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CASE SERIES

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Type 3B jejunoileal atresia management at a tertiary hospital in northern Tanzania: A report of three cases

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Key Clinical Message

Jejunalileal atresia is a cause of intestinal obstruction in the newborn, hence a surgical emergency. Prenatal diagnosis can be made by simple obstetric ultrasound and postnatal by plain abdominal x-ray to plan a multidisciplinary approach to reduce morbidity and neonatal mortality.

Abstract

Atresia can occur anywhere along the intestines and is a common cause of intestinal obstruction in neonates. Jejunoileal atresia (JIA) is a rare disease occurring in 2.1 per 10,000 live births. Type 3b jejunoileal atresia occurs in 11% of all small bowel atresia. We present three cases of type 3b jejunoileal atresia. They were all missed by prenatal ultrasonography, and presented with features of intestinal obstruction. The diagnosis was confirmed by plain abdominal x-rays and ultrasound, followed by laparotomy. Postoperative care was given in the neonatal unit according to local protocols. One recovered, however, two succumbed from neonatal infection. Jejunoileal atresia requires surgery and long postoperative care, with outcomes associated with numerous prognostic factors including multidisciplinary care and neonatal intensive care. Jejunoileal atresia is less commonly associated with other congenital anomalies, unlike duodenal atresia. Efforts are needed to scale up prenatal diagnosis of jejunoileal atresia, and therefore to plan for appropriate care after delivery. Also, further studies are needed to understand neonatal sepsis in the postoperative period and ways to improve outcomes.

KEYWORDS

atresia, case series, gastrointestinal tract, intestinal obstruction, neonate

1 | INTRODUCTION

Atresia is a congenital defect of a hollow organ in which there is complete obstruction of the lumen of the organ. Atresia can occur anywhere along the intestines and is a common cause of intestinal obstruction in newborns. About 50% of atresia of the intestine occur in the duodenum, with a global incidence of approximately 1 to 2 neonates per 10,000 births.[1] Jejunal or ileal atresia occurs in approximately 20% of all small intestine atresia with an

incidence of around 0.7 per 10,000 births. Colon atresia is the least common type of intestinal atresia occurring in about 10% of all intestinal atresia with an incidence of about 0.25 per 10,000 births.[2,3]

Jejunoileal atresias were first classified by Grosfeld et al into types 1, 2, 3a, 3b, and 4 (Table 1).[4] Type 3b, also known as apple peel small bowel syndrome, are the least common, yet the most severe congenital jejunoileal malformations.[4]

2 | CASE REPORTS

2.1 | Case report 1

2.1.1 | Case history/examination

A 7-day-old female baby presented with abdominal distension 3 days after birth. It was of gradual onset, worsening with time, and associated with vomiting bilious contents after feeds. Yellowish discoloration of the skin was reported for 1 day, started from the eyes, then spread to the extremities; however, fever was denied. Mother reported the baby to have passed scanty amount of greenish mucus per rectum 4 days after birth. The baby was born by spontaneous vaginal delivery, weighing 2700 g at 37 weeks of gestation, cried immediately after birth and was breastfed during the first hour of life. Mother booked antenatal clinic at 25 weeks of gestation where she attended three visits, received all supplements with an uneventful pregnancy. Mother's age was 38 years and this was her fourth child.

On examination, the baby was jaundiced, with orogastric tube drainage in situ draining bilious content. Body temperature was 37.7°C, respiratory rate was 42 breath/min, pulse rate of 162 beats/min, saturating 94% on room air, and random blood glucose was 5.1 g/dL. The abdomen was symmetrically distended, and the umbilicus was clamped, hyperemic and wet. The abdomen was nontender, organomegaly was hard to ascertain and normal findings in other systems.

He had a leukocyte count of 13.89×10^9/L (4.00–11.00×10^9/L), hemoglobin of 19.8 g/dL (10.7–17.1 g/dL), platelet count of 378×10^9/L ($150–500\times10^9$/L), human immunodeficiency virus (HIV) was negative, blood group A Rh-positive, serum creatinine of 348 µmol/L (44–88 µmol/L), blood urea nitrogen (BUN) of 23.09 mmol/L (0.00–8.30 mmol/L), serum potassium and sodium within normal range, total bilirubin of 385.0 µmol/L (<17 µmol/L), and conjugated bilirubin of 104.97 µmol/L (<5 µmol/L). Abdominal-pelvic ultrasound showed gaseous abdomen with dilated bowel loops, and peristalsis was not appreciated. Plain abdominal x-ray concluded features of jejunoileal atresia (JIA) (Figure 1).

2.1.2 | Methods

After thorough counseling, the baby was taken for laparotomy whereby type 3b JIA was found. End-to-side

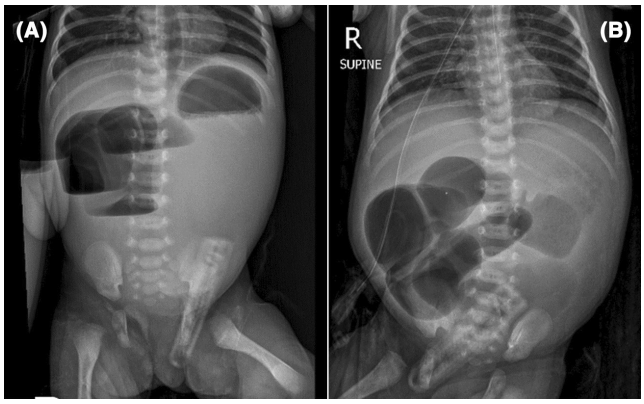

FIGURE 1 (A) Erect abdominal x-ray showing multiple air-fluid levels. (B) Supine x-ray showing "double-bubble" sign.

TABLE 1 Table describing Louw and Barnard (1955) classification of jejunoileal atresia.

Type	Description
Type 1	Intestine looking completely intact but with a membrane obstructing the lumen
Type 2	Small intestine is divided into blind ending proximal and distal parts connected by a fibrous cord
Type 3a	Two completely separate blind ending loops of small intestine
Type 3b	Defect of mesentery with the distal loop coiled into apple peel shape and absence of distal portion of superior mesenteric artery
Type 4	Multiple atretic segments with the appearance of multiple sausages

anastomosis was done and Bishop–Koop stoma of the distal limb was raised (Figure 2). No bowel resection was carried out hence the length of the small bowel was approximately 110 cm and colon was 50 cm as the whole length of bowel was found viable.

2.1.3 | Conclusion and results

The baby was initiated on small oral feeds, stoma care, wound care, intravenous (IV) antibiotics and maintenance fluids. The baby succumbed on Day 23, with the possible cause of death being late onset neonatal sepsis.

2.2 | Case report 2

2.2.1 | Case history/examination

A female baby delivered at our facility at 32 weeks gestation age, cried immediately, with APGAR score of 8 and 9 at 0 and 5 min, respectively, weighed 1900 grams. Mother received dexamethasone before delivery for lung maturation. Mother denied history of premature rupture of membranes. Mother was tested IgG positive for toxoplasmosis and rubella. Mother was normotensive and normoglycemic throughout pregnancy. This was the second pregnancy.

Upon examination, the baby was alert, active, and pink on room air. She was afebrile, not pale, not jaundiced but slightly dyspneic with a respiratory rate of 63 per minute, saturation of 98%, and a temperature of 36.7°C. She had normal abdominal contour, soft, clean clamped umbilical cord, normal female genitalia, and patent anus. On Day 2 of life, she started vomiting after breastfeeding and developed jaundice. Total bilirubin was 215 μmol/L, conjugated bilirubin was 1.45 μmol/L, and C-reactive protein was 1420 mg/L (<10 mg/L). Bowel was not distended, soft, and bowel sounds were present. She was started on IV ampicillin and gentamicin according to local protocol. Orogastric tube (OGT) was inserted and was kept on IV 10% dextrose.

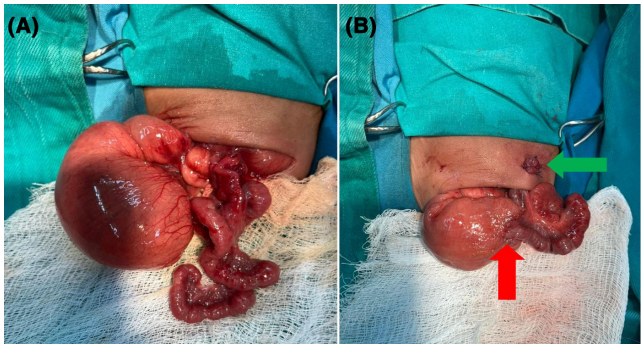

FIGURE 2 (A) Operative photograph showing JIA 3b. (B) Anastomosis (red) and Bishop–Koop stoma (green).

2.2.2 | Methods

On Day 3, the baby continued to drain bilious contents with mild abdominal distention; hence a plain abdominal X-ray was done that revealed "double-bubble" sign (Figure 3). The mother was counseled on the condition and baby was taken for emergency laparotomy, where intraoperatively type 3b JIA was appreciated, and the colon was collapsed. The length of the small intestine from the ligament of Treitz to ileocecal junction was approximated to be 100 cm with no necrotic segments. Side-to-side anastomosis was carried out of the proximal distended jejunum with distal collapsed ileum, patency was established, and the abdomen was closed (Figure 3).

Postoperatively she was kept on IV ceftriaxone and metronidazole for 10 days, nil orally for 48 h, and IV dextrose normal saline (DNS). Colostrum swabs were started on the third day postsurgery, and expressed breast milk was started Day 4 post-surgery at a rate of 2 mL every 3 h.

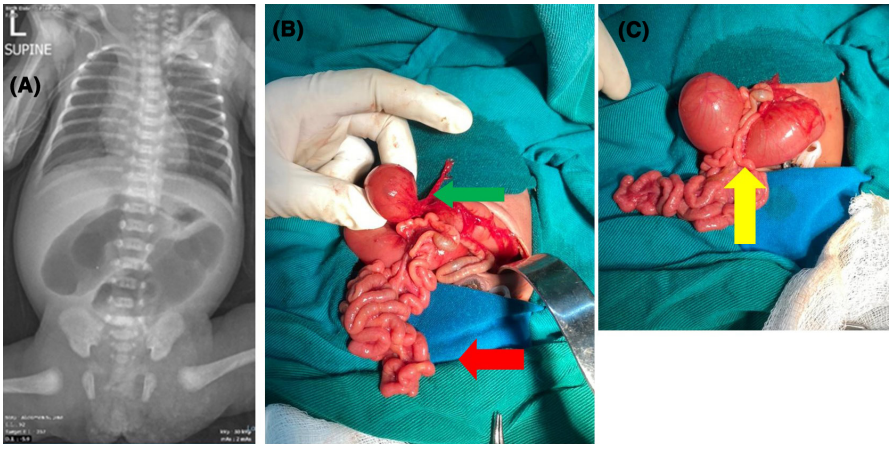

FIGURE 3 (A) Plain abdominal x-ray showing "double-bubble" sign. (B) Operative photograph showing JIA type 3b with proximal distended limb (green) and distal collapsed bowel loops (red). (C) Side-to-side anastomosis (yellow).

2.2.3 | Conclusion and results

On Day 8 postsurgery, the jaundice resolved, and she was tolerating 5 mL/3 hourly of expressed breast milk per oral and passed small amounts of stools rectally. She had developed a superficial surgical site infection and hence was dressed daily with saline and mupirocin topical cream. On Day 20 postsurgery, the surgical site had healed, was tolerating 25 mL of expressed breast milk, and was passing stools. However, on Day 21 after surgery (25 days of life), she started to desaturate, efforts to intubate and resuscitate were not successful and she succumbed. The cause of death documented to be respiratory failure due to septicemia and prematurity.

2.3 | Case report 3

2.3.1 | Case history/examination

A 1-day male baby was referred to our center due to abdominal distention and vomiting. Caretaker reported the baby passed meconium and cried immediately after delivery by caesarian section at 36 weeks gestation age due to non-reassuring fetal status with premature rupture of membranes and severe oligohydramnios. He weighed 2400 g at birth. There was neither history of cyanosis nor difficulty in breathing reported. Antenatally, the mother was clinically stable with normal blood pressures and glycemic control.

Upon examination, the baby was active and alert, pink, not jaundiced, and not cyanotic. His vital signs were within normal range, saturating at 96% on room air, and blood glucose of 3.9 mmol/L. His abdomen was moderately distended on the upper quadrants, soft to touch with normal bowel sounds. The umbilical cord was clamped, clean, and dry. Other systems were unremarkable. The baby was kept nil per oral and initiated on intravenous antibiotics and maintenance fluids, and orogastric tube was inserted and was draining bilious contents.

2.3.2 | Methods

C-reactive protein was negative, normal serum sodium level, moderate hyperkalemia of 6.14 mmol/L (3.5–5.1 mmol/L) (hence managed), and hemoglobin of 17.5 g/dL. Plain abdominal x-ray revealed a double-bubble sign suggestive of jejunal atresia (Figure 4). Abdominal ultrasound showed dilated bowel loops with reduced peristalsis. The baby was taken for an emergency laparotomy whereby intraoperatively a type 3b JIA was found. The blind ends of the bowels were resected (approximately 2 cm from each end), and end-to-side jejuno–jejunal anastomosis was performed to establish gastrointestinal patency leaving about 100 cm of remaining small bowel length (Figure 4).

2.3.3 | Conclusion and results

The baby recovered well postoperatively and was tolerating feeds well gradually. On Day 4, he sustained a burst abdomen. He underwent an emergency exploratory laparotomy and found to have anastomotic leaked. The anastomosis was revised and the abdomen was closed with tension sutures. The baby then fared well pos-operatively in the neonatal ICU where feeding was progressed gradually and intravenous albumin was infused. The baby progressed well and was discharged on Day 16. He was then reviewed at the surgical outpatient clinic on Days 14 and 30 postdischarge, respectively, where was tolerating feeds well and the incision had healed. He also had an adequate weight gain.

3 | DISCUSSION

Type 3b jejunoileal atresia is the least common, most severe form of intestinal malformations, and with poorest outcome compared to other types.[4] Type 3b JIA occurs in 11% of all small bowel atresia. Jejunoileal atresia is caused by an ischemic insult to a segment of the developing fetal bowel during pregnancy. Because fetal bowels are sterile, the gangrenous portion is resorbed into the body along with its necrotic mesentery leaving proximal and distal ends. This vascular insult in the fetus can be caused by intussusception, segmental midgut volvulus, strangulated internal hernia, or thromboembolism.[5]

Maternal risk factors associated with the condition include cigarette smoking and cocaine use and use of vasoconstrictive medications, such as ephedrine and pseudoephedrine, during the first trimester.[6] A fetus with cystic fibrosis may develop jejunal or ileal atresia as a result of segmental small bowel obstruction by the thickened meconium. Fetus with gastroschisis is also more likely to develop jejunoileal atresia from strangulation of intestines in the abdominal wall fascial defect or volvulus of the herniated midgut.[6] Fetal inherited clotting disorders leading to thrombus formation and family history of jejunoileal atresia have been reported as fetal risk factors.[7] No risk factors were identified among the patients we present.

Clinical features of jejunoileal atresia may be picked during prenatal ultrasound by observing signs of fetal intestinal obstruction associated with polyhydramnios and

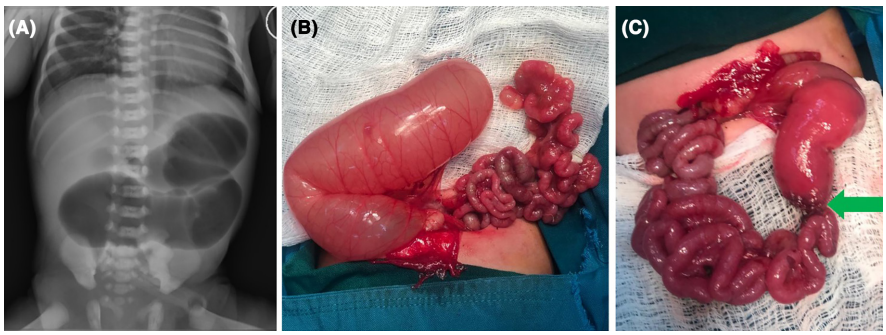

FIGURE 4 (A) Supine abdominal x-ray showing "double-bubble" sign. (B) Photograph showing JIA type 3b with distended proximal jejunum and collapsed distal bowel loops (ileum). (C) Anastomosis done (green) and note the filling of the distal bowel loops.

ascites or become apparent on the first or second day of life. Postnatal signs and symptoms include abdominal distension, bilious vomiting, failure to pass stool, and irritability. A history of maternal polyhydramnios is common.[8] All cases that we present were diagnosed in the postnatal period, despite attending obstetric clinics and doing obstetric ultrasonography. They were presenting with features of intestinal obstruction. In Case 3, there was maternal severe oligohydramnios rather than polyhydramnios.

Fetal congenital gastrointestinal anomalies represent one of the common causes of maternal polyhydramnios.[9,10] About 5% of pregnant women with polyhydramnios deliver babies with polyhydramnios.[9] The severity of polyhydramnios correlates with the degree of congenital malformation.[9,10] The presence of polyhydramnios on prenatal ultrasound should raise suspicion on the presence of congenital gastrointestinal, central nervous system, and skeletal system anomalies.[9,10]

Premature rupture of membranes (often times missed by the mother) and placental insufficiency are common causes of maternal polyhydramnios.[11,12] The presence of premature rupture of membranes in Case 3 of the cases reported in this manuscript explains the presence of oligohydramnios in pregnancy that resulted in delivery of a baby with a congenital jejunoileal atresia.

Plain abdominal x-ray will reveal air-fluid levels with absence of air in the colon and classical "double-bubble" sign as evident in our cases. Barium enema will reveal a microcolon. Upper gastrointestinal contrast study should be carried out to rule out malrotation with midgut volvulus which is a true surgical emergency.[13]

The definitive management of JIA involves surgical repair. Prior to surgery, neonates with jejunoileal atresia should be placed nil orally, nasogastric/orogastric tube should be inserted for gastric decompression, and they should be hydrated with intravenous fluids.[2] During surgical repair, the entire small intestine should be inspected for areas of atresia. Atretic segments should be resected and primary anastomosis should be considered. If the proximal segment is edematous, ileostomy may be raised and anastomosis deferred to a later time.[14] In Cases 2 and 3, primary anastomosis was done. In Case 1, end-to-side anastomosis was performed and Bishop–Koop stoma was raised. The baby was able to tolerate oral feeds postsurgery.

In Case 2, healing was delayed due to surgical site infection. However, the patient succumbed to neonatal sepsis that was not directly related to the surgery. Prematurity and low birth weight as were in this case, and the presence of other congenital cardiac and abdominal wall defects are associated with high mortality.[1,15] Late presentation and postoperative complications are associated with increased mortality.[14] In Case 3, primary anastomosis was complicated by anastomotic leak and burst abdomen that required re-operation, thereafter recovered.

Prognosis depends on length of viable bowel that remains after surgical resection of the atretic segment and the presence of ileocecal valve. Neonates who develop short bowel syndrome postoperatively need to be placed on total parenteral nutrition. Subsequently, they should be started on enteral feeds to facilitate adaptation to the remaining segment of bowel and to maintain sucking and swallowing reflexes. Ultrashort bowel syndrome requires surgical lengthening procedures such as serial transverse enteroplasty (STEP) or small bowel transplantation.[16] We managed three patients who were placed on dextrose containing intravenous fluids for nutrition postoperatively. Simultaneously, these patients were started on colostrum swabs Day 2 postsurgery, and enteral feeding per orogastric tube on Day 3. Feeding started with 2 mL of expressed breast milk per 3 h and was gradually increased. They both were able to tolerate oral feeds.

In low- and middle-income countries, jejunoileal atresia is still associated with high morbidity and mortality because of late presentation and limited access to neonatal intensive care unit services and parenteral nutrition.[14] Two out of three patients died in the neonatal intensive care unit on Days 23 and 25 of life from causes related to sepsis. The first case was referred to our center 7 days after birth, with clinical features suggestive of intestinal obstruction that had become apparent 4 days prior to referral. This patient was relatively unstable throughout her stay in the neonatal intensive care unit (NICU). The

second case was delivered at our center at a premature gestational age. The diagnosis of intestinal obstruction was established early, and the baby was operated and progressed well postoperatively. However, while in the ward, after completion of postoperative antibiotics, and at a time, she was tolerating enteral feeds, she developed features of sepsis and succumbed from septicemia. A third case was received on the first day of life. Diagnosis of intestinal obstruction was established on the same day and he was operated urgently, after which he recovered although with some complications.

Management of jejunoileal atresia is still associated with high morbidity and mortality. This is attributed to inadequate close follow up to this delicate group of patients. Efforts are needed to scale up prenatal diagnosis of jejunoileal atresia, and therefore to plan for appropriate care after delivery. Also, further studies are needed to understand neonatal sepsis in the postoperative period and ways to improve outcome.

AUTHOR CONTRIBUTIONS

Evance Salvatory Rwomurushaka: Writing – original draft. **David Msuya:** Data curation; supervision. **Robert Mbwambo:** Methodology. **Jay Lodhia:** Conceptualization; writing – original draft; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors declare they have no competing interests.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

ETHICS STATEMENT

Ethical approval was obtained from the Department of General Surgery, Kilimanjaro Christian Medical Centre.

CONSENT

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

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