CHAPTER 61
Neonatal Intestinal Obstruction

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Introduction

Neonatal intestinal obstruction (NIO) is one of the most common emergency conditions a paediatric surgeon is called upon to assess during the neonatal period. Successful management of NIO depends on timely diagnosis and referral for therapy. The diagnosis is based on history (symptoms) and physical examination (signs) confirmed by some investigations such as radiographic and histopathological studies. Catastrophic events such as volvulus, ischaemic loop of bowel, pneumoperitoneum, and/or pneumonia from aspiration and malnutrition could be overcome through efficient and timely resuscitation and urgent transport to a specialised unit.

The desired goal of healthy survival of neonatal intestinal obstruction requires a coordinated interaction of medical, nursing, and rehabilitative specialties in an organised team. Early surgical intervention is paramount and may mean all the difference between intestinal salvage and crippling short gut syndrome. The typical case of neonatal bowel obstruction is generally straightforward, and the outcome is potentially excellent. Only very preterm babies and those of extremely low birth weight may succumb. However, in Africa, late presentations and poor resources lend to a mortality of up to 50%.[1] Few dedicated paediatric hospitals exist in developing countries.[2]

Intestinal obstruction can be complete (atresia, anorectal malformation (ARM)) or incomplete (stenosis, web). Obstruction may be intraluminal (meconium ileus or meconium plug syndrome) or functional (Hirschsprung's disease (HD)). Proximal obstruction presents with earlier vomiting and less abdominal distention, whereas distal bowel obstruction lends itself to late emesis and greater abdominal distention. This chapter provides an overview on neonatal bowel obstruction. Each specific condition is covered in chapters elsewhere in this book, specifically, Chapter 58 (inguinal and femoral hernias and hydroceles), Chapter 62 (duodenal atresia and stenosis), Chapter 63 (intestinal atresia and stenosis), Chapter 65 (intestinal malrotation and midgut volvulus), Chapter 67 (meconium disease), Chapter 76 (Hirschsprung's disease), and Chapter 77 (anorectal anomalies).

Demographics

The incidence of NIO is approximately 1 in 5,000 live births. The true incidence in Africa is unknown, but a recent report from Tanzania has shown that it is still the most common neonatal surgical emergency.[1] Many cases still die undiagnosed and untreated.

Aetiology/Pathophysiology

Neonatal intestinal obstruction has varied aetiology, so the pathophysiology is diverse.

The gastrointestinal tract (GIT) arises from the yolk sac. At 3 to 4 weeks' gestation, it becomes a distinct entity. A connection, the vitelline (omphalomesenteric) duct, may persist as a Meckel's diverticulum. The alimentary tube is divided according to its blood supply into the foregut, midgut, and hindgut.

The foregut comprises the oesophagus, stomach, and duodenum. These are vascularised by multiple sources—the thyrocervical, intercostal, celiac axis, and superior mesenteric vessels.

The midgut comprises the jejunum and ileum as well as the ascending and proximal transverse colon. These are supplied by the superior mesenteric vessels.

The hindgut comprises the distal colon, which is supplied by the inferior mesenteric vessels, and the rectum, which is supplied by the internal iliac vessels.

Aberrations of foregut formation include duodenal stenosis, duodenal atresia, and annular pancreas. Maldevelopment of the midgut includes malrotation as well as jejunal and ileal atresia. Meconium ileus involves the distal ileum. Hirschsprung's disease (aganglionic megacolon), meconium plug syndrome, and imperforate anus involve the hindgut. Enteric duplications occur in all three locations.

Jejunoileal atresia is a condition acquired during foetal development due to disruption of the mesenteric blood supply. In their classic work on foetal dogs in 1955, Louw and Barnard from Cape Town, South Africa, clarified the pathophysiology of jejunoileal atresia.[3] Other abdominal conditions occurring in utero, such as gastroschisis, volvulus, or intussusception, may be associated with intestinal atresia due to kinking, stretching, or otherwise disrupting the blood flow to the foetal bowel. Chromosomal anomalies are rare (<1%) in babies with jejunoileal atresia.

Meconium ileus is the earliest manifestation of cystic fibrosis (CF), an autosomal recessive condition characterised by abnormalities in cellular membrane physiology and chloride ion transport that contribute to progressive respiratory failure, derangements in cellular secretory patterns, and diminished mucosal motility. In developed countries, 10–20% of newborns with CF present with meconium ileus, an association first described by Landsteiner in 1905.[4] A cystic fibrosis gene that is different from that in the caucasian population has been identified in the black African population;[5] however, the incidence in Africa is much reduced, and in many sub-Saharan countries, the disease is not reported.[6]

Abnormalities in the cystic fibrosis transmembrane regulator (CFTR) disrupt transmembrane flux of the chloride ion, which subsequently affects sodium transport as well. The meconium of affected babies is thick and sticky; this, coupled with the poor motility of an immature intestine, leads to intraluminal obstruction of the terminal ileum. A contrast enema might reveal the characteristic finding of a microcolon. It is essential to push the contrast up to the level of obstruction, namely, the distal ileum to be therapeutic by flushing out the meconium plugs (Figures 61.1 to 61.4).

Meconium plug syndrome refers to inspissated meconium obstructing the colon; it may denote HD but not CF. Conditions that predispose to dysmotility of the neonatal bowel (e.g., maternal pre-eclampsia, diabetes mellitus, administration of magnesium sulfate, prematurity, sepsis, and hypothyroidism) may be responsible for the formation of the meconium plug. A water-soluble contrast enema can be both diagnostic and therapeutic for this condition.

Hirschsprung's disease is a disorder of the neuroenteric pathways in the distal colon that results in a bowel that is tonically contracted. Bowel peristalsis is controlled by neuroenteric ganglion cells, which

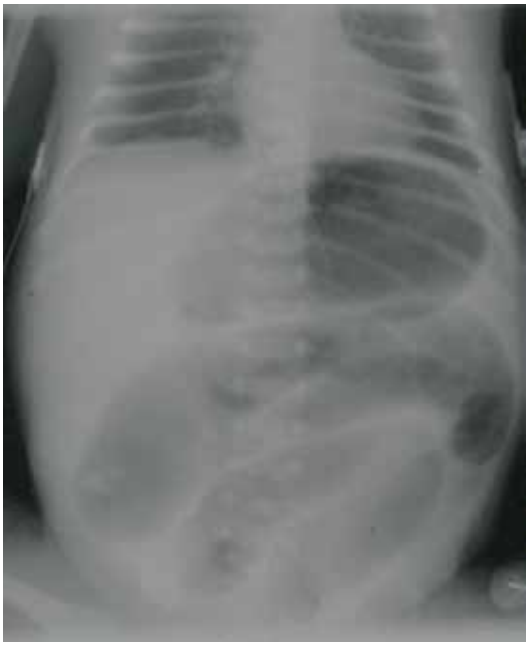

Figure 61.1: Radiograph showing ground-glass appearance of meconium ileus.

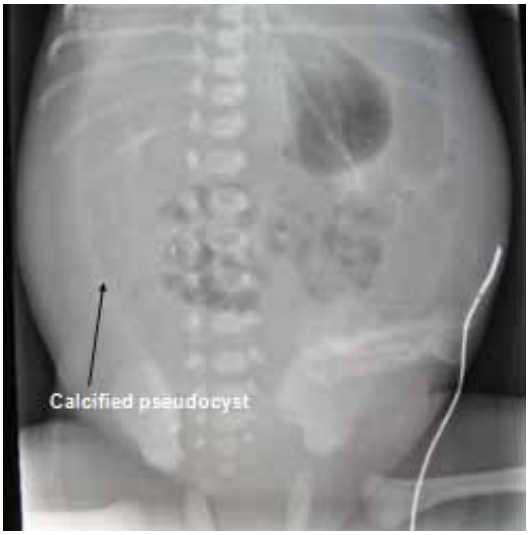

Figure 61.2: Meconium peritonitis with calcification and pseudocyst due to in utero perforation.

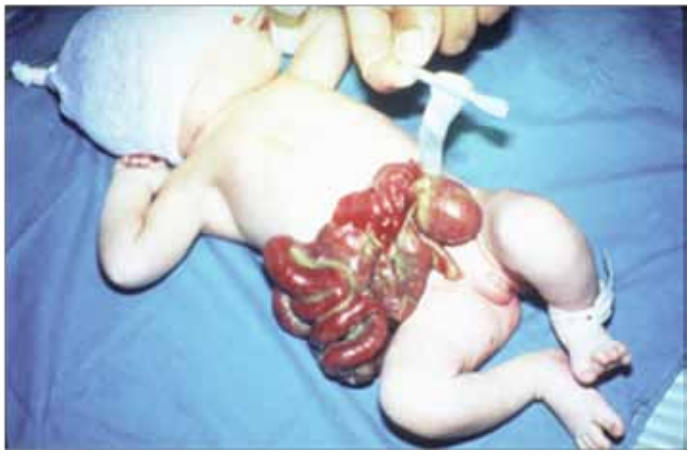

Figure 61.3: Meconium ileus with perforation.

are present in the submucosal layer of the intestine and migrate from the neural crest distally along the bowel to reach the rectum at about 7–10 weeks' gestation. HD is the congenital absence of neuroganglion cells; consequently, the peristaltic relaxation phase is absent distally, and the affected intestine does not appropriately relax, causing a functional obstruction. The extent of the aganglionic segment varies with each patient, but extends from the distal rectum proximally. The level at which the proximal but healthy bowel starts to dilate is called the transition zone (Figure 61.5).

The genetic defects responsible for HD consist of abnormalities on more than one chromosome and include the RET proto-oncogene, located at chromosome 10q11.21. RET interacts with a protein termed EDNRB, encoded by the gene EDNRB, which is located on chromosome 13.

Anorectal malformation

At 4 to 6 weeks' gestation, the hindgut separates into the urogenital sinus and the anorectum, which then undergoes canalisation. The distal third of the anus develops from ectoderm and becomes the anal pit, whereas the proximal portion of the anal canal is derived from mesoderm. An anal membrane covers the canal until 8 weeks' gestation, when it perforates and becomes a patent anus. Imperforate anus results if this sequence of events occurs improperly.

In summary, conditions of NIO include:

- hernia (inguinal, internal);
- atresia, stenosis, web (oesophageal, duodenal, jejunoileal, colonic);
- anorectal malformation;
- Hirschsprung's disease;
- meconium ileus or plug; and
- malrotation with midgut volvulus.

Clinical Presentation

History

The history of NIO is typical for the level (high/ low) and type (mechanical, functional) of obstruction. High intestinal obstruction presents with early vomiting, whereas low intestinal obstruction presents with abdominal distention and later onset vomiting. Feeding intolerance with bile-stained vomiting, absent meconium, and abdominal distention are therefore paradigmatic. With late presentation, the symptom presentation might change due to complications and malnutrition.

On antenatal ultrasound, polyhydramnios is a common feature. Intrauterine bowel dilatation may also be noted if scanned after 24 weeks' gestation.

Physical Examination

Depending on the level of intestinal obstruction, the physical finding is that of a distended abdomen. Within the African context, these babies often present late with aspiration pneumonia, malnutrition, and final events such as intestinal perforation and sepsis. A careful examination and search for typical signs (e.g., abdominal distention, peristaltic waves across the abdomen, absent anus or perianal fistulas, severe distention, and malnutrition in HD) usually reveal the appropriate suspected diagnosis.

Investigations

The most important and useful test in any NIO is the abdominal radiograph (AXR). A single supine and lateral shoot-through is usually sufficient. The distribution of the air contrast directs one to the appropriate diagnosis. Very distended loops of bowel are indicative of atresia. Long-standing drainage from a nasogastric tube (NGT), however, could make such a diagnosis difficult. A duodenal atresia could in this way be missed purely by the duodenal bulb being collapsed from the ongoing drainage. A repeat radiograph with injection of some air through the NGT facilitates the diagnosis.

Once a diagnosis is suspected, contrast studies may help in assessing the rest of the bowel and/or be therapeutic. Such investigation is paramount for the demonstration of the anatomy in ARM after a colostomy has been performed and to assess the length and level in HD (see Figure 61.5). A water-soluble contrast enema will help to clear the thick meconium in meconium ileus.

Further radiological studies have to be requested to assess associated abnormalities such as those included in the acronym VACTERL (vertebral, anorectal, cardiac, tracheo-oesophageal, renal, and limb).

Blood tests are needed to facilitate and modulate resuscitation. Depending on the severity of the condition and its delayed presentation, blood products might be needed for the surgery.

Management

Preoperative Treatment

All conditions need fluid resuscitation and nasogastric decompression. Broad-spectrum antibiotics should be started prophylactically.

Condition-Specific Management

Duodenal atresia

Evaluate for trisomy 21. Because duodenal atresia is considered a midline defect, an evaluation for associated anomalies should include echocardiography, head and renal ultrasonography, and vertebral skeletal radiography.

Jejunoileal atresia

Intraoperatively distal atresias can be identified by flushing the distal intestinal lumen with warm saline to confirm intestinal continuity down to the level of the rectum.

Meconium ileus

The traditional gastrografin enema has been replaced with a water-soluble contrast enema, which is equally effective in loosening the meconium impaction. The enema fluid must be refluxed into the terminal ileum.

N-acetylcysteine may be administered by NGT to further loosen the meconium.

Hyperosmolar enemas may increase the risk of hypovolaemic shock and injury to the intestine with perforation. The risk of perforation is reportedly 3–10%.

Meconium plug syndrome

A gentle rectal washout with temperate normal saline might alleviate the obstruction immediately. A rectal suction biopsy and or a contrast enema should rule out HD.

Full-thickness rectal muscle biopsy is recommended where there is no frozen section or histochemical assay available

Hirschsprung's disease

Initial rectal washout will alleviate the obstruction. Rectal suction biopsy or full thickness biopsy will confirm the diagnosis. A contrast enema will show the level of disease.

Imperforate anus

An 18-hour plus AXR, which is the time required for swallowed air to reach the level of obstruction, will help to show the level of abnormality.

Operative Therapy

Duodenal atresia

A diamond-shaped or side-to-side duodenoduodenostomy is an easy procedure to bypass the obstruction.

Malrotation with volvulus

Malrotation with midgut volvulus (Figure 61.6) is a true surgical emergency in the newborn. Delay in operation may result in catastrophic loss of the bowel and death.

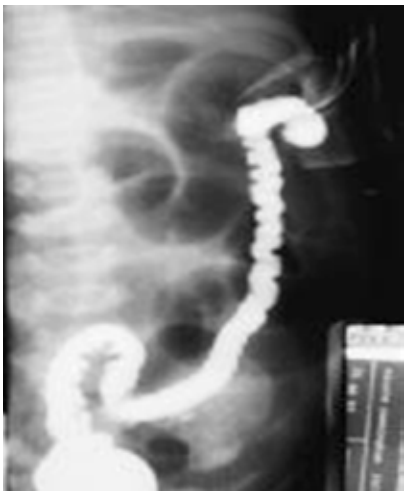

Figure 61.4: Contrast enema study showing microcolon.

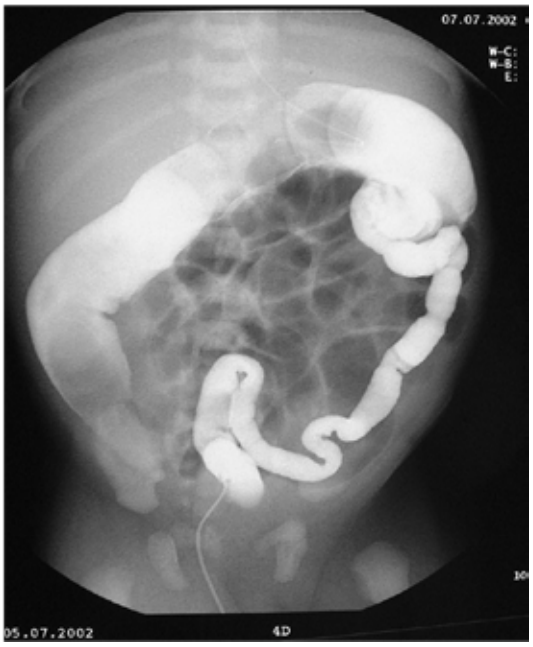

Figure 61.5: Contrast enema in Hirschsprung's disease showing diseased narrow bowel, transition zone, and dilated normal bowel.

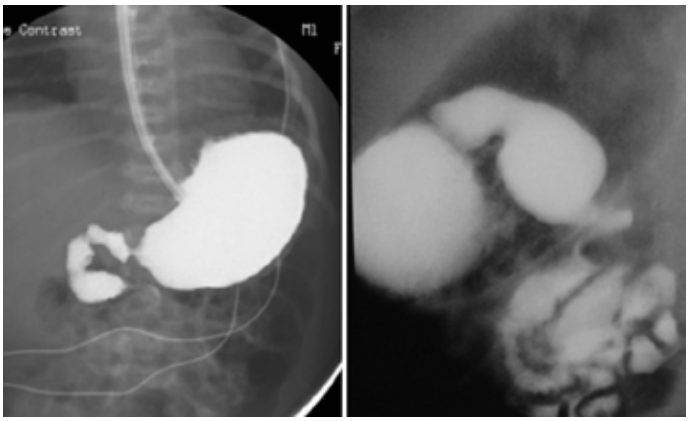

Figure 61.6: Contrast study showing malrotation with volvulus.

Assess the stage of ischaemia and derotate counterclockwise if the bowel seems viable. Inform the anaesthetist about the manoeuvre because there will be a flush of intravenous endotoxins.

Depending on the viability of the bowel, continue on to a Ladd's procedure, in which an extensive mobilisation of the mesentry is performed.

Removal of the appendix is controversial and a surgical choice.

Jejunoileal atresia

Surgery for jejunoileal atresia involves resection of the most distended proximal bowel and primary anastomosis. A diverting ostomy is avoided if possible. As with surgery for duodenal atresia, resection or tapering of the proximal dilated segment is occasionally necessary to limit the dysmotility that occurs in grossly dilated bowel. The ileocaecal valve is preserved if possible because this prevents egress of bacteria from the colon into the small intestine with resultant bacterial overgrowth and malabsorption.

Meconium ileus

Calcification on AXR (see Figure 61.2) indicates that an intestinal perforation occurred in utero and spontaneously sealed; if not, the extruded meconium is walled off by adjacent intestine to form pseudocysts.

These babies have meconium peritonitis, and their appearance is unmistakable; these are babies who are born with (as opposed to those who develop) a distended, erythematous abdomen.

A laparotomy is undertaken with drainage of the meconium pseudocyst and identification of the site of the perforation, which is converted to an enterostomy.

In uncomplicated meconium ileus, an enterostomy with irrigation of the bowel contents may successfully loosen the meconium and permit its evacuation and facilitate the closure of the enterostomy over a t-tube. Postoperatively, after a contrast study shows distal patency, the t-tube can be removed for the controlled fistula to close without further need of surgery. Rarely, some patients might need an ostomy for diversion and access for proximal and distal irrigation with N-acetyl cysteine.

Hirschsprung's disease

The treatment of Hirschsprung's disease is primarily surgical, except in instances of enterocolitis.

Patients with HD are treated with a colostomy near the transition zone (level of beginning of dilatation). If histological leveling is not possible in emergency cases, a right transverse diverting colostomy is safe. It is sometimes difficult to visualise the transition zone in a neonate.

A pull-through procedure is performed after the child is feeding and gaining weight or at least 6 weeks after enterocolitis.

Different procedures have been described as one- or two-stage procedures and are increasingly performed at a younger age.

The most recent innovations include minimally invasive techniques, such as the transanal pull-through using laparoscopy in cases in which the transition zone is not located in the distal sigmoid colon.

Imperforate anus

Low lesions with fistulous connections to the perianal skin can be repaired primarily by anoplasty.

If the fistula runs from the rectum to the vagina or urethra or urinary bladder, the imperforate anus is classified as high, and the infant should undergo a colostomy.

Definitive repair of the imperforate anus is classically performed by posterior sagittal anorectoplasty, in which the rectum is situated within the striated muscle complex and anal sphincter.[7]

The laparoscopic pull-through using three ports has become a favoured procedure for high anorectal malformations.

Increasingly, laparoscopic techniques have been used to repair the above-mentioned conditions. A good alternative for Africa is the minimal approach described by Banieghbal and Beale,[8] whereby access is gained through the umbilicus. Such an approach leaves a virtually unrecognisable scar.

General Postoperative Care

Modern supportive care in the intensive care unit (ICU) with continuing fluid resuscitation, parenteral nutrition, and respiratory support have been the bases for the increased survival rate. In countries where parenteral nutrition is not available, transanastomotic tubes have been tried with indefinite success for the purpose of early feeding. This postoperative management will make all the difference to the survival of children in Africa.

Two weeks after anorectoplasty, serial anal dilatations should start by using anal dilators of increasing size. Within Africa, the child should be kept hospitalised until the mother is comfortable with digital dilatations.

In all of these conditions, the neonatologist and paediatric surgeon must work together in a coordinated fashion, allowing the diagnosis to be quickly established and therapy to be rapidly implemented. In conditions of the intestine that are known to be associated with systemic disease, such as duodenal atresia (trisomy 21) and meconium ileus (cystic fibrosis), appropriate consultation should be obtained early, and the continued involvement of appropriate specialists may be warranted long after the baby has recovered from the initial hospitalisation.

Postoperative Complications

Postoperative complications pertain to factors of

- total parenteral nutrition (cholestasis and hyperalimentation hepatitis);
- central venous access (pneumo/hemothorax, catheter embolus); and
- catheter sepsis.

Postoperative Stricture and/or Adhesions

Anastomotic stricture is a complication after surgery. Postoperative adhesions can occur after any laparotomy. They may be caused by peritonitis from leaking anastomosis. A recent study of 1,541 children who had intestinal surgery showed an adhesion rate of almost 10% in the operative site and a rate of approximately 5% elsewhere.[9]

Decreased Gut Motility

Poor motility is often observed following repair of atresias. Chronic dilatation of the intestine proximal to the obstruction may alter normal peristalsis across that segment of bowel, even after the obstruction has been relieved.

Malabsorption

Short gut syndrome results when the length of intestine that remains postoperatively cannot sustain normal absorption of nutrients. The normal length of the small bowel in a term infant is approximately 250 cm. The estimated minimum jejunoileal length for sufficient bowel function in a term infant is around 75 cm. Resection of more than 60% of the small bowel or resection that removes crucial anatomic segments, such as the ileocaecal valve, predisposes to malabsorption.

Bacterial overgrowth may contribute to malabsorption and subsequent failure to thrive. Probiotics have been shown in some studies to normalise bowel flora and improve outcomes. Bowel-lengthening procedures and hormonal bowel manipulation may help wean the patient with short gut syndrome from dependence on parenteral nutrition.

Newer techniques, such as the serial transverse enteroplasty procedure (STEP), may offer improved bowel function and length in some patients. Small bowel transplant, with or without other viscera such as liver and pancreas, is being performed in select centres in the United States and United Kingdom with varying results.

Prognosis and Outcomes

The prognosis for babies with these conditions depends entirely on the delay at presentation, appropriateness of resuscitation, operative and anaesthetic expertise, and most of all on the postoperative care available. Unless there is a neonatal ICU (NICU), outcome is inevitably poor. All efforts in Africa should be spent on improving transport to hospital and postoperative care.

Prevention

The improvement of antenatal care and early transport to a tertiary centre may improve the management and outcome of babies with the above-mentioned congenital abnormalities. There seems to be some evidence that folic acid may decrease the incidence of ARM.

Evidence-Based Research

Table 61.1 presents a comparative analysis of neonatal patients with intestinal obstruction in two groups treated during five-year consecutive periods to track the trends in management.

Table 61.1: Evidence-based research.

Title	Trends in neonatal intestinal obstruction in a developing country, 1996–2005
Authors	Ekenze SO, Ibeziako SN, Ezomike UO
Institution	Department of Surgery, University of Nigeria Teaching Hospital, Enugu, Nigeria
Reference	World J Surg 2007; 31:2405–2409
Problem	Outcome of neonatal intestinal obstruction is poor in Africa compared to the rest of the world.
Intervention	Better resources and expertise and referral to a tertiary centre may improve results.
Comparison/ control (quality of evidence)	A comparative analysis was performed involving 128 consecutive NIO cases managed from January 1996 to December 2005 at the University of Nigeria Teaching Hospital, Enugu, in southeast Nigeria. Fifty-five (43.0%) neonates were managed in the first five years (group A), and 73 (57.0%) in the last five years (group B). The aetiology of obstruction did not vary significantly in the two groups. Average duration of symptoms before presentation fell from 5.9 days (group A) to 4.7 days (group B). With the exception of Hirschsprung's disease, all other cases required operative treatment. In HD, the colostomy rate declined from 44.4% (group A) to 26.7% (group B). More neonates in group B were managed with general anaesthesia and perioperative third-generation cephalosporin antibiotics ($p = 0.01$). Although the complication rate did not vary significantly in the two groups (group A, 42%; group B, 40.3%), survival improved (group A, 61.8%; group B, 72.6%). Earlier presentation, improved manpower, and use of potent antibiotics may have contributed to the improved outcome.
Outcome/ effect	Challenges in the form of lack of neonatal intensive care facilities and dearth of qualified personnel persist. There is a trend toward earlier presentation and increased survival of babies with NIO. Improving the existing facilities and trained manpower, and establishing collaboration with centres that have excellent results may further encourage the trend.
Historical significance/ comments	Survival of neonates in Africa with intestinal obstruction can improve from 50% to above 90%, as reported in the well-resourced part of the world, if the above challenges are met.

Key Summary Points

1. Neonatal intestinal obstruction is one of the most common neonatal surgical emergencies.
2. Successful management of NIO depends on timely diagnosis and referral for therapy.
3. The diagnosis is made based on clinical findings of bile-stained vomiting, degrees of abdominal distention, and failure to pass meconium.
4. Plain radiographs assist in most diagnoses.
5. Radiological contrast and histopathological studies further aid in the diagnoses.
6. Catastrophic events such as volvulus, ischaemic loop of bowel, pneumoperitoneum, and or pneumonia from aspiration and malnutrition, could be overcome through efficient and timely resuscitation and urgent transport to a specialised unit.
7. Outcomes are resource- and expertise-dependent.

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