

**COLONIC DUPLICATION CYSTS AS A CAUSE OF BLEEDING PER RECTUM IN
INFANTS: A CASE REPORT AND REVIEW OF LITERATURE*****Kirsty Wallace and James Lucocq**

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ABSTRACT

Enteric Duplication Cysts may be located anywhere along the gastrointestinal tract, with 75% located in the intra-abdominal cavity. Of these the most common site is the small bowel, with colonic duplication cysts being less common. They can present in multiple different ways, but are often asymptomatic. In this study, we report a case of a colonic duplication cyst in an infant which presented with bleeding per rectum, and review the literature surrounding enteric duplication cysts.

INTRODUCTION

Enteric Duplication Cysts (EDCs) are rare gastrointestinal tract lesions, initially reported in 1733.^[1] They are most commonly discovered in the first year of life, and are more common in children than adults.^[2] They are non-malignant, but have the potential for malignant transformation in the adult population.^[3] EDCs may be found anywhere along the gastrointestinal tract from mouth to anus, with the most common site being the ileum.^[2] Histologically, they can contain tissue from any part of the gastrointestinal tract, surrounding a fluid-filled sac.^[4] Presentation may be via a range of gastrointestinal symptoms, making diagnosis challenging.^[5]

In this case we will discuss an infant who presented with bleeding per rectum, on a background of previously discovered colonic EDC. A review of the current literature surrounding diagnosis and the management of EDCs will be undertaken.

CASE

A 9 week old male infant was admitted with a 1 day history of acute abdominal pain, 3 weeks of dark stools and fresh red clots per rectum. The parents noted poor feeding, increased agitation, lethargy, and frequent bowel movements.

The infant was born at term, with nil complications either in utero or following delivery. At birth they had the following malformations: microphthalmia, cleft palate, and unilateral deafness. A neonatal ultrasound was undertaken due to the presence of a mass felt on abdominal palpation when they were born. They were being investigated for cow's milk protein allergy, and

were maintained on a diet of breastfeeding supplemented with altered formula milk.

Observations on admission were unremarkable: HR 118bpm, T 37.2°C, RR 46bpm, O₂ saturations 100% on air. He was irritable throughout examination, which demonstrated a mildly distended abdomen in the absence of any palpable masses. Stool was thick in consistency, very dark coloured, with some clots and fresh red blood present. Bloods taken on admission showed a mild anaemia (Hb 88g/L), but otherwise were within normal limits (liver function tests, urea and electrolytes, coagulation screen, and inflammatory markers).

An ultrasound was reported as showing a complicated enteric duplication cyst measuring 1.9cm at its maximum, communicating with a bowel loop, likely small bowel (Figure A). There was surrounding thickened and echogenic mucosa indicating associated inflammation and enlarged mesenteric lymph nodes (approximately 5mm in diameter). The cyst appeared to be the lead point of a partial intussusception of the bowel.

He was made nil by mouth, commenced on maintenance fluids intravenously and taken to theatre the following day for laparoscopy. This revealed a haemorrhagic cyst adhered to the descending colon, approximately 3cm distal to the splenic flexure (Figure B and C). The cyst wall was inflamed and adhered to the bowel wall rendering the cyst excision difficult via laparoscopy. Conversion to laparotomy was therefore performed and the cyst was excised. Due to cyst location and size, coupled with its haemorrhagic nature the decision was made to resect a segment of the descending colon (Figure

D). Primary anastomosis of the bowel was then performed, with minimal leakage.

Post-operatively, he was commenced on 48hrs of intravenous antibiotics (cefuroxime and metronidazole), and continued to be nil by mouth for the 24hrs following procedure. Intravenous fluids and analgesia were prescribed. He had one self-terminating pyrexial episode in the immediate post-operative period, but was clinically well throughout and subsequently remained afebrile.

Repeat bloods undertaken on the day following surgery showed a decreased haemoglobin of 83g/L, likely a dilutional drop, indicated by a haematocrit of 0.24. White blood cells were raised at $19.3 \times 10^9/L$. All other parameters remained normal (liver function tests, urea and electrolytes, coagulation screen).

He remained clinically stable on the ward, and had no further episodes of abdominal pain or blood in stools. He was therefore deemed medically safe for discharge home three days following surgery with outpatient follow-up.

Appendix

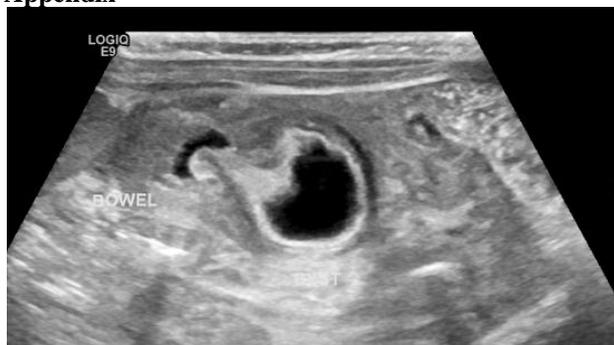


Figure A: Abdominal ultrasound image of EDC attached to the bowel wall.



Figure B: Enteric duplication cyst viewed via laparoscopy.

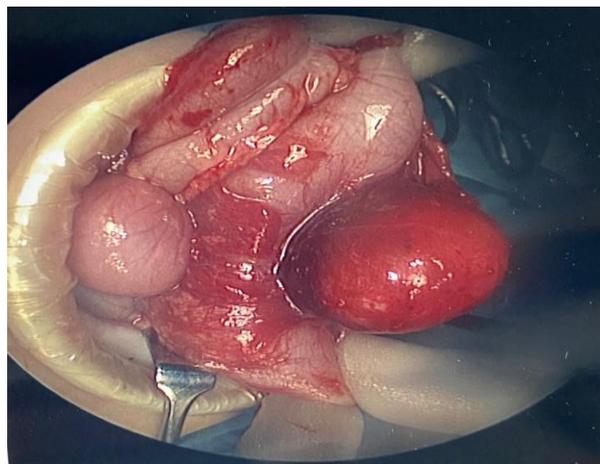


Figure C: Enteric duplication cyst attached to bowel prior to excision.



Figure D: Resected enteric duplication cyst and resected descending colon.

DISCUSSION

Enteric duplication cysts (EDCs) are rare fluid filled sacs of the gastro-intestinal (GI) tract, which are more commonly found in children than in adults.^[6] Approximately 67% of EDCs are discovered within the first year of life.^[2] They can contain tissue from anywhere along the GI tract, but to be classified as an EDC it must have the following features: contain epithelial lining with mucosa from the GI tract, have a smooth muscle coating, and be closely adhered to, but not communicating with, the GI tract (share a tract wall).^[4] Of note, the tissue contained within the cyst is often not the same as the structure to which the cyst is attached.^[2] In these cases, the cyst is named for its location, not its histological structure.^[2]

EDCs form between weeks 4 and 8 in utero, but precise formation cause is currently unknown.^[4] Several theories exist, one of the which is the 'Abnormal Recanalization Hypothesis' where it is believed that epithelial cells may proliferate in an abnormal pattern during development and cause deformity/occlusion of the GI tract.^[7] Another

theory suggests that additional foregut buds forms in utero, which become de-functioned and relocate as the GI tract grows and develops.^[8]

EDCs may occur anywhere along the GI tract from mouth to anus.^[2] with 75% being located within the abdominal cavity.^[9] The most common site of EDC location is the small intestine (approximately 50%), in particular the ileum.^[6] The next most common location is the colon, where approximately 6-13% of EDCs are located.^[4,6]

Many EDCs are found incidentally (especially in adults), but some may present symptomatically when the cyst fills with fluid.^[7] Approximately 30% of EDCs contain gastric mucosa,^[9] which may secrete enzymes into cystic fluid leading to inflammation and subsequent pain.^[7] Presenting symptoms vary depending on location, for example dysphagia if it is located in the oesophagus or meleana if it is located more distally.^[5] Although GI bleeding is a rare presenting symptom for EDCs due to the lack of communication between the cyst and the GI tract, it should be noted that some EDCs may have their own blood supply.^[10] As was the case with our patient, haemorrhage into the cyst can cause leakage at the site of communication with the bowel, and as such the patient may present this way.^[7]

The wide range of presentations can make diagnosis challenging.^[5] EDCs are usually diagnosed either pre-natally or in the first year of life,^[2] with an increasing number of cases being diagnosed in utero, as improved screening images evolve.^[9] Currently, however, it remains that only 20-30% of cases are diagnosed prior to birth.^[11] Ultrasound (US) imaging is the imaging modality of choice in children,^[4] but some challenges still exist with distinguishing an EDC from other cystic structures.^[7] The presence of an echogenic centre to the cyst with a regular internal muscular layer of hypoechoic tissue, suggests an increased probability of EDC.^[2] Other features assessed for on imaging include the 'gut signature' sign (the presence of a double walled muscular rim), and the 'Y' sign (splitting of the muscular layer as the bowel tissues develop into the attached cyst).^[4] If there is any diagnostic uncertainty in adults, or if the management plan remains unclear, a CT scan or MRI scan may be considered.^[6]

There is the potential for malignant transformation in some of these cysts.^[2,3,10] with one study in adults showing that 23% of resected EDCs in the ileum were found to be ileal neoplasms.^[1] Other complications seen include intestinal obstruction via external compression, which is the most common surgical emergency encountered in neonates,^[9] and intussusception, as was seen with the patient discussed in this report. Intussusception occurs as the site of cyst location on the bowel acts as a 'lead point' to draw the bowel back in on itself.^[4] The presence of gastric mucosa in the cyst can also lead to ulceration and rarely perforation – a life

threatening emergency.^[4] Early intervention should therefore be undertaken to avoid development of these complications.^[11]

Treatment is via early surgical resection of the cyst.^[4] This is the case for all EDCs, both symptomatic and asymptomatic, as complications can arise as detailed above which could prove life threatening.^[2,4] Early intervention also decreases the hospital stay, and limits the need for additional treatments.^[2,4] Laparoscopy is considered the 'gold standard', as this is associated with less abdominal trauma and therefore a quicker recovery than abdominal laparotomy.^[3] If the EDC is large, or there is complex involvement of the bowel then laparoscopy may be converted to laparotomy, as was the case with the patient discussed in this report. In our case, the complex involvement of the descending colon meant that this had to be resected in addition to the cyst – another recognised complication.^[9]

CONCLUSION

In conclusion, EDCs are a relatively rare finding but are more common in children than adults. It should be considered as a differential in children presenting with any GI symptoms, and can be investigated for using ultrasound scanning. Treatment is via surgical resection, with laparoscopic procedure being the gold standard. EDCs should be resected in all cases, regardless of the presence of symptoms, as there is a high risk of future complications such as obstruction, intussusception, or malignant transformation in adults.

Consent

Consent was gained verbally from the mother of the child via phone call at time of writing report. Consent for photograph and medical record use was also obtained at time of surgery via consent form.

Conflicts of Interest

There are no conflicts of interest to declare.

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