CASE REPORT

Early onset necrotising enterocolitis in association with congenital diaphragmatic hernia in a term baby

Yu Han Koh,¹ Rebecca Cooksey,² Peter Prager³

¹Adelaide Medical School, The University of Adelaide, Adelaide, South Australia, Australia

²Paediatric Surgical Unit, Women's and Children's Hospital Adelaide, North Adelaide, South Australia, Australia

³Department of Critical Care Medicine, Women's and Children's Hospital Adelaide, North Adelaide, South Australia, Australia

Correspondence to
Mr Yu Han Koh,
kohyuhan1@gmail.com

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SUMMARY

We report a case of a male, term newborn with known left congenital diaphragmatic hernia (CDH) who presented with circulatory compromise requiring maximal inotropic support in the first 24 hours of life. Repeat X-ray at 24 hours showed pneumatosis intestinalis. Emergency laparotomy was performed for suspected necrotising enterocolitis. The terminal ileum to the sigmoid colon were frankly necrotic with multiple perforations. Subtotal colectomy was performed. Although the recovery was protracted, the baby had a favourable outcome with progressive weight gain on follow-up at 3, 6 and 9 months of age.

This case of CDH had a postnatal systemic instability that was more severe than predicted, and as well as assessing for persistent pulmonary hypertension of newborn should trigger immediate suspicion for other organ involvement.

BACKGROUND

Early onset necrotising enterocolitis (NEC) in association with congenital diaphragmatic hernia (CDH) in a term baby is rare.^{1 2} Given the significant mortality and morbidity from each of these conditions separately, the combination would be expected to have a poor outcome. We report a case with favourable outcome due to early intervention.

CASE PRESENTATION

A newborn male baby, with known left CDH, presented with circulatory compromise requiring maximal inotropic support in the first 24 hours of life.

The baby was diagnosed with left CDH on morphology ultrasound at 21 weeks' gestation. Antenatal MRI showed no liver involvement. Lung-to-head ratio was 1.26, suggesting an estimated survival of 61%.³ The pregnancy was otherwise uncomplicated.

The baby was electively induced at 37 weeks and delivered via forceps. Foetal tracing showed no intrapartum distress. He was crying at birth, with Apgar score of 8+9. Birth weight was 4000 g. Immediately post-delivery, he was electively intubated, ventilated and a nasogastric tube was passed. Initial ventilator parameters were peak inspiratory pressure at 24 cmH₂O, peak end expiratory pressure at 5 cmH₂O, fraction of inspired oxygen (FiO₂) at 0.35 and ventilator rate of 32 breaths per minute. Initial chest X-ray showed a right shift of mediastinal structures with the left hemi-thorax containing abdominal contents including a visible

loop of bowel (figure 1). Bedside echocardiography showed a structurally normal heart with a large patent ductus arteriosus (PDA).

At 16 hours of age, he developed persistent tachycardia, hypotension and worsening mixed acidaemia. Findings were consistent with circulatory compromise. He required inotropic support with norepinephrine at 0.5 µg/kg/min and epinephrine at 0.2 µg/kg/min. Hydrocortisone was commenced to support the ongoing hypotension. Nitric oxide was started at 10 ppm for presumed worsening of pulmonary hypertension (lowest preductal SpO₂ 85%, lowest postductal SpO₂ 76%). In view of the suspected septicæmia, gentamicin and benzylpenicillin were added. Conventional ventilation was switched to high-frequency oscillation ventilation with the following settings, frequency at 7 Hz, mean airway pressure at 20 cmH₂O, FiO₂ at 1.00 and amplitude at 50 cmH₂O to support the mixed acidosis. Despite these measures, the baby continued to deteriorate.

INVESTIGATIONS

Repeat X-ray showed pneumatosis intestinalis with associated further worsening of left lung compression and further right mediastinal shift (figure 2). Refractory clinical deterioration was attributed to the ongoing gastrointestinal insult secondary to NEC.

Figure 1 Chest X-ray performed at birth, showing the right shift of mediastinal structures with the left hemithorax containing abdominal contents.

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Figure 2 Abdominal X-ray performed at 24 hours of age, showing pneumatosis intestinalis.

DIFFERENTIAL DIAGNOSIS

Ischaemia of incarcerated bowel, NEC.

TREATMENT

Emergency laparotomy was performed at 24 hours of age. The entire small bowel, transverse colon and spleen were herniated into the left hemithorax through the diaphragmatic defect. No anatomical cause for obstruction was found. The terminal ileum to the sigmoid colon were frankly necrotic with multiple perforations. Subtotal colectomy via a left subcostal laparotomy was performed, ligating remaining bowel ends. Thoracic contents were reduced, meconium in the left hemithorax and peritoneal cavity was washed out, and primary repair of the diaphragm defect was performed. The diagnosis of NEC was confirmed on surgery and on histopathology (figure 3). Laparostomy was formed with a plan to relook when the baby's condition stabilised.

OUTCOME AND FOLLOW-UP

Postoperatively, ischaemia and multiple perforations resulted in multiorgan failure, including persistent lactic acidosis, fluid overload, coagulopathy (requiring multiple blood products), acute kidney injury and liver dysfunction. There were difficulties with ventilation and oxygenation due to the above in combination with hypoplastic left lung, pulmonary haemorrhage post suctioning, persistent pulmonary hypertension of newborn (PPHN) with left to right shunt and congestive cardiac failure.

Haemodynamic stability required norepinephrine at 0.8 µg/kg/min, epinephrine at 0.6 µg/kg/min and hydrocortisone.

Figure 3 Histopathology of the resected sigmoid colon: (A) extensive necrosis with loss of nuclear staining within the mucosa, (B) numerous gas cysts in the submucosa.

PPHN was difficult to treat, requiring prostin infusion, nitric oxide, bosentan and sildenafil. His recovery was further complicated by multiple episodes of likely sepsis, ventilator-associated pneumonia (methicillin-susceptible staphylococcus aureus and klebsiella) and klebsiella bacteraemia. Antibiotic coverage was switched to piperacillin/tazobactam.

Four days after surgery, an acceptable heart rate, mean arterial pressure and urine output (augmented with albumin infusion and furosemide diuresis) were achieved. Coagulopathy and metabolic acidosis subsequently resolved. A relook laparostomy was done. Small bowel was healthy up to terminal ileum ligated previously. The remaining sigmoid up to peritoneal reflection appeared non-viable and was resected. Ileosigmoid anastomosis and loop ileostomy performed. Laparostomy was closed.

Parenteral nutrition was introduced at day 4 of life, with commencement of enteral feeds 4 days later. Eventually, the baby's large PDA (>4.2 mm on echocardiography) was ligated with thoracotomy, in attempt to improve his pulmonary hypertension. The baby was discharged home at 3 months of age on low flow oxygen (0.5 L/min) and weaning dose of sildenafil. An uneventful elective ileostomy closure was performed at 9 months of age.

DISCUSSION

NEC in the setting of CDH is rare with only one other study describing such phenomenon, although with poor outcome and occurring postsurgical repair of diaphragmatic defect.⁴

NEC in the setting of CDH is a challenging diagnosis even for experienced clinical teams. In our case, the occurrence of NEC did not follow the time pattern described previously, typically after 5 days and after introduction of feeds.⁵ Systemic manifestations of NEC can be difficult to differentiate from the postnatal cardiorespiratory instability secondary to pulmonary hypoplasia encountered in the setting of CDH.⁶ In addition, early radiographic findings of NEC can be non-specific and even harder to appreciate in the setting of altered anatomy in CDH.⁷

Patient's perspective

Interviewed patient's parents when baby was 9 months old.

Although he had a major setback at birth, both my partner and I are very amazed by his progress thus far. During his last routine follow-up, he was tracking along the 20th percentile for his weight, 90th percentile for length and head circumference. He is catching up with the other kids of his age. At home, he is starting to walk unsupported, calling out 'mama' and 'dada', feeding well, gaining weight and his bowel frequency has slowed down. The cardiologist and respiratory doctors are also happy with his heart and lungs. These have been very reassuring for us, as parents.

The care that he received so far has been very thorough and we would like to express our gratitude to the doctors who have made today possible. We hope that this case report can help share valuable information to the medical community.

Learning points

- ▶ As demonstrated, this case of congenital diaphragmatic hernia (CDH) had a postnatal systemic instability that was more severe than predicted, and as well as assessing for pulmonary hypertension of newborn, should trigger immediate suspicion for other organ involvement.
- ▶ Necrotising enterocolitis as a differential diagnosis should be considered in a haemodynamically unstable baby with CDH despite adequate cardiopulmonary stabilisation.
- ▶ Emergency laparotomy should not be delayed. This minimises both the loss of 'functional' bowel and the subsequent impact of peritonitis secondary to perforation in necrotic bowel on overall haemodynamic.

Current approach for CDH defect repair favours surgery after the degree of pulmonary hypertension improves.^{8 9} In our case, the repair of the diaphragmatic defect occurred earlier than expected due to the ongoing gastrointestinal insult, which

affected the management of the cardiopulmonary function. Considering the risk of further bowel loss and worsening cardiopulmonary function, we feel that early intervention is indicated to minimise both the loss of 'functional' bowel and the subsequent impact of peritonitis secondary to perforation in necrotic bowel on overall haemodynamic.

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