

Surgical Management of Necrotizing Enterocolitis following Congenital Diaphragmatic Hernia Repair

Nida Fatima Sakrani, MbBCh, BAO* Eman Hamza, MbBCh, BAO** Hussain Ahmed, MRCS, FEBPS*** Martin Corbally, FRCSI, FRCSEd, FRCS (Paed Surg) MRCPI (Assoc)****

Necrotizing Enterocolitis (NEC) is a significant cause of in-hospital mortality. The pathogenesis remains unclear, but may be associated with Staphylococcus epidermidis related sepsis, hypertonic feeds or other stress. It is also associated with Abdominal Compartment Syndrome (ACS) as documented after complete closure of gastroschisis. While the incidence of NEC is rare, the associated mortality is significant.

We report a case of NEC following the repair of a congenital diaphragmatic hernia (CDH); an unusual yet serious complication. A literature search revealed only one similar case which resulted in mortality. The possibility of serious postoperative complications following the repair of CDH must be considered in any neonate who exhibits deterioration in their general condition.

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NEC is a leading cause of in-hospital mortality¹. Its causes appear to be multiple with an unclear pathogenesis but may be associated with an infectious etiology when it occurs in full-term infants². Spontaneously occurring NEC is not primarily the result of increased intra-abdominal pressure but may occur in primary repair of gastroschisis. Surgical repair of congenital diaphragmatic hernia (CDH) involves restoration of large volumes of the intestine into the abdominal cavity, which would result into an increase in intra-abdominal pressure. Despite this, the reported incidence of NEC is extremely rare in this subset of patients and appears to be associated with a high mortality.

The aim of this presentation is to report a rare case of left-sided CDH in a full term neonate complicated postoperatively by NEC. Only one such case has been recorded in the literature to date.

THE CASE

A full-term male infant was diagnosed with CDH within 3 hours of birth. Imaging revealed the stomach, spleen and a large portion of the small and large intestine in the left chest, see figure 1. Following three days of standard stabilization with ventilator support, the diaphragm was repaired through an open left upper quadrant laparotomy. The large defect was closed primarily following reduction of the spleen, stomach and small intestine. The abdominal closure was not excessively tight. The patient was kept in the Neonatal Intensive Care Unit, intubated, ventilated and given prophylactic antibiotics: Ampicillin,

Cefotaxime and Metronidazole intravenously. Postoperatively, he was stable with a good urine output and acceptable blood pH levels.



Figure 1: The Stomach, Spleen and a Large Portion of the Small and Large Intestine in the Left Chest

On the second postoperative day, tight abdominal distension was noted and urgent abdominal radiography showed a shadow over the liver with free air in the peritoneum, see figure 2. The patient was otherwise stable, and blood pH level was 7.33. An urgent laparotomy revealed frank blood in the peritoneal cavity, a small bowel perforation and a mesenteric tear, which were

* Intern
** Senior House Officer
Department of General Surgery
*** Senior Registrar
**** Chief of Medical Staff
Consultant Pediatric Surgeon
Professor and Chairman, Department of Surgery, RCSI-MUB
King Hamad University Hospital
Kingdom of Bahrain
Email: eman.hamza@khuh.org.bh; nfs090430@rcsi-mub.com

repaired with Maxon sutures. Portions of the small bowel were diffusely dusky; however, visible peristalsis was reassuring. A peritoneal drain was inserted, and the abdomen was closed with a decision to perform a second look laparotomy after 24 hours.

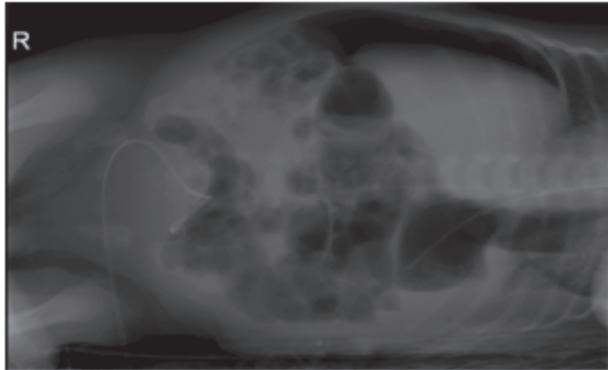


Figure 2: Shadow over the Liver with Free Air in the Peritoneum

After 24 hours, the abdomen was noted to remain distended and tense with wall edema but urine output was satisfactory, and meconium had also been passed. At re-exploration, the diaphragmatic repair was intact but the proximal jejunum was unhealthy with questionable viability for a length of approximately 30 centimeters. Necrotizing Enterocolitis was presumed, and thirty centimeters of the small bowel was resected with consequent jejunostomy and mucous fistula formation. The diagnosis of Necrotizing Enterocolitis was retrospectively confirmed by the histopathology report.

Following a period of 24 days of total parenteral nutrition, enteral feeds and stomal re-feeding were initiated. However, in view of high stomal output, the baby suffered from malabsorption which resulted in poor weight gain. Therefore, it was necessary to reverse the stoma at an earlier stage; the enterostomies were closed uneventfully eight weeks later.

During hospitalization, the patient had two occasions of sepsis, both of which were successfully treated. The first deterioration was due to *Staphylococcus epidermidis* and treated with Cefotaxime, Vancomycin and Meropenem. *Enterobacter cloacae* was the second culprit and was treated with Amikacin and Cloxacillin.

At one month follow-up, the infant was well and gaining weight with no intestinal or respiratory symptoms.

DISCUSSION

There are few documentation of serious postoperative intestinal complications following CDH repair². Through an extensive literature search, we found seven cases of NEC after CDH repair; however, only one was comparable to ours^{2,3}. Blaise et al reported NEC which was diagnosed postoperatively and treated, but mortality occurred later². Ruttenstock et al provided a detailed discussion on the topic; ACS was the cause of morbidity in their case⁴.

NEC should be considered as the cause of deterioration after any neonatal surgery. The first case was documented

in 1976 following cardiac surgery⁵. A study revealed that the leading surgical cause of NEC was myelomeningocele repair and the ventriculoperitoneal shunt procedure used to treat hydrocephalus³. Multiple factors may contribute to the development of this condition including bowel ischemia during surgery due to the manipulation, enteral feeding toxicity and infection or sepsis². The typical clinical picture of NEC is not a reliable diagnostic indicator in a postoperative patient because abdominal distension, bile-stained gastric content and diarrhea could occur due to operative stress³. The most reliable confirmatory signs are pneumatosis intestinalis and portal vein gas seen using radiographic imaging⁶.

The second and equally possible diagnosis is ACS. The premise is that an increase in intra-abdominal pressure (IAP) may occur when a large quantity of organs are placed into an underdeveloped abdominal cavity; it has been defined as prolonged IAP >20 mm Hg (normal 0-5mm Hg) which is associated with new organ dysfunction or failure⁴. Organ dysfunction and irreversible damage occur over 20 mmHg because the increased pressure compresses the inferior vena cava, thereby reducing cardiac preload and urine output⁷.

ACS is a well-recognized complication in trauma patients after receiving excessive fluid resuscitation, following primary surgical treatment of gastroschisis or omphalocele and in adults who have had delayed presentation and repair of a Bochdalek hernia^{7,8,9}. It could occur in other conditions that constrain the peritoneal space such as pneumoperitoneum, pancreatitis, retroperitoneal hemorrhage or a neoplastic mass. A study of 150 patients admitted to neonatal and pediatric intensive care units by Divarci et al found that ACS or intra-abdominal hypertension occurred in approximately one-tenth of the patients; NEC, abdominal wall defects, and diaphragmatic hernia were found to be high-risk factors¹⁰.

Several methods of measuring IAP are known but are difficult to accomplish in a neonate⁴. The methods include measuring bladder pressure via a Foley's catheter as well as respiratory monitoring and the use of manometry or pH studies to approximate intragastric pressure^{7,8}. Ultrasound imaging displaying fluid in the abdominal cavity could apparently be highly suggestive of IAP¹¹.

Only one case of ACS following right-sided CDH hernia repair has been reported in the literature, and the authors found that the clinical indicators of a disturbance in perfusion should act as early warning signs of ischemia⁴. Perfusion abnormalities are indicated by a sudden difficulty with ventilation or deranged blood pH and lactate level⁴. However, it may be challenging to identify the cause of changes in blood gases and increased lactic acid levels since these changes could be due to ischemia to an organ, cardiopulmonary pathology or a cerebral hypoxic-ischemic process⁴. Therefore, Doppler ultrasound is recommended to quantify perfusion to abdominal viscera; however, the most accurate and definitive diagnostic tool is urgent re-laparotomy⁴.

CONCLUSION

Serious postoperative complications following the repair of CDH are rare, such as NEC, but must be considered in any

infant who exhibits abdominal signs such as abdominal wall distension, edema and erythema postsurgical. Although abnormal blood gases and gas exchange are usually respiratory in origin; ACS could be one of the infrequent causes. Therefore, there should be a low threshold to consider exploratory surgery if there is a deterioration in vital signs and blood gases associated with abdominal signs.

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