Bilious vomiting in the newborn

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Abstract

Bilious (dark green) vomiting in the newborn is a surgical emergency as the underlying diagnosis may be volvulus of the entire midgut secondary to malrotation. This diagnosis is time critical as, untreated, volvulus will lead to midgut necrosis, resulting in short gut syndrome or death. While a range of other diagnoses are possible, transfer to a paediatric surgical centre should be undertaken urgently so that malrotation/volvulus can be excluded or treated. This review focuses on the causes, investigation and management of proximal bowel obstruction in the neonate that present primarily with bilious vomiting.

Keywords Duodenal atresia; malrotation; small bowel atresia; volvulus

Introduction

Bilious (dark green) vomiting in a newborn is a concerning feature as it indicates mechanical intestinal obstruction at or distal to the ampulla of Vater in the second part of the duodenum. Such obstruction occurs in approximately 6 per 10,000 live births. Malrotation/volvulus is the working diagnosis until proven otherwise. Neonates with distal bowel obstruction have bilious vomiting as a later sign, but also have features such as failure to pass meconium and abdominal distension. Bilious nasogastric aspirates in a premature infant are relatively common, and are usually due to dysmotility and do not necessarily require such rapid investigation.

Initial management is directed at intravenous fluid resuscitation and naso-gastric decompression (aiming for a 8 Fr tube). Antenatal history may be consistent with bowel atresia if, for example, a 'double—bubble' typical of duodenal atresia has been seen on the antenatal scan. Examination should include the groin to exclude incarcerated inguinal hernia, and the perineum to confirm patency of the anus. A careful history of timing of passage of meconium should also be elucidated. In cases of proximal bowel obstruction, there may be little accompanying abdominal distension. A plain abdominal radiograph is undertaken to assess the bowel gas pattern, but this may be misleadingly normal in malrotation/volvulus and should not in itself be considered reassuring. Urgent referral and transfer to a paediatric surgery centre should be undertaken at this stage. Unless there is clear clinical/radiological evidence of a bowel atresia or distal obstruction, or the neonate is seriously unstable, then an urgent upper GI contrast study should be performed to assess bowel rotation.

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Malrotation

Malrotation is a congenital anomaly of intestinal position and presents clinically in around 1 in 2500 live born infants (under 1 year of age). However, as an anatomical entity, it is much more common than this, occurring in 0.2%−1% of the normal population. The importance of considering a diagnosis of malrotation in any baby presenting with bilious vomiting cannot be overemphasized, as malrotation can lead to volvulus of the mid-gut around a narrow-based mesentery, which if left untreated, midgut infarction can rapidly occur (<6 hours). There is a high rate of other congenital anomalies associated with malrotation. These include cardiac anomalies, ano-rectal malformation, duodenal web and trisomy 21 (Down syndrome). Intestinal rotation anomalies are also seen in association with heterotaxia. Type IIIb small bowel atresia may be a secondary outcome of antenatal volvulus.

Patho-embryology

Physiological herniation of the embryonic intestinal loop into the umbilical cord occurs in the 4th week of gestation. The mid-gut returns to the peritoneal cavity between 8 and 10 weeks and during this process is associated with a total of 270° counterclockwise rotation, leading to the normal anatomical arrangement of the duodenal—jejunal (DJ) flexure being situated on the left of the midline and the caecum being within the right iliac fossa. Malrotation results from a failure of this process. The most common abnormal configuration of malrotation has the DJ flexure lying to the right of the midline, the caecum to the left of the midline, a narrow mesentery lacking fixation to the posterior wall of the peritoneal cavity, and peritoneal (Ladd's) bands passing from the caecum to the right side across the duodenum. Ladd's bands, while often present, do not usually themselves contribute to duodenal obstruction. Mid-gut volvulus occurs due to torsion of the narrow 'mobile' mesentery, leading to venous obstruction and subsequent failure of arterial inflow.

A wide range of other rotational anomalies occur. '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 may also be considered a subgroup of rotational anomalies.

Presentation

The most common presentation of malrotation is bilious vomiting during the first month of life (>50% cases). This may be accompanied by variable degrees of abdominal distension. Babies with malrotation volvulus are usually hypovolaemic and can be profoundly unwell. Presentation subsequent to this age becomes more varied and may include episodic vomiting (often non-bilious), recurrent abdominal pain, or growth failure, previously attributed to gastro-oesophageal reflux[1]

Diagnosis of malrotation is usually confirmed by means of an upper GI contrast examination. Plain radiographs in established volvulus classically demonstrate a 'gasless' appearance; air in the stomach and proximal duodenum only. Bilious vomiting and peritonitis with a 'gasless abdomen' on plain radiograph is sufficiently concerning that immediate laparotomy without further investigation being undertaken. Conversely, a normal plain radiograph does not exclude malrotation and should not be considered reassuring.

The hallmark features of malrotation on an upper GI contrast study are the DJ flexure not crossing the midline to lie to the left of the L2 spinous process, and the low position of the pylorus positioned inferiorly to the L2 horizontal plane. With established volvulus, there may be a 'bird-beak' cut-off at the mid-duodenum, or a spiralling appearance as the contrast passes through the volved small bowel as it passes posteriorly to anteriorly (Figure 1). Ultrasound may demonstrate a reversed configuration of the superior mesenteric vein and artery. However, this is not diagnostic, and is a supplementary investigation rather than one that can rule out malrotation in isolation. Occasionally, malrotation may be detected incidentally, in which case semi-urgent surgical correction is recommended.

Surgical management

Surgical correction of malrotation was first described by William Ladd in 1936 and is still termed Ladd's procedure. The first step is to de-rotate the midgut (if twisted) in a counter-clockwise direction. The right colon is mobilized laterally and reflected medially. This allows exposure of the duodenum, whose lateral attachments can be divided allowing the duodenum to be straightened. The root of the small bowel mesentery is 'widened' by carefully incising the peritoneal layer on its anterior surface. To minimize the risk of recurrent volvulus, the small bowel is placed back in the right side of the abdominal cavity and the colon on the left side. The aim is that the widened mesentery is more stable and postoperative adhesions will partially anchor the replaced bowel. As the caecum will now lie in the left upper quadrant an incidental appendicectomy is usually performed to reduce future diagnostic confusion. Some neonatal surgeons perform this by inverting rather than excising the appendix.

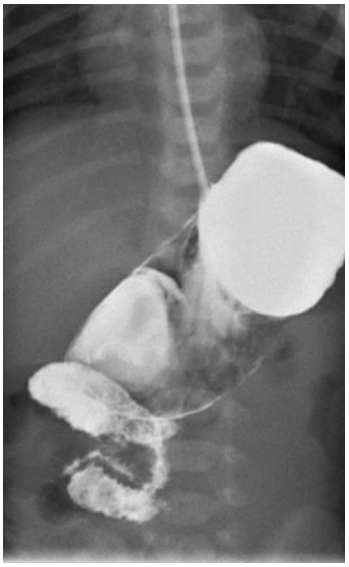

Figure 1 Appearances of malrotation/volvulus on upper GI contrast examination. The duodeno–jejunal flexure is lying to the right of the L2 pedicle with a corkscrew appearance of the distal bowel.

Established midgut necrosis presents a difficult surgical and ethical challenge. Resection of the entire small bowel results in short-gut syndrome, a life-long dependence on parenteral nutrition or subsequent need for small bowel transplantation. A full discussion (and multidisciplinary approach) with the infant's family is required to allow informed choice regarding active management or consideration of withdrawal of care. Ideally, the possibility of finding extensive necrosis will have been discussed with the family at the time of consent, so that this is not being approached for the first time postoperatively. If there is any question regarding viability, the bowel is returned to the abdominal cavity and a 'second-look' laparotomy performed at 24–48 hours. Successful resolution of apparent established midgut infarction with postoperative systemic thrombolysis has been reported in two infants and may be a promising option.[2]

Laparoscopic Ladd's procedure is now well-established, but may not be appropriate in the emergency setting. Laparoscopic assessment of bowel rotation in non-emergency cases can be performed if there is diagnostic doubt from the imaging. Complications of Ladd's procedure include adhesive small bowel obstruction (6%) and recurrent volvulus (about 1%). Malrotation volvulus accounts for a significant proportion of children with short-gut syndrome. Mortality from malrotation volvulus usually reflects whether bowel ischaemia is present or not, and is approximately 3%.[3]

Duodenal atresia

Duodenal atresia (DA) occurs in 1 in 10,000 live births, representing 60% of all intestinal atresias.[4,5] The congenital obstruction most commonly occurs in the second part of the duodenum, giving rise to bilious vomiting if the obstruction occurs distal to the ampulla of Vater. There is a slight male preponderance and many infants will have associated conditions including trisomy 21 (30%), and structural cardiac defects (25%). Up to 10% of affected children will have malrotation and a proportion will have the VACTERL association.[4]

Patho-embryology

Intrinsic congenital duodenal obstruction may occur as a result of an atresia, stenosis or web. Such anomalies used to be believed to occur as a result of failure of recanalization of the duodenal lumen by the 8th week of gestation, but most embryologists no longer believe that the duodenum completely occludes during normal human development.[6] The current theory is that hypertrophy of the primitive duodenal villi leads to occlusion. This is in contrast to the ischaemia theory that is well recognized as the aetiology of other intestinal atresias.[7] Extrinsic duodenal obstruction may occur as a result of anatomical abnormalities such as an annular pancreas (pancreatic tissue surrounding the duodenum), a pre-duodenal portal vein, or malrotation.

Classification

Duodenal atresia is classified into three distinct types:

