

Delayed finding of Congenital Duodenal Obstruction following Congenital Diaphragmatic Hernia repair

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Abstract

Congenital diaphragmatic hernia (CDH) is encountered in just under 1 in 6000 live births whilst congenital duodenal obstruction (CDO) is slightly less common and is seen once every 8000 live births. These congenital anomalies have only been reported to occur together as part of the VACTERL syndrome and therefore in isolation represent an incredible rare occurrence. This is a case report of a baby girl born at 34 weeks gestation who had an antenatal diagnosis of left CDH. Five days following operative repair of this there was extensive pneumoperitoneum and left sided pneumothorax. Upper gastrointestinal contrast study showed a perforation of the duodenum and at laparotomy a duodenal web was found in the fourth part of the duodenum with perforation immediately proximal. Following duodenoduodenostomy the post operative recovery was good. She has achieved full enteral feeds and has been discharged from the neonatal unit. This case highlights the importance of considering rare associations if post operative recovery is not as expected to prevent a delay in undergoing definitive treatment.

Case report

Baby Z was born at 34 weeks gestation via emergency caesarean section due to foetal distress weighing 1.13kg. She had an antenatal diagnosis of left sided congenital diaphragmatic hernia (CDH) and her parents underwent counselling for this at a tertiary surgical neonatal unit. No other anomalies including a “double bubble” sign were seen on antenatal ultrasonography. She was born in poor condition and intubated by the third minute of life. She was commenced on high frequency oscillated ventilated on admission to the neonatal intensive care unit with 100% FiO₂ and nitrous oxide. She required extensive inotropic support initially with pre and post ductal saturations of 90% and 84% respectively.

Echocardiography revealed a patent ductus arteriosus and ventricular septal defect. There were no other congenital anomalies detected and genetic sequencing showed 46XX.

By day 9 of life she was receiving no inotropic support and required 25% oxygen on convention ventilation with pressures of 23/6. She weighed 1.53kg and was taken to theatre on this day for a thoracoscopic left CDH repair which revealed a 5x4cm left posterior lateral diaphragmatic defect containing small bowel, colon, spleen and left lobe of liver. There was difficulty reducing the contents of the hernia and the procedure was converted to open via a

subcostal laparotomy. After returning the contents of the hernia to the abdomen a polytetrafluoroethylene patch (Gore-Tex, W. L. Gore & Assoc Inc, Flagstaff, USA) was used to close the defect with posterior and lateral fixation around the rib. A 10Fr chest drain was left in-situ.

On day 14 of life on the neonatal unit, which was day 5 post op, she was seen to have a pneumoperitoneum and persistent pneumothorax (figure 1). This was initially thought to be an air leak from the left hemi-thorax and therefore an abdominal drain was placed in addition to the chest drain. Drainage was of air only.

Despite chest and abdominal drainage there was no improvement in the appearance of the radiographs with persistent pneumoperitoneum which re-collected following aspiration and drainage. Given the suspicion of visceral perforation she underwent an upper gastrointestinal water soluble contrast study (figure 2). This showed contrast leak consistent with visceral perforation.

She was taken back to theatre on day 22, day 13 post op, for a laparotomy where a perforation was found in the fourth part of the duodenum proximal to a duodenal web. The small and large bowel were inspected in entirety and no distal atresia or web was discovered. She had non-rotation of the midgut with a wide based mesentery (figure 3). The CDH repair remained intact. Given this finding the existing drains were removed and 3cm of duodenum containing the web was resected and she underwent duodenoduodenostomy.

She was commenced enteral feeding on day 14 post duodenoduodenostomy and established full enteral feeds on day 23. She required decreased respiratory support over the coming days and was extubated on day 60 of life. She has not experienced any further complications and was discharged from the neonatal unit on day 82 of life.

Review of literature

This is the first reported case of congenital duodenal obstruction (CDO) associated with congenital diaphragmatic hernia (CDH) in an infant without a syndrome or chromosomal anomalies. The importance of this, particularly in the era of thoracoscopic CDH repair, is that this association should be considered if there is intra-abdominal concerns following CDH repair. Here we demonstrate a late, atypical presentation identified due to duodenal perforation and pneumoperitoneum.

Population based study within the United Kingdom and Ireland found that the incidence of CDH is 1 in 5880 live births and the majority of infants had an antenatal diagnosis.(1) These babies are typically critically unwell and as many as 44% required high frequency oscillated ventilation and 61% required inotropic support. Survival to surgery was around 80% of infants with early mortality mostly due to severe pulmonary hypertension. Thoracoscopic repair of CDH has gained popularity in recent years and has similar results to the open procedure without the need for an intra-abdominal procedure.(2) A disadvantage of this approach is that there is no opportunity to discover rare intra-abdominal associated anomalies using this technique. However, intra-abdominal associated anomalies requiring intervention are unusual.(3)

CDO, consisting of a duodenal web or atresia, is rarer than CDH and is seen in 1 in 8197 live births in the UK and Ireland.(4) The diagnosis is most commonly made antenatally and associated anomalies are common. Around 50% of these anomalies are cardiac however in the largest population based study to date there were no reported cases of CDH.(5) The finding of a duodenal web in the fourth part of the duodenum is particularly unusual as these

are usually seen in more proximal duodenum. Moreover, duodenal webs occur in only 38% of CDO. Outcomes of CDO are very good with 98% survival at 28 days post-surgical repair.(5) Although the use of laparoscopy for these repairs has been reported, 95% of these procedures are still repaired using an open technique.(5) Laparoscopic repair does offer similar outcomes with improved cosmesis.(6)

Duodenal or gastric perforation secondary to CDO is rarely reported.(7-9) This is likely due to early detection of the anomaly, gastric decompression and prompt surgical repair. In our case there was initial uncertainty of the source of pneumoperitoneum and pneumothorax. It only became apparent following an upper gastrointestinal contrast study that clearly demonstrated an extra-luminal leak of contrast. Human factors training appears much more commonly on undergraduate and post graduate medical curriculums in recent years.(10)-statement unclear. Without the co-existing CDH in our patient or the presence of pneumothorax, there may have been an opportunity for earlier intervention. It is apparent that diagnostic confusion was caused by cognitive bias in this incredibly rare association. Our infant had a very good outcome however with no adverse effects of these two significant congenital anomalies.

Existing reports of this rare association, of CDH and CDO, are limited. In a child with VACTERL association right CDH and duodenal atresia has been reported.(11) Extrinsic duodenal obstruction due to a wandering spleen in CDH has also been described.(12) Our case is an intrinsic CDO in a child without a chromosomal anomaly and highlights the importance of considering this association in any infant that fails to tolerate enteral feeds or develops pneumoperitoneum post-operatively.

Figure legends

Figure 1 - Chest and abdominal radiograph on day 1 of life demonstrating left congenital diaphragmatic hernia and absence of dilated bowel loops.

Figure 2 – Chest and abdominal radiograph on day 14 of life demonstrating left pneumothorax and pneumoperitoneum.

Figure 3 – Image taken from upper gastrointestinal tract contrast study demonstrating extra-luminal leak of contrast in the duodenum.

Figure 4 – Intraoperative image of perforation proximal to duodenal web.

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