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Best practice guidelines

Malrotation and intestinal atresias

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ABSTRACT

This review encompasses four congenital conditions which present with symptoms of bowel obstruction in the neonatal period. The antenatal and postnatal features of malrotation, jejuno-ileal atresia, duodenal atresia and colonic atresia are discussed. Each condition is outlined including the classification, epidemiology, aetiology and presentation, and a summary of the surgical management is described.

Contents

1. Introduction

Bowel obstruction due to malrotation or bowel atresia occurs in around 6 per 10,000 live born infants. This article reviews the major congenital obstructive pathologies, presenting a summary of the condition in each case and a guide to management.

2. Malrotation

Malrotation is a congenital anomaly of intestinal position and may lead to volvulus of the mid-gut around a narrow-based mesentery. The importance of considering this diagnosis cannot be over-emphasised when an infant presents with bilious vomiting, as left untreated, mid-gut infarction rapidly occurs. Malrotation presents in around 1 in 2500 live born infants (under 1 year of age), however as an anatomic entity it is much more common than this, occurring in 0.2%–1% of the normal population.

Normal intestinal rotation is established initially by physiological herniation of the embryonic intestinal loop into the umbilical cord in the 4th gestational week, associated with a 90° anti-clockwise rotation. The mid-gut returns to the peritoneal cavity by the 8–10th week. As it does so, it rotates a further 180°, completing a total of 270° anticlockwise rotation, such that the duodenal–jejunal (DJ) comes to lie on the left of the midline and the caecum in the right iliac fossa. This standard description has been challenged by more recent experimental evidence [1]. The relative rotation is likely to be due to rapid differential growth, the most important feature of which is development of the duodenal loop. It may be that the return of the rest of the intestine occurs by a more passive mechanism, rather than being driven by rotation.

The classical picture of malrotation results from an error in the normal process outlined. The commonest end-result is that of the caecum lying to the left of the midline, the DJ flexure lying to the right of the midline, narrow mesentery which lacks fixation, and peritoneal (Ladd's) bands passing from the caecum to the right side across the duodenum. There is much discussion of Ladd's bands in the literature, these being the result of the caecum's attempts at peritoneal fixation. They may be present but only occasionally give rise to duodenal obstruction. The primary concern is that mid-gut volvulus may occur

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following twisting of the narrow mesentery and thus threaten the venous outflow and subsequently the arterial inflow to the intestine.

Other rotational anomalies are described — non-rotation is seen in the context of abdominal wall defects and congenital diaphragmatic hernia. The duodenum passes straight inferiorly on the right side; the colon lies on the left. Reverse rotation is described where the duodenum passes in front of the superior mesenteric artery, with the colon lying in front of the mesentery. Internal small bowel hernias are also considered a subgroup of rotational anomalies.

There is a high rate of associated anomalies reported in infants with malrotation. Type IIIb small bowel atresia may be a secondary outcome of antenatal volvulus. Other common associations include cardiac anomalies, ano-rectal malformation, duodenal web and trisomy 21. Non-rotation is almost ubiquitous in children with abdominal wall defects or congenital diaphragmatic hernia. Interestingly, the association of volvulus is rather less with this group, perhaps due to the contribution of post-operative (or congenital) bowel adhesions. Intestinal rotation anomalies are seen in association with heterotaxia. It is debated whether elective investigation and surgical correction of malrotation are merited in the context of heterotaxia, in particular when significant cardiac anomalies may be present. Recent evidence suggests that the morbidity from an elective procedure, when the infants cardiac status is relatively stable, may be low, however, the overall risk of volvulus appears to be low and screening in all cases may be misleading.

The most common clinical presentation is with bile vomiting during the first month of life (>50% cases); this may be accompanied by abdominal distension or bleeding per rectum as a late sign. Presentation subsequent to this age becomes more varied and may include episodic vomiting (often non-bilious), recurrent abdominal pain or simply failure to thrive, all of which may have been attributed to gastro-oesophageal reflux [2].

The diagnosis of malrotation is usually made by means of an upper GI contrast examination. Plain radiographs in cases of established volvulus typically show air in the stomach and proximal duodenum and a gasless appearance beyond. If the presentation is of an infant with peritonitis, then immediate laparotomy is mandated without the luxury of contrast examination first. Otherwise, plain XRs can be normal and do not exclude malrotation. The hallmark feature on upper GI contrast examination is a duodeno-jejunal flexure that does not pass to the left of the spinous process of the 2nd lumbar vertebra, together with a low-lying pylorus. If volvulus is present this may show as complete obstruction ('bird-beak') at the level of the proximal to mid-duodenum or indeed as a 'cork-screw' type appearance as the small bowel passes from posterior to anterior (Fig. 1). Ultrasonographic features have been described, in particular reversal of the position of the superior mesenteric artery and vein, although this modality would not be considered reliable enough to exclude malrotation when clinically suspected. Occasionally malrotation may be detected incidentally, for example on CT scan. For truly incidental older children (>2 years), some authors describe avoiding operation and advise urgent investigation and treatment should any gastro-intestinal symptoms occur. The authors' practice is to recommend semi-urgent surgical correction.

The surgical procedure to correct malrotation was described by William Ladd in 1936 and still remains in widespread use. The premise is to de-rotate the mid-gut if it is twisted, straighten the duodenum, widen the mesenteric root and place the intestine in a stable position. To minimise the risk of recurrent volvulus, the small bowel is placed on the right side of the peritoneal cavity and the colon to the left. The caecum is positioned in the left upper quadrant thus an incidental appendicectomy may be performed to avoid future diagnostic confusion; inversion appendicectomy is the authors' preference but this step remains controversial. The operating surgeon may be faced with the situation of volvulus and total mid-gut infarction at initial laparotomy. If mid-gut resection is under taken at this stage, the infant is committed to short-gut syndrome with the possibility of intestinal transplantation. A difficult ethical consideration arises which should be managed with full discussion with the family to allow informed choice. If there is any question regarding viability, the bowel is returned to the abdominal cavity and a second-look laparotomy can be performed at 24–48 h. A successful reversal of apparently established infarction of the mid-gut using post-operative systemic thrombolysis has been reported in 2 infants and appears to be a promising therapy [3]. Laparoscopic Ladd's procedure is now well-established, but may not be appropriate in the emergency setting.

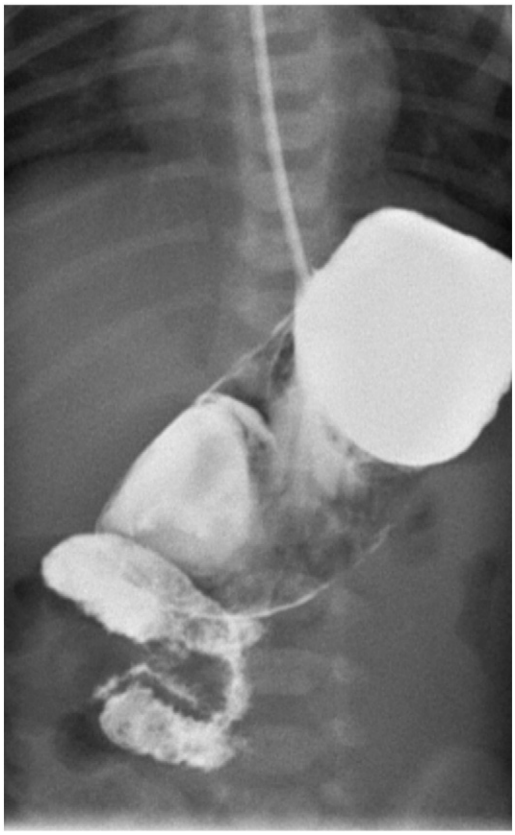

Fig. 1. Radiograph from an upper GI contrast examination demonstrating classical appearances of malrotation with volvulus of the mid-gut. The DJ flexure lies to the right and there is a corkscrew appearance beyond.

Complications of Ladd's procedure include adhesive small bowel obstruction (6%), incisional hernia, perforated viscus and recurrent volvulus (around 1% each). Malrotation still accounts for a significant proportion of children with short-gut syndrome. Mortality depends on whether bowel ischaemia is present and is approximately 3% [4].

3. Jejuno-ileal atresia

Jejuno-ileal atresia (JIA) is the commonest type of intestinal atresia and accounts for around 1 in 5000 to 1 in 14,000 live births. Over one third of affected children are born prematurely, there is no sex predominance and usually there are no associated chromosomal abnormalities (<1%).

Small bowel atresias are classified using Louw's classification (Table 1, Fig. 2) and the relative frequency spreads reasonably evenly between 4 groups. Type III is further subdivided (as per Grosfeld et al. [14]) to account for IIIb — the apple peel atresia which is present in 7% of JIAs. The apple peel deformity results in an atresia just beyond the DJ flexure, with the remaining small bowel coiled around the ileocolic artery. The superior mesenteric artery is absent beyond the origin of the middle colic vessel. Type IIIb and IV atresias are more commonly