying a thin layer of the agent, such as Silvadene, once daily and covering the area with a sterile gauze. Over the first four weeks, the omphalocele sac is noted to be replaced by granulation tissue, which then slowly becomes an epithelialized scar over the next four to twelve months [9]. Overall, the "paint and wait" technique is typically utilized to allow for gradual abdominal distention and delayed surgical closure [10]. Studies have evaluated the effectiveness of different agents to apply to the omphalocele sac, including the use of Acacia nilotica paste compared to povidone-iodine, with no significant difference noted in duration of hospital stay and overall mortality [6]. Of the available topical agents, Silvadene is most commonly used, followed by povidone iodine [8].

Given the increased incidence of other anomalies, including chromosomal abnormalities with omphalocele, mortality is higher in these infants compared to those born with gastroschisis [3]. It is estimated that only 10% of all omphaloceles diagnosed with prenatal ultrasound have no long-term issues including no other significant structural or chromosomal abnormalities [1]. Additionally, complications can arise during the "paint and wait" period that can contribute to overall mortality. Necrotizing enterocolitis, spontaneous fistulization of bowel into the omphalocele sac, bowel ischemia, and sepsis have occurred during the period of epithelialization [1,2]. Patients with large omphaloceles often suffer from gastroesophageal reflux, pulmonary insufficiency, recurrent lung infections, and feeding difficulties [1], as described in the cases presented above. Many patients with giant omphaloceles report cosmetic problems as a major long-term issue, with both abdominal scarring and the lack of an umbilicus lowering patient satisfaction [4].

Deficits in developmental achievements in many children with giant omphaloceles have been found, with cognition, language, and motor abilities all being impacted. A study of children at one year of age found the delays ranged from mild to profound, with 40% having severe delays for cognition, language, and motor outcome [11]. The cases presented above include children with prolonged hospitalizations, periods of hypoxia, and multiple surgical and invasive interventions. All of these factors increase the risk of abnormal neurodevelopmental outcomes [12].

While the incidence of omphaloceles has remained constant [1], as various surgical techniques are investigated, the presence of morbidity and mortality within this population persists. Continued research is needed to minimize the risk of complications that arise, while working to decrease the duration of hospital stay to allow for the best possible outcomes in these children.

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