Bilious vomiting in the newborn

Lucinda GC Tullie
Michael P Stanton

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 may indicate 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, 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 an 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.

Lucinda GC Tullie *MRCS(Ed) is a Specialist Registrar in Paediatric Surgery at University Hospital Southampton, UK. Conflicts of interest: none declared.*

Michael P Stanton *MD FRCS (Paed Surg) is a Consultant Paediatric and Neonatal Surgeon at University Hospital Southampton, UK. Conflicts of interest: none declared.*

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. If left untreated, mid-gut 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° counter-clockwise 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.

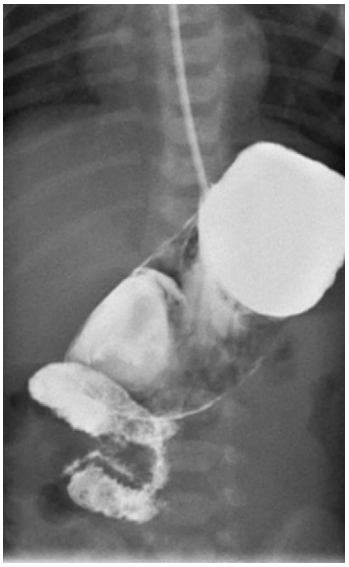

Figure 1 Appearances of malrotation/volvulus on upper GI contrast examination. The duodeno–jejunal flexure is lying to the right of the L2 pedicle with a corkscrew appearance of the distal bowel.

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:

- **Type I** involves an intraluminal membrane or web causing obstruction, and accounts for up to 90% of cases. In this type of duodenal atresia, there is no disruption of muscular or serosal layers, and hence the external appearance of the bowel, with the exception of the proximal dilatation and distal collapse, is normal. Type I defects may lead to development of a 'windsock' anomaly where the obstructing membrane distends distally, leading to the external appearance of dilatation beyond the actual level of obstruction.
- **Type 2** defects arise from obliteration of a duodenal segment with a fibrous cord connecting blind-ending proximal and distal segments.
- **Type 3** duodenal atresia involves complete separation of the proximal and distal segments and is associated with a typical 'V' shaped mesenteric defect.

Presentation

Almost half of cases of duodenal atresia are diagnosed antenatally, initially with polyhydramnios and later with a classic 'double bubble' appearance on ultrasound caused by the presence of amniotic fluid in the stomach and the duodenum proximal to the obstruction. Post-natally, infants may present with vomiting in the first 48 hours of life, with the majority being bilious if the atresia is situated distal to the ampulla. A plain radiograph showing a 'double bubble' (resulting from swallowed air in the stomach and proximal duodenum) with absence of distal gas, is diagnostic (Figure 2).

As outlined above, malrotation and midgut volvulus is an important differential, and may be excluded by clues in the antenatal history and radiographic features. However, any suspicion of malrotation/volvulus mandates emergency investigation and/or operative intervention. Distal gas beyond the 'double bubble' may indicate malrotation/volvulus, but can also occur in the rare anatomical variant of a bifid common bile duct (or pancreatic duct) inserting simultaneously into the proximal and distal duodenal segments, allowing air to pass between the two.

Management

Initial management is with supportive treatment comprising intravenous fluids and nasogastric decompression, and examination/investigation to assess for the presence of associated anomalies. Surgical correction is not usually undertaken as an emergency. Operative correction typically involves a duodeno-duodenostomy, whereby the proximal and distal duodenal pouches are opened and joined, bypassing the atretic segment. The classic teaching is that the anastomosis is performed as a 'diamond' shape to improve the patency of the join. This procedure can be performed either open or laparoscopically. For a duodenal membrane, a vertical duodenotomy, web resection and transverse closure may be performed. Particular care should be taken to avoid damage to the common bile duct, pancreatic duct or ampulla. If duodeno-duodenostomy is not feasible duodeno-jejunostomy may be performed. However, there is a recognized risk of blind-loop syndrome following this procedure. At operation, the surgeon should also assess for distal atresias and confirm the position of the DJ flexure, given the concurrent incidence of further atresias and of malrotation. Parenteral nutrition can usually be avoided with placement of a trans-anastomotic tube to enable early jejunal feeding while normal gastroduodenal function resumes.[8]

The mortality rate of around 5% in duodenal atresia is related to the high rate of associated anomalies and prematurity. Long-term outcome is usually very good in terms of intestinal function. Rarely, gross dilatation of the proximal duodenum may occur which is amenable either to re-anastomosis or tapering duodenoplasty.

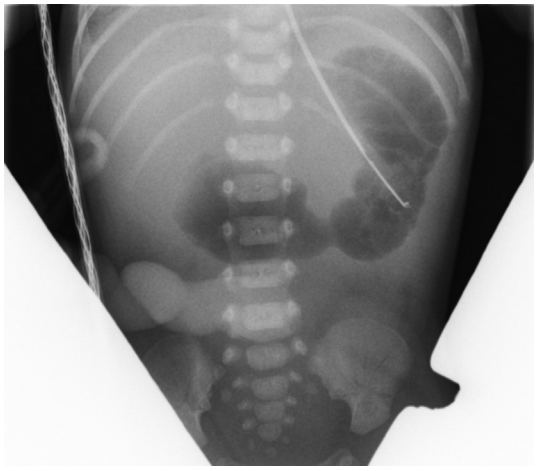

Figure 2 'Double bubble' sign on plain abdominal radiograph suggestive of duodenal atresia.

Jejuno–ileal atresia

Jejuno–ileal atresia, the most common intestinal atresia, occurs in around 1 in 5000 live births. Over one-third of affected children are born prematurely. There is no gender predominance and associated chromosomal abnormalities are rare.

Fifteen per cent of all jejuno-ileal atresias occur in infants with gastroschisis. Malrotation is present in 20% infants with jejuno-ileal atresias, and it has been postulated that an antenatal volvulus may give rise to the atresia. Concurrent duodenal or colonic atresia may occur in up to 6% of patients.

Cystic fibrosis has been reported in 10% of infants with jejuno-ileal atresia. Some clinicians therefore advocate testing these patients for cystic fibrosis, either via genetic or sweat tests. Cystic fibrosis screening in the UK is now routine for all newborns and has 98% sensitivity for common mutations.

Embryology

Jejunal and ileal atresias have long been considered to occur as a result of an antenatal vascular insult. Experiments in the 1950s, by Louw and Barnard, demonstrated that mesenteric vessel ligation resulted in atresia, with the degree of atresia dependent upon both the site and extent of the vascular insult.[7] Analysis of post-atretic intestinal content demonstrated bile salts, epithelial cells and ingested lanugo hair suggesting that this insult occurs late in fetal development.[9]

Classification

Small bowel atresias are classified using the modified Louw and Barnard classification (Figure 3):[10]

- **Type I**: a mucosal web/stenosis.
- **Type II**: atretic bowel ends separated by a fibrous cord.

- **Type IIIa**: atretic bowel separated by a 'V'-shaped mesenteric gap.
- **Type IIIb**: a tortuous atresia with a classic 'apple peel' appearance. This atresia occurs just beyond the DJ flexure, with the distal atretic bowel coiled around the ileocolic artery.
- **Type IV**: multiple atresias. Overall, jejunal atresias are more common than ileal atresias, with incidence decreasing the more distal the atresia.[11]

Presentation

Approximately one-third of cases are detected antenatally, with sonographic features of multiple dilated bowel loops and polyhydramnios. Post-natal presenting features depend upon the level of the atresia but include bilious vomiting, abdominal distension and delayed passage of meconium. Presentation may be similar to that of malrotation/volvulus and must therefore be investigated as an emergency.

Plain abdominal radiography alone is usually diagnostic, with swallowed air acting as a contrast medium (Figure 4). In proximal atresias, a small number of dilated loops are seen with an absence of distal air. The presence of calcification implies prenatal bowel perforation of the proximal atretic segment. An upper GI contrast can be utilized to exclude malrotation. Contrast enema may be required to differentiate distal atresia from meconium ileus, Hirschsprung's disease or small left colon syndrome. Atresia is excluded if contrast refluxes into dilated gas-filled loops of bowel.

Once the diagnosis is confirmed, nasogastric decompression and fluid resuscitation are instituted. At laparotomy, the bowel is inspected to confirm the position of the DJ flexure, and catheterized and irrigated to assess for distal atresias. Resection of the dilated proximal end, and primary anastomosis to the distal portion, is usually undertaken. Multiple atresias can be anastomosed, but performing more than three anastomoses at the same time is not advised, and a temporary covering stoma may be required. Preservation of bowel length is vital, and therefore, at operation, careful measurement and documentation of bowel length is important. Postoperatively short-term parenteral nutrition (PN) is usually required while normal intestinal function resumes.

Overall survival for these patients is in the region of 90%. Outcomes are generally related to the total length of the remaining intestine, as morbidity and mortality stems from short-gut syndrome and complications associated with parenteral nutrition and/or further surgery (intestinal lengthening and liver or small bowel transplantation).

Distal bowel obstruction

Presentation

Distal bowel obstruction, causes of which include Hirschsprung's disease, meconium ileus, distal ileal or colonic atresias, and small left colon, may also lead to bilious vomiting, particularly if the obstruction has been prolonged. However, infants commonly present with other symptoms/signs first, including delayed passage of meconium and abdominal distension.

Confirmation of the position of the anus is an important first step to exclude an anorectal malformation. Once the overall state

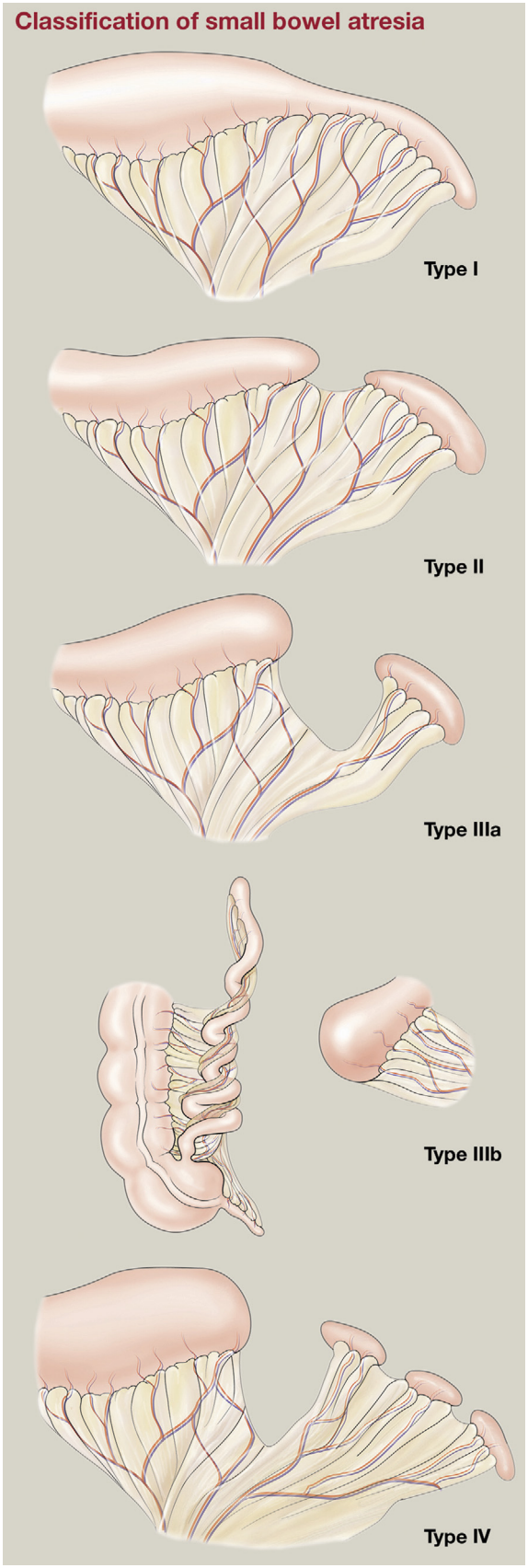

Figure 3

of the baby has been assessed and supportive measures introduced, a rectal washout is performed if the baby is stable. This serves as both an 'investigation' and potential management. Successful washout may both decompress the abdomen, relieving the obstruction, and identify the cause. As intraluminal air is only present from swallowing, passage of air alongside stool during washouts suggests bowel continuity, excluding a distal atresia. Decompression with passage of meconium 'plugs' points toward a diagnosis of meconium plug syndrome. Further investigation with a contrast enema may be required if a rectal washout fails to decompress the bowel fully. This may confirm a number of pathologies depending upon the cause of the obstruction.

Differential diagnosis of distal bowel obstruction

Hirschsprung's disease: In Hirschsprung's disease there is absence of ganglion cells in the myenteric and submucosal intestinal plexuses of the rectum and variable lengths of the colon. The length of bowel affected by the disease varies significantly. In the majority of infants, Hirschsprung's disease affects the recto-sigmoid (75%), but approximately 10% will have total colonic involvement. Hirschsprung's disease affects approximately 1 in 5000 live births and has a male preponderance.

Contrast enema may demonstrate a 'transition zone' between the aganglionic and normal bowel. However, if a diagnosis of Hirschsprung's disease is suspected, rectal biopsy is the gold standard to confirm the diagnosis. In neonates this is usually done as a suction rectal biopsy at the bedside. Histological features of Hirschsprung's disease include the absence of ganglion cells, hypertrophied nerve roots, and a characteristic pattern of acetyl cholinesterase staining.

Once the diagnosis is confirmed, initial management involves continued regular rectal washouts. However, if these fail to achieve adequate decompression then a stoma may be required. Definitive surgical management is in the form of a bowel 'pull-through' operation where the affected segment is resected and bowel continuity is restored by joining the innervated bowel to the anus. A covering stoma may be required, which is closed at a later date. (See also *Hirschsprung's Disease* on pages 640—645 of this issue).

Meconium ileus (MI) involves inspissated meconium adhering to the inside of the bowel wall leading to an ileal obstruction. Infants may present antentally with sonographic evidence of dilated bowel or with distension at birth. It accounts for up to 30% of cases of neonatal intestinal obstruction with an incidence of 1 in 3000 live births.[12] There is a strong association with cystic fibrosis, with at least 95% of patients with MI also having this disease.[12]

MI is classified as being 'simple' (50%) or 'complex' (50%). In 'simple' MI, there is obstruction by meconium only. In 'complicated' MI, the meconium obstruction is associated with atresia, volvulus or perforation. A plain radiograph may demonstrate features suggestive of MI; gastrografin contrast enema typically shows a redundant microcolon and may outline pellets of meconium more proximally. Contrast enema may be therapeutic and, if gas filled dilated loops of bowel are reached, may negate the need for a laparotomy in simple meconium ileus. Success rates vary between 16% and 50%.[13] If unsuccessful, a laparotomy is required, with enterotomy and 2% N-acetylcysteine washout. This acts by breaking the disulphide bonds in the inspissated meconium. In complicated meconium ileus, laparotomy is almost always required and surgical management may include bowel resection and stoma formation depending upon the intraoperative findings.

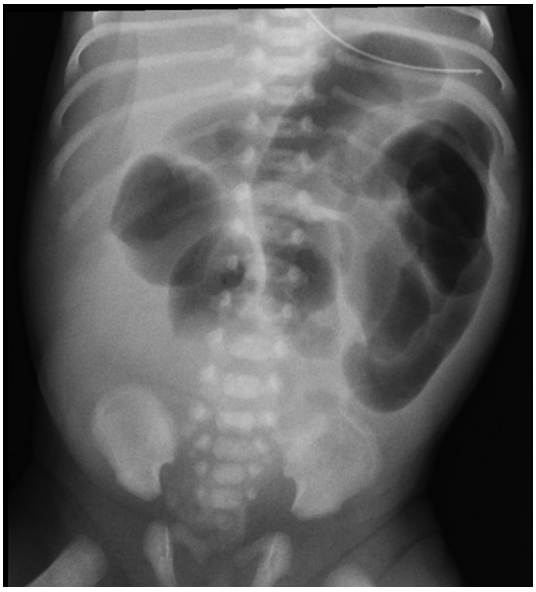

Figure 4 Multiple dilated bowel loops in the upper abdomen on plain radiograph, consistent with a diagnosis of ileal atresia.

Colonic atresia is rare, comprising 2—15% of all intestinal atresias. It is classified according to the same system as other bowel atresias and is likely to share a common aetiology. The estimated incidence varies between 1 in 10,000 and 1 in 66,000. The right colon is more frequently affected than the left, and a type III atresia is the most common.

Antenatal bowel dilatation may be detected in distal bowel atresia in general, but definitive antenatal diagnosis of colonic atresia has only once been described.[14]

The proximal colon is often hugely dilated and thus palpable on examination. Plain abdominal radiograph frequently demonstrates very dilated loops of bowel. Contrast enema may confirm the diagnosis, although clinical findings and features on plain radiograph will often be sufficient to merit laparotomy. Surgical management is either primary excision and anastomosis, or stoma formation and delayed repair. Due to its rare association with Hirschsprung's disease, it is important to exclude distal aganglionosis before primary anastomosis is undertaken.[15]

Conclusions

Bilious vomiting is neonates can be caused by a number of different conditions. However, the diagnosis is malrotation with volvulus until proved otherwise, and this diagnosis must be urgently made and managed, or ruled out. ◆

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