Intestinal Obstruction in Neonatal/Pediatric Surgery

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Intestinal obstruction in the newborn infant and older child may be due to a variety of conditions, including atresia and stenosis, annular pancreas, malrotation, duplication cyst, meconium ileus, meconium plug syndrome and neonatal small left colon syndrome, Hirschsprung's disease, neoplasia, trauma, and other rarer causes. The mode of presentation can be acute or more chronic with systemic upset due to shock. Neonates, more so than older children, with unrecognized intestinal obstruction deteriorate rapidly, show an increase of associated morbidity and mortality and appropriate surgical treatment becomes more hazardous. Early diagnosis depends largely on the prompt detection of obstructive manifestations by the clinician and the subsequent accurate interpretation of radiographic findings and other investigations, leading to definitive treatment, which should always be preceded by appropriate resuscitation/ preparation of the infant/child. Management of intestinal obstruction will almost always be surgical, apart from some notable exceptions and all are discussed in more detail. With the advent of pediatric and neonatal intensive care and multidisciplinary care, the morbidity and mortality of cases of intestinal obstruction reported in current series is generally extremely low and mainly determined by the coexistence of other major congenital anomalies (eg, cardiac), delays in diagnosis and treatment or coexisting medical conditions. Newer treatments and future developments may reduce the residual mortality in such cases as ultra-short-bowel syndrome.
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INTESTINAL OBSTRUCTION IN the newborn infant and older child may be due to a variety of conditions, including atresia and stenosis, annular pancreas, malrotation, duplication cyst, meconium ileus, meconium plug syndrome and neonatal small left colon syndrome, Hirschsprung's disease, neoplasia, trauma, and other rarer causes.[1]

This section will review and revisit the different common and uncommon causes of intestinal obstruction in neonates and children and summarize their treatment and other salient features. Other sections will focus appropriately on other issues, eg, investigations, etc.

PRESENTATION/GENERAL MANAGEMENT OF INTESTINAL OBSTRUCTION

The mode of presentation can be acute, with obvious features of obstruction (vomiting, which will invariably become bile-stained); pain (usually colicky), with or without features of peritonism and perforation; and severe systemic upset due to shock. The presentation can also be more subtle and chronic in cases of incomplete or recurring/resolving bouts of obstruction.

Neonates, more so than older children, with unrecognized intestinal obstruction deteriorate rapidly, show an increase in associated morbidity and mortality, and appropriate surgical treatment becomes more hazardous. Early diagnosis depends largely on the prompt detection of obstructive manifestations by the clinician and the subsequent accurate interpretation of radiographic findings and other investigations, leading to definitive treatment (usually surgical), which should always be preceded by appropriate resuscitation/preparation of the infant/child. Conservative management has a much lower chance of success in acute cases and cannot be advocated generally.

Detailed history, careful clinical examination, and early instigation of general resuscitation measures (correction of hypovolemia, oxygen administration, intravenous antibiotics, gastric decompression) and baseline radiological[1] and laboratory investigations (biochemistry, hematology, blood crosshatch) are considered at least as important as detailed "second line" investigations, which will eventually direct definitive treatment.

Management of intestinal obstruction will almost always be surgical, apart from some notable exceptions (including among others some cases of postoperative adhesion obstruction, meconium ileus, neonatal narcotizing enterocolitis [NNEC], colonic volvulus, neoplasia—notably lymphoma, inflammatory bowel disease), discussed in more detail in subsequent sections of this article.

GASTROINTESTINAL TRACT OBSTRUCTION ANATOMY/PATHOGENESIS/ETIOLOGY/TREATMENT

There are many ways to classify intestinal obstruction, perhaps the most widely used being the classical way, describing the origin of the obstruction: extrinsic, mural, or intraluminal. Another mnemonic, which attempts to classify according to etiology appears in Fig 1.

Obstructions at each anatomical region will be described in more detail from stomach to the anorectum.

Lesions of the Stomach

Pyloric stenosis (Fig 2) is the commonest cause of gastric outlet obstruction. The repeated vomiting leads to

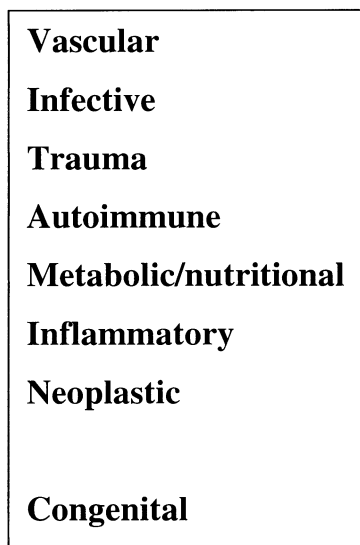| Vascular |
| :--- |
| Infective |
| Trauma |
| Autoimmune |
| Metabolic/nutritional |
| Inflammatory |
| Neoplastic |
| |
| Congenital |

Fig 1.   Mnemonic for classifying causes of intestinal obstruction (or indeed most other conditions).

dehydration and frequently hyponatremic, hypochloremic, hyperkalemic metabolic alkalosis. Rarely, in the latest stages of the disease, paradoxical aciduria ensues in an attempt to conserve potassium at the expense of hydrogen excretion by the kidney. These need to be corrected prior to surgery, which involves the classical Ramstedt's pyloromyotomy with virtually no mortality or recurrence.

Other rarer causes include pyloric web or atresia, infective[2] or syndromic conditions, eg the junctional epidermolysis bullosa-pyloric atresia syndrome, which is recognized as a distinct autosomal recessive entity. Affected infants present with skin fragility and inability to feed due to intestinal obstruction. Despite successful surgical repair of the anatomical defect, the outcome is poor owing to poor feeding, malabsorption, failure to thrive, and sepsis, with death occurring before age 11 months. The poor prognosis of this condition must be considered when decisions are made regarding surgical correction and, although attempting surgical correction may be warranted in individual circumstances, withholding surgical intervention and providing palliative support may be an acceptable alternative.[3]

Intestinal Atresia and Stenosis

Atresia is the most common cause of congenital intestinal obstruction and accounts for approximately one third of cases of obstruction in the newborn with incidence of approximately 1:2700 births. The mortality of the condition was 90% prior to 1950, but this declined sharply and remains at approximately 10% (Table 1).

Duodenal Atresia[4]

Duodenal obstruction is most commonly due to complete atresia of the postampullary segment of the duodenum and presents with bile-stained vomiting and a classic "double-bubble" appearance of the plain abdominal x-ray. It commonly coexists with an annular pancreas due to the associated embryological defect. Treatment is by duodeno-duodenostomy (Figs 3 and 4), either linear or diamond-shaped, and—although classically described, a transanastomotic tube is no longer advocated as it delays recovery and increases the time to full feeding.[5] A duodenal web (which may confuse the diagnosis with that of proximal jejunal atresia due to a "windsock effect") is much rarer and is treated by duodenotomy, excision of the web, and duodenoplasty.

Jejunoileal Atresia

Intestinal atresia (Fig 5) is classified into 5 types: Type 1 denotes an obstructing membrane and Type 2 a cord anomaly at the site of the atresia. Type 3A involves complete separation of the 2 ends by a mesenteric defect and Type 3B is described as the apple-peel deformity. Type 4 describes multiple intestinal atresias. Treatment is, whenever possible, by primary anastomosis with a general principle being the preservation of as much bowel length as possible.

Type 3A or "apple-peel" atresia presents a special case, where the bowel is very abnormal, shortened, and revolves round and entirely supplied by the ileocolic artery (Fig 6). Radiographic evidence of high small-
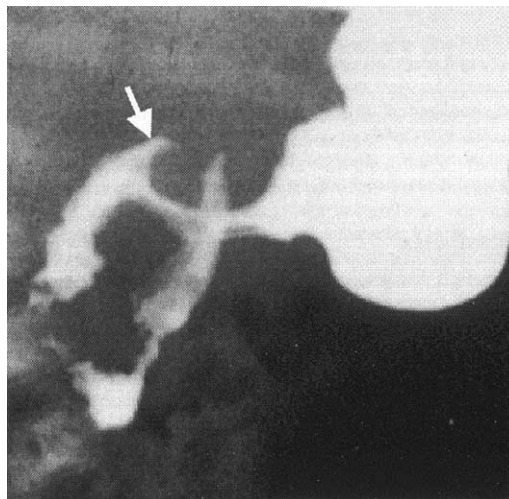

Fig 2.   Contrast meal showing pyloric stenosis. Note the "shouldering" of duodenal mucosa towards the pylorus (arrow), which puts it in peril during surgical incision of the pyloric muscle. Test feed and, in difficult cases, ultrasound scan of the pylorus should have a virtually 100% diagnostic accuracy making contrast examination unnecessary.

Table 1. Review of Cause, Intervention, and Outcome in 227 Neonates with Intestinal Atresia[73]

Level of Obstruction (227 neonates)	Associated Anomalies/Features	Surgical Management	Operative Mortality	Long-term Survival (Morbidity and Mortality Due to)
Duodenal (n = 138)	Prematurity, 46%	Duodeno-duodenostomy, 86%	4%	86% (cardiac anomalies)
	Polyhydramnios, 33%	Duodenotomy and web excision, 7%		
	Down syndrome, 24%	Duodeno-jejunostomy, 5%		
	Annular pancreas, 33%			
	Malrotation, 28%			
Jejunoileal (n = 128)	Intrauterine volvulus, 27%	Resection with (70%) or without (30%)	0.8%	84% (ultrashort-bowel syndrome)
	Gastroschisis, 16%	primary anastomosis 76%		
	Meconium ileus, 12%	Temporary stoma without resection, 20%		
		Web excision, 4%		
		Bianchi procedure, 1 case		
Colonic (n = 21)		Temporary stoma and delayed anastomosis, 86%	0%	100%
		Resection and primary anastomosis, 14%		
Multiple (n = 10)				

bowel obstruction and a malrotated microcolon on preoperative roentgenogram with contrast enema should suggest this diagnosis. It has been suggested that this disorder is transmitted by either an autosomal recessive gene in some cases or that there may be a more complex spectrum of genetic transmission in others.[6]

Fifty-seven cases of apple-peel jejunal atresia have been reported in the English literature. Patients with this anomaly have a high incidence of prematurity (70%), malrotation (54%), short-gut syndrome (74%), multiple atresias (15%), complications (63%), and mortality (54%). Mortality has decreased from 63% to 47% since 1970.[6]
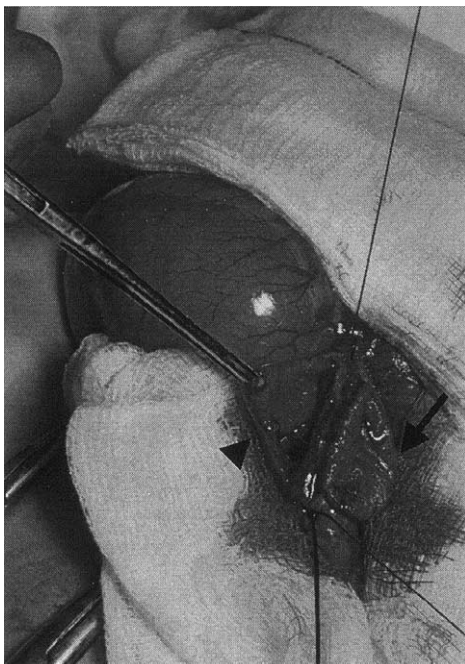

Fig 3. Duodeno-duodenostomy for duodenal atresia (arrowhead: proximal duodenotomy, arrow: distal duodenotomy).

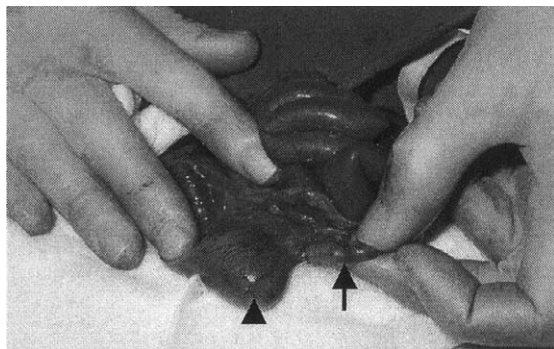Fig 4. Duodeno-duodenostomy suture line completed. No transanastomotic tube is used.[5]

Fig 5.   Ileal atresia. Note the grossly distended blind ending proximal loop (arrowhead) and the collapsed distal end containing meconium (arrow).

Colonic Atresia

Colonic atresia is rare. Treatment is generally by segmental resection with or without primary anastomosis, but because of the association of colonic atresia with Hirschsprung's disease, a careful examination of the resected specimen to rule out Hirschsprung's disease is recommended. Performing a rectal biopsy must be considered for patients who initially were treated for colonic atresia and who have a slow return of normal gut function. All colonic atresias in a reported series were diagnosed neonatally; however, there was mean delay of 15.4 months (range, 1 month to 5 years) in diagnosing associated Hirschsprung's disease.[7]

Colonic Stenosis

Colonic stenosis may be congenital[8] or acquired.[9] Acquired colonic stricture in older patients has been reported in association with cystic fibrosis (cystic fibrosis colonopathy[10]) and may be caused by excessively high
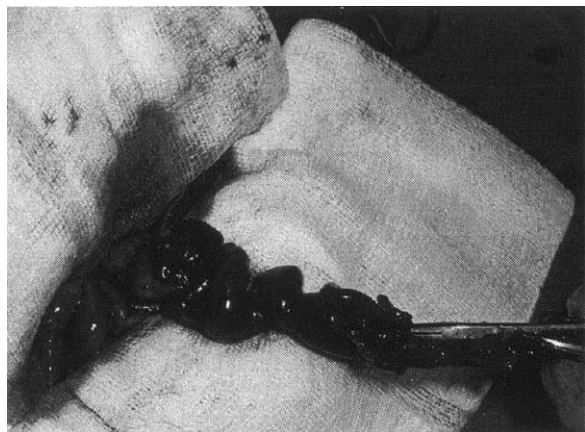

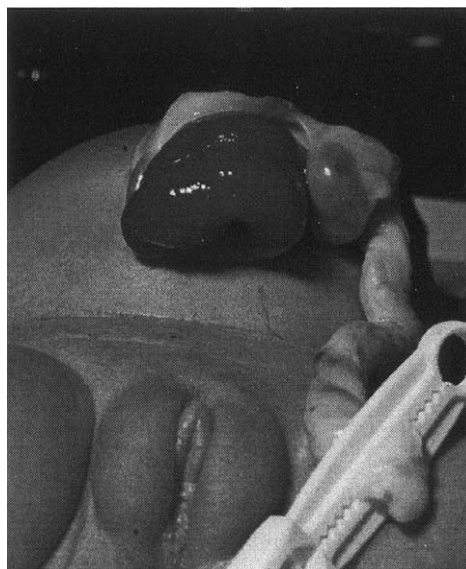**Fig 6.   "Apple peel" atresias and intestinal ischemia. The patient did not survive.**

Fig 7.   Prolapsed obstructing vitellointestinal tract remnant through umbilical defect.

doses of pancreatic supplements. The treatment principles are the same as those for colonic atresia.

Omphalomesenteric Tract Remnants

Omphalomesenteric duct malformations comprise a wide spectrum of anatomic structures and associated symptoms (or no symptoms). They may range from a completely patent omphalomesenteric duct at the umbilicus to a variety of lesser remnants, including the diverticulum of Meckel. Symptoms may involve intestinal fistulas at the umbilicus, intussusception/prolapse of ileum at the umbilicus (Fig 7), intestinal obstruction from a variety of causes, melena and anemia, abdominal pain and inflammation. Symptoms occur most frequently during childhood years (especially in the first 2 years of life).[11]

Duplications

Intestinal duplication may occur at any level of the gastrointestinal tract from mouth, esophagus, stomach,[12] ileum,[13] colon, cecum[14] to rectum.[15] They may contain heterotopic tissue (eg, pancreas[13]) and may rarely be associated with intraspinal communications (residual neuroenteric canal-split notochord syndrome) causing high morbidity and mortality.

Duplications may be fusiform, tubular, or cystic and usually present with obstruction or bleeding. Treatment is by complete excision and primary anastomosis where possible. Where the cyst cannot be safely excised (usually because of the sharing of a common wall with the normal bowel segment, or because complete excision may endanger the bile ducts) every attempt should be
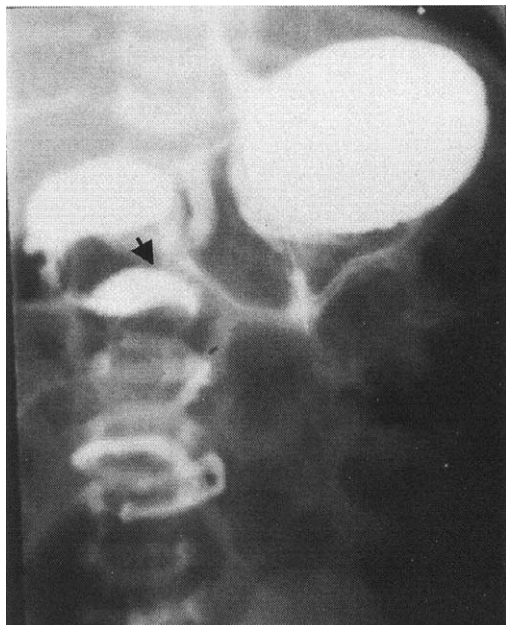
Fig 8. Upper gastrointestinal meal and follow-through of a 4-day-old infant with bile-stained vomiting showing malrotation with volvulus. Note the duodenojejunal (DJ) flexure (arrow) does not cross the midline and lies at a level below the pylorus. Contrast then flows into the proximal jejunum in a "corkscrew" manner.

made to completely excise its mucosal lining as duplication cysts are associated with late malignant transformation.[16-21]

Malrotation

Malrotation of the intestinal tract is the result of a well-defined aberrant sequence of embryological events. Because the consequences of malrotation associated with a midgut volvulus may be so catastrophic, an understanding of the anatomy, diagnostic criteria, and appropriate therapy for this putative emergency illness is imperative. More than half of the patients present during the first month of life, and virtually all have bile-stained vomiting. However, symptoms may first become manifest in teenagers and adults, often with disastrous outcomes.[22] Most investigators, therefore, recommend surgical correction of all cases of malrotation.[23,24]

Upper gastrointestinal contrast series (Fig 8) is the preferred diagnostic study, being both sensitive (18/19, 95%) and accurate (18/21, 86%).[25] A Ladd procedure is the preferred treatment, which typically includes evisceration and inspection of the mesenteric root; derotation of a midgut volvulus (which has always been reported to occur in a clockwise direction); lysis of Ladd's bands with Kocherization of the duodenum along the right abdominal gutter, with or without appendectomy; and placement of the cecum into the left side of the abdomen

(Figs 9 and 10). A high index of suspicion in the neonate with vomiting, rapid diagnosis, and appropriate operative therapy results in a predictable favorable outcome for children with malrotation with or without volvulus.[25]

Volvulus Without Malrotation

A literature review indicates a significant relation of intrauterine midgut volvulus without malrotation to preterm birth with low-birthweight infants, usually presenting in the immediate postnatal period. The speed of presentation and intervention can ensure a favorable outcome.[26]

Volvulus may also occur round an intraabdominal fibrotic or omental band (Fig 11) or present as an isolated entity with axial rotation around a Meckel's diverticulum.[27]

Colonic Volvulus

Colonic volvulus can also occur due to the idiosyncrasies of the peritoneal fixations/or nonfixation of segments of the colon and may involve the cecum,[28] transverse colon,[29,30] splenic flexure,[31] or sigmoid.[32,33] It is usually preceded by recurrent low-grade symptoms and commonly associated with intestinal dysmotility and constipation.[31] Presentation is usually acute, with associated systemic symptoms and shock.

Cecal Volvulus

Resection and primary end-to-end ileocolic anastomosis is advocated in the presence of gangrene or perfora-

Fig 9. Operative findings in patient from Fig 8, showing the complete volvulus around the foregut mesentery carrying the mobile caecum and appendix (arrow) to the left upper quadrant (small bowel at the top of the picture and large bowel at the bottom).

Fig 10. Ladd's procedure completed. After full kocherization of the duodenum, the jejunum and small intestine are placed in the right-hand side of the abdomen and the large intestine on the lower left. The appendix is generally left behind, with clear notes and instructions that it does not lie in the right iliac fossa.

tion of the cecum. In noncomplicated cases of cecal volvulus, detorsion, and cecopexy is also an option because of low mortality, morbidity, and recurrence rates.[28]

Transverse Colon Volvulus

Resection and primary end-to-end anastomosis is the treatment of choice in this condition,[29,30,34] as operative or colonoscopic management by detorsion[34] can be associated with recurrence. Transverse colon appears to be the only part of the bowel reported to volve in a counterclockwise direction.

Splenic Flexure Volvulus

Resection and primary end-to-end anastomosis is also the treatment of choice in this condition.[31]

Sigmoid Volvulus

Sigmoid volvulus, although not uncommon in the adult population,[33] is an exceptionally rare and potentially life-threatening condition in the pediatric age group. In a review of 63 cases of sigmoid volvulus in children since 1940 (median age was 7 years), two distinct presentations (acute and recurrent) were identified. Abdominal symptoms dominated the clinical picture. Barium enemas either confirmed or were highly suggestive of sigmoid volvulus,[32,33] however the classic roentgenographic omega sign of volvulus was present on plain films in only 29% of the cases.[33] Reduction by barium enema was successful in 77% (10 of 13) of the attempts. Forty-nine patients underwent operative treatment, with sigmoidectomy (with or without primary anastomosis) being the most common. The overall mortality rate was 6%, operative mortality was 8.1%, and neonatal mortality was 14%. It was associated with Hirschsprung's disease in 17% of cases (11 of 63 patients).[32]

Sigmoid resection is the definitive treatment for children as well as adults, but nonoperative decompression to allow for elective resection could be attempted in patients who have no evidence of peritonitis.[33]

Meconium Disease of Infancy

Meconium ileus. Intestinal and pancreatic dysfunction, which in most cases results from the autosomal recessive disease cystic fibrosis, can result in intestinal obstruction due to meconium ileus (Fig 12), which is the result of the accumulation of sticky and inspissated intraluminal meconium. Both nonoperative and operative therapies can be effective in relieving this small-bowel obstruction. In the past, although less so today, a successful nonoperative treatment was associated with a more favorable outcome. Once the meconium ileus is
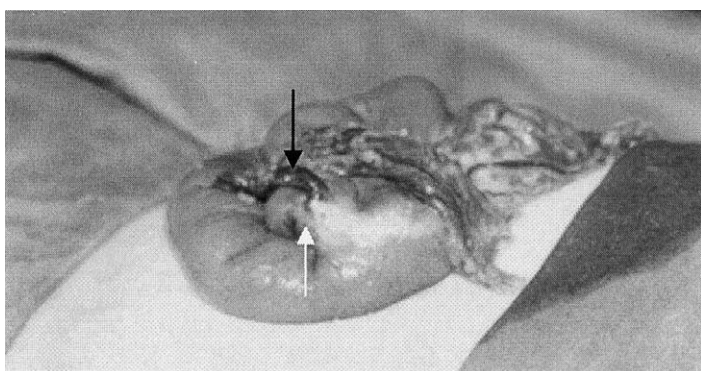

Fig 11. Inflamed Meckel's diverticulum (white arrow) with omental band adhesion (black arrow) acting as a pivot, round which small intestine volved causing closed loop obstruction. Treatment was by segmental resection and primary anastomosis.
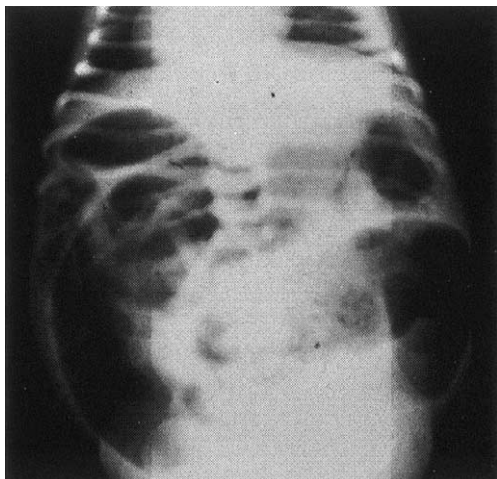
Fig 12.   Plain abdominal radiograph of a neonate with intestinal obstruction due to meconium ileus. Note the grossly distended intestinal loops and bowel wall edema.

relieved successfully, and the diagnosis of cystic fibrosis is established, the treatment for the intestinal manifestations focuses on nutritional supplementation, and pancreatic enzyme replacement. Simultaneously, the treatment of the life-threatening pulmonary disease focuses on mucous retention and chronic infection in the lungs.[35]

Gastrointestinal manifestations of cystic fibrosis include, in addition to neonatal meconium ileus, distal intestinal obstruction syndrome, constipation and acquired megacolon, rectal prolapse, and rarely pancreatitis. If the intestinal malabsorption is well controlled with an effective pancreatic enzyme preparation, distal intestinal obstruction syndrome, constipation, and rectal prolapse are infrequent. Persisting gastrointestinal symptoms should be investigated thoroughly to exclude other disorders not directly related to the cystic fibrosis.[36,37]

Survival at 1 year was 92% in patients with uncomplicated meconium ileus and 89% for those with complicated meconium ileus. The therapy of choice for uncomplicated meconium ileus is nonoperative hyperosmolar (Gastrograffin or Omnipaque) enema (Fig 13), with enterotomy and irrigation reserved for enema failures. Complicated cases require exploration and, in the absence of giant cystic meconium peritonitis, are usually amenable to bowel resection and primary anastomosis.[38] Injection of a solution of n-acetyl cysteine in the inspissated meconium helps to loosen it and facilitate its removal from the bowel lumen.

Meconium plug syndrome. Meconium plug syndrome or meconium obstruction in very low birth weight (VLBW) infants does not appear to be associated with cystic fibrosis or Hirschsprung's disease.[39] It is usually relieved by rectal stimulation (eg, glycerin suppository, rectal washouts, or contrast enema).

Milk Curd Syndrome

Intestinal obstruction caused by the inspissation of formula feeds should be considered in any infant presenting with distal small-bowel obstruction. There are typical radiographic and ultrasonographic changes that suggest the diagnosis. Relief of the obstruction is often possible without surgery.[40]

Intussusception

Intussusception is the invagination of one portion of the intestine into another. It is the most common form of intestinal obstruction in infants and accounts for about 700 hospital admissions each year in England and Wales.[41] It rarely occurs out with the first 5 years of life and is classically associated with intense intermittent abdominal pain, vomiting, bloody mucoid diarrhea, and a palpable abdominal mass. Etiologies of childhood intussusception differ depending on age at presentation: in younger infants (<18 months of age) the cause appears to be an inflamed enlarged intestinal lymph tissue patch acting as the lead point, whereas in older children the lead point may be, amongst others,[42] a hemangioma or other mural neoplasm or a Meckel's diverticulum. Plain abdominal x-ray may or may not be diagnostic. Improved results of treatment have followed recent technological developments, which include ultrasonographic imaging and pneumatic reduction techniques. Contrast or, preferably, air enema can be both diagnostic and therapeutic. Most intussusceptions can be reduced successfully without the need for operation but close cooperation between surgeon and radiologist is essential. In cases of prolonged symptoms, evidence of small-intesti-
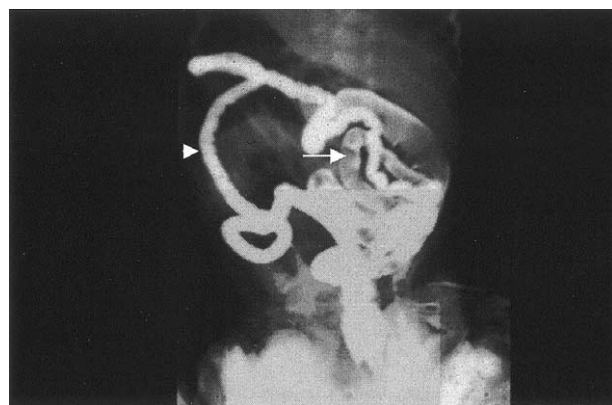

Fig 13.   PA view of the above patient after contrast enema. Note microcolon (arrowhead), reflux of contrast into distended bowel loops and outlines of meconium plugs in lumen (arrow).
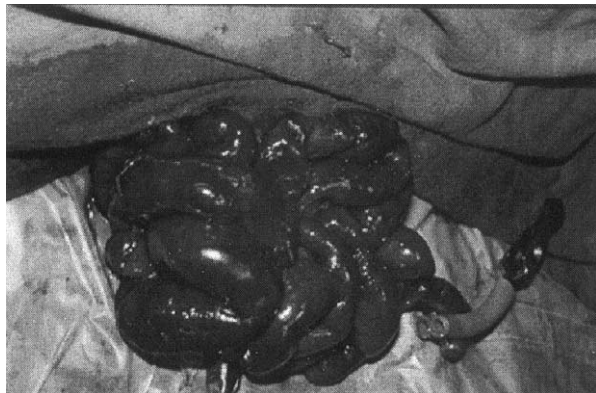
Fig 14. Operative findings in a case of severe NNEC, showing bowel gangrene and ischemia. Rubber shod clamp at level of resected perforated intestinal segment.

nal obstruction or peritonitis surgical treatment may be required.[43]

It is exceedingly important to emphasize that these children can be extremely ill at presentation, with few typical manifestations of the gravity of their clinical condition until they reach a point of decompensation and rapid deterioration. The immediate management, therefore, should center on the full and effective resuscitation of the child prior to any radiological or operative manipulation. Mortality and morbidity rates from the condition have progressively declined in recent decades but avoidable deaths still occur.[41]

Internal Hernia

An acute intestinal obstruction with strangulation in the absence of an external hernia and with no history of previous surgical procedures must suggest the possibility of an internal hernia,[44,45] especially if the patient has a history of chronic intermittent abdominal distress and a palpable abdominal mass is found on examination.[44]

Neonatal Necrotizing Enterocolitis

NNEC is one of the most common gastrointestinal emergencies observed in neonatal intensive care units. Despite extensive research efforts, the etiology and pathogenesis of necrotizing enterocolitis remain unclear. Unfortunately, the most sensitive and specific tests detect only advanced disease and perforation (Fig 14).

Intestinal stenosis or stricture occurs in approximately one third of medically treated infants surviving the acute phase of NNEC. Identification of these lesions by the use of routine contrast enemas has been advocated as a means of decreasing potential morbidity from delayed diagnosis, especially those infants not residing near pediatric surgical facilities.[46]

The mainstay of surgical treatment is resection with enterostomy, although resection and primary anastomosis is useful in selected cases. In addition, some neonates may benefit from peritoneal drainage, second-look procedures, or proximal diversion.[47]

Hirschsprung's Disease

Timely passage of the first stool is a hallmark of the well-being of the newborn infant. Failure of a full-term newborn to pass meconium in the first 24 hours may signal intestinal obstruction. Lower intestinal obstruction may be associated with disorders such as Hirschsprung's disease, anorectal malformations, meconium plug syndrome, small left colon syndrome, hypoganglionosis, neuronal intestinal dysplasia, and megacystis-microcolon-intestinal hypoperistalsis syndrome.

Hirschsprung's disease is a relatively common cause of intestinal obstruction in the newborn. It is characterized by an absence of ganglion cells in the distal bowel beginning at the internal sphincter and extending proximally for varying distances.[48] It may be considered to be a "neurocristopathy"—an abnormality of migration of neural crest cells and is associated with many other conditions, notably sensorineural deafness and Down syndrome (Fig 15).

Radiologic studies and rectal biopsy are usually required to make the diagnosis.[49] The mainstay of treatment initially involves the performance of bowel washouts (Fig 15) to ensure bowel remains empty and decompressed (not effective in total colonic disease) until the infant begins to thrive and is able to tolerate definitive treatment. The performance of colostomy and subsequent Duchamel procedure has now been replaced by 1-stage endorectal pull-through techniques and, more recently, laparoscopically assisted pull-through procedures. It is important to remove the affected bowel completely and this is facilitated by intraoperative availabil-

Fig 15. Bowel washout in a patient with Hirschsprung's disease and Down syndrome.

Fig 16. The characteristic appearance of a transitional zone in Hirschsprung's disease.

Fig 18. Anoscrotal fistula in a male infant.

ity of frozen-section biopsy reporting to ensure that the transition zone is totally removed (Fig 16).

Anorectal Atresia Spectrum

Anorectal atresia (Figs 17 and 18) may present with intestinal obstruction, but most often presents with failure to pass meconium and an easily recognizable anatomical aberration of the perineum. Rectal atresia and stenosis are rare and peculiar anorectal malformations for which many and varied surgical procedures have been described.[50] The mainstay of treatment involves large-bowel decompression by a defunctioning dismembered sigmoid colostomy (allowing sufficient length of distal colon) to allow full anatomical investigation/evaluation followed by an anorectal pull-through procedure.

Trauma

The most commonly reported intestinal injury from seat belts or other blunt abdominal trauma in children is perforation. A rarely reported late sequel following this type of injury is posttraumatic intestinal stricture.[51]

Inflammatory Bowel Disease

Crohn's disease is a chronic, transmural inflammatory disease of the intestinal tract most frequently involving the terminal ileum and colon. It is a disorder of undetermined etiology that shares many clinical aspects with chronic ulcerative colitis. Intestinal obstruction can be caused by stricture formation or involvement of bowel loops in an inflammatory mass. Surgical treatment of Crohn's disease continues to be generally limited to the treatment of the complications of the disease. The surgical principle generally adhered to, is to remove only enough intestine to relieve the complication but to maintain as much intestinal length as possible. Because of this requirement, strictureplasty has become a worthwhile surgical adjunct, especially when dealing with multiple minimally inflamed fibrotic strictures in the small intestine.[52]

Tropical Infections: Ascariasis

Ascaris lumbricoides, the most frequent human intestinal nematode, is the causative agent of ascariasis, with an estimated worldwide prevalence of over 1 billion cases. Although characterized with low morbidity and mortality rates, the global prevalence of ascariasis still results in approximately 20,000 deaths annually, primarily as a consequence of intestinal obstruction. In humans, transmission usually occurs by hand-to-mouth route by way of contaminated agricultural products and food, or from dirty hands.
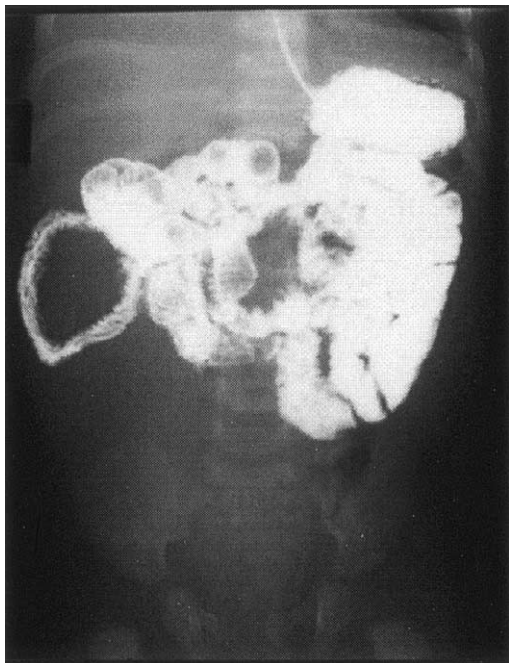

Fig 17. Anoperineal fistula in a female infant.

Fig 19. Contrast follow-through in an infant with abdominal distention and feed intolerance showing collapsed and elongated bowel loop in right flank, with impression of mesenteric filling defect.

Plain abdominal x-ray may suggest the diagnosis, but contrast radiography and ultrasonography, and sometimes endoscopic retrograde cholangiopancreatography, may be required. Ultrasonography can detect worms in the biliary tract, pancreas, and intestine and is a useful noninvasive technique for diagnosis and follow-up of such patients.

Although generally asymptomatic, heavy infestation (more than 60 organisms) may cause partial or complete obstruction of biliary or intestinal tracts. The organism load is about 10-fold higher in fatal cases.[53] Infestation can be complicated by intussusception, perforation, and gangrene of the bowel; acute appendicitis; and appendicular perforation.[54] Anthelminthic chemotherapy is required to eradicate the parasites and prevent potentially serious complications. Mebendazole, albendazole, pyrantel, and levamisole are the most widely used agents to treat ascariasis and can be delivered to communities in endemic regions, serving as an affordable and cost-effective strategy to reduce the prevalence and morbidity in these regions.[54,55]

Surgical treatment involves enterotomy and removal of the parasites or treatment of other complications, eg, intestinal gangrene. Endoscopic retrograde cholangiopancreatography can be used to extract worms from the biliary and pancreatic ducts when indicated.

Other Rare Causes of Intestinal Obstruction

Ingested foreign bodies. Typical patterns of symptoms of intermittent obstruction resembling the adult gallstone ileus syndrome can be the presenting features of foreign body ingestion (peanut, phytobezoar,[56] or pica). Therapeutic approach includes conservative measures, endoscopic or operative removal of foreign body. Pica is a serious health risk for mentally handicapped patients: 75% of cases required surgical intervention, with a 30% complication rate and 11% death rate.[57]

Mesenteric lymphatic cyst. Cystic mesenteric lymphatic malformations are uncommon abdominal masses, which can cause abdominal signs and symptoms for a wide variety of reasons (Figs 19 and 20). Also known as mesenteric, omental, or retroperitoneal cysts, they can present in a variety of ways, including intestinal obstruction, volvulus,[58] nonspecific abdominal pain, intracystic hemorrhage, or as an asymptomatic abdominal mass. Abdominal ultrasound scan provides a definitive diagnosis in most suspected cases. Complete resection is possible in most patients, except those with extensive retroperitoneal involvement. Recurrence is unusual when complete resection is accomplished.[59]

Solitary intestinal fibromatosis. Solitary intestinal fibromatosis is a very rare condition, which may present with intestinal obstruction. It appears to be a condition of infancy and carries a very good prognosis after segmental resection.[60]

Eosinophilic enteritis. Eosinophilic enteritis or gastroenteritis is a rare disease characterized by tissue eosinophilia, which can affect different layers of the bowel wall. It can affect any area of gastrointestinal tract from the esophagus to the rectum, although the stomach and small intestine are the sites most frequently affected.[61] It may present as childhood gastrointestinal obstructive
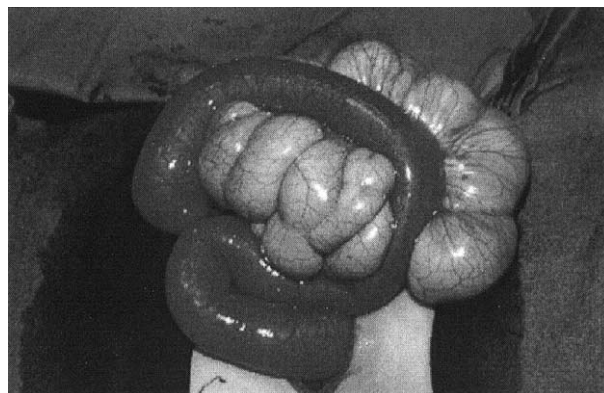

Fig 20. Operative picture of above, demonstrating a mesenteric lymphangioma containing chyle. Definitive treatment was by complete excision of the cyst with segmental bowel resection and primary anastomosis.

disease with involvement of the colon but sparing of the stomach.[62] Symptoms can be controlled with steroid therapy.

Benign intestinal tumors. Intestinal tumors are rare in children. However, an intestinal tumor can cause obstruction and would usually be treatable by segmental resection and primary anastomosis. An inflammatory myofibroblastic tumor of the jejunum is such an entity. Histological characterization is essential, as potential recurrence would necessitate long-term follow-up.[63]

Malignant intestinal tumors. Malignant intestinal tumors are also, thankfully, very rare in children. The most common tumor is intestinal lymphoma, which is very responsive to chemotherapy. Primary intestinal leiomyosarcomas are other rare tumors in children. Visceral metastases from these neoplasms are atypical and, with complete excision, recorded long-term prognosis may be favorable, unlike their counterpart in the adult population.[64]

Adenocarcinoma of the colon has also been reported in a 10-year-old boy presenting with history of lower gastrointestinal bleeding for 1 year and acute intestinal obstruction.[65] Although short-term follow up was favorable after radical colectomy, these tumors are usually of poor prognosis.

The abdominal cocoon. This is a rare cause of intestinal obstruction most often found in adolescent girls from tropical and subtropical countries. It is characterized by a thick fibrotic sac covering the small bowel partially or completely, the etiology of which is unknown. A correct diagnosis is not often made preoperatively; however, following simple surgical release of the entrapped bowel, these patients usually do well.[66]

Chronic intestinal pseudo-obstruction.[67] This condition may present with symptoms and signs of intestinal obstruction and evidence of gross intestinal dilatation on abdominal x-ray, but the gastrointestinal tract is fully patent. It is a very difficult clinical condition to treat and both medical (eg, prokinetic agents) and surgical methods (laparotomy, adhesion lysis, enteropexy, temporary placement of long intestinal tube) are generally ineffective. The diagnosis is usually made after a negative laparotomy and is an important one to make in order to avoid further unnecessary explorations.

AIDS. The pathologic changes in the gastrointestinal tract of children with AIDS are variable, clinically significant, and reflect multisystemic disease processes, involving any combination of inflammation, changes in the lymphoid tissue, miscellaneous lesions, AIDS associated arteriopathy, and tumors. Cytomegalovirus infection of the gastrointestinal tract can be associated with ulceration, bleeding, perforations, and intestinal obstruction and carries a high morbidity and mortality. The remaining infections are generally not life-threatening. Both lymphomas and smooth muscle tumor in children with AIDS can cause intestinal obstruction and are related to Epstein-Barr virus infection. The smooth muscle tumors are frequently malignant and multiple.[68]

Investigations. Imaging plays a major role in most neonatal/pediatric gastrointestinal emergencies. The role may vary from helping to establish a diagnosis, to the evaluation of associated abnormalities, to surgical planning, or to therapy for some conditions like meconium ileus or meconium plug syndrome. Plain film of the abdomen is often helpful in determining the level of obstruction[1] and usually dictates, together with clinical symptoms, the choice of further investigations (with ultrasound, computed tomography scan, and magnetic resonance imaging playing roles in more complex cases[69,70]). Other investigative modalities may be required to evaluate
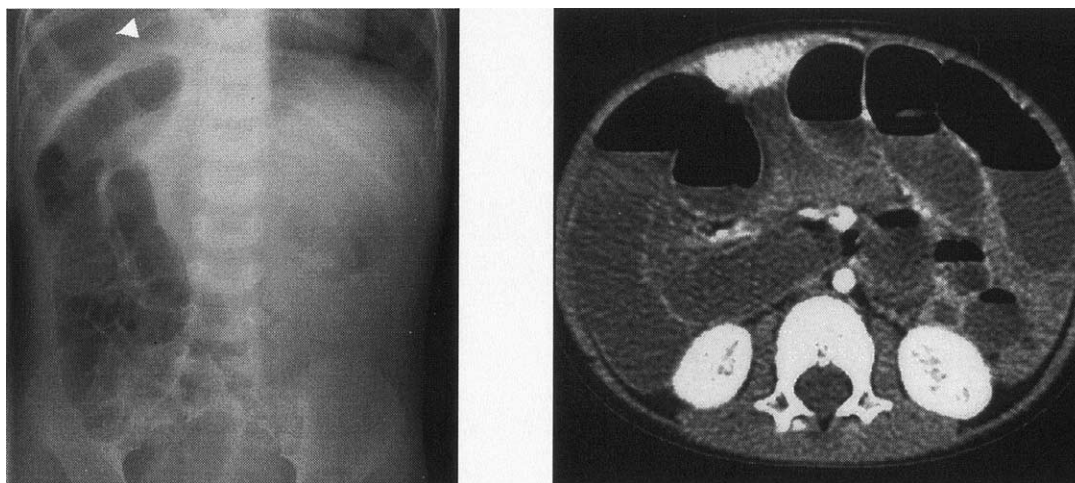

Fig 21. Plain x-ray and intravenous contrast-enhanced computed tomograph showing bowel ischemia, in a case of late presentation of incarcerated diaphragmatic hernia.

complex cases with concomitant congenital and chromosomal anomalies.[71]

In cases of suspected small-bowel obstruction, computed tomography may confirm the diagnosis and demonstrate the cause of obstruction, preventing a delay in surgical treatment. In addition, it may have a role in differentiating simple from strangulated small-bowel obstruction[72] (Fig 21).

Detailed review of the investigation of the vomiting child will be presented in another article.

CONCLUSIONS/THE FUTURE

With the advent of pediatric and neonatal intensive care and multidisciplinary care the morbidity and mortality of cases of intestinal obstruction reported in current series is generally extremely low and mainly determined by the coexistence of other major congenital anomalies (eg, cardiac), delays in diagnosis and treatment or coexisting medical conditions[73] (Table 1).

Future therapies for patients with gastrointestinal disease secondary to cystic fibrosis include lung transplantation, pharmacologic manipulation of the epithelial cell abnormality, and gene transfer therapy.[35]

When, by the nature of the intestinal disorder (eg, multiple jejunoileal atresias, NNEC), the patient suffers from ultrashort-bowel syndrome (<40 cm) requiring long-term total parenteral nutrition, which can be complicated by liver disease, the morbidity and mortality can be prohibitive. Use of growth factors to enhance adaptation, probiotics and selective substrates (glutamine, raftulose), and advances in small-bowel transplantation may improve long-term outcomes.[73]

Finally, the exciting developments in laparoscopic surgical technique and refinements in the instrumentation involved (the first report of a successful diamond-shaped duodeno-duodenostomy for duodenal atresia has been reported by Bax[74]) are further complementing the treatment modalities for intestinal obstruction.

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