Neonatal abdominal conditions: a review of current practice and emerging trends

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Abstract

In the UK, NHS maternity statistics show that the number of preterm deliveries is increasing. Premature neonates have an increased incidence of congenital anomalies and these are an independent risk factor for a premature birth. Therefore, the population of neonates with gestational prematurity and congenital anomalies requiring the attention of a surgeon is rising.

Many conditions affecting neonates (e.g. gastrochisis) can now be diagnosed antenatally with ultrasound. Defects of the abdominal wall are obvious at birth. Babies with intestinal obstruction will present with the four cardinal signs: vomiting, abdominal distension, pain and constipation. Abdominal X-Ray and contrast studies are useful studies in identifying the cause and level of obstruction.

In the UK, neonatal surgery is carried out by specialist paediatric surgeons in a few tertiary referral centres. The timing of transfer is a key consideration in managing neonates with surgical conditions.

This article reviews current practice and emerging trends in the management of several neonatal abdominal conditions that continue to provide a challenge to paediatric surgeons.

Keywords exomphalos; gastrochisis; Hirschprung’s disease; intestinal atresia; malrotation; meconium ileus/obstruction; necrotising enterocolitis; newborn; neonatal hernia; neonate/newborn; pyloric stenosis; short gut syndrome; surgery; volvulus

Introduction

Neonatal deaths account for more than half of deaths in children aged less than 1 year. Premature neonates have an increased incidence of congenital anomalies and major congenital anomalies are an independent risk factor for pre-term birth. In the UK, NHS maternity statistics show that the number of preterm deliveries is increasing — in 2007–08, ~ 7.5% of live singleton births were pre-term. The population of neonates with gestational prematurity and congenital anomalies requiring the attention of a surgeon is, therefore, rising. This article reviews current practice and emerging trends in the management of several neonatal abdominal conditions that continue to provide a challenge to paediatric surgeons worldwide.

Index cases such as Hirschprung’s disease and anorectal malformations have an incidence of around 1 per 5000 live births. A specialist paediatric surgical centre will, therefore, admit 6–12 babies with each condition per year, but a large District General Hospital may expect to see only one.

The majority of neonatal surgical conditions present shortly after birth or may be diagnosed antenatally by ultrasound scanning (gastrochisis, exomphalos, meconium ileus etc). This permits antenatal counselling and planning for the birth. For many such cases, a decision will be made to deliver the baby where there is a specialist paediatric surgeon, although the evidence that this would reduce morbidity is unclear for the majority of conditions.

Defects of the abdominal wall are obvious at birth. Babies with intestinal obstruction will present with the four cardinal signs: vomiting, abdominal distension, pain and constipation. Commonly, a baby may take initial feeds well, but then refuses feeds and starts to vomit — milk initially, becoming green (bile stained). (Green vomit in a baby must be investigated as a matter of urgency lest malrotation is missed with subsequent loss of small bowel.) Abdominal distension and constipation follow, depending on the level of the bowel obstruction. Tenderness of the abdomen is unusual, but may signal ischaemic bowel and the need for emergency laparotomy.

After careful history taking and examination, the most useful investigation for such babies is a plain abdominal X-ray. (Figure 1) Many of the index conditions have characteristic radiology appearances. When one congenital anomaly has been identified, investigations for associated anomalies should be performed. For example, in the VACTERL association, a number of serious surgical conditions may co-exist (e.g. vertebral, anal, rectal, cardiac, tracheo-esophageal, renal and limb) and babies with Duodenal Atresia may have Down syndrome with associated cardiac conditions.

Following urgent resuscitation including nasogastric drainage, the child should be referred to the regional specialist paediatric surgical centre — only specialist surgeons now manage neonates with surgical conditions in the United Kingdom.

Abdominal conditions of the neonatal period (Table 1)

Gastrochisis

Gastro (stomach) schisis (rent) — is a condition where a defect in the abdominal wall (almost always to the right) allows the bowel to eviscerate antenatally (Figure 2). Most cases are now diagnosed antenatally on ultrasound and provision can be made for appropriate delivery in a centre with specialist neonatal surgery. Repair of the defect is straightforward in 90% of cases, but damage to the bowel by the amniotic fluid usually results in prolonged ileus requiring intravenous feeds for 3–6 weeks or longer.

The incidence of gastrochisis is increasing — from 2.5 per 10,000 total births in 1994 to 4.4 per 10,000 in 2004 in the U.K. (Figure 3). The aetiology and pathogenesis remain unclear. Associations with maternal age (less than 20 years), low maternal BMI, recreational and non-recreational drugs and

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genitourinary infections have been posited. A major UK wide study is presently underway.

The rise in reported incidence cannot be explained by rising genetic frequency alone, particularly considering the low recurrence rate within families. This suggests that environmental factors play a role. The existence of “gastroschisis clusters”, for example that associated with the Nant-y-Gwyddon landfill site in Newport, Wales further support this impression.

Although induction of labour between 35–37 weeks gestation has been advocated, a recent review of evidence casts doubt on any advantage (time to oral feed, incidence of bowel obstruction) associated with this practice.

At birth, the surgeon must decide whether to opt for primary or staged closure. Primary closure may be surgical — with replacement of the bowel and closure of the small defect, or following several days retaining the bowel within a pre-formed silo. In studies where neonates have been randomised to silo versus primary closure, outcomes are similar. It is our experience that small defects, with bowel with little amniotic damage are more amenable to primary closure. Use of the preformed silo may allow immature lungs to improve and bowel oedema to reduce, but there have been reported problems with bowel ischaemia and poorer umbilical cosmesis.

### The incidence of abdominal conditions of the neonatal period

<table>
<thead>
<tr>
<th>Condition</th>
<th>Incidence (live births)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdominal wall</td>
<td></td>
</tr>
<tr>
<td>Gastrochisim</td>
<td>1 in 2–4000</td>
</tr>
<tr>
<td>Exomphalos</td>
<td>1 in 4000</td>
</tr>
<tr>
<td>Inguinal hernia</td>
<td>2 in 1000</td>
</tr>
<tr>
<td>Foregut</td>
<td></td>
</tr>
<tr>
<td>Pyloric Stenosis</td>
<td>2 in 1000</td>
</tr>
<tr>
<td>Duodenal Atresia/Stenosis</td>
<td>1 in 10,000</td>
</tr>
<tr>
<td>Midgut</td>
<td></td>
</tr>
<tr>
<td>Malrotation and volvulus</td>
<td>2 in 1000</td>
</tr>
<tr>
<td>Necrotising Enterocolitis</td>
<td>1 in 3000</td>
</tr>
<tr>
<td>Meconium Obstruction</td>
<td>Rare</td>
</tr>
<tr>
<td>Intestinal duplication</td>
<td>10 in 1000</td>
</tr>
<tr>
<td>Hindgut</td>
<td></td>
</tr>
<tr>
<td>Intestinal Atresia</td>
<td>1 in 10,000</td>
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<tr>
<td>Duodenal atresia</td>
<td>3 in 10,000</td>
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<tr>
<td>• Jejuno-ileal</td>
<td>1 in 66,000</td>
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<tr>
<td>• Colonic</td>
<td></td>
</tr>
<tr>
<td>Hirschsprung’s disease</td>
<td>1 in 5000</td>
</tr>
<tr>
<td>Anorectal Malformations</td>
<td>1 in 5000</td>
</tr>
</tbody>
</table>

Table 1

![Figure 1](image1.png) **Figure 1** “Surgical X-Ray”. There is sparse proximal gas and a “ground glass” appearance. This is very suggestive of a mid bowel obstruction — meconium ileus in this case.

![Figure 2](image2.png) **Figure 2** Gastrochisis.

![Figure 3](image3.png) **Figure 3** Number of reported cases of gastrochisis between 1994 and 2004. Donaldson L. Gastrochisis: a growing concern. London: Department of Health, 2004. This diagram arises from the Donaldson DOH report (www.dh.gov.uk/assetRoot/04/11/57/82/04115782.pdf).
In around 10% of cases, primary closure is only possible with an unacceptable rise in ventilation pressures. For such babies, the bowel is initially placed in a silo and manually reduced over 7–10 days, followed by surgical closure. When surgical closure has been achieved, a low threshold for repeat laparotomy reduces the risk of bowel necrosis. Physical signs such as poor urine output post operatively should not be ignored.

An increased mortality has been reported for premature babies, and when there is a pre-existing bowel atresia or perforation. Rare cases of “closed” gastroschisis occur where the defect has closed over antenatally resulting in loss of the majority of small bowel and extreme short gut.

Post-operative morbidity is due to prolonged ileus, TPN-associated cholangitis, end stage liver failure, gastro-intestinal failure or necrotising enterocolitis. The overall mortality rate remains around 10%.

**Exomphalos**

Early reports suggested that gastroschisis was a result of rupture of an exomphalos; however the differing nature of the two defects and their incidence refutes this. Exomphalos (omphalocele) is a defect of the abdominal wall where somatic mesoderm has failed to migrate between the exoderm and endoderm, leaving a wide defect centred on the umbilicus. Usually the bowel and liver is covered with a membrane, although this may rupture before, during or after birth.

This condition affects 1 in 4000 live births. Compared to gastroschisis, neonates with exomphalos have a higher incidence of associated chromosomal, syndromic and structural anomalies. The minor variant (defect less than 5 cm, not containing liver) is, counter-intuitively, associated with more chromosomal and syndromic abnormalities. While exomphalos minor carries the better outcomes, the major variant has higher mortality and morbidity associated with pulmonary complications. Isolated exomphalos seems to carry the best prognosis.

Antenatal detection has changed the landscape of this condition by providing the possibility of karyotyping, surveillance and termination of pregnancy. For example, in a retrospective review of 445 cases, an abnormal karyotype was identified in 56% (n = 250) with subsequent termination of pregnancy in 99% of this group (n = 248).

For exomphalos minor, primary closure is the mainstay of treatment and is relatively straightforward. For exomphalos major, achieving adequate skin and muscle cover may prove more of a challenge. Conservative management — application of sulfadiazine or jelonet gauze — allows epithelialisation of the defect. The resulting large “incisional” hernia can be closed when the child is older.

Alternatively, primary closure may be attempted utilising biological implants e.g. Permacol®, or a silo followed by reduction over a number of days.

**Inguinal hernia**

Inguinal hernias are a common condition affecting 1–4% of infants and children and up to 30% of premature infants. Herniotomy forms a significant bulk of the workload of a neonatal surgeon. The risk of incarceration within a month of diagnosis is approximated at 10%.

Minimal access surgery in the paediatric population is still undergoing evolution. We expect the progression to laparoscopic herniotomy in the neonate as laparoscopy continues to be more widely adopted by paediatric surgeons. There are indeed published series of successful herniotomy in infants. There are several descriptive series describing outcomes with laparoscopic herniotomy (LH) versus open herniotomy (OH) in older children (mean age 3.7 ± 2.3 years). On the whole, LH delivers similar outcomes to OH. However, others have found that whilst LH delivers similar outcomes, it demands more theatre time and recovery time. LH has also been associated with more post-operative pain and a slightly higher recurrence rate. The laparoscopic approach has also resulted in a new controversy i.e. the utility of contra-lateral patent processus vaginalis ligation in the absence of frank hernia. A systematic review of this very question found that the number needed to treat to prevent one metachronous hernia in both boys and girls with either left or right hernia was 14.

**Pyloric stenosis**

Pyloric stenosis occurs 2 in every 1000 children usually between the 3rd to 6th weeks of life — although it has been reported in preterm neonates and children over 1 year of age. The diagnosis is suspected in a hungry neonate who has increasingly projectile and non-bilious vomits after feeds. Although male preponderance and increased incidence in rural communities is well recognised, a theory of aetiology explaining these phenomena is yet to emerge.

Ramsted’s pyloromyotomy has been the mainstay of surgical management since its description in 1912. The thickened circular muscle is divided down to, but excluding, the mucosa. Evidence from retrospective reviews and single centre prospective cohort studies has been equivocal regarding the merits of a laparoscopic versus a circumumbilical surgical approach. While some single-centre randomised control trials have demonstrated a benefit in decreased analgesic requirement and decreased post-operative nausea and vomiting, others highlight the increased risk of incomplete pyloromyotomy associated with the laparoscopic procedure.

**Malrotation and midgut volvulus**

The small bowel mesentery normally arises along a relatively broad diagonal base from the duodeno-jejunal flexure to the ileocaecal valve. When the “C” loop of the duodenum fails to cross right to left across the midline, the whole of the small bowel originates from a narrow pedicle. This non-rotation of the bowel is prone to volvulus at any age from prenatally to late adulthood. The catastrophe of midgut volvulus relates to the amount of bowel that becomes ischaemic — up to the whole midgut (Figure 4). The highest risk of mortality is in neonates with gut necrosis at laparotomy. For the neonates who survive resection of an ischaemic midgut, the sequelae of short bowel syndrome continue to threaten survival.

The prevalence of malrotation is not known. However, an incidence of 1 in 500 live births is commonly quoted. Approximately 40–60% of cases of midgut volvulus secondary to malrotation in infants and children occur within the first month of life. Acute or intermittent and bilious vomiting in a previously health neonate is the cardinal sign.

Historically, malrotation was diagnosed using contrast enema to assess caecal position. However, this practice has been wholly
superseded by upper gastrointestinal (UGI) contrast due to the mobility and variability of the caecal position in children. UGI contrast is used to the position of the duodeno-jejunal flexure and has a reported sensitivity of 96%.

In addition, in non-rotated bowel, the superior mesenteric artery and vein are abnormally oriented. This can usually be detected on duplex ultrasound (US). US diagnosis is attractive where the neonate is too ill to transfer for UGI contrast. However, while negative predictive value was high (96.3%), the positive predictive value was rather low (42.1%), rendering it useful tool only when the diagnosis unlikely.

Since its description in 1936, Ladd’s procedure remains the operative intervention of choice for malrotation with mid-gut volvulus. The Ladd’s bands, which obstruct the duodenum, are divided and the mesentery widened. The small bowel is replaced in the right of the abdomen and the large bowel to the left. Inversion appendicectomy is usually also undertaken.

The laparoscopic Ladd’s procedure has also been described and applied in infants as young as 5 days old with malrotation but no volvulus. Laparoscopic correction of volvulus has also been described. During an open Ladd’s procedure, is thought that the handling of the bowel leads to formation of adhesions which, in turn, prevents recurrence of volvulus. It is yet to be seen if the laparoscopic Ladd’s procedure will be associated with a higher incidence of recurrence of volvulus.

**Necrotising enterocolitis**

This condition affects up to 10% of pre-term, very low-birth weight neonates. Increased preterm survival has increased the population at risk of NEC.

The pathophysiology of NEC remains a matter of debate. Recent advances in our knowledge include the role of the neonatal gastrointestinal vasculature and immature immune defences. Exposure of immature gut to gastrointestinal flora is key, hence the care taken in establishing the optimal feeding regime. Evidence-based preventative strategies include antenatal glucocorticoid, delayed feeding, standardised feeding regimens and the use of probiotics. A recently published randomised control trial has shed some doubt on the value of donor milk as opposed to pre-term formula as a substitute for maternal milk in extremely premature neonates (less than 30 weeks). While the lowest incidence of NEC and gram-negative bacteraemia was found in the group receiving maternal milk, the group receiving donor milk had slower weight gain despite greater supplement and milk intake, when compared to the group receiving pre-term formula. There was no difference in the incidence of NEC, other infections, mortality or length of hospital stay between the donor milk and preterm formula groups.

Indications for surgery, as always, are “failure of maximal medical therapy” and/or “complications of the disease”. Surgery may be required for acute bowel perforation, inflammatory mass (phlegmon) or development of a stricture. The timing of surgical management and the optimal operation remain a matter of great debate. The surgeon is rightly more reliant on experience and acumen than laboratory indicators e.g. platelets, CRP, metabolic acidosis. Laparotomy and resection of necrotic bowel is the basic standard for isolated NEC. Where NEC is extensive, patch or primary closure (“patch, drain and wait”) with a second look in 24–48 hours may enable bowel of initial dubious vascularity to be preserved, thus preventing “short gut syndrome”.

In unstable or very low birth weight neonates (less than 1500 g), the debate concerning laparotomy versus primary peritoneal drainage continues. A recent randomised control trial comparing laparotomy and bowel resection with primary peritoneal drainage in neonates less than 34 weeks gestation and weighing less than 1500 grams showed no difference on survival to 90 post-operative days.

Half the neonates who survive primary peritoneal drainage will then go on to have laparotomy, whilst half will require no further treatment — the overall survival being 67%. NEC carries a significant surgical morbidity and risk of mortality. For those neonates who survive surgery, sequelae include failure to thrive, short bowel syndrome neuro-developmental impairment.

**Meconium obstruction**

Meconium, the dark first stools of the neonate, is usually passed within the first 24 hours in term neonates. Meconium obstruction presents as a triad of delayed passage of meconium, abdominal distension and emesis. There are several distinct syndromes that present with meconium obstruction. In the absence of intestinal atresia, delayed clearance of meconium is due to abnormal intestinal motility or abnormally thickened meconium. Abnormal motility leading to meconium obstruction is seen in very low birth weight, and often premature infants. The obstruction is typically an ileal obstruction.

Delayed passage of meconium is also seen in anomalies of gut innervations e.g. Hirschsprung’s disease. In a recent large series (n = 77) of neonates diagnosed with ‘meconium plug syndrome’ on contrast enema, Hirschsprung’s disease was identified in 13% of the study population.

Neonates with cystic fibrosis can present with obstructive symptoms – meconium ileus.

To avoid unnecessary operative intervention, it is important to differentiate between these syndromes.
Meconium plug syndrome

| Meconium obstruction in low birth weight neonates | Cystic fibrosis associated meconium ileus (CFMI) |

| Gestation Overall condition | Term Systemically well | Preterm Systemically well | Term Respiratory symptoms Failure to thrive |

| Level of obstruction | Colonic Ileal and colonic Uniformly dilated loops without air fluid levels |

| Plain XR | Uniformly dilated loops of bowel, paucity of rectal gas |
| Contrast enema | Meconium plugs |
| Treatment | Resolves with rectal stimulation +/- enema |
| Meconium findings | Pellet-like or snake-like plugs followed by normal meconium |

A neonate with meconium ileus will have a gradual onset of abdominal distension. The abdomen remains soft and nontender, distinguishing meconium obstruction from necrotising enterocolitis. Bilious vomiting is a late sign. An abdominal radiograph will demonstrate uniformly dilated loops of bowel. Others advocate the use of a decubitus plain radiograph, suggesting that the absence of air fluid levels excludes colonic atresia and obviates the need for a fluoroscopic-guided contrast enema. A contrast enema is diagnostic and may be therapeutic. Our agent of choice is Gastrografin. Others have reported success with enema of the mucolytic N-acetyl-L-cysteine. Indeed, antegrade orogastric N-acetyl-L-cysteine is also reported as a successful treatment modality. We perform contrast enemas under fluoroscopic control. There are reports, however, of contrast enemas without fluoroscopic guidance. Three cases of delayed colonic perforation are also reported but these authors report it as unrelated to the enema.

Contrast enema acts as an osmotic laxative and leads to defecation of meconium plugs and relief of obstruction in most cases. Neonates with CFMI may require regular washouts until their hyper-viscous meconium has been cleared. Rarely, meconium obstruction may be complicated by necrotising enterocolitis, intussusception, fulminant obstruction and bowel perforation.

Imminent perforation is an indication for laparotomy. An enterostomy is formed in a dilated segment with a mucous fistula at the level of distal collapse. It is our practice to perform on-table expression of distal meconium as well as post-operative saline washouts per mucous fistula.

In our experience, enteral feeding can be restarted as soon as obstruction is resolved. Decreasing abdominal distension, non-bilious gavage and a regular stooling habit are all positive signs.

A large proportion (13%–43%) of patients with meconium obstruction are ultimately diagnosed with cystic fibrosis. Therefore, definitive diagnosis of cystic fibrosis should be sought in all neonates presenting with meconium obstruction.

Intestinal atresia

Intestinal atresia is a rare condition whose aetiology remains largely unknown. Tandler suggested that failure of the gastrointestinal tract to recanalise completely is following epithelial proliferation is a cause of intestinal atresia. There is also evidence to support the theory of Louw and Barnard's: that interruption of intestinal vasculature during embryonal development results in an atretic segment. Increasingly, there is evidence to suggest that duodenal, jejuno-ileal and colonic atresia should be considered as three separate entities. Consider, for example, that these conditions vary in frequency. Duodenal atresia is extremely rare, occurring in 1.3 per 10,000 births. Jejuno-ileal atresia occurs in 2.9 per 10,000 births (Figure 5). Colonic atresia is even rarer, occurring in 1 per 66,000 live births. Duodenal atresia is associated with associated anomalies, particularly cardiac (38%) and renal (14%). However, most distal atresias will be isolated.

![Figure 5](jejunal atresia. Proximal bowel is grossly dilated and adynamic. Distal bowel has multiple atresias and a precipitous blood supply. The risk of volvulus and short gut is clear.)
There is also a difference in genetic associations. Familial duodenal atresia appears to be inherited in an autosomal recessive manner. Patients with Down syndrome have a higher incidence of duodenal atresia/stenosis. There is no chromosomal syndrome association associated with jejuno-ileal or colonic atresia. Case reports are accumulating of an autosomal recessive apple peel atresia most commonly observed at the duodeno-jejunal junction. Familial colonic atresia, with a possible X-linked pattern has also been described.

Increasingly, atresia is being diagnosed antenatally on routine fetal monitoring ultrasound. However, reported sensitivity rates are poor (51%). Antenatally diagnosed duodenal atresia may suggest the need to exclude Trisomy 21.

Post-natally, symptoms and signs are dependent on the level of obstruction, with more distal obstruction corresponding to more abdominal distension. The diagnosis is confirmed radiologically with gaseous distension and air fluid levels to the atresia and a distal paucity of gas. Distal contrast study is useful in colonic atresia but is of limited benefit in jejuno-ileal atresia.

In cases of isolated atresia, primary anastomosis is often possible and the post-operative course is benign. Overall survival with this condition has improved with time (Table 2). It is easy to observe that the more distal the atresia, the better the survival outcomes. This is likely related to the higher incidence of associated anomalies seen particularly with duodenal. Or perhaps it reflects a temporal span of cranio-caudal development of the gastro-intestinal tract i.e. the more proximal the atresia, the earlier the gestational insult, the greater the impact on this insult on organogenesis.

Post-operative surgical challenges are the management of neonates with short bowel syndrome. Time to enteral feeding, TPN dependence and growth are key outcome measures in this group.

Intestinal duplication

Intestinal duplication is a rare anomaly. Most duplications are found in the midgut. With advances in antenatal diagnostics, however, we predict that the observed incidence is set to increase. Therefore, the neonatal surgeon will inherit the dilemma of what to do with a potentially asymptomatic condition diagnosed at birth. Evidence is limited to case series and reviews. When symptomatic, patients most often presents with nausea and vomiting and hence operative intervention can be justified. Where duplication has been incidentally detected on laparotomy or laparoscopy, the rationale for operative management is to prevent the risk of rarely reported obstruction, volvulus, intussusception, malignant transformation and the risk of bleeding from heterotopic gastric mucosa.

Hirschsprung’s disease

In the neonatal period, Hirschsprung’s disease (HD) presents with delayed passage of meconium (more than 24 hours), progressing to symptoms and signs of bowel obstruction (Figure 6).

The cause of HD is thought to be failure of cranio-caudal migration of neural crest cells into the mesenchymal myenteric and sub-mucosal plexus during the 4th to 12th of gestation. The cranial crest cells are of vagal origin. Ganglion cells also arise from migration of caudal sacral plexus neurons in a cranial direction. Most commonly, a short segment from the recto-sigmoid junction and following on distally is affected.

For the neonatal surgeon, the challenge of this condition is timely diagnosis and early decompression. Lорjin et al prospectively assessed various anorectal manometry, contrast enema and rectal suction biopsy as initial tests for HD. They confirmed

<table>
<thead>
<tr>
<th>Location of atresia</th>
<th>Source, y</th>
<th>Survival (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duodenal</td>
<td>deLorimer et al,6 1969</td>
<td>60</td>
</tr>
<tr>
<td></td>
<td>Nixon and Tawes,7 1971</td>
<td>60</td>
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<td></td>
<td>Stauffer and Irving,64 1977</td>
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<td></td>
<td>Kullendorf,65 1983</td>
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<td></td>
<td>Grosfeld and Rescorla,20 1993</td>
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<td></td>
<td>Mooney et al,29 1987</td>
<td>95</td>
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<td></td>
<td>Current study</td>
<td>86</td>
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<td>Jejunoileal</td>
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<td>Martin and Zerella,16 1976</td>
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<td>Cywes et al,46 1980</td>
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<td>Colon</td>
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<td></td>
<td>DeFore et al,41 1976</td>
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<td>91</td>
</tr>
<tr>
<td></td>
<td>Current study</td>
<td>100</td>
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</tbody>
</table>


Table 2
rectal suction biopsy (first described in 1969) was the most sensitive (93%) and specific (100%) and therefore, the most appropriate screening tool for HD.

Surgical correction of HD involves a single stage or multi-stage procedure. Procedures described by Swenson, Duhamel and Soave vary in the treatment of the rectal cuff. However, the principles of early proximal diversion, resection of aganglionic segments of bowel followed by primary anastomosis of proximal ganglionic bowel to the rectum remain the same. Regardless of operative method, constipation and faecal incontinence remain a problem for 30–50%. This reinforces the impression that residual ganglionic bowel may be innervated, but not normally so. Future directions for HD may include cell replacement therapy. Already, transplant of both human and murine enteric nervous system stem cells into human, murine and chick aganglionic bowel has been described. The identification of serum markers for HD may lead to a "protein chip" diagnostic tool that may result in HD being identified earlier and with greater sensitivity.

Anorectal malformations

Anorectal malformations occur in about 1 in 5000 live births. There have been a number of attempts to classify the various degrees of anomaly. The movement from the W ringspread classification to the Krickenberg classification reflects the need to standardise descriptions into a system allowing comparable follow-up that also includes rare anomalies e.g. pouch colon. Authors find greatest utility in the classification offered by Levitt and Pena.

Antenatal sonographic diagnosis will undoubtedly be superseded by the more accurate foetal magnetic resonance imaging. However, at present most anorectal malformations are discovered post-natally. Once a diagnosis has been reached, the principles of post-natal management remain the same. For low malformations, early anoplasty provides good results. Early division and with delayed anorectoplasty and secondary anastomosis is the standard for high and intermediate malformations.

The posterior sagittal anorecto-plasty (PSARP) has achieved prominence over the past 25 years. This procedure offers better continence than the preceding abdomino-perineal pull-through approach without Pena stimulation. A more anatomical repair is achieved by pulling the rectum through the levator- sphincter complex. Laparoscopic assisted anorectal pull-through (LAARP) is gaining popularity in repair of high and intermediate malformations. Proponents report a good visualisation of perineal muscles, allowing dissection of thelevator complex. The LAARP can be applied as part of a single-stage procedure in the neonatal period. Similar to the PSARP, electro-stimulation is used to identify the median point through which the dissected rectal fistula will be passed. Although constipation and soiling remain a major problem in the mid- to long-term, they do seem to improve with age.

Neonatal surgical principles

Laparoscopy has yet to prove an advantage for most neonatal conditions. At open laparotomy, a primary anastomosis is performed where safe and feasible. In most cases, however, a stoma will be formed. If the stoma is a distal ileostomy or colostomy, the baby should thrive and definitive surgery/closure can wait until the infant has gained weight and conditions such as Hirschsprung’s disease or distal bowel obstruction have been excluded (by suction biopsy or distal contrast radiology respectively).

Proximal jejunostomies are more challenging and may result in failure to thrive, sodium deficit and para-stoma skin excoriation. For such cases, recycling stomal losses distally or early closure may be required.

Short gut syndrome

A baby with short gut syndrome is unable to maintain homeostasis on enteral feeds alone. The length of foetal small bowel doubles in the final trimester (115 to 250 cm), and some babies may adapt to tolerate enteral feeds in time. However, for a small number of surgical babies, the length of bowel remains below that which allows enteral independence (estimated at 40 cm without or 20 cm with a functioning ileo-caecal valve). Such babies present difficult ethical, medical and surgical challenges and all efforts in the care of neonates must centre around prevention of short gut syndrome where possible. Children with this condition may undergo years of invasive and expensive treatment even before a bowel transplant is considered.

Surgical care may include preservation of bowel where possible, surgically inserted central feeding lines, restoration of bowel continuity and operative reduction of atonic extreme dilated bowel.

A number of ingenious procedures have been described to try to reduce the diameter of adynamic bowel loops and at the same time increase the intestinal length. The most commonly employed are Bianchi’s L.I.L.T. and the S.T.E.P. procedure. A number of techniques exist to slow bowel transit time or redirect the intestinal flow, but they frequently lead to bowel obstruction etc.

Small bowel transplant (usually included with a liver transplant) has now entered mainstream therapy, but many children do not survive to transplant and the ongoing post transplant management remains considerable.

Conclusion

Each index neonatal surgical condition has an incidence of around one per 5000 live births. Although a non specialist hospital may see only a few cases per year, every neonatal unit and paediatric centre should be able to identify a baby with a potential surgical problem to allow urgent referral and management in the regional unit.

The majority of surgical conditions are either identified antenatally by ultrasound scan or very shortly after birth. Suggestive symptoms include food refusal, vomiting — becoming bile stained, abdominal distension, constipation and pain. Plain abdominal radiography helps diagnose most conditions. Where malrotation or bowel ischaemia is suspected (tender abdomen) emergency referral should take place to reduce the risk of the catastrophe of short gut syndrome.

There is a familial element to some of these conditions and with successful treatment the incidence is likely to increase. In addition, the increase in premature births will lead to more neonatal surgical challenges.
Conflict of interest statement

None.

FURTHER READING


Practice points

- Improvements in neonatal intensive care has increased the survival of pre-term neonates and, in turn, increased the population of neonates surviving with congenital anomalies.
- Many neonatal abdominal conditions can now be identified antenatally by ultrasound scan, or by radiography soon after birth.
- Early diagnosis and referral to specialist surgical centre is key.
- Morbidity following surgery includes short-gut syndrome, stoma complications, adhesional obstruction, dysmotility, malabsorption and the requirement of long-term parenteral nutrition.