Midgut Atresia: Diagnostic and Management Challenges From Northern Tanzania

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ABSTRACT: Intestinal obstruction is one of the most common surgical emergencies in the neonatal period. Early diagnosis is vital for proper management and good outcome. Intestinal obstruction can be divided into high, for example, duodenal atresia and jejunal atresia, or low, for example, ileal atresia, colonic atresia, and Meckel's diverticulum. The most common cause of intestinal obstruction in neonates is midgut atresia. Surgical correction is needed and is a challenge in the developing countries where there is lack of pediatric surgeons, anesthesiologists, and intensive care. More research and data is also needed across countries to show the uneven distribution of the available resources.

KEYWORDS: Duodenal atresia, intestinal obstruction, jejuno-ileal atresia, neonatology

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Introduction

Intestinal obstruction (IO) is the most common surgical emergency in the neonatal period accounting for 1 in 1500 live births.[1] This can be divided into high (occurring proximal to the ileum) or low intestinal obstruction (occurring in the ileum or distal to it).[2] Atresia, a congenital complete obstruction of the intestinal lumen, usually resulting from failed recanalization or vascular anomalies in fetal life, accounts for approximately 22% to 49% of intestinal obstructions in neonates.[3,4] Duodenal atresia is not a rare occurrence among pediatric surgical conditions. It has an incidence as high as 1 in 5000.[5] Duodenal atresia often occurs concurrently with other malformations such as Verbal, Anorectal, Cardiac, Esophageal, Renal & Limb (VACTERL) and is more common in males than females.[5] Jejunoileal atresia occurs in about 1 in 5000 live births with equal male-to-female distribution.[1] About 1 in 3 affected infants are premature.[1] Other common causes of neonatal IO include meconium plug, Hirschprung's disease, anorectal malformations (ARM), malrotation, Meckel's diverticulum, and colonic atresia.[6] In developing countries, where pediatric surgical services are few and far between, there is a paucity of published data regarding the epidemiology of neonatal intestinal obstruction.

Congenital malformations of GI systems are rare, but more than half can be detected in utero. Early diagnosis and surgical correction are important for achieving positive long-term results with low morbidity and mortality.[7] Surgical correction was a challenge for pediatric surgeons in the past due to inadequate pediatric anesthesia and pediatric intensive care. However, improvements in surgical techniques and post-operative care have improved the survival rate and the quality of life.[7] Herein we present 5 cases of newborns with midgut atresia managed surgically at our tertiary level facility and the challenges faced.

Case Presentation

Case 1

A 10-day-old male baby was referred to our center due to vomiting since birth. This baby was born at home by spontaneous vaginal delivery at term. Birth weight at delivery and Apgar score were hence not documented. At admission, examination showed the baby was sick-looking, wasted, dehydrated, pink on room air, and saturating at 98%. His temperature was 37°C, respiratory rate was 80 breaths/minute. His random blood glucose on admission was 9.2 mmol/L. On respiratory system examination, he had marked lower chest indrawing, nasal flaring, and grunting. His abdomen was not distended; it was soft, with a palpable mass in the supra-umbilical region. The digital rectal exam was normal, and the child had normal male external genitalia. His abdominal ultrasound showed poor peristalsis and minimal dilation of the bowel bilaterally, suggesting partial intestinal obstruction. Abdominal X-ray showed dilated bowels with multiple air-fluid levels and an empty rectum, features suggestive of intestinal obstruction. His blood workup showed a low serum sodium level of 117.82 mmol/L, potassium of 5.87 mmol/L, urea of 34.92 mmol/L, serum creatinine of 317 µmol/L, AST 29.66 U/L, and ALT 14 U/L. Serology for human immunodeficiency virus was negative. We reached a diagnosis of intestinal obstruction (IO), late-onset neonatal sepsis, and acute kidney injury secondary to dehydration. The baby was kept nil orally and started on intravenous (IV) fluids. He received 30 mls bolus of ringer's lactate for 30 minutes as shock management. He was thereafter kept on dextrose normal saline as maintenance and 3% hypertonic saline to correct the hyponatremia over 24 hours.

A urethral catheter was inserted to monitor urine output. The child was also started on IV Ceftriaxone 255 mg once

daily. After stabilizing and correcting electrolytes, on day three post-admission, laparotomy was done, and intra-operatively, there was a distended loop of the small intestine about 70 cm from the Treitz ligament with a blind end. The distal loop was collapsed with a meconium plug at its blind loop (Figure 1). Patency was established by an end-to-end ileo-ileal anastomosis of the blind loops by increasing the distal caliber using a 2-cm slit on the ant-mesenteric side. Distal loop patency was confirmed by injecting saline. The mesenteric vent was closed, one drain inserted, and the abdomen closed in layers after thorough abdominal lavage with saline. On the second day post-laparotomy, the baby was still very sick and on maintenance IV fluids. Control lab results revealed elevated serum creatinine of 244 µmol/L, serum sodium of 160.06 mmol/L, serum potassium of 2.87 mmol/L, serum urea of 34.33 mmol/L, total protein of 27.7 g/L, and albumin 15.94 g/L. He sustained multiple cardiac arrests over 3 days, remaining in critical condition after each resuscitation. On day 3 post-surgery, the child succumbed.

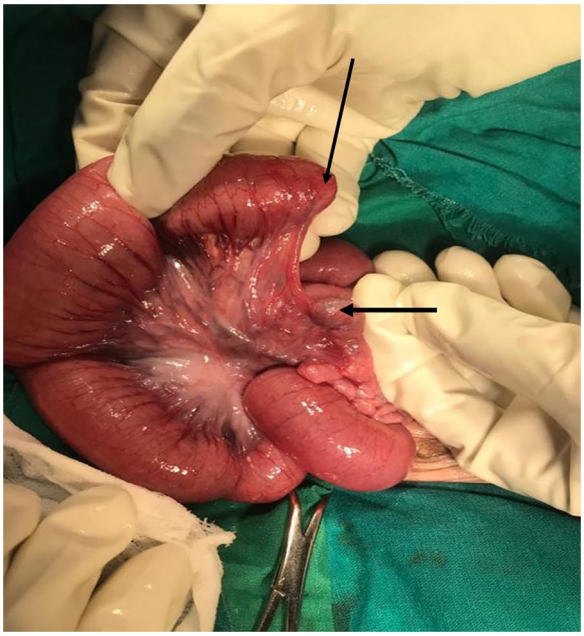

Figure 1. Atretic bowel loops; type 3A.

Case 2

A 5-day-old male baby was referred to our center with the chief complaint of vomiting since birth. The vomitus was greenish with the smell of feces and was mainly after being breastfed. He did not pass meconium until the third day of life, when he passed a small amount of dark mucoid stool. He had no fever and was passing urine normally. The baby was delivered vaginally at term with a birth weight of 3900 g. He cried immediately after birth and was breastfed within the first hour after delivery. The mother started antenatal clinic at 12 weeks of gestation, made 5 visits, and received supplements according to the national guidelines. She tested negative for HIV and syphilis. Mother was normotensive with normal blood sugar levels throughout her pregnancy. On examination, the baby had grade-5 jaundice, dehydrated with clubbed feet and his vitals were stable. The baby weighed 3100 g on admission. Abdominal examination showed a symmetrically distended abdomen with a hyper-tympanic percussion note and exaggerated bowel sounds. The rectum had little soft fecal matter. Duodenal atresia was suspected.

His abdominal X-ray showed dilated bowels with multiple air-fluid levels (Figure 2). Lab workup showed hemoglobin of 14.8 g/dL, potassium of 3.8 mmol/L, and sodium of 135 mmol/L. He was kept nil orally, given intravenous fluids, and scheduled for an emergency laparotomy. Intra-operatively, a completely atretic ileum approximately 45 cm proximal to the ileocecal junction was found (Figure 3). The proximal ileum was dilated while the distal ileum was collapsed. An ileal-ileal side-to-side anastomosis was done, and the small bowel

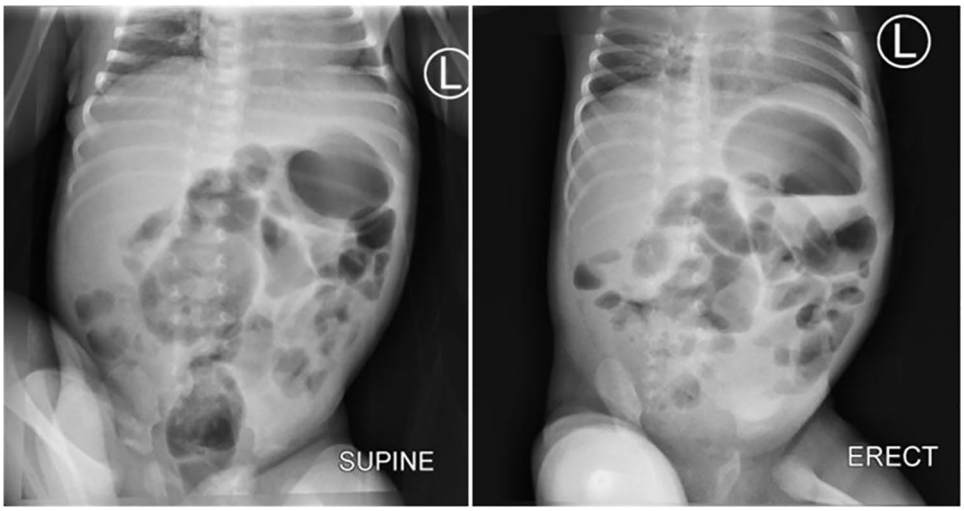

Figure 2. Plain abdominal X-ray showing double-bubble sign.

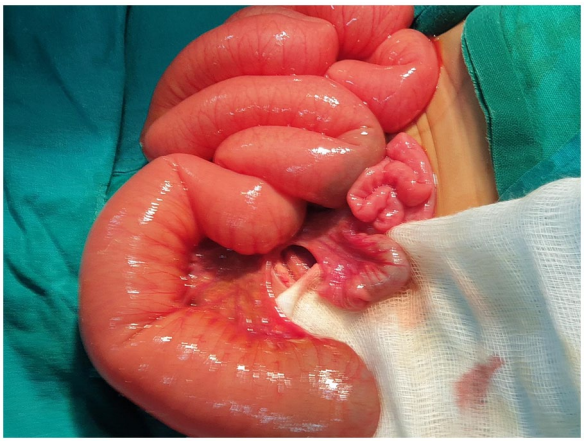

Figure 3. Type-3A jejunal ileal atresia.

continuity was established. The post-operative diagnosis was Type-3A mid-ileal atresia. The baby was kept nil per oral, given IV fluids; dextrose normal saline, IV antibiotics, and analgesia. Post-operatively, the baby continued to do well clinically, started to tolerate oral feeds, and passed stools normally. The jaundice gradually subsided. On day four, the child began having serous discharge from the abdominal incision with hyperemic skin; hence, surgical site infection was diagnosed. Culture of the wound discharge showed *Escherichia coli.* Debridement of the wound and daily dressing were done, and the patient was started on intravenous antibiotics. On the 20th post-operative day, the baby was discharged with oral cod liver oil according to local protocol. The mother was counseled to continue dressing at a nearby health center. He was reviewed 2 weeks later at the surgical outpatient unit, where the wound had healed fully, and the baby was tolerating feeds well. At 6 months of age, the baby had reached 9 kg and started weaning with no complaints.

Case 3

A 1-hour-old baby was brought from the labor ward with a complaint of difficulty in breathing. The baby was delivered by cesarean section due to a breech presentation. The mother reported experiencing vaginal leakage 3 days prior, which was non-foul-smelling and clear in color. The mother had an otherwise non-eventful pregnancy. The baby had an Apgar score of 4 and 6 in the first and fifth minute; resuscitation was done with a bag valve mask and suction. The baby was kept on oxygen and transferred to the neonatal unit. On examination, the baby was ill-looking, dyspneic on oxygen, not pale, not jaundiced, with a temperature of 36°C, pulse 160 bpm, respiratory rate 50 bpm, saturating 98% on oxygen. The baby had normal bronchial and vesicular breath sounds, normal heart sounds, and normal primitive reflexes on systemic examination. Examination of the abdomen found normal movements with respiration, no distention, and no organomegaly. The provisional diagnosis was a preterm baby at 33 weeks of gestation with appropriate weight for gestation age and early-onset neonatal sepsis. Two weeks later, when the patient was initiated on oral feeds, it was seen that she vomited everything and had abdominal distension. The vomitus contained bilious material and food contents. Hence workups to rule out intestinal obstruction were initiated.

Abdominal X-ray showed a double-bubble sign suggestive of duodenal atresia. The patient was scheduled for emergency laparotomy in which a type-C proximal duodenal atresia, a distended stomach, and collapsed bowel loops were found. Gastrojejunostomy was done to shorten the operating time, hemostasis was achieved, the abdomen closed in layers, and the wound dressed. The baby was nursed at the neonatal intensive care unit and maintained on IV fluids post-operatively. The baby was initiated oral feeds on day three but could not tolerate hence continued on IV dextrose. On day 5 post-surgery, the baby developed biliary leakage from the incision site and showed signs of disseminated intravascular coagulation (DIC). The baby succumbed to DIC and sepsis before revision of the surgery could be done.

Case 4

A preterm baby delivered at a gestation age of 33 weeks due to premature rupture of membranes was referred to on the third day of life due to vomiting. The baby had a birth weight of 2.3 kg and had Apgar scores of 8 and 10. The mother reported that the baby started vomiting yellowish material after breastfeeding from the first day of life. The vomitus was non-foul swelling, non-projectile, and contained mother's milk and yellow secretions. This condition was associated with abdominal distention, and the baby had not passed stools since birth. The pregnancy had been uneventful save for the premature rupture of membranes.

On examination, the baby had grade-1 jaundice, was not pale, not cyanosed, had a temperature of 34.5°C, pulse rate of 134 bpm, respiratory rate of 41 breaths/min, saturating 94% on room air, with a random blood glucose of 4.2 mmol/L. Abdominal examination found a dry umbilicus, decreased bowel sounds (1 per minute), and a patent anus on digital rectal exam. The sucking reflex was weak. The provisional diagnosis was a preterm baby at 33 weeks gestation by date with early-onset neonatal sepsis and pyloric stenosis.

An X-ray revealed a double-bubble sign (Figure 4), suggestive of duodenal atresia. The patient was booked for emergency explorative laparotomy. An upper transverse incision was made, which revealed proximal ileal atresia type-II with a dilated proximal segment of the duodenum and collapsed distally. Side-to-side ileal anastomosis bypassing the atretic area was done to establish patency, hemostasis achieved, and the abdomen closed in layers. The baby was kept on IV fluids, and oral feeds were initiated on day 3. The baby progressed well and by

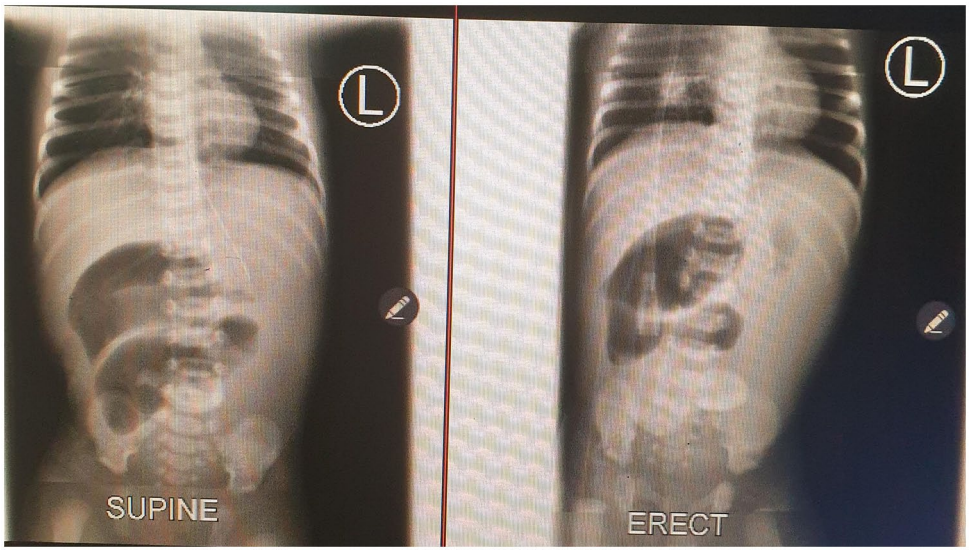

Figure 4. Double-bubble sign.

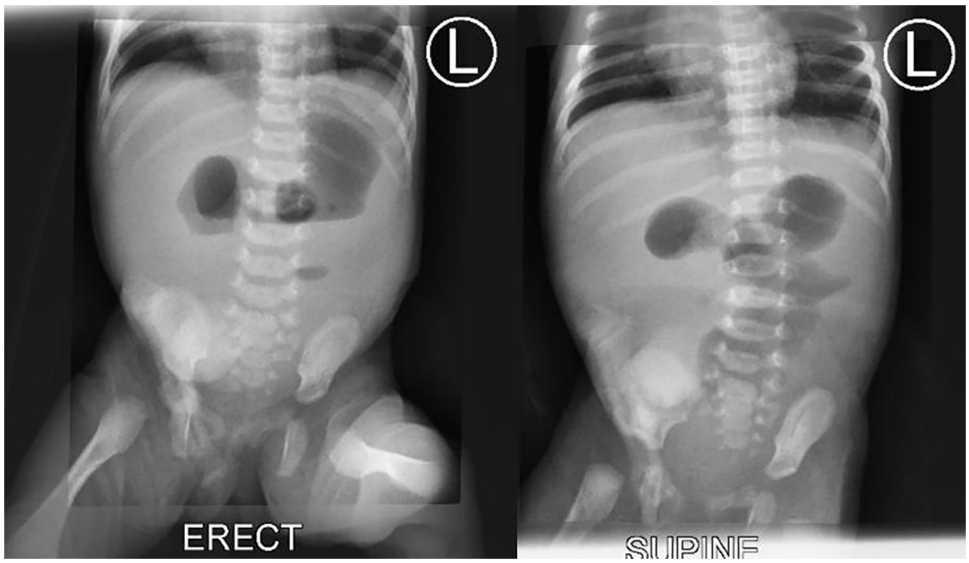

Figure 5. Plain abdominal X-ray suggestive of duodenal atresia.

the second week was able to breastfeed and was discharged. Follow-up in clinic until 6 months of age found the baby to be progressing well and had started weaning.

Case 5

A 3-day-old preterm baby was referred to us with a complaint of vomiting and not passing stool since birth. She had been born at a gestational age of 31 weeks and had a birth weight of 1.9 kg. The mother had an uneventful pregnancy but went into spontaneous labor at 31 weeks of gestation.

On initial examination, the baby was ill-looking, not pale, not cyanosed, mild dyspneic, and with a distended abdomen. Other vitals were within normal range. The provisional diagnosis was intestinal obstruction secondary to duodenal atresia. The patient was transferred to the neonatal intensive care unit (NICU), where IV fluids were initiated. Her plain abdominal X-ray (Figure 5) was suggestive of duodenal atresia. Emergency laparotomy was planned in which intraoperatively Type-1 jejunal atresia was identified (Figure 6) approximately 10 cm distal to the ligamentum Treitz. A longitudinal incision was made over the membranous area, the membrane was excised, and the bowel was repaired in a transverse manner to maintain patency. The patient was kept nil per oral for the first 24-hours after surgery and then initiated on oral feeds, which were gradually increased. She however could not tolerate feeds greater than 4 mls. On day 4, the baby developed a burst abdomen (Figure 7) and was taken to theater for an emergency laparotomy. The abdomen was cleaned thoroughly, and the repair was found to be intact; hence abdomen was closed in layers. Post-operatively

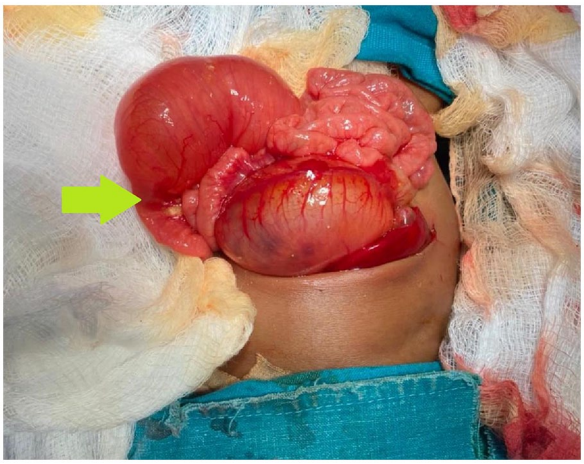

Figure 6. Type-1 JIA (green arrow).

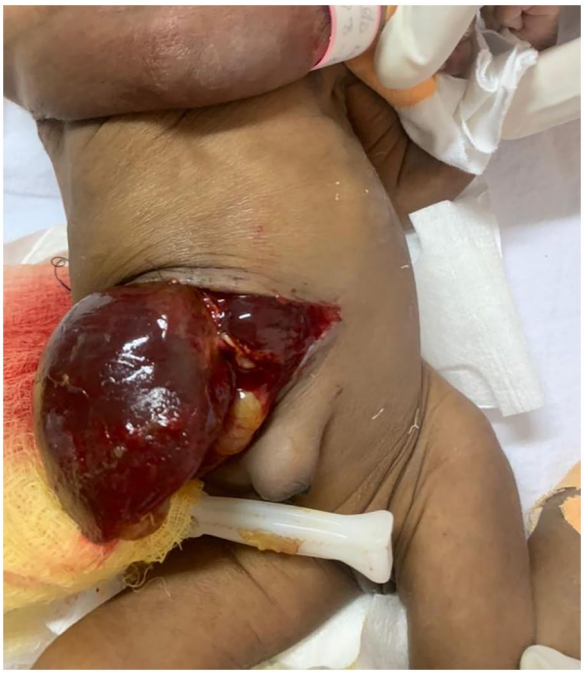

Figure 7. Burst abdomen with evisceration.

the baby started to tolerate up to 6 mls of expressed breast milk. The patient passed away suddenly from an unknown cause 4 days after the second surgery.

Discussion

The gastrointestinal tract arises via gastrulation from the endoderm of the trilaminar embryo during the third week of gestation and extends from the buccopharyngeal membrane to the cloacal membrane. During the fourth week, the 3 different portions (fore-, mid-, and hind-gut) extend the length of the embryo and will contribute to various components of the gastrointestinal tract.[8] Basic knowledge of gastrointestinal embryology is important to understand different types of congenital malformations. Failure of any of the embryological steps leads to predictable and often visible abnormalities on fetal scans.[8]

Gastrointestinal atresia is the most common cause of IO in neonates, duodenal atresia being the commonest type of atresia with an incidence of 1 in 6000 to 10 000 live births with a male predominance.[2,9] This gender predominance is seen in the index case series. Duodenal atresia can be diagnosed by a plain abdominal X-ray showing a "double-bubble" sign, as reported in case 3.[2] The incidence of jejunal-ileal atresia is approximately 1 in 5000 live births and 3 times more common in premature infants, as evident from cases 4 and 5 in the index series.[2] Colonic atresia is relatively uncommon, contributing 1.8% to 15% of all intestinal atresia with an incidence of 1 in 40 000 live births.[2] Contrast enema is needed to diagnose colonic atresia.[10] Hirschsprung's disease (HD) is usually present concomitantly in 2% of these cases; hence some authors suggest ruling out HD before establishing continuity of the colon.[2]

Gastrointestinal atresia is usually not inherited but can be associated with other congenital anomalies like VACTERL association, laryngotracheoesophageal cleft, or trisomy 21.[8] In all the index cases, other malformations were ruled out. Two proposed theories for the etiology are "imperfect recanalization" and "vascular insufficiency."[8,9] Polyhydramnios is a common feature seen on obstetric ultrasound, but is not always the case.[8]

Most neonates present with bilious vomiting and abdominal distention and should be considered mechanical obstruction until proven otherwise. Other features include jaundice in about a third of the cases, as in cases 2 and 4, and failure to pass meconium in the first 24 hours of life.[11] Radiological evaluation is required, and early surgical management improves the outcome and minimizes complications.[11] Surgical management depends on the type of atresia.[12] From our experience, there is a delay in presentation to tertiary centers, leading to poor outcome and increased mortality. This is partly due to a lack of knowledge of the referring medical attendants at the primary care centers. Poor health-seeking behaviors from the caretakers due to tradition and culture also play a role. In the rural areas, where the majority of the population lives, there is also limited access to pediatric surgeons, who are mainly in the major cities.[13]

Antenatal sonography and magnetic resonance imaging (MRI) can be used to diagnose intestinal atresia in the fetal period by the presence of dilated bowel loops and polyhydramnios; hence it is important to elicit such history from the caretakers if they underwent obstetric ultrasound during their pregnancy.[11,14] From our experience, none of the caretakers could give the details of the obstetric ultrasound. Abdominal ultrasound and plain abdominal X-rays can aid in confirming the diagnosis of atresia by the presence of "double-bubble" sign or "triple-bubble" sign on an erect plain abdominal X-ray as evident in cases 3, 4, and 5.[11]

A review by Miscia et al showed that open surgery was the preferred mode of surgical intervention in correcting duodenal atresia compared to the laparoscopic method. This is because it

is easier to look for missed intestinal atresia in open surgery than in laparoscopy.[15] In our setting, with the most available technique being open surgery, a comparative outcome could not be established as was the only available technique.

We followed the cases for up to 6 months, during which 3 of the patients died, and 2 survived. Delays in presentation, as well as failure to diagnose and manage associated anomalies, might have played a role in the poor outcome of our cases. It highlights the need for early detection and training health care providers on the proper use of imaging in early pregnancy to detect such abnormalities so that early referral to a tertiary center for management can be planned. Generally, pediatric surgery in Africa is practiced in a resource-limited environment with suboptimal facilities and limited trained human resources, particularly pediatric surgeons; hence many emergency pediatric surgeries are performed by general surgeons.[13,16] This paper also highlights the need to train medical personnel in rural health centers on emergency conditions and to create awareness of such conditions though they are not as common as other surgical conditions like trauma.[16]

Conclusion

Intestinal atresia is common cause of intestinal obstruction and is a common neonatal surgical emergency. Early diagnosis allows clinicians adequate antenatal care by a multidisciplinary team, improving prenatal outcome and prognosis. It should be kept in mind as a possibility whenever there is poly-hydramnios. More data is needed in the field of pediatric surgical conditions in sub-Saharan African countries and the scale-up of pediatric surgery services across the countries.

Declarations

Ethical approval and consent to participate

Approval was obtained from the Department of General Surgery (Kilimanjaro Christian Medical Center), and the appropriate hospital institutional review board has approved the publication of this case series. Consent to participate in this case series was obtained from the parents of each patient.

Consent for publication

Written informed consent was obtained from the patient's parent(s) for publication for this case series; additionally, accompanying images have been censored to ensure that the patients cannot be identified. A copy of the consent(s) is available on record.

Author contribution(s)

Jay Lodhia: Conceptualization; Project administration; Supervision; Writing—original draft; Writing—review & editing. Hilary Chipongo: Conceptualization; Data curation; Writing—original draft. Beatrice Mathew: Writing—original draft. David Msuya: Data curation; Methodology; Writing—review & editing. Samwel Chugulu: Investigation; Methodology; Writing—review & editing. Rune Philemon: Conceptualization; Writing—original draft; Writing—review & editing.

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Availability of data and materials

Not applicable

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REFERENCES

1. Rao LD, Rao KV, Kameswari K. Neonatal intestinal obstruction. *J Dent Med Sci*. 2018;17:50-53.
2. Singh V, Pathak M. Congenital neonatal intestinal obstruction: retrospective analysis at tertiary care hospital. *J Neonatal Surg*. 2016;5:49.
3. King A. *Intestinal Atresia*. UpToDate; 2022. Accessed 1 November, 2022. https://www.uptodate.com/contents/intestinal-atresia
4. Verma A, Rattan KN, Yadav R. Neonatal intestinal obstruction: a 15 year experience in a Tertiary Care Hospital. *J Clin Diagn Res*. 2016;10:SC10-SC13.
5. Chiarenza SF, Bucci V, Conighi ML, Zolpi E, Costa L, Fasoli L, et al. Duodenal atresia: open versus MIS repair—analysis of our experience over the last 12 years. *Biomed Res Int*. 2017;2017:1-7.
6. Tsitsiou Y, Calle-Toro JS, Zouvani A, Andronikou S. Diagnostic decision-making tool for imaging term neonatal bowel obstruction. *Clin Radiol*. 2021;76:163-171.
7. Wessel LM, Fuchs J, Rolle U. The surgical correction of congenital deformities: the treatment of diaphragmatic hernia, esophageal atresia and small bowel atresia. *Dtsch Arztebl Int*. 2015;112:357-364.
8. Tonni G, Grisolia G, Granese R, et al. Prenatal diagnosis of gastric and small bowel atresia: a case series and review of the literature. *J Matern Fetal Neonatal Med*. 2015;29:1-9.
9. Osmulikevici O, Renji E, Jaffray B, Embleton N. Isolated ascites in a newborn with 'apple peel' jejunal atresia. *BMJ Case Rep*. Published online October 3, 2017. doi:10.1136/bcr-2017-219781
10. El-Asmar KM, Abdel-Latif M, El-Kassaby A-HA, Soliman MH, El-Behery MM. Colonic atresia: association with other anomalies. *J Neonatal Surg*. 2016;5:47.
11. Sukewanti NM, Artana IWD, Putra PJ, Kardana IM, Sukmawati M, Ariyanta KD. Jejunal atresia in the newborn: three cases after resection and end-to-end anastomoses. *Intisari Sains Medis*. 2020;11:211.
12. Kumar P, Kumar C, Pandey PR, Sarin YK. Congenital duodenal obstruction in neonates: over 13 years' experience from a single centre. *J Neonatal Surg*. 2016;5:50.
13. Chirdan LB, Ameh EA, Abantanga FA, Sidler D, Elhalaby EA. Challenges of training and delivery of pediatric surgical services in Africa. *J Pediatr Surg*. 2010;45:610-618.
14. Rubio EI, Blask AR, Badillo AT, Bulas DI. Prenatal magnetic resonance and ultrasonographic findings in small-bowel obstruction: imaging clues and postnatal outcomes. *Pediatr Radiol*. 2017;47:411-421.
15. Miscia ME, Lauriti G, Lelli Chiesa P, Zani A. Duodenal atresia and associated intestinal atresia: a cohort study and review of the literature. *Pediatr Surg Int*. 2019;35:151-157.
16. Mhando S, Young B, Lakhoo K. The scope of emergency paediatric surgery in Tanzania. *Pediatr Surg Int*. 2008;24:219-222.