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Introduction

Congenital anomalies of the gastrointestinal (GI) tract commonly present as neonatal bowel obstruction. Clinical and radiological features help to distinguish proximal and distal obstruction. Malrotation/volvulus is one of the causes of bilious (dark green) vomiting in neonates and infants. For this reason, bilious vomiting is a time-critical surgical emergency.

Development

- The embryological gut tube is formed by the end of the fourth week of gestation and is supplied by three main arterial trunks arising from the aorta (coeliac trunk, superior mesenteric artery, inferior mesenteric artery).
- The primitive foregut gives rise to the oesophagus, stomach, duodenum (proximal to the ampulla of Vater), liver, gallbladder, and pancreas.
- The midgut consists of the distal duodenum, jejunum, ileum, caecum, ascending colon, and the proximal two-thirds of the transverse colon.
- The hindgut gives origin to the rest of the colon; the rectum and urogenital sinus separate from an expansion called the cloaca.
- The rotation of the midgut is of particular importance. The primary intestinal loop (ileum) forms a physiological herniation into the umbilicus during the sixth embryonic week, returning to the peritoneal cavity during weeks 8–10. During this process there is a total of a 270-degree counter-clockwise rotation, such that the caecum lies in the right iliac fossa and the duodenojejunal (DJ) flexure is to the left of the midline.
- Malrotation (as well as 'non-rotation') occurs due to a failure in this process.

Abdominal wall defects

Gastroschisis and exomphalos result in the neonatal appearance of exteriorized abdominal contents. Both are diagnosed antenatally in the majority of cases and delivery is planned at a neonatal surgical centre.

In gastroschisis, there is no covering membrane over the bowel, although there may be significant 'peel' on the surface.

Gastroschisis

Gastroschisis is associated with preterm delivery (up to 25%) and has an incidence of 1 in 2000. Gastroschisis can be classified as 'simple' (normal bowel) and 'complex' where there can be associated volvulus or atresia. Other than intestinal atresia (10%), infants with gastroschisis usually have no other associated anomalies. Newborn management involves covering the exteriorized bowel with, e.g. cling film, placement of a nasogastric tube, and intravenous (IV) fluid resuscitation. It is common practice currently to attempt placement of a pre-formed spring-loaded silo, the base of which fits inside the umbilical defect, to cover the bowel. Sequential reduction of the external contents can then be performed over the next few days. Either sutureless umbilical closure or primary surgical closure is usually possible. A small proportion of cases may require surgical silo formation and delayed primary closure.

Long-term outcome is usually good, and survival is >95%.

Nutrition—essentially all infants will require long line placement and parenteral nutrition as return of gut function takes a median of 20 days (BAPS-CASS 2011 data). Feed intolerance is relatively common and specialized formula may be required if breastfeeding is not possible.

Exomphalos

Exomphalos (1 in 8000) results in exteriorized liver and/or bowel and can be considered as 'minor' (defect <5cm), major (defect >5cm), and 'giant'. In giant exomphalos, there is an obvious disproportion in the size of the external organs and the abdominal cavity, usually precluding abdominal closure in the neonatal period.

Neonatal management also involves covering the external contents, but surgery is not required urgently if the covering amniotic sac remains intact. Associated anomalies include cardiac, renal, and chromosomal anomalies. Beckwith–Wiedemann syndrome may result in neonatal hypoglycaemia. Echocardiography and renal tract ultrasound are undertaken. As exomphalos has significant associated anomalies, the survival rate is much lower than for gastroschisis (as low as approximately 10% in some series from the time of antenatal diagnosis). Long-term problems include gastro-oesophageal reflux and respiratory difficulties.

Minor exomphalos can usually be closed primarily in the first few days of life. Major exomphalos may require staged abdominal closure. Giant exomphalos is usually managed conservatively, with dressings to allow epithelialization over the external contents. Delayed closure can be attempted at 9–12 months.

Malrotation

Bilious vomiting in neonates/infants must be managed as a time-critical surgical emergency as the cause may be malrotation/volvulus. Left untreated, irreversible midgut infarction may occur resulting in short-gut syndrome or death. Volvulus of the bowel occurs due to twisting around the narrow mesentery, causing twisting of the entire midgut.

Incidence

1 in 2500.

Pathology

Failure of the normal process of intestinal rotation (see 'Development', p. 2). The end result of failed normal intestinal rotation is that the DJ flexure is found to the right of the midline (and below the trans-pyloric plane), with the caecum high in the midline. Ladd's bands may cross the duodenum from the caecum but are usually non-obstructive.

A narrow midgut mesentery results, which is prone to volvulus.

Associations

Trisomy 21, anorectal malformations, and cardiac anomalies.

Heterotaxy

This is abnormal positioning of thoracoabdominal organs on the left–right axis. There is debate as to whether elective correction of intestinal rotation anomalies is merited, in particular when atrial isomerism is present. The risk of elective surgery in infants with a major cardiac anomaly should be balanced with the risk of midgut volvulus.

Presentation

Bilious vomiting is the key feature. Abdominal distension may be present, with systemic upset.

Investigation

Plain abdominal X-ray—classically shows dilated stomach and duodenum, with an otherwise gasless pattern. Immediate laparotomy is indicated in this scenario if there is any associated peritonism and/or cardiovascular instability.

Upper GI series—demonstrates DJ flexure to the right of the midline, together with a 'bird-beak' narrowing at the distal duodenum, and/or corkscrew appearance of small bowel volvulus.

Ultrasound—may demonstrate inversion of the usual superior mesenteric artery/vein relationship (SMA/SMV). The SMV lies on the right in the normal anatomical situation. A corkscrew may be observed if volvulus is present. Ultrasound is not diagnostic in itself and should not be used to exclude malrotation/volvulus.

Management

Laparotomy and Ladd's procedure—bowel which has undergone volvulus is de-rotated (counter-clockwise). The duodenum is straightened, the caecum is placed in the left upper quadrant, and the small bowel on the right.

As the appendix will lie in the right upper quadrant, appendicectomy is usually removed (inversion or open excision).

If the viability of the bowel is questionable, it is de-rotated and observed to see if normal colour is restored.

In the situation of complete midgut necrosis, the options are resection (committing the infant to short-gut syndrome) or a re-look laparotomy 48 hours later.

Laparoscopic Ladd's procedure is undertaken at some centres although there is general agreement that this is not appropriate for neonates with suspected established volvulus. Laparoscopy may be useful for assessing cases where malrotation is radiologically equivocal.

Complications

- Adhesion obstruction (6%).
- Recurrent volvulus (1%).
- Short-gut syndrome.

Duodenal atresia

The incidence of duodenal atresia is 1 in 10,000, and accounts for 60% of intestinal atresias.

Presentation

- Detected at antenatal sonography in approximately 50% of cases—'double bubble' and polyhydramnios.
- Postnatally, vomiting occurs in the first 48 hours and is bilious if the obstruction is distal to the ampulla of Vater (two-thirds of cases), or non-bilious if the obstruction is proximal.

Differential diagnosis

Malrotation/volvulus—this should be suspected if there is distal gas beyond the dilated stomach and duodenum on X-ray.

Associations

Trisomy 21 (30%) and structural cardiac abnormalities (25%).

Management

Nasogastric decompression and IV fluid resuscitation. Surgery is usually not an emergency.

Surgery

Duodeno-duodenostomy (laparoscopic or open). The proximal and distal duodenal pouches are opened and joined—bypassing the atretic segment. Placement of a trans-anastomotic (nasojejunal) tube allows early enteral feeding, and avoids the need for long line/parenteral nutrition.

Long-term outcome

Usually very good in the absence of other co-morbidity. Rarely tapering duodenoplasty is required if severe proximal dilatation occurs.

Small bowel (jejuno-ileal) atresia

Four types (I–IV) are described, type IV is multiple atresias; types IIIb and IV are associated with significant loss of bowel length.

Associations

Cystic fibrosis—producing antenatal segmental volvulus (10%), prematurity, and gastroschisis.

Presentation

Antenatal (dilated bowel loops, polyhydramnios) in one-third.

Postnatally, abdominal distension, bilious vomiting, and failure to pass meconium occur.

Management

Plain X-ray is usually diagnostic—multiple dilated bowel loops with no distal gas. Nasogastric decompression and IV fluid resuscitation are commenced.

Laparotomy and primary resection/anastomosis is usually possible. Occasionally stoma formation is necessary (e.g. if multiple atresias). Postoperative parenteral nutrition is often required while gut function returns.

Long-term outcome

Usually good if bowel length is sufficient to allow enteral autonomy.